Cardiac Arrhythmia Management
A Practical Guide for Nurses and Allied Professionals
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To my husband Boris, my son Feliks, and my parents for their encouragement, love, support, and many sacrifices. To all my teachers for their wisdom and inspiration.

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Paul J. Wang
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Foreword by N.A. Mark Estes III

Over the last three decades, we have witnessed a remarkable expansion and maturation of the fields of cardiac electrophysiology, ablation, pacing, and defibrillation. This has resulted in a core of highly skilled nurses, physician assistants, technicians, and industry-employed allied professionals assuming a larger and increasing important role in the clinical care of patients with arrhythmias. Given the considerable progress in the clinical evaluation and management of patients, the need has emerged for a comprehensive educational resource to ensure the highest quality of care. Cardiac Arrhythmia Management: A Practical Guide for Nurses and Allied Professionals merits particular recognition as it uniquely fulfills this need. It represents a timely and novel contribution that should be considered essential for all health care professionals involved in the care of patients with heart rhythm disorders.

Amin Al-Ahmad, MD, Paul J. Wang, MD, Andrea Natale, MD, Angela Tsiperfal, RN, NP, Linda Ottoboni, RN, MS, and Salwa Beheiry, RN, as editors, have masterfully selected topics and authors to produce an essential educational resource. All sections, including those on anatomy, physiology, arrhythmia mechanisms, pacemakers, defibrillators, pediatric arrhythmias, syncope, sudden death, and ethical issues, are superbly written by leading clinical educators. The case-based approach supplements the didactic materials. This allows the practical application of both clinical and technical knowledge to the individual case. As always, this markedly enhances information retention and clinical utility.

The editors and authors are to be congratulated for producing this unique, practical, and comprehensive book. All interested in improving their knowledge and skills related to arrhythmias, ablation, pacing, and defibrillation should consider it an essential resource. With mastery of its content, all health care professionals will meaningfully improve their ability to ensure optimal patient outcomes.

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The role of allied professionals in providing care to patients with cardiac rhythm management devices is constant yet dynamic. While this statement may seem paradoxical, its truth is self-evident to all who work in this rapidly growing subspecialty within cardiology.

One has only to look at the history of cardiac electrophysiology to discern the truth of constancy amid innovation. Today’s devices are a testament to the visions of scientists who, working in concert with engineers and medical professionals, have produced devices that are life saving and life enhancing. Innovative catheter-based technologies and cardiac rhythm management devices present new challenges. As visions become reality and theory is applied to clinical practice, new operational features are assessed, evaluated, and integrated to ensure the provision of safe, optimal patient care.

Our goal always has and always shall be providing safe and optimal patient care. Daily, we strive to incorporate how to best assess available evolving therapies and an ever-expanding array of physiologically based device features and device-based diagnostic data as we evaluate patients “in person” and “remotely.” Efforts to contain health care costs impact us and we are expected to be not only proficient but also efficient. We seek to apply scientific principles as we navigate and plumb the depths of devices, their programmers, and remote Web sites to evaluate data and ensure devices are optimally programmed to meet the needs of individual patients. How to assess and address the unique challenges of device-based care permeate this text, which integrates the theoretical and the practical. In this regard, the text for allied professionals espouses and exemplifies the standards of professional practice for allied professionals in pacing and electrophysiology (Gura et al. 2003).

Devices represent only one of the varied therapy options. The challenges to understand and apply technology to the management of life-threatening/life-altering arrhythmias in patients are vast. A thorough understanding of arrhythmia mechanisms provides an essential foundation for identifying the most appropriate technique for patient treatment. Advanced diagnostic testing creates additional patient-specific information that results in an optimal treatment decision. Within the electrophysiological procedure and ablation, innovative technological achievements have simplified
arrhythmia location, improved catheter ablation therapy techniques, and reduced patient complications. All of these are outlined in the text so that caregivers can deliver improved patient care with an understanding of the pathophysiology and biomedical technology. One must also consider the “care” in patient care. We strive to do so while acknowledging the unique human qualities and quirks of our patients, recognizing that the ordinary and mundane to one person may represent the unique, provoking anxiety and distress to others.

The authors and editors of this text provide exemplary material designed to teach us how to utilize technology to enable each of our patients to derive maximum benefit.

Rosemary S. Bubien, RN, MSN, FAHA, FHRS, CCDS

REFERENCE

The field of cardiac arrhythmias has evolved greatly over the past several decades. This field involves direct patient management of patients who have implantable cardiac devices such as pacemakers or implantable cardioverter defibrillators. In addition, the role of catheter ablation in these patients has expanded over the past few years for arrhythmia management. The front line in management of arrhythmia patients is often nursing or allied professional staff that works closely with cardiologists and cardiac electrophysiologists.

This aim of this book is to be a comprehensive reference for allied professionals in a very specialized field. The book is divided into six sections that cover the variety of topics in the field of arrhythmia management, from the most basic to the complex. Each chapter was written and edited by experts in the field and was the collaboration of electrophysiologists and allied professionals.

Our goal is to provide the fundamentals and nuances in management of cardiac arrhythmia devices, as well as arrhythmia management for patients who undergo radiofrequency ablation procedures.

We hope that this book will be used by both experienced and novice nurses and allied professionals. We also hope this book may be useful for those preparing for any examination of competency in the field and will be valuable as a learning guide as well as a useful resource on a day-to-day basis. We hope that this book will contribute to the improvement in care of the arrhythmia patient.

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Cardiac Arrhythmia Management

A Practical Guide for Nurses and Allied Professionals
Section 1
Basics of Cardiac Anatomy and Electrophysiology
Electrical stimulation is the key in initiating the sequence of events that result in cardiac contraction, the ultimate measure of cardiac performance. The inherent pacing properties that are required to generate an electrical impulse, the intrinsic conduction pathways that move depolarization from the initial impulse throughout the entire cardiac muscle, and finally, the patterns of depolarization that create an optimal squeeze of the cardiac muscle are the result of the electrical conduction system and mechanical system functioning synchronously. Impulse generation and dispersion to all areas of the heart muscle via cell-to-cell activation and via electrical pathways must be well understood to comprehend the complexity of electrical conduction and the strategies for treating conduction abnormalities. This chapter will provide an overview of cellular physiology, electrical physiology, the anatomy of the conduction system, and the medications that can be used to treat conduction abnormalities. A thorough understanding of the normal anatomy and physiology of the conduction system will enable the allied professional to understand the rationale for utilizing specific arrhythmia treatment modalities, whether it be medications, ablations, or devices.

ANATOMY OF THE CARDIAC CONDUCTION SYSTEM

The anatomy of the conduction system is composed of electrical tracts within the myocardium. This electrical network is strategically arranged in the nodes, bundles, bundle branches, and branching networks of fascicles. The cells that form these structures lack contractile capability but can generate spontaneous electrical impulses and alter the speed of electrical conduction throughout the heart. The sinoatrial (SA) node, internodal tracts, atrioventricular (AV) node, bundle of His, right
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within the heart and is composed of cells capable of impulse formation or “pacing.” Pacing cells within the SA node independently move to a threshold potential, thereby initiating depolarization. The SA node establishes the intrinsic heart rhythm between 60–100 pulses per minute but is influenced by the autonomic nervous system to meet the changing requirements of the body (Fig. 1.1.2). The region of the sinus node has numerous nerve endings and is predominantly regulated by the parasympathetic system or acetylcholine at rest and the sympathetic tone is mediated with the release of norepinephrine to meet increased energy requirements.

Anatomically, the SA node is subepicardially located in the left upper corner of the right atrium, near its junction with the superior vena cava. The SA node is the native pacemaker site

bundle, left bundle, anterior and posterior fascicles, and the Purkinje fibers are all the necessary conduction routes established throughout the cardiac muscle (Fig. 1.1.1). Normal conduction utilizes this electrical conduction system to expedite transmission of the electrical impulse from the top of the heart to the bottom. Abnormal conduction or arrhythmias are the result of an arrhythmogenic site or region that interferes, alters, or bypasses the normal conduction circuit. Therefore, a comprehensive understanding of normal conduction provides a foundation for better understanding the mechanisms present in abnormal conduction or arrhythmias.

Figure 1.1.1 Anatomy of the conduction system.
flow (Anderson et al. 1979). The function of the sinus node may be jeopardized if the blood supply is reduced due to coronary artery disease or an increase in fibrous tissue with maturity, resulting in fewer SA cells available for impulse formation within the sinus node (Davies and Pomerance 1972).

Once the impulse is initiated within the SA node, it not only travels cell to cell through the atrium but also utilizes more specialized, expedient pathways known as internodal tracts (Fig. 1.1.1). The Bachmann’s bundle moves away from the SA node anteriorly around the superior vena cava and then bifurcates with one branch leading from the right to the left atrium, while the other branch descends along the interatrial septum into the anterior portion of the AV node (fast pathway). The Wenckebach’s tract transfers the stimulus from the superior region of the SA node, posterior to the superior vena cava, and travels through the atrial septum to the AV node, while the third pathway (Thorel’s) is responsible for moving the impulse inferiorly and posteriorly along the coronary sinus, arriving into the posterior portion of the AV node (slow pathway).

Once atrial depolarization is completed, depolarization moves into the AV node via the internodal tracts previously described or via cell-to-cell conduction. Normally, the structure of the AV node is the only conduction route from the atrium to the ventricle because the chambers are separated by fibrous and fatty tissue that is nonconductive. The primary function of the AV node is to slow electrical conduction adequately to synchronize atrial contribution to ventricular systole. The AV node is also capable of rescue pacing when the SA node fails and will provide a heart rate of 40–60 bpm (Fig. 1.1.2). By contrast, an ectopic
site within the AV node is capable of pacing competitively against the SA node to produce arrhythmias or junctional tachycardias greater than 100 bpm.

The fast and slow pathways of the AV node are anatomical as well as functional structures. Slow pathway physiology is not seen in every individual. The fast pathway conducts more quickly but has a longer refractory period or recovery period. By contrast, the slow pathway conducts more slowly but has a shorter refractory or recovery period. Conducted impulses commonly travel along the fast pathway through the AV node, but with increased heart rates or the presence of a premature stimulus, the fast pathway may be unable to transmit because it is unable to recover fast enough to transmit the stimulus or be “refractory.” Because the slow pathway has a shorter effective recovery time or is able to recover more quickly, it is able to transmit a signal down the slow pathway while the fast pathway is still recovering. The timing of recovery and the ability or inability to transmit a signal can result in a reentrant tachycardia (Fig. 1.1.3). Reentry is the result of a circuit that is initiated by a signal, often early, being blocked and forced to move in the opposite direction. When the electrical signal conducts back toward the area of block, the structure has had time to recover and is now able to transmit the signal in the opposing direction. Hence, the critical timing sequence of the signal being transmitted creates an independent reentrant circuit.

Once the activation through the AV node occurs, depolarization travels to the common bundle of His (also called His bundle or common bundle). The region where the AV node (node of Tawara) and the His bundle join can be termed the triangle of Koch. Anatomically, the triangle of Koch includes the coronary ostium, the tendon of Todaro, and the tricuspid valve annulus along the septal leaflet. The AV node is approximately 5–6 mm long and 2–3 mm wide, and 0.5–1.0 mm thick, although there is some discrepancy in what is included in the AV node (Hecht et al. 1973; Becker and Anderson 1976). The blood supply of the AV node is the AV nodal artery and is usually dual supplied by the right coronary artery in 90% of the patients and the remaining 10% receive blood from the left circumflex coronary artery. Similar

*Figure 1.1.3*  Reentry of the fast and slow pathways.
to the SA node, there is evidence of a generous autonomic innervation of the AV node, and therefore, the autonomic nervous system influences the rate of conduction through the AV node. AV nodal conduction abnormalities arise from altered blood supply, change in autonomic tone, increased fibrous tissue replacing AV nodal tissue, and an alteration in the normal conduction route.

Once depolarization moves through the bundle of His, it branches out to the right and left bundle branches. The right bundle branch remains compact until it reaches the right distal septal surface, where it branches into the interventricular septum and proceeds toward the free wall of the right ventricle. Because the left ventricle is larger in size, the left bundle branch moves conduction down the left septum and then bifurcates into a posterior and anterior descending fascicle. The left fascicles extend to the base of the papillary muscles and the adjacent myocardium, while the right bundle stays along the interventricular septum superficially within the endocardium (see Fig. 1.1.1).

The final destination is the arrival into the complex network of the specialized Purkinje fibers, capable of independently pacing at a rate of 20–40 bpm if needed along with rapid conduction (Fig. 1.1.2). Once the impulses arrive at the Purkinje fibers, they proceed slowly from the endocardium to epicardium throughout the left and right ventricles. This assures earlier activation at the apex of the heart, the sequence necessary to achieve the most efficient cardiac pumping, which is the intended outcome of cardiac depolarization.

**CARDIAC ACTION POTENTIAL**

The conduction system is composed of two distinctly different cells, pacing cells and nonpacing cells. “Pacing” cells are specialized cells with automaticity, meaning that they can move to a threshold potential independently and propagate or spontaneously initiate an impulse. The specialized cells with automaticity reside within the SA node, AV node, and the Purkinje fibers. All the rest of the cardiac cells, myocytes, are “nonpacing cells” or conducting cells, which means they can be stimulated by an electrical impulse arriving at the cell and then conduct or transmit the impulse from one cell to another cell once the cell is stimulated. Therefore, cardiac cells are unable to initiate an impulse contrary to pacing cells.

Cells have the property of pacing or conductivity due to the electrical charge or voltage on the inside of the cell compared with the voltage on the outside of the cell. If the electrical charge inside the cell is less than the charge on the outside, the transmembrane potential is “negative.” By contrast, if the electrical charge is greater inside the cell than outside the cell, the transmembrane potential is “positive.” Depolarization occurs when the transmembrane potential is positive, while repolarization restores the cell to its negative state, making it available to accept an electrical stimulus in its negative or resting state. Pacing cells are able to depolarize independently, in contrast to a nonpacing cell, which is dependent on an outside stimulus to initiate depolarization.

The transmembrane potential is altered by ions moving in and out of the cell across the cellular membrane. Ion movement is the result of the selective permeability of ion channels distributed along the cell membrane. The movement of the Na⁺, K⁺, and Ca²⁺ ions are the most predominant throughout the cardiac action potential. These ions move in or out of the cell as a result of a change in concentration gradient, electrical gradient, ion pumps, and altered membrane permeabilities (Table 1.1.1). Alterations in permeability to specific ions are most often regulated by voltage-gated channels that will open or close depending on the current measured between the inside and the outside of the cell, but there are additional properties that are responsible for moving ions in and out of the cell (Table 1.1.2). Some of these ion shifts occur passively, while other transport...
Table 1.1.1  Fundamentals of ion transport.

I. Passive ion movement—no energy requirement
   A. Concentration gradient
      Ions shifting from an area of greater concentration to an area of lesser concentration in an effort to equalize the two sides
   B. Electrical gradient
      Ions shifting from an area of greater electrical charge to an area of lesser electrical charge in an effort to equalize the two sides

II. Active ion pumps or transporters (require energy!)
   A. Sodium/potassium pump (sodium ATPase)
      \( \text{Na}^+ \) and \( \text{K}^+ \) transported against their concentration gradients and sodium moves out of the negatively charged interior
      \( \rightarrow \) Three \( \text{Na}^+ \) ions OUT of the cell
      \( \leftarrow \) Two \( \text{K}^+ \) move INTO the cell
   B. Calcium pump
      \( \text{Ca}^{2+} \) removal from inside cell during repolarization
      \( \rightarrow \) \( \text{Ca}^{2+} \) to OUTSIDE of the cell
   C. Sodium/calcium exchange (NCX)
      A small ionic gradient current is generated resulting in the transport of three \( \text{Na}^+ \) ions in exchange for one \( \text{Ca}^{2+} \) ion; the direction of the ion transfer is dependent on the electrical charge of the cell
      1. Repolarization
         \( \leftarrow \) Three \( \text{Na}^+ \) move INTO the cell
         \( \rightarrow \) One \( \text{Ca}^{2+} \) moves OUT of the cell
      2. Depolarization phase
         \( \rightarrow \) Three \( \text{Na}^+ \) move OUT of the cell
         \( \leftarrow \) One \( \text{Ca}^{2+} \) moves INTO the cell

III. Ion channel properties
   A. Ion permeability
      The selective permeability that allow ions to move through the open channel at specific times
   B. Gating—opening/closing of ion channels
      1. Voltage gated
         Ion permeability enhanced or decreased based on the measured voltage of the membrane potential
      2. Ligand-dependent gating
         The opening of the channel is dependent on activation of a protein along the binding site (i.e., \( I_{\text{ACh}} \) → acetylcholine binds to M-2 receptor → activates G protein-signaling pathway → activates inward rectifying \( \text{K}^+ \) channel)
      3. Mechanosensitive gating
         A physical input transfers into an electrical signal, that is, stretch → electrical signal
         Least studied but responsible for arrhythmias associated with dilatation
Table 1.1.2  Ion-specific channel characteristics.

I. Sodium channels
   a. Voltage gated
      \( I_{Na} \)—fast inward current
      Increased transmembrane potential of \(-90 \) to \(-60\) mv
      Inactivation—rapid response followed by no \( Na^+ \) entry
   b. Target for antiarrhythmics—class I
      Block occurs when \( Na^+ \) channel is either open or inactivated during the action potential
      Increased heart rate (increase in number of action potentials) with reduced recovery time results in an accumulation of block that is use-dependent
   c. Abnormalities causing arrhythmias
      Inactivation of the sodium channel does not occur, but continues with brief bursts of Na channel openings; this phenomenon is the basis for the subgroup of long QT syndromes (LQT3)
      Mutations of the sodium channel gene, SCN5A, is associated with LQTS, Brugada syndrome, and primary cardiac conduction disease (Wang et al. 1990)

II. Potassium channels
   a. Voltage gated
      \( I_o \)—transient outward current in phase 1
      Rapid activation that provides a transient outward current
      \( I_{Kur}, I_{Kr}, \) and \( I_{Ks} \)—delayed rectifiers—phase 3
      \( I_{Kur} \)—ultrarapid
      \( I_{Kr} \)—rapid
      \( I_{Ks} \)—slow
      Voltage-gated channels open in response to membrane depolarization, which generate a current to restore resting potential
      Slowly activating outward current (moves \( K^+ \) outside the cell) during repolarization
      \( I_{Ko} \)—inward rectifier
      Moves \( K^+ \) into cell in phase 4
   b. Ligand gated
      \( I_{Koa} \)—activated by muscarinic receptors
      May cause hyperpolarization and shorten APD
      Outward current
      \( I_{Kub} \)—activated by adenosine
      Outward current
      \( I_{KATP} \)—blocked by ATP
      Activated when ATP is low (ischemia)
      Shortens APD when activated
   c. Voltage and ligand
      \( I_i \)—nodal tissue during phase 4
      Activated by hyperpolarization (about \(-40\) mV)
   d. Target for antiarrhythmics—class III

III. Calcium channels
   a. Two types
      \( I_{Ca} \)—slow, inward calcium current
      Low-threshold type with long-lasting openings
      Contributes to cardiac cell phase 2 (plateau)
      \( I_{Cat} \)—transient inward current
      Found principally in pacemaker cells
      Opens transiently in phase 4
   b. Target for antiarrhythmics—class IV
mechanisms require energy at the cellular level. The ion “pumps” or ion transfers that require energy will be at risk in the event that the cell does not have an energy source or is oxygen deprived, for example, ischemia provides an opportunity for arrhythmias to occur.

Phases of the Cardiac Action Potential

The cardiac action potential of the “nonpacing” cell consists of five phases:

- Phase 0—rapid depolarization
- Phase 1—early rapid repolarization
- Phase 2—plateau phase
- Phase 3—repolarization
- Phase 4—resting phase

The cell moves from one phase to another very quickly with the entire process occurring within milliseconds. Although we describe each specific phase, the transition from one phase to another is dynamic and seamless. The action potential takes a round-trip journey in that the signal is able arrives at baseline (phase 4) and is able to travel to the destination (depolarization—phase 0). Then, the action potential is able to return back to home (repolarization—phases 1–4) and prepare to depart from home or baseline (resting—phase 4) once again. What actually occurs at each phase is described below.

Phase 0—Rapid Depolarization

When an electrical impulse arrives at the cell, the membrane potential shifts from approximately −90 to −60v and reaches “threshold” potential. The shift in voltage triggers the “voltage-gated” sodium channels to open and the permeability of the plasma membrane to sodium ions ($P_{Na^+}$) increases, thereby resulting in rapid movement of sodium ions from extracellular to intracellular along their electro-mechanical gradient. Positively charged Na$^+$ ions shift from the outside of the cell to the inside of the cell, causing the membrane potential to become more positive, now to approximately 0mV (Fig. 1.1.4). The “fast” sodium channels inactivate within a few milliseconds, decreasing permeability of the cellular membrane to Na$^+$ and preventing any further voltage increase.

![Figure 1.1.4](image)

**Figure 1.1.4** Phase 0—rapid depolarization. Sodium moving into the cell quickly increases the intracellular charge, creating a positive transmembrane potential.
Phase 1—Early Rapid Repolarization

Transient outward K+ current, $I_{to}$, is turned on briefly by depolarization and drives the potassium out of the cell. This transient outward current rapidly inactivates, so the rapid outward current is brief, resulting in a slightly reduced intracellular charge as the positively charged K+ ions move outside of the cell (Fig. 1.1.5).

Phase 2—Plateau Phase

The following ions are in motion in phase 2:

- Calcium moves slowly to the inside of the cell through the $I_{Ca-L}$ channel (inward calcium channel).
- Potassium moves to the outside of the cell with the voltage and concentration gradient in an effort to equalize the voltage and the concentration of K+ within the inside and the outside of the cell.
- Three sodium ions are moving into the cell in exchange for one calcium ion moving out of the cell.

The cumulative, simultaneous movement of these ions results in a stable voltage along the membrane or a “plateau phase” (Fig. 1.1.6).

Phase 3—Final Repolarization

In final repolarization, potassium diffuses to the outside of the cell with the increased permeability along the cell membrane with potassium channels opening and due to the movement caused by the concentration gradient. These voltage-dependent potassium channels are delayed rectifier currents and are slowly activating outward currents ($I_{Kur}$, $I_{Kr}$, $I_{Ks}$). Concurrently, Ca$^{2+}$ channels close, so the inward movement of calcium stops, while potassium continues to move outside of the cell and allows the membrane potential to go back to a negative resting membrane potential. Ion movement includes the following:

- Inactivation of the $I_{Ca-L}$ stops Ca$^{2+}$ entry into cell.
- Delayed rectifier K+ currents, $I_{Ks}$ (slow), $I_{Kr}$ (rapid), and $I_{Kur}$ (ultrarapid), moving K+ to the outside of the cell, while inwardly...
rectifying currents, \( I_{Ki} \) and \( I_{KCh} \), result in the movement of positive charges out of the cell.

- Potassium conductance falls to plateau levels as a result of the inward rectification, membrane conductance changes with voltage (\( K^+ \) channels are open at negative potentials but closed at less negative or positive voltages) (Fig. 1.1.7).

**Phase 4—Resting Membrane Potential**

The cardiac action potential relies on the cell to adequately prepare for depolarization in the resting phase. It is during the cardiac cell resting phase that the intracellular potential is approximately \(-90\) to \(-100\) mV relative to the measured voltage outside of the cell, making it negative. During resting phase, there are more potassium ions within the cellular membrane (intracellular), while the majority of sodium and calcium ions are kept on the outside of the cell membrane (extracellular).

Although phase 4 is referred to as “resting” phase, the negative intracellular voltage is the result of ion movement related to a combination of complex systems that include the opening of selective ion channels, altering membrane permeability, concentration gradients, electrogenic gradients, and active ion pumps (Fig. 1.1.8). This phase includes the sodium-potassium pump, which requires energy, thus it relies on oxygenation to the area to maintain resting phase. Maintaining the resting membrane potential of \(-90\) to \(-100\) mV allows the cell to be ready to accept an outside stimulus or to be depolarized.

**Action Potential of Pacemaker Cells (Slow Response)**

The unique quality of the pacing cells is that they have the capability of reaching depolarization independently. Therefore, they can initiate a stimulus as opposed to being able to only...
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Figure 1.1.7 Phase 3—final repolarization. Potassium efflux is the essential ion movement.

Figure 1.1.8 Phase 4—resting phase. 1. Open, inward rectifying K⁺ channels, $I_{K1}$, moving potassium to the inside of the cell. 2. Three Na⁺ ions move to the outside of the cell while two K⁺ ions are transferred to the inside of the cell by the active Na⁺/K⁺ pump. 3. With the negative transmembrane potential, the Na⁺/Ca²⁺ exchanger is exchanging three Na⁺ ions to the inside of the cell while moving one Ca²⁺ ion outside the cell. 4. Plasma membrane calcium (PMCA) pump removes calcium.

Conduct or transmit a stimulus. The specialized cells with automaticity reside within the SA node, AV node, and the Purkinje fibers. Their inherent pacing rate of the specialized cells is most rapid in the sinus node while slowest in the ventricles. This provides rescue pacing if the higher pacing sites fail, for example, the AV node will pace at a rate of 40–60 bpm in the absence of the sinus node firing at a rate of 60–100 bpm (Fig. 1.1.2). As mentioned
Cardiac Arrhythmia Management

The voltage of the cell at the onset of phase 4 is $-40$ to $-70$. This is the result of the presence of the $I_f$ channel, pacemaker or “funny” current, which is a current activated by hyperpolarization and causes $\text{Na}^+$ and $\text{K}^+$ to enter the cell, thus allowing the cell to independently move to depolarization. Automaticity is dependent on a combination of the $I_f$ channel, the deactivation of $I_{\text{Kl}}$ current, and the transient inward calcium current, $I_{\text{CaT}}$. The $I_f$ channel moves $\text{Na}^+$ and $\text{K}^+$ into the cell to offset the deactivation of $I_{\text{Kl}}$ current, which causes an inward $\text{K}^+$ current. The $I_{\text{CaT}}$ current is limited to pacing cells exclusively and the opening of this calcium current allows calcium to move slowly into the cell, moving the charge inside of the cell to $-30$ and $-40$, resulting in “threshold” potential, and finally, opening of the fast $\text{Na}^+$ channels for depolarization to occur.

Phase 4—Diastolic Depolarization of the Pacing Cell

Automaticity of the pacing cell is the result of ions shifting to achieve a net gain in intracellular positive charges during diastole. This ion movement allows the cell to independently reach a “threshold” potential. There are a number of differences between the action potential of the pacing cell and the cardiac cell that allow this to be achieved. First, the transmembrane potential of the pacing cell does not return to the same negative membrane potential as the cardiac cell in resting phase. Instead, the voltage of the cell at the onset of phase 4 is $-40$ to $-70$. This is the result of the presence of the $I_f$ channel, pacemaker or “funny” current, which is a current activated by hyperpolarization and causes $\text{Na}^+$ and $\text{K}^+$ to enter the cell, thus allowing the cell to independently move to depolarization. Automaticity is dependent on a combination of the $I_f$ channel, the deactivation of $I_{\text{Kl}}$ current, and the transient inward calcium current, $I_{\text{CaT}}$. The $I_f$ channel moves $\text{Na}^+$ and $\text{K}^+$ into the cell to offset the deactivation of $I_{\text{Kl}}$ current, which causes an inward $\text{K}^+$ current. The $I_{\text{CaT}}$ current is limited to pacing cells exclusively and the opening of this calcium current allows calcium to move slowly into the cell, moving the charge inside of the cell to $-30$ and $-40$, resulting in “threshold” potential, and finally, opening of the fast $\text{Na}^+$ channels for depolarization to occur.

Phase 0—Depolarization of the Pacing Cell

The significant contrast of the pacemaker cell and the cardiac cell during phase 0 is the absence of a stimulus to alter the transmembrane potential in the pacing cell. The cell itself
moves from a transmembrane potential of \(-60\) to “threshold potential” by a slow, inward current rather than a fast inward Na current, as described above. The discharge rate of the sinus node normally exceeds the discharge rate of the other potentially automatic pacemaker sites, and therefore, maintains the dominant rate. It is also more sensitive to the effects of norepinephrine (sympathetic) and acetylcholine (parasympathetic) so it provides the best physiological heart rate. The lower, alternative pacing sites in the AV node and Purkinje fibers provide an electrical stimulus in the absence of an intact sinus node. The complex intrinsic pacing capability of the heart is essential in providing optimal blood flow and meeting the oxygen demands of the body during times of increased physical activity and/or increased stress.

**DRUGS FOR CARDIAC ARRHYTHMIAS**

Cardiac arrhythmias generally result from an abnormality in the rate, rhythm, or conduction of an electrical impulse in the heart (Perry and Illsley 1986). These abnormalities are disturbances in normal impulse initiation (automaticity), impulse conduction, or both. Various antiarrhythmic agents affect intracellular and extracellular concentrations of sodium, potassium, calcium, and magnesium. The balance of all these molecular components have varying effects on the electrophysiology of the heart and are critical to controlling arrhythmias with antiarrhythmic medications. In general, antiarrhythmic medications are available to treat tachyarrhythmias. There are no currently available medications to treat bradyarrhythmias effectively, particularly in oral form.

The classification of antiarrhythmic agents is discussed below, with emphasis on the particular electrophysiological action of each drug classification. Several of the drugs studied had more than one of the four actions, so that it deserves emphasis that the classification is not so much categorization of drugs in accordance with chemical structures or physical properties, but describes four ways in which abnormal cardiac rhythms can be corrected or prevented (Vaughan Williams 1984). Based on the Vaughan Williams classification, there are four main classes of antiarrhythmic medications (Tables 1.1.3 and 1.1.4). Although much maligned, the Vaughan Williams classification system is still the most commonly used by those in the medical field worldwide. Because the antiarrhythmic drugs usually target a specific ion and either block or enhance its movement in or out of the cell, there are electrocardiogram (ECG) changes that may be evident as a result of that (see Table 1.1.5).

The classes are further simplified and subdivided based on the primary electrophysiological effect of either their ability to convert the rhythm or control the rate (Table 1.1.6). Class I and class III drugs are more effectively utilized to prevent arrhythmias and maintain sinus rhythm. Class IV drugs provide rate control with the primary goal of reducing conduction through the AV node, while class II drugs are used to reduce heart rate and maintain sinus rhythms in those patients who have arrhythmias that are triggered by catecholamines. The discussion below describes each group in more detail.

**Class I Drugs: Sodium Channel Blockade**

The class I drugs act by modulating or blocking the sodium channels, thereby inhibiting or altering phase 0 depolarization (Fig. 1.1.4). Their dominant electrophysiological property has been related to their ability to reduce the maximal rate of depolarization in cardiac muscle. A reduction in the rate of depolarization by therapeutic concentrations of these drugs has been found to be associated with an increase in the threshold of excitability, a depression in conduction velocity, and a prolongation in the effective refractory period (Singh 1978). Three different subgroups, class IA, IB, and IC, have been identified because
Table 1.1.3 Drug effects on ECG.

<table>
<thead>
<tr>
<th>Drug</th>
<th>Effects on ECG</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amiodarone</td>
<td>Bradycardia, prolongs PR, QRS, and QT</td>
</tr>
<tr>
<td>Acebutol, esmolol, metoprolol, propanolol</td>
<td>Bradycardia, prolongs PR</td>
</tr>
<tr>
<td>Diltiazem, verapamil</td>
<td>No change in QRS</td>
</tr>
<tr>
<td>Adenosine</td>
<td>Prolongs PR, heart block (transient)</td>
</tr>
<tr>
<td>Sotalol</td>
<td>Bradycardia, prolongs PR and QT</td>
</tr>
<tr>
<td>Dofetilide, ibutilide</td>
<td>Prolongs QT</td>
</tr>
<tr>
<td>Flecainide, propafenone</td>
<td>Prolongs PR and QRS</td>
</tr>
<tr>
<td>Digoxin</td>
<td>Prolongs PR, depresses ST segment, flattens T wave</td>
</tr>
<tr>
<td>Lidocaine, mexilitine</td>
<td>No significant change</td>
</tr>
<tr>
<td>Dronedarone</td>
<td>Bradycardia, prolongs QT</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Class</th>
<th>Effect on repolarization/ depolarization</th>
<th>Phase of cardiac action potential</th>
<th>Effect on action potential duration</th>
<th>Effect on ECG</th>
</tr>
</thead>
<tbody>
<tr>
<td>IA: sodium channel blockade</td>
<td>Prolongs repolarization</td>
<td>Phase 0</td>
<td>Depression</td>
<td>Prolongs</td>
</tr>
<tr>
<td>IB</td>
<td>Shortens depolarization</td>
<td>Phase 0</td>
<td>Weak phase 0 depression</td>
<td>Decrease</td>
</tr>
<tr>
<td>IC</td>
<td>No effect</td>
<td>Strong phase 0</td>
<td>Depression</td>
<td>No effect or mildly prolongs</td>
</tr>
<tr>
<td>II: beta adrenergic blockade</td>
<td>Enhanced depolarization</td>
<td>Enhanced phase 4</td>
<td></td>
<td></td>
</tr>
<tr>
<td>III: potassium channel blockers</td>
<td>Prolongs repolarization</td>
<td>Phase 3</td>
<td>Prolongs</td>
<td>QT interval is longer at slower heart rates, decreases as heart rate increases</td>
</tr>
<tr>
<td>IV: calcium channel blockers</td>
<td>Slows depolarization</td>
<td>Phase 4</td>
<td>Prolongs</td>
<td>Slows the sinus rate and increases PR</td>
</tr>
</tbody>
</table>

Their mechanism or duration of action is somewhat different due to variable rates of drug binding to and dissociation from the channel receptor (Snyders et al. 1991).

The major drugs with class IA classification are quinidine, procainamide, and disopyramide. These drugs depress phase 0 (sodium-dependent) depolarization, thereby slowing conduction. They also have moderate potassium channel blocking activity (which tends to slow the rate of repolarization and prolong action potential duration [APD]), anticholinergic activity, and depress myocardial contractility. At slower heart rates, when use-dependent
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Table 1.1.4  Specifics of each drug classification.

<table>
<thead>
<tr>
<th>Class</th>
<th>Effect</th>
</tr>
</thead>
<tbody>
<tr>
<td>Class I: sodium channel blockers</td>
<td>Slow down the depolarization</td>
</tr>
<tr>
<td>Class IA, IB, IC</td>
<td>Slow down the depolarization</td>
</tr>
<tr>
<td>Class II: beta adrenergic blockers</td>
<td>Slow depolarization by blocking the beta receptors in the parasympathetic nervous system</td>
</tr>
<tr>
<td>Class III: potassium ion channel blockers</td>
<td>Prolong phase 3 (action potential duration) and lengthen the QT interval</td>
</tr>
<tr>
<td>Class IV: calcium channel blockers</td>
<td>Block the influx of calcium into the cell, Shorten depolarization, prolong repolarization, slow down conduction through the AV node</td>
</tr>
<tr>
<td>Class V: miscellaneous (none of the above)</td>
<td>Adenosine: slows sinus node automaticity and AV conduction, Digoxin: increases phase 4 slope and decreases resting membrane potential, decreases conduction velocity, increases vagal tone</td>
</tr>
</tbody>
</table>

blockade of the sodium current is not significant, potassium channel blockade may become predominant (reverse use dependence), leading to prolongation of the APD and QT interval and increased automaticity.

The class IB drugs include lidocaine, mexiletine, and tocainide. They have less prominent sodium channel blocking activity at rest but effectively block the sodium channel in depolarized tissues. This group tends to bind in the Na⁺ channel inactivated state (which follows the fast channel opening in phase 0 depolarization) and dissociate from the sodium channel more rapidly than other class I drugs. As a result, they are more effective with tachyarrhythmias than with slow arrhythmias.

The class IC drugs, flecainide and propafenone, block both the open and inactivated sodium channels and thus, slow conduction. They dissociate slowly from the sodium channels during diastole, resulting in increased effect at more rapid rate (use dependence). This characteristic is the basis for their antiarrhythmic efficacy.

Table 1.1.5  Antiarrhythmic drug effects on the ECG.

<table>
<thead>
<tr>
<th>Antiarrhythmic medication</th>
<th>PR effect</th>
<th>QRS effect</th>
<th>QT effect</th>
<th>ST effect</th>
<th>T wave effect</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amiodarone</td>
<td>Bradycardia</td>
<td>Prolonged</td>
<td>Prolonged</td>
<td>Prolonged</td>
<td></td>
</tr>
<tr>
<td>Acebutolol, esmolol, metoprolol, propanolol</td>
<td>Bradycardia</td>
<td>Prolonged</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Diltiazem, verapamil</td>
<td>Transient heart block</td>
<td>Prolonged</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Adenosine</td>
<td>Prolonged</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sotalol</td>
<td>Bradycardia</td>
<td>Prolonged</td>
<td>Prolonged</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dofetilide, ibutilide</td>
<td>Prolonged</td>
<td>Prolonged</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Flecainide, propafenone</td>
<td>Prolonged</td>
<td>Prolonged</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Digoxin</td>
<td>Prolonged</td>
<td></td>
<td></td>
<td>Depressed</td>
<td>Flattens</td>
</tr>
<tr>
<td>Lidocaine, mexiletine</td>
<td>No significant change</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Table 1.1.6  Common drugs for atrial fibrillation and supraventricular arrhythmias.

<table>
<thead>
<tr>
<th>Arrhythmia</th>
<th>Drug of choice</th>
<th>Alternative</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atrial flutter/fibrillation</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Acute management</td>
<td>Rate control: verapamil, diltiazem, beta blocker, or digoxin</td>
<td></td>
</tr>
<tr>
<td>Chronic treatment</td>
<td>Rhythm conversion: DC cardioversion</td>
<td>Catheter ablation to eliminate arrhythmia</td>
</tr>
<tr>
<td></td>
<td>Rate control: oral verapamil, diltiazem, beta blocker, or</td>
<td></td>
</tr>
<tr>
<td></td>
<td>digoxin</td>
<td></td>
</tr>
<tr>
<td>Other supraventricular tachycardias</td>
<td>Maintain sinus rhythm: amiodarone, sotalol, flecainide, propafenone, or dolefilide</td>
<td>Quinidine, procainamide, disopyramide, amiodarone (may require drug loading)</td>
</tr>
<tr>
<td>Acute management</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Long-term suppression</td>
<td>Beta blockers, verapamil, diltiazem, flecainide, propafenone,</td>
<td>Vagotonic maneuvers (such as carotid sinus massage, gagging, or the Valsalva maneuver) that impair AV nodal conduction may be tried first</td>
</tr>
<tr>
<td></td>
<td>amiodarone, sotalol, or digoxin</td>
<td>Catheter ablation can cure most patients</td>
</tr>
</tbody>
</table>

DC, direct current; IV, intravenous.

especially against supraventricular arrhythmia. Use dependence may also contribute to the proarrrhythmic activity of these drugs, especially in the diseased myocardium, resulting in incessant ventricular tachycardia.

- Flecainide was first introduced in 1985 for treatment of ventricular arrhythmias then subsequently for oral use to prevent supraventricular arrhythmias. The indications for using flecainide to treat ventricular arrhythmias were limited after a controlled trial found that postmyocardial infarction patients with asymptomatic ventricular arrhythmias who took the drug had twice as high a mortality rate as patients who took placebo (Echt et al. 1991). Flecainide decreases the rate of cardiac conduction in all parts of the heart. In animals, at normal resting heart rates, the drug causes only a small increase in the refractory period, but at the rapid rates typical of atrial fibrillation, flecainide markedly increases atrial APD and refractoriness (Wang et al. 1990). The drug is metabolized in the liver and excreted in the urine. Rarely, patients may be deficient in the enzyme system required for metabolism of the drug. Flecainide is effective for prevention of paroxysmal supraventricular tachycardia, moderately effective for suppression of paroxysmal atrial fibrillation, and is generally well tolerated. Because of its proarrrhythmic effects, however, use of the drug should be restricted to patients without clinically significant structural heart disease who have disabling symptoms refractory to other drugs.

- Propafenone, much like flecainide, markedly decreases cardiac conduction velocity. And like flecainide, it can also aggravate existing arrhythmias or precipitate new ones, especially in patients with underlying heart disease and sustained ventricular tachycardia. Propafenone has a low degree of beta blocking activity in some patients.
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Class II Drugs: Beta Blockade (Antagonists)

Hyperactivity of the sympathetic nervous system has been recognized for many years as a factor in the genesis of cardiac arrhythmias. The class II drugs, such as atenolol, metoprolol, carvedilol, act by inhibiting sympathetic activity, primarily by causing beta blockade. Their principal electrophysiological effect on heart muscle in clinically relevant concentrations is the depression of phase 4 depolarization (see Fig. 1.1.8), resulting in a reduced heart rate. Only in very high concentrations do these drugs exert effects on other parameters, such as the upstroke velocity of the phase 0 of the action potential (Singh 1978). Beta agonists or catecholamines (i.e., epinephrine and norepinephrine) are endogenous, neurohormonal substances that mediate diverse physiological and metabolic responses in man by interaction with adrenergic receptors (beta receptors) in various tissues. As a result of this, beta agonists potentiate positive chronotropic (increased heart rate) and inotropic (increased contractility) actions. By contrast, beta adrenergic antagonists’ main therapeutic effect is to slow the heart rate and decrease myocardial contractility. They reduce sinus rate, especially when sympathetic control of the heart is dominant, as during exercise. They have less effect on heart rate in an individual at rest. They also decrease the rate of spontaneous depolarization of ectopic pacemakers, slow conduction in the atria and AV node, and increase the refractory period of AV node.

Class III Drugs: Potassium Channel Blockade

Class III drugs, amiodarone, ibutilide, dofetilide, sotalol, azimilide, and dronedarone, block the potassium channels, thereby prolonging repolarization, the APD, and the refractory period (Arnsdorf et al. 2009; see Fig. 1.1.7). These changes are manifested on the surface ECG by prolongation of the QT interval, providing the substrate for torsade de pointes, a polymorphic ventricular tachycardia. Amiodarone is an exception, with very little proarrhythmic activity. Amiodarone has since been found to be a potent antiarrhythmic drug in the clinic, but although it does prolong the QTc (corrected QT) interval on the ECG in patients, ventricular arrhythmias have not been encountered during prolonged periods of treatment in large numbers of patients (Singh 1978; see Table 1.1.5).

- Amiodarone—Among available antiarrhythmics, amiodarone (Cordarone and others) is the most effective for prevention of atrial fibrillation and of ventricular tachycardia or fibrillation. The antiarrhythmic actions of amiodarone can be attributed to its property of inhibiting adrenergic stimulation (alpha and beta blocking properties), its effects on sodium, potassium, and calcium channels, its ability to prolong the action potential with consequent lengthening of the effective refractory period in myocardial tissue and decreasing AV nodal conduction and sinus node function. Multiple clinical trials have indicated that amiodarone is the most potent antiarrhythmic agent for the control of refractory ventricular tachyarrhythmias and for the prophylaxis of recurrent supraventricular tachyarrhythmias, including atrial fibrillation or flutter complicating the Wolff–Parkinson–White syndrome. Amiodarone is well tolerated by most patients, but there are several potential side effects that need to be monitored for closely.
- Dronedarone—This is one of the newer class III antiarrhythmic drugs and is a “cousin” to amiodarone; it is indicated for the treatment of atrial arrhythmias. The primary differences compared with amiodarone are attributable to the lack of iodine in the molecular structure, along with a reduced half-life due to its less hydrophobic nature. As a result, it may be associated with fewer long-term
complications compared with amiodarone. Its efficacy can be evaluated more quickly, as achieving therapeutic levels is not reliant on a “loading” regimen; rather it is given twice daily, with steady state achieved usually within 3–7 days.

• Sotalol—Sotalol is a racemic mixture of d-sotalol and l-sotalol; both isomers have similar class III antiarrhythmic effects, while the l-isomer is responsible for virtually all of the beta blocking activity. Sotalol contains both beta adrenoreceptor blocking (class II) and cardiac APD prolongation (class III) properties. The noncardioselective beta-blocking effect of sotalol (increased sinus cycle length, slowed heart rate, decreased AV nodal conduction, and increased AV nodal refractoriness) occurs at oral doses as low as 25 mg/day. The class III effects (prolongation of the atrial and ventricular monophasic action potentials, and effective refractory prolongation of atrial muscle, ventricular muscle, and AV accessory pathways in both the antegrade and retrograde directions) are seen only at oral doses ≥160 mg/day. Sotalol should be initiated and doses increased in a hospital with facilities for cardiac rhythm monitoring and assessment, as proarrhythmic events can occur after initiation of therapy and with each upward dosage adjustment.

• Dofetilide—Dofetilide has no effect on sodium channels, adrenergic alpha receptors, or adrenergic beta receptors. It increases the monophasic APD and effective refractory period of the myocyte, thereby terminating reentrant tachyarrhythmias and preventing their reinduction (Roukoz et al. 2007). The increase in the QT interval is a function of prolongation of both effective and functional refractory periods in the His-Purkinje system and the ventricles. Changes in cardiac conduction velocity and sinus node function have not been observed in patients with or without structural heart disease. PR and QRS width remain the same in patients with preexisting heart block and or sick sinus syndrome. Dofetilide is generally well tolerated but like other antiarrhythmic agents in its class, torsades de pointes may be induced as a consequence of therapy. Therefore, it should be initiated and doses titrated while in a hospital with facilities for cardiac rhythm monitoring and assessment.

Class IV Drugs: Calcium Channel Blockade (Antagonists)

As a class, calcium channel antagonists do not increase the effective refractory period of the atria, ventricle, His-Purkinje fibers, or the accessory pathways in the heart. The dominant effect of calcium channel antagonists is slowing of conduction in the AV node with the prolongation of the AV nodal refractory period (Singh et al. 1983). Selective calcium channel antagonists, such as verapamil and diltiazem, have been found to have some antiarrhythmic activity. They preferentially affect slow-response myocardial tissue rather than fast-response tissue. Slow-response tissues (the SA and AV nodes) depend on calcium channel currents to generate slowly propagating action potentials. By contrast, fast-response myocardial tissues (the atria, specialized infranodal conducting system, the ventricles, and accessory pathways) depend on sodium channel currents. Verapamil is the prototype calcium antagonist and has the most clearly defined antiarrhythmic properties (Singh et al. 1983). Verapamil, as well as diltiazem, terminate paroxysmal supraventricular tachycardia and slow the ventricular response in atrial flutter and fibrillation. They also have prophylactic value in preventing recurrences of paroxysmal supraventricular tachycardia and controlling the ventricular response in atrial flutter and fibrillation during long-term oral therapy. They play a much more limited role in the treatment of ventricular arrhythmias (Singh et al. 1983).
Antiarrhythmic drugs are available as one treatment option for controlling arrhythmias. As you can see, there are a variety of medications available to treat the full spectrum of tachyarrhythmias. Each clinician may prefer one agent over another, and a particular patient’s arrhythmia control and tolerance of medications may vary considerably. Therefore, the use of antiarrhythmics in patient management may not be straightforward and may require increased patient surveillance. Additional methods of treatment, that is, ablation, may be utilized as an adjunct to medication and provides the patient with additional options for controlling arrhythmias.

REFERENCES


RESOURCES


INTRODUCTION: APPROACH TO THE ARRHYTHMIA PATIENT

The diagnosis of cardiac rhythm disorders depends on the accurate documentation of the abnormal rhythm, usually in association with symptoms. Rhythm disorders can be very infrequent, occurring a few times a year or can be constant, such as in persistent atrial fibrillation. Thus, the challenge of diagnosis of arrhythmias depends a great deal on the appropriate selection of diagnostic tools. An accurate diagnosis of arrhythmia allows prompt and appropriate treatment. While the “gold” standard for obtaining a diagnosis is demonstrating a rhythm abnormality associated with symptoms, this is not always possible. Often, the use of multiple diagnostic tests in addition to the history and physical examination may yield a likely diagnosis. In addition, in some cases, the risk of waiting to document an arrhythmia may be too great. For example, patients with syncope who may be at high risk for ventricular arrhythmias may be best served by placement of an implantable cardioverter defibrillator (ICD; based on the appropriate guidelines) rather than waiting to document the ventricular arrhythmia that likely caused the syncope and placing the patient at significant risk (Miller et al. 2004; Zipes and Miles 2004).

Evaluation of arrhythmia should always start with a detailed history and physical assessment. Careful assessment of the key information from history and physical will help guide further diagnostic testing. History is an important initial component of evaluation of a patient with arrhythmia. It can provide key pieces of information that help determine the diagnosis or guide the type and sequence of diagnostic testing. Common symptoms include palpitations, light-headedness, presyncope, syncope, and shortness of breath. Patients
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should be asked when the date of onset, frequency, and duration of symptoms, as well as the presence of any associated symptoms such as nausea, shortness of breath, or chest pain. In addition, it is important to identify triggers such as exercise, stress, alcohol, caffeine, and so on that may provoke the arrhythmia; and a detailed history of these potential triggers is warranted. In collecting the patient’s past medical history, it is important to identify the presence of any structural heart disease, thyroid problems, pulmonary disease, or other systemic problem that may contribute to a potential arrhythmic disorder. In addition, a detailed family history is important to elucidate any familial predisposition to arrhythmias, particularly those that may be life threatening and result in sudden cardiac death (Barsky 2001). Family history is often also used for risk stratification in some conditions such as hypertrophic cardiomyopathy when attempting to determine the risk of mortality due to lethal arrhythmias. Patients with strong family history of sudden cardiac death who present with unexplained syncope may need more urgent and, potentially, more invasive evaluation. If the family history of sudden death is present, it is important to find out the circumstances of the death. It may even be necessary to obtain an autopsy report if available. Medication and dietary history should also be taken. Current medications may explain some arrhythmias. For example, decongestants with beta adrenergic agonist activity may provoke tachycardia episodes, while beta blockers, calcium channel blockers, and digoxin may result in bradycardia. Some prescription and even nonprescription drugs may cause prolongation of the QT interval and cause potentially lethal arrhythmias in patients with an underlying predisposition.

Information gathered from the review of patient history may provide important diagnostic clues, but ultimately, the documentation of the actual arrhythmia episode has the best diagnostic value. There are several testing modalities used for this purpose (Table 1.2.1).

**Rhythm Assessment and Diagnosis**

**Electrocardiography**

Electrocardiography is the most frequently used, relatively inexpensive, and easily available noninvasive test used in arrhythmia evaluation. Using the electrocardiogram or ECG, a clinician cannot only determine the rhythm and the heart rate at the time of the ECG, but other information can be gleaned including evidence of conduction system disease or presence of ongoing ischemia, old myocardial injury, pre-excitation, hypertrophy, pulmonary abnormalities, electrolyte abnormalities, or QT prolongation. The ECG is very useful diagnostic test but can be limited in that it may be normal in patients with sporadic symptoms or sporadic arrhythmias. However, despite not having an active arrhythmia at the time of the ECG, clues on the ECG may still be useful in management of patients. For example, a patient with syncope with Mobitz type II second-degree heart block or significant conduction system disease may be appropriate for implantation of a permanent pacemaker. In addition, it is often worthwhile to have a good baseline ECG for future reference. When interpreting the ECG, it is important to confirm that the leads are placed correctly.

**Ambulatory Monitoring Systems**

While it is not always possible to document an arrhythmia using an ECG, there are a number of ambulatory electrocardiographic monitoring systems available to evaluate the patient with intermittent symptoms. Prolonged electrocardiographic recording in ambulatory patients is
a very useful method to document an arrhythmia or to evaluate the extent of arrhythmia in terms of frequency and duration and correlate the arrhythmia with the patient’s symptoms. It can also be used to evaluate the effect of antiarrhythmic drug therapy. There are many different types of ambulatory monitors that vary in terms of the duration of each recorded episode, presence of automatic detection of arrhythmias, and the total duration of the monitoring period. Table 1.2.2 lists these different monitors and examines the differences between them.

Table 1.2.1 History and physical assessment.

<table>
<thead>
<tr>
<th>History and physical assessment</th>
<th>Pertinent data</th>
<th>Examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>History of present illness (HPI)</td>
<td>• Mode of onset of the episode</td>
<td>Mode of termination: If palpitations can be terminated by vagal maneuvers, such as breath holding or Valsalva maneuver, it is likely to be AVNRT or AV reciprocating tachycardia utilizing an accessory pathway</td>
</tr>
<tr>
<td></td>
<td>• Mode of termination</td>
<td></td>
</tr>
<tr>
<td></td>
<td>• Frequency</td>
<td></td>
</tr>
<tr>
<td></td>
<td>• Duration of symptoms</td>
<td></td>
</tr>
<tr>
<td></td>
<td>• Associated symptoms, such as chest pain or shortness of breath</td>
<td></td>
</tr>
<tr>
<td></td>
<td>• Triggers</td>
<td></td>
</tr>
<tr>
<td></td>
<td>• What treatment modalities were already used and how effectively</td>
<td></td>
</tr>
<tr>
<td>Past medical history (PMH)</td>
<td>• Coronary artery disease</td>
<td>Wide complex tachycardia in a patient with history of myocardial infarction is more likely to be ventricular in origin than supraventricular.</td>
</tr>
<tr>
<td></td>
<td>• Congestive heart failure</td>
<td></td>
</tr>
<tr>
<td></td>
<td>• Chronic obstructive pulmonary disease</td>
<td>In a patient with a history of corrective surgery for congenital heart disease, scar may lead to reentrant tachycardias.</td>
</tr>
<tr>
<td></td>
<td>• Thyrotoxicosis</td>
<td></td>
</tr>
<tr>
<td></td>
<td>• Hypertension</td>
<td></td>
</tr>
<tr>
<td></td>
<td>• Diabetes</td>
<td></td>
</tr>
<tr>
<td></td>
<td>• Congenital heart disease</td>
<td></td>
</tr>
<tr>
<td></td>
<td>• Valvular disease</td>
<td></td>
</tr>
<tr>
<td>Past family history (PFH)</td>
<td>• Sudden death</td>
<td>Patients with a family history of hypertrophic cardiomyopathy are at a higher risk for sudden death and may need to have an ICD implanted</td>
</tr>
<tr>
<td></td>
<td>• Arrhythmia</td>
<td></td>
</tr>
<tr>
<td></td>
<td>• Long QT</td>
<td></td>
</tr>
<tr>
<td></td>
<td>• Hypertrophic cardiomyopathy</td>
<td></td>
</tr>
<tr>
<td></td>
<td>• Muscular or myotopic dystrophies</td>
<td></td>
</tr>
<tr>
<td>Social history</td>
<td>• Alcohol or caffeine consumption</td>
<td>Alcohol and caffeine are common triggers of atrial fibrillation or AVNRT episodes.</td>
</tr>
<tr>
<td></td>
<td>• Use of street drugs</td>
<td></td>
</tr>
<tr>
<td></td>
<td>• Occupation</td>
<td>Patients who work in areas with significant electromagnetic interference and require an implantable pacemaker or defibrillator should be evaluated for risk of pacing inhibition or inappropriate shock</td>
</tr>
</tbody>
</table>

AVNRT, atrioventricular nodal reentrant tachycardia; AV, atrioventricular.
Table 1.2.2  Ambulatory monitor selection.

<table>
<thead>
<tr>
<th>Type of monitor</th>
<th>Duration of use</th>
<th>Advantages</th>
<th>Disadvantages</th>
<th>Patient selection</th>
</tr>
</thead>
<tbody>
<tr>
<td>Holter monitor</td>
<td>24–48 hours</td>
<td>• Full disclosure</td>
<td>• Retrospective data</td>
<td>• Patients experiencing daily symptoms</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Provides arrhythmia counts (number of PVCs in 24–48 hours)</td>
<td>• Difficult to correlate to symptoms that are not daily</td>
<td>• Precise quantification of arrhythmias, heart</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Heart rate and AF burden in a graph format</td>
<td>• Single lead transmission</td>
<td>rate, or response to medications</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>• Short duration</td>
<td></td>
</tr>
<tr>
<td>Event monitor</td>
<td>1–30 days</td>
<td>• Ease of use</td>
<td>• Requires patient intervention to transmit</td>
<td>• Patients with infrequent symptoms lasting</td>
</tr>
<tr>
<td>(nonlooping memory)</td>
<td></td>
<td>• Lack of skin irritation</td>
<td>• No trending data provided</td>
<td>more than 5 minutes</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>• Compliance: If patient forgets the monitor at home, he or she will be</td>
<td>• Patients with allergy to electrode patches</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>unable to record when symptoms occur</td>
<td></td>
</tr>
<tr>
<td>Loop monitor</td>
<td>1–30 days</td>
<td>• Looping memory</td>
<td>• Patient must press record button to capture</td>
<td>• Patients with infrequent symptoms, particularly those that are brief</td>
</tr>
<tr>
<td>(looping memory)</td>
<td></td>
<td>• Ability to capture the initiation and termination of the episode</td>
<td>• Requires patient intervention to transmit</td>
<td>• For evaluation of drug effectiveness or</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>• No trend data</td>
<td>assessment of arrhythmia frequency and burden</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>• Skin irritation and compliance issues</td>
<td></td>
</tr>
<tr>
<td>Implantable loop monitor</td>
<td>Up to 2–3 years</td>
<td>• Continuous monitoring for prolonged period of time.</td>
<td>• Need for invasive procedure to implant</td>
<td>• Patients with have severe and infrequent</td>
</tr>
<tr>
<td>(ILR)</td>
<td></td>
<td>• Can be automatically or manually activated</td>
<td></td>
<td>symptoms such as syncope and negative EP study</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Extended monitoring period</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Table 1.2.2  (Continued)

<table>
<thead>
<tr>
<th>Type of monitor</th>
<th>Duration of use</th>
<th>Advantages</th>
<th>Disadvantages</th>
<th>Patient selection</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mobile cardiac outpatient telemetry (MCOT)</td>
<td>1–30 days</td>
<td>• Beat-to-beat analysis&lt;br&gt;• Symptom correlation&lt;br&gt;• Heart rate and AF trending&lt;br&gt;• Ability to contact patient real time&lt;br&gt;• Ability to report arrhythmias that meet MD notification criteria&lt;br&gt;• 96 hours of retrievable memory</td>
<td>• Skin irritation</td>
<td>• Patients with infrequent symptoms&lt;br&gt;• Patients who require monitoring and assessment of arrhythmia burden for known arrhythmias&lt;br&gt;• Patients with potentially more acute arrhythmias in which response time might be important&lt;br&gt;• Postablation arrhythmia burden&lt;br&gt;• Drug management</td>
</tr>
</tbody>
</table>

PVCs, premature ventricular contractions; AF, atrial fibrillation; MD, medical doctor.

The Ambulatory ECG (Holter) Monitor

Invented by Norman Holter, the Holter monitor is a battery-operated portable device that continuously records two or three electrocardiographic channels for 24–48 hours. The heart rhythm is recorded onto flash card technology and then processed. A wide variety of information can be obtained from this recording including heart rates during day and night, abnormal heart beats, and recording of the rhythm during symptoms. A diary comes with the Holter for the patient or caregiver to write down the time and symptoms so it can be correlated to the tracing.

Some Holter monitoring systems have the capability of recording or reconstructing a full 12-lead ECG for the duration of the 24–48-hour recording. This can be utilized to examine the morphology of arrhythmic beats. An example where a 12-lead Holter monitor may be useful is in the setting of a patient with frequent ventricular premature complexes (VPCs). It is often beneficial to have the full morphology of these beats to determine if they are monomorphic as well as to determine the site of origin as this may have implications in terms of treatment strategy.

Event Monitors

In many cases, 24–48-hour monitoring is not sufficiently long to document the cause of the patient symptoms or to detect sporadic arrhythmic events, as they may occur infrequently. Event or loop monitors are used by patients for longer periods of time, usually for 3–4 weeks. These recorders can be loop recorders, meaning that the device is always recording, but the device only saves or captures a single-lead or two-lead recording when the device is activated to save the data by depressing a button on the device. This allows for some ECG recording prior to when the button is pressed and is valuable for very transient symptoms. A typical recording will show when the trigger was activated. Some of these recorders store more than
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30 seconds of the ECG before the patient activates the recording and continues recording for another 30–60 seconds after the trigger ends. Some devices have the ability to record when an automatic programmable trigger is activated, for example, the heart rate is less than 50 beats per minute or greater than 150 beats per minute. In addition, devices have automatic algorithms that can detect atrial fibrillation and record when the rhythm converts to atrial fibrillation.

Nonlooping event monitors only record when activated and are not continually recording. These can be as small as a credit card with two electrodes and can be pressed to the skin during symptoms (Fig. 1.2.1).

Nonlooping monitors are not particularly useful for very transient symptoms or arrhythmias as the arrhythmia may be missed in the time needed to place the device on the chest wall. One potential advantage of nonlooping monitors is that they do not require any adhesive electrodes that can sometimes cause skin irritation.

Once the event is recorded, patient needs to transmit it to a central processing station over a standard telephone line. There are some disadvantages to these systems. For example, not all patients are compliant with the instructions to wear the monitor all the time. If the episode occurs when the monitor is off, the data cannot be collected. In addition, if the patient has symptoms but does not activate the recording by pressing the button and the automatic algorithms are not activated, there will be no valuable diagnostic data.

**Mobile Cardiac Outpatient Telemetry (MCOT)**

Another type of outpatient monitoring system incorporates cell phone technology so that the patient does not have to transmit the recordings using a standard telephone line, rather the recordings are automatically transmitted wirelessly to the central monitoring system. In addition, because all the beats are recorded and analyzed, the data from these systems includes the average heart rate, minimum and maximum heart rates, as well as the amount of time the patient spends in atrial fibrillation. Another advantage of cell phone technology is that dangerous rhythms can be identified quickly, and with the patient in possession of a cell phone the patient can be contacted if needed and emergency services could also be activated if needed (Fig. 1.2.2).

**The Implantable Loop Recorder (ILR)**

Some patients may have infrequent symptoms that cannot be recorded by regular event recorders because they occur very rarely. In these cases, it may be necessary to implant an ILR. An ILR may be a device that is rectangular in shape about the size of a pack of chewing gum or more oval similar in shape to many pacemakers. The recorder is inserted under the skin along the left sternal border or in a standard position above the pectoral muscle (Fig. 1.2.3).

The battery lasts up to 3 years. This device can be automatically or manually activated to store patient-activated episodes. The recordings may be downloaded using a programmer that is used to interrogate pacemakers and defibrillators. Previous investigators have examined the diagnostic utility of the ILR. In one study of 24 patients implanted with ILR for recurrent syncope of unclear etiology, 21 patient had syncope within 5 months after the implantation and in 18 of those patients an accurate cause of the syncope was defined using the ILR.

For example, this rhythm strip was recorded in a patient during an episode of light-headedness (Fig. 1.2.4).

**Signal-Averaged ECG (SAECG)**

SAECG is a method of filtering the surface ECG signal in order to detect late ventricular poten-
Figure 1.2.1  Event monitor recording in a patient shows an episode of ventricular tachycardia. © CardioNet. Reprinted with permission.

Figure 1.2.1  Event monitor recording in a patient shows an episode of ventricular tachycardia. © CardioNet. Reprinted with permission.

Image not available in the electronic edition

Prospects for the Noninvasive Detection of Late Potentials that can be usually present in patients with the substrate for reentrant ventricular arrhythmias. These late potentials are of very low amplitude and cannot be detected by routine electrocardiography. The SAECG method improves signal-to-noise ratio and provides filtering of noise, permitting detection of ventricular potentials of less than 1 mV. These late potentials have been recorded in 70–90% of patients with spontaneous sustained and
inducible ventricular tachycardia (VT) after myocardial infarction (Berbari 2004). Unfortunately, despite the presence of late potentials in patients at risk for ventricular arrhythmias, the utility of this test to risk-stratify patients for placement of an ICD is poor and routine use of the SAECG is no longer common for these patient groups (Kudaiberdieva et al. 2003).

At the current time, the use of the SAECG is limited to patients with suspected arrhythmogenic right ventricular dysplasia (ARVD), a condition where the normal ventricular myocardium of the right ventricle is replaced by a fibro-fatty infiltrate. These patients commonly have ventricular arrhythmias and may even have had an episode of sustained VT or cardiac
Figure 1.2.3  The implantable loop recorder. Reprinted with permission of Medtronic, Inc.

Figure 1.2.4  The implantable loop recording in a patient with syncope.
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Figure 1.2.5 Abnormal SAECG; shows late potential.

arrest. The SAECG can also be abnormal in cases of ARVD and may be a helpful diagnostic tool in these cases (Fig. 1.2.5).

Cardiac Structural and Functional Assessment

Part of the comprehensive assessment of the patient undergoing evaluation for arrhythmia is a structural assessment of cardiac function. This can be accomplished with different imaging modalities, most commonly echocardiography. Other imaging modalities such as computed tomography (CT) or nuclear magnetic resonance imaging (MRI) may also be used in arrhythmia patients to gain insight on the anatomy and possible structural abnormalities. In addition, nuclear imaging can be used to evaluate for coronary artery disease and evaluate cardiac function (Strickberger et al. 2006).

Echocardiography

When evaluating a patient with an arrhythmia, an echocardiogram can be a very effective tool to assess cardiac function, and evaluate for chamber enlargement or hypertrophy and valvular heart disease. In addition, an echocardiogram may also add insight to the presence of hemodynamically significant problems such as pulmonary hypertension. Identification of a low ejection fraction (EF) may identify patients with indications for the implantation of an ICD, such as in patients with a low left ventricular EF. Determination of a low EF may identify patients that may benefit from medications
such as angiotensin-converting enzyme (ACE) inhibitors or beta blocking therapy. In addition, in patients with low EF, antiarrhythmic medications such as class IC agents should be avoided.

In patients being considered for catheter ablation or surgery for atrial fibrillation, assessment of the left atrial size by echocardiography may be used to predict procedural success. Although routine transthoracic echocardiography is unable to detect the presence of left atrial thrombus in most patients, another form of echocardiography, the transesophageal echocardiogram (TEE), accurately detects atrial thrombus and may be used prior to electrical cardioversion or catheter ablation.

Combining cardiac echocardiography with exercise stress testing is also commonly used to assess for the presence of coronary artery disease, as this has a significant impact on the types of arrhythmias that the patient may at risk for. Stress echocardiogram provides information on left ventricular wall motion with exercise. Patient who are not able to exercise may have pharmacological stress test using echocardiography (Fig. 1.2.6).

**Cardiac CT**

Cardiac CT allows tomographic imaging of the heart and coronary arteries. It allows precise quantification of left ventricle (LV) volumes, EF, LV mass, and segmental images of the coronary arteries. This noninvasive test can be used to screen for the presence of coronary artery disease. In addition, cardiac CT is used to acquire images before pulmonary vein isolation procedures as there can be a significant amount of variation in the anatomy of the pulmonary veins. The CT image of the left atrium can be used during the procedure as a guide as to optimal ablation targets. In addition, electroanatomical mapping systems that are commonly used during ablation procedures allow the CT scan of the left atrium to be imported into the electroanatomical mapping system software. This allows the clinician performing the ablation to have an accurate representation of the heart and have a better understanding of the catheter position within the heart (Fig. 1.2.7).

**Cardiac MRI**

Cardiac MRI is used to evaluate cardiac structure, function, perfusion, and myocardial viability. By using delayed enhancement, cardiac MRI is able to detect scar due to myocardial infarction or nonischemic cardiomyopathy. The presence of scar tissue makes improvement in cardiac function less likely. In addition, the amount of scar due to myocardial infarction may be related to the likelihood of inducing

![Figure 1.2.6 Intracardiac echocardiogram.](image)

![Figure 1.2.7 Cardiac computed tomography (CT).](image)
ventricular arrhythmias during electrophysiological testing. Cardiac MRI can also provide valuable diagnostic information in patients with suspected ARVD, as the MRI may be able to detect the presence of abnormal fibro-fatty infiltrate in the right ventricle as is seen in this condition (Fig. 1.2.8).

Exercise Stress Test

The exercise stress test can be combined with imaging modalities such as echocardiography or nuclear imaging to test for the presence of significant coronary disease. In addition, this test may be of value in arrhythmia patients without additional imaging. For example, stress testing may be used to reproduce exercise-induced arrhythmias or to evaluate the effectiveness of rate control medications. Exercise stress test is also used for patients with pacemaker or ICD to make adjustments to the sensor programming or to determine how to best program the upper tracking rate. For example, a young, very active patient with an ICD who may be inappropriately shocked for sinus tachycardia may benefit from a stress test to evaluate what the maximal sinus rates are during exertion. A clinician can then make the appropriate adjustments to the ICD programming. Stress testing may also be combined with oxygen consumption measurement to assess the metabolic changes during exercise. Such data may be used to assess the severity of left ventricular dysfunction and distinguish functional limitations due to cardiovascular causes from respiratory causes.

Case 1.2.1

A 34-year-old Asian man is evaluated in clinic for a syncopal episode. He states he passed out while talking on the phone but spontaneously recovered. He states this was his first and only syncopal episode. He denies any associated symptoms, including chest pain, shortness of breath, dizziness, light-headedness, or nausea. He denies any recent illness or any other similar episodes. He is not sure how long he was unconscious. He denies loss of bowel or bladder control. He states he did not injure himself during the fall.

He is not allergic to any medications. He takes ibuprofen occasionally for headaches. His past medical history is significant for tonsillectomy as a child. His mother is 76 years old and has diabetes and glaucoma; his father is 82 and has atrial fibrillation and gout. He has three younger siblings without any known cardiac history. His older brother died in his sleep at the age of 30. He is an engineer, recently married. He smoked occasionally while in college. He drinks socially. He denies using illicit drugs.

An ECG obtained in a clinic showed concave ST elevation in leads V1, V2, and V3. His echocardiogram shows no evidence of structural abnormalities and normal function (Fig. 1.2.9).

Discussion

In this patient, the likely reason for his syncope can be made just from the ECG findings. Above-
mentioned ECG findings are typical for patients with Brugada syndrome. It is manifested by right bundle branch block, concave ST segment elevation, and T wave inversion in the right precordial leads V1 through V3, with no reciprocal ST segment depression. The concave pattern of the ST segment has contributed to premature repolarization and/or conduction delay in the right ventricle.

Approximately 5% of patients who experience sudden cardiac death have no structural heart disease or obvious cause and are diagnosed as having idiopathic ventricular fibrillation (Kurita et al. 2002). The Brugada syndrome, first described by Dr. P. Brugada in 1992, is a genetic disorder that predominantly affects Asian and Caucasian men (Kurita et al. 2002). Because it is often a hereditary syndrome, family members should be screened for the disease.

This patient underwent an electrophysiology study that revealed easily inducible polymorphic VT. An ICD was placed. During follow-up, he was successfully treated by his device for an episode of fast polymorphic VT.

**REFERENCES**


RESOURCES
Section 2

Supraventricular Tachycardia: Diagnosis and Management
INTRODUCTION

Supraventricular tachycardia (SVT) is the most common symptomatic arrhythmia affecting patients. Approximately 2.3 of every 1,000 persons in the general population will experience an SVT in their lifetime (Orejarena et al. 1998). When analyzing the reasons for 1.1 billion emergency room visits over a 10-year period, 550,000 were found related to SVT (not including atrial fibrillation or flutter). About a quarter of these emergency visits resulted in hospitalization (Murman et al. 2007).

Given the multiple potential for health care workers to interface with patients who suffer from SVT, allied professionals are called on to provide quality care to patients with these arrhythmias. To provide such care, a comprehensive understanding of the underlying mechanism of SVT, appreciation for the clinical presentation, and knowledge of the methods presently available to treat SVT is essential.

What is SVT?

Technically, any arrhythmia that involves part or all of the atrial myocardium or the atrioventricular (AV) junctional region is an SVT. Atrial fibrillation, atrial flutter, atrial tachycardia, sinus tachycardia, and inappropriate sinus tachycardia are all examples of SVT that require the atrium for initiation or sustenance. Junctional tachycardia is an arrhythmia of the AV junction tissue that occurs with or without AV involvement. AV reciprocating tachycardia (AVRT) is an SVT that requires both atrial and ventricular tissue to occur (see below). Finally, the most common of the SVTs, AV nodal reentrant tachycardia (AVNRT), is a complex...
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Arrhythmia that can occur at times independent of the atria, compact AV node, and ventricular tissue. The critical component of this arrhythmia appears to be a portion of atrial tissue in the vicinity of the compact AV node. Several mechanisms of arrhythmogenesis may be operative in the various SVTs. AVNRT and AVRT are classical examples of reentrant arrhythmias, whereas most forms of atrial tachycardia involve enhanced automaticity.

In this review, we will discuss three common forms of SVT; namely, AVNRT, AVRT, and atrial tachycardia. Atrial fibrillation and other complex atrial arrhythmias are discussed elsewhere.

**CLINICAL AND ELECTROCARDIOGRAPHIC PRESENTATION OF SVT**

The typical patient presenting with SVT is young and otherwise healthy. The usual scenario is one of an abrupt onset of tachy palpitations that is uncomfortable but rarely results in loss of consciousness. The offset (termination) is also usually abrupt and the patient is able to return to normal activities fairly soon. Usually, a specific trigger or circumstance for the arrhythmia cannot be identified; however, some patients may note a particular position, excessive caffeine intake, or exercise initiating the arrhythmia and mild fatigue along with an urge to micturate when the arrhythmia subsides. SVT presents electrocardiographically usually as a regular narrow complex tachycardia. When bundle branch block is present or antegrade conduction over an accessory pathway is occurring during the tachycardia, a wide QRS complex results. The reader is referred to several established criteria that help distinguish SVT from ventricular tachycardia when wide QRS tachycardia is seen (Wellens et al. 1978; Akhtar et al. 1988).

Of the three main causes of SVT (AVNRT, AVRT, and atrial tachycardia), the most common in all age groups is AVNRT. There is a slight preponderance of occurrence in females, and with advanced age, AVNRT and atrial tachycardia become more prevalent, whereas AVRT is relatively more common in the young (Lockwood et al. 2004; Porter 2004; McElderly and Kay 2006).

The least common of the tachycardias discussed in this review, focal atrial tachycardia, accounts for approximately 5–15% of SVTs in adults undergoing electrophysiology study (Ellenbogen et al. 2004; Porter 2004; Roberts-Thomson et al. 2006a,b). However, atrial tachycardia may often coexist with atrial fibrillation or atrial flutter and in fact may be the primary arrhythmia initiating or maintaining atrial fibrillation (Shah et al. 2002; Asirvatham 2007a,b).

In this chapter, the three forms of SVT discussed will be individually reviewed. The underlying anatomy and physiology, clinical and electrocardiographic presentation, medical management, and invasive diagnosis and ablative therapy are elaborated. Following this, a suggested algorithmic approach for differential diagnosis and management is presented.

**AVNRT**

AVNRT is likely the first of the SVTs that is easily learned by the beginner. The student quickly appreciates the workings of the arrhythmia, rationale for treatment, and methods. This rhythm at first glance appears to be the most straightforward to ablate. On looking further, however, AVNRT is extremely complex, with many unsolved riddles. Experts in the field, even today, are unclear as to the exact mechanism of the arrhythmia and some of its variants, and there is considerable debate as to why radiofrequency energy is beneficial when delivered in certain locations. Despite the lack of exact understanding, however, once the caregiver is adept at recognizing this arrhythmia, excellent treatment options that are reviewed
Anatomy and Physiology

Electrophysiologists have been intrigued by the circuit responsible for the maintenance of AVNRT. The debate centers on whether atrial myocardium is primarily responsible for this arrhythmia (thus making it a type of atrial tachycardia) or the AV junction alone is capable of maintaining this arrhythmia (thus making it a type of junctional tachycardia). Initial observations that continue to be made today in the electrophysiology (EP) laboratory show that this tachycardia can routinely be dissociated from the ventricle, His bundle, and infra-Hisian conduction. In addition, at times, the arrhythmia has also been clearly shown to be dissociated from the atrium. Some of the early leaders of electrophysiology concluded that AVNRT is a form of junctional tachycardia based on these findings. However, other observations including the fact that the arrhythmia could sometimes be dissociated from the AV node and be reset or terminated with pacing or other maneuvers in the atrium have led others to believe the circuit is mostly or entirely in the atrium.

Dr. Sunao Tawara, in his original work (Tawara 1906), confirmed the presence of “specialized” myocardium that connected not only into the atrium but also into the AV node. The compact AV node anatomy itself was first characterized by the German pathologist Wilhelm Karl Koch as being situated in what is now termed Koch’s triangle. The anterior border of the triangle is formed by septal leaflet of the tricuspid valve and the posterior border by tendon of Todaro, which in turn is a continuation of the valve of the inferior vena cava (Eustachian valve). The apex of this triangle is directed superiorly and towards the ventricle and becomes continuous with the membranous portion of the interventricular septum. Here the compact AV node connects to the penetrating bundle of His as it becomes encased within the central fibrous body. The base of Koch’s triangle (inferior portion) is bounded by the ostium of the coronary sinus (CS) and the septal portion of the cavitricuspid isthmus. The compact AV node itself is a histologically discrete group of cells located within this triangle in the atrium with connections to the His bundle at the membranous septum and right and leftward attachments toward the tricuspid and mitral valve, respectively. In addition, cells that have features of both node and atrium (transitional cells) extend beyond the compact node even further along the tricuspid annulus and in contact with the left atrial septum in the region of the CS (Anderson and Ho 1994) (Fig. 2.1.1). Thus, it was established that discrete atrial inputs into the AV node from the atrial myocardium existed. It was immediately appreciated that when there is more than one pathway (AV node atrial connections), the potential for reentrant arrhythmia existed.

Dr. Gordon Moe (Mendez and Moe 1966) observed that critically timed premature atrial impulses were shown to block in one part of a continuous circuit (later called the fast pathway) and conducted over a separate portion of the circuit (slow pathway) with shorter effective refractory periods. The delay in conduction along the second limb of the circuit allowed recovery in the initial limb that blocked and subsequent retrograde activation. The resultant circuit’s activity produced sustained AVNRT. The exact anatomic localization of these separate pathways in the nodal region was explored by Sung et al. (1981). They demonstrated that the slow pathway input was located in the region of the CS ostium within the septal isthmus (it later became appreciated that the CS myocardium itself connects to this tissue, lengthening this extension). The fast pathway consistently originated on the atrial wall anterior (in front) of the fossa ovalis just posterior to the compact AV node (behind the tendon of Todaro) (Anderson and Ho 1994; Lockwood et al. 2004) (Fig. 2.1.2). It was the
later appreciation of the anatomical discreetness of the two limbs of the AVNRT circuit that allowed successful ablation therapy targeting one of these limbs without affecting the compact AV nodal tissue at all.

**Terminology**

It is prudent for the learner to be completely familiar with some of the common terms used in describing the physiology of AVNRT.

- **Fast and slow pathways:** These structures are distinguished not by the speed of conduction through them but rather by their discrete anatomical locations. Portions of the fast pathway may or may not conduct faster than slow pathway tissue, but it is always located anterior and behind the tendon of Todaro. The slow pathway is defined by its posterior location related closely to the ostium of the CS. Thus, during ventricular pacing with retrograde conduction via the AV node,
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Figure 2.1.2 Circuit for AVNRT. The slow pathway region denoted in green is located in the region of the CS ostium in the septal isthmus. The fast pathway exit site, denoted in blue, is located just behind the tendon of Todaro. During typical AVNRT, antegrade conduction occurs via the slow pathway and retrograde via the fast pathway. CT, crista terminalis; EV, Eustachian valve; ER, Eustachian ridge; CS, coronary sinus; IVC, inferior vena cava; FO, fossa ovalis; TCV, tricuspid valve. (With permission from the Mayo Foundation for Medical Education and Research. All rights reserved.)

if the site of earliest activation is noted to be in the CS ostial region rather than behind the tendon of Todaro, then slow pathway conduction is said to exist.

- **Typical AVNRT:** Earliest atrial activation in the region of the fast pathway (behind the tendon of Todaro) identifies the type of AVNRT to be “typical.” Antegrade conduction to the AV node occurs through a slow pathway and retrograde conduction via the fast pathway. The circuit is then completed using variable portions of atrial myocardium (Lockwood et al. 2004).

- **Atypical AVNRT:** Whenever the site of earliest conduction during tachycardia is not the fast pathway region, then the AVNRT is called atypical. The retrograde limb may be one of several slow pathway inputs related to the inferior right and leftward extensions of the AV node and the continuous atrial myocardium.

Atypical AVNRT is sometimes further distinguished into fast-slow and slow-slow variants. Earlier, it was felt that in fast-slow AVNRT, the antegrade limb was made up of the fast pathway and thus was a mirror image of typical AVNRT. More recent thought based on a significant body of evidence has shown that the fast pathway is a bystander and is not a limb of the fast-slow
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It is useful for the beginner to visualize AVNRT as a relatively simple circuit. The AV node has two or more extensions, with the superior extension exiting behind the tendon of Todaro being the fast pathway and the more inferior extensions (right or leftward) being the slow pathway. The circuit can be visualized as conduction proceeding antegrade through the slow pathway and retrograde to the fast pathway and then atrial myocardium being used to complete the circuit. When visualized, it is easy to appreciate that ablation or other maneuvers targeting the anatomically distinct slow pathway will result in termination and potentially cure this arrhythmia.

While useful, this simple construct does have several flaws that require a far more complex and intricate set of explanations. These detailed descriptions of AVNRT variants and further concepts on the underlying anatomy and physiology of the upper and lower common pathways are beyond the scope of this chapter but can be found in several excellent references (Lockwood et al. 2004; Gonzalez and Rivera 2006; Nakagawa and Jackman 2007).

**Diagnosis**

A characteristic clinical presentation and electrocardiogram (EKG) typically suggest the diagnosis of AVNRT. During the EP study, a limited amount of mapping and dynamic pacing maneuvers usually quickly identify this arrhythmia and distinguish it from AVRT and atrial tachycardia.

**Clinical Presentation**

All age groups can be affected by AVNRT. There is a slight female preponderance. Patients describe sudden, regular, strong palpitations (out of the blue) that seem to disappear as abruptly as they started. Most patients who experience one episode will eventually get another, and over time, the episodes become
frequent. This typically gives rise to lifestyle modifying symptoms.

The onset can occur either at rest or with exercise, but in a given patient, one situation may provide a more constant consistent trigger. Frequently, patients feel fatigued for a few hours after the termination of arrhythmia and some will give a history of having the urge to micturate soon after the offset of arrhythmia.

Some patients may complain of an uneasy sensation in the neck from the Cannon “A” waves, and in the elderly, syncope may be the presenting feature. Often, when syncope occurs, it is not the tachyarrhythmia itself that leads to fainting, but when the arrhythmia terminates, a sinus pause (reflecting underlying sinus node dysfunction) may result giving rise to presyncope or frank syncope. Because of resulting amnesia, the patient may not remember the palpitation and the presentation and workup becomes that of syncope.

**Electrocardiographic Characteristics**

Whenever possible, a 12-lead EKG should be obtained during tachycardia, as in most instances it can be diagnostic. A regular narrow complex tachycardia is seen often with rates between 150 to 250 beats per minute. The P waves are usually negative and narrow in the inferior leads (reflective of the exit on the interatrial septum behind the tendon of Todaro). The P waves may occur before, after, or during the QRS complex. Usually, they are seen just following the QRS complex (short RP tachycardia) and may produce a terminal positive deflection on the QRS in lead V1 called the pseudo-R’ or an additional pseudo-S wave in the inferior leads (Fig. 2.1.3). The RP interval can be any value in AVNRT; however, when the RP interval is very short (≤100 ms), AVNRT is far more likely than AVRT, although atrial tachycardia is still possible (see below).

![Figure 2.1.3](image-url) Comparison of EKG during SR and typical AVNRT. The left side of the panel shows a 12-lead ECG obtained during normal sinus rhythm (NSR). The right side of the panel shows a 12-lead EKG during typical AVNRT taken in the same patient. Arrows on the left side of the panel indicate the presence of a pseudo-S wave in lead III and a small deflection in the terminal portion of lead V1 (pseudo-R’) not visible during NSR.
Electrophysiological Findings

The electrophysiology findings that lend support to the diagnosis of AVNRT include maneuvers performed during arrhythmia as well as pacing maneuvers when the patient is in normal rhythm. The pacing maneuvers primarily identify the presence of dual AV nodal physiology and with ventricular pacing determine that no accessory pathway is present and that ventriculoatrial (VA) conduction is through the AV conduction apparatus.

**Dual AV Nodal Physiology**

With ventricular pacing, the presence of retrograde conduction occurring either through the fast or slow pathway is relatively straightforward. As described above, if the earliest atrial activation is behind the tendon of Todaro, then retrograde fast pathway conduction is present, and similarly, if earliest conduction is in the CS ostial region or the posteroseptal tricuspid annulus, then retrograde conduction is through the slow pathway AV nodal input.

It is much more difficult to demonstrate that fast and slow pathways are present during sinus rhythm or pacing the atrium since, regardless of which pathway is used, the first ventricular activation is still at the same site via the infra-Hisian conduction system. However, the difference in the refractory periods and conduction properties of the two pathways usually allows demonstration of the phenomenon of dual AV nodal physiology.

Dual AV nodal physiology is defined as an increase in 50 ms or greater in the interval between an atrial extra stimulus and its associated His bundle electrogram (A2H2 interval) that occurs when the coupling interval between the drive train and the placed extra stimulus (A1A2) has been shortened by a 10-ms decrement or less. In other words, an abrupt increase in the AH interval, termed a “jump,” occurs, signifying that the fast pathway is blocked and now conduction is via the antegrade slow pathway (Fig. 2.1.4). The reason the abruptness of the jump is carefully defined and important is because some increase in the AH interval will

![Figure 2.1.4](image)

**Figure 2.1.4** Dual AV node physiology. The left side of the panel demonstrates pacing in the high right atrium (HRA) at a cycle length of 600 ms followed by a single atrial premature extra stimulus delivered at an interval of 530 ms. The corresponding A2H2 interval as measured on the distal His bundle electrogram (His 1) is 560 ms. During the subsequent atrial drive train of 600 ms, a single atrial premature stimulus is delivered at 520 ms (right side of panel). The corresponding A2H2 interval measures 620 ms. The prolongation in the A2H2 interval of 60 ms meets the criteria for dual AV node physiology.
occur simply as a result of decrement in the AV node. Some patients may have more than one “jump,” possibly signifying multiple slow pathways. Conversely, some patients with AVNRT do not demonstrate dual AV nodal physiology, and in these patients the use of isoproterenol or adenosine may be required (Belhassen et al. 1998). The student must recognize that VA conduction does not have to be present for AV node reentry to occur. In other words, if there is complete VA block during ventricular pacing, while an accessory pathway mediated tachycardia can be excluded, AV node reentry remains the diagnosis. This is because tachycardia does not require participation of the majority of AV nodal tissue and the infranodoseptal system where retrograde block may be occurring.

**Intracardiac Activation Patterns**

Intracardiac electrograms during typical AVNRT characteristically demonstrate near simultaneous AV activation (very short VA times). As mentioned previously, atrial activation may precede, occur during, or succeed ventricular activation, but the very short (sometimes zero) VA times distinguish this arrhythmia from AVRT. The reason for the possible very short VA time is that there is really no VA conduction occurring during AVNRT. When the wavefront comes down the slow pathway, it is from a common turnaroundpoint that activation now simultaneously precedes antegrade to the ventricle and retrograde back up to the atrium. Thus, the His to atrial (HA) and VA times are both “pseudointervals.” This is an important concept to remember since one should not be surprised when atrial activation occurs even before the His bundle electrogram during tachycardia (Fig. 2.1.5).

Retrograde activation is said to be concentric (septum earlier than free wall). It is more useful to think about retrograde activation based on the earliest site of recorded atrial electrograms. In most EP studies, a His bundle and CS catheter will be placed. When retrograde activation is earliest to the CS ostium, the retrograde slow pathway activation (atypical AVNRT) may be present (Fig. 2.1.6). On the other hand, earliest recorded atrial electrograms on the proximal His bundle catheter suggest retrograde fast pathway activation consistent with typical AVNRT. It should be remembered, however, that the fast pathway locations are behind the tendon of Todaro. We typically place the His bundle catheter in front of the tendon of Todaro. Thus, when doubt exists, a specific mapping catheter should be placed behind the tendon to ascertain that fast pathway activation is earliest.

Sometimes, in typical AV node reentry, the CS activation occurs from the lateral to septal manner (so-called eccentric activation). This is because when early activation occurs behind the tendon of Todaro, there may be block across the Eustachian ridge preventing wavefront propagation to the CS ostium. In these instances, activation proceeds to the left atrium and from the left atrium through muscular connections to the CS at variable distances including its mid and distal portions and then the wavefront propagates in the CS toward the ostium, giving the appearance of early lateral activation. This situation will be clarified based on dynamic maneuvers such as para-Hisian pacing and the fact that the fast pathway sites will still be activated earlier than the earliest location in the CS.

It should be noted that typical and atypical AVNRT cannot be reliably distinguished based on the VA time. Sometimes, the shortest VA times will be seen in atypical AVNRT. Again, this is a consequence of the VA interval being a pseudointerval, essentially a race, between antegrade conduction from the common turn-around point (which may be proximal to a lower common pathway or close to the AV node) to the ventricle and retrograde to the atrium. However, the arrhythmias are easily distinguished from each other by simple analysis of the earliest site of atrial activation as noted above.
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Figure 2.1.5 Intracardiac electrograms (EGMs) obtained during typical AVNRT. Pictured from top to bottom are surface leads II and V1 followed by EGMs recorded from the high right atrium (HRA), His bundle proximal (His 4) to distal (His 1), RV apex, and coronary sinus proximal (CS 19/20) to distal (CS 1/2). Note the appearance of near simultaneous ventricular and atrial activation (VA time <80 ms). The red arrow denotes atrial activation in the His bundle EGM, which precedes ventricular activation.

Dynamic Maneuvers to Diagnosis Tachycardia Mechanism

The principle maneuver to define the mechanism of SVT is the placement of sensed premature ventricular contractions (PVCs). The sensed PVCs are first placed with a very late coupling interval. These PVCs occur at the time of His bundle refractoriness (after antegrade conduction through the His bundle as just occurred). Thus, if the PVCs are able to pre-excite the atrium, then they must have traveled through an extra nodal pathway (accessory pathway) (Fig. 2.1.7). Since in AVNRT there is no mechanism other than the AV node or VA conduction, only PVCs with a short coupling interval (high preexcitation index) can pre-excite the atrium.

With carefully placed catheters and recorded His bundle electrograms, it would be noted that PVCs pre-excite the atrium only when advancing (pre-exciting) the retrograde His bundle deflection. The magnitude of atrial pre-excitation can never be more than the magnitude of retrograde His bundle pre-excitation since it is only through pre-exciting the His that the A can come earlier. However, to demonstrate pre-excitation via the AV node with PVCs
Electrophysiology dictum #1: An early coupled sensed PVC that pre-excites the atrium by pre-exciting the His by an equivalent or greater amount did so without a change in activation sequence and reset the tachycardia. What is meant by reset is that the pre-excited A in turn perturbs (pre-excites or delays) the antegrade limb of the circuit, which in turn affects the subsequent retrograde limb, and so on. The extent to which the retrograde His needs to be pre-excited is different in the various forms of AVNRT. Typically, the His needs to be pre-excited only by about 10 ms to pre-excite the A in typical AVNRT, but pre-exciting (pulling in) the retrograde His by as much as 40–50 ms may be required to demonstrate pre-excitation of the A and subsequent reset in atypical AVNRT.

**Electrophysiology Diagnosis of Retrograde Conduction**

An essential component of the EP study to distinguish AVNRT from AVRT is to define when...
arrhythmia is not present. One critical determinant is whether VA conduction is via the AV node or through an accessory pathway. Several maneuvers can be helpful in this regard.

- Para-Hisian pacing: This is an extremely useful maneuver that is based on the fact that the penetrating bundle of His is insulated, and high output pacing near this structure is required to directly capture the His bundle. The pacing catheter is placed close to the distal His bundle region at low-/regular output pacing. Only the local ventricular myocardium is captured, resulting in a wide QRS complex. For His bundle activation to occur, propagation of this wavefront needs to precede antegrade toward the apex where the distal right bundle can be entered and then there is retrograde propagation from the right bundle to the His bundle itself. Thus, the VH interval is long when only the ventricular myocardium is captured (wide QRS). Since the VH interval is long, the VA interval is also long when AV nodal conduction is the mechanism of retrograde VA activation. In contrast, when high-output pacing from the same location and same rate is performed, not only is the local ventricular myocardium captured (as it was with low output pacing) but now there is also
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VA times without the change in the activation sequence when compared to high-output pacing (short VA and VA times), then retrograde conduction is through the AV node.

Since retrograde accessory pathway activation is independent of His bundle direct capture of the His bundle; thus, the VH interval is short, and if retrograde conduction is via the AV node, then the VA interval is short as well.

**Electrophysiology dictum #2:** With para-Hisian pacing, if, with lowoutput stimulation, a wide QRS results with long VH and

**Figure 2.1.8** Placement of sensed PVCs during tachycardia. Pictured from top to bottom are surface leads II and V1 followed by EGMs recorded from the high right atrium (HRA), His bundle proximal (His 4) to distal (His 1), RV apex, and coronary sinus proximal (CS 19/20) to distal (CS 1/2). The tachycardia cycle length is 330 ms. The first PVC, indicated by the large pacing artifact closest to the left hand margin, occurs at a time when the His bundle is refractory to stimulation. The first two red arrows show that the His timing remains unperturbed at 330 ms as it remains activated in an antegrade fashion. A second PVC follows, which pre-excites the His (His-His interval now 280 ms). This in turn is followed by pre-excitation of the atrium (AA interval now 295 ms). Note the identical atrial activation sequence throughout the tracing. Note also that tachycardia is reset. This confirms tachycardia as typical AVNRT.
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Activation, as long as the local ventricular myocardium is captured (both with low- and high-output pacing), pathway activation occurs and the VA intervals are the same.

Electrophysiology dictum #3: If during para-Hisian pacing, regardless of whether a wide QRS (low-output pacing) or a narrow QRS (high-output pacing) occurs, the VA conduction time is fixed and there is no change in the activation sequence, then retrograde VA conduction is via an accessory pathway (Fig. 2.1.9).

- **Decremental pacing**: Pacing from anywhere in the right ventricle at gradually shorter cycle lengths will show decremental conduction and eventual Wenckebach to the atrium with AV nodal conduction. With accessory pathway conduction, there is usually no discernable decrement and VA block occurs abruptly. While this maneuver is simple to perform, care is needed since some pathways show decremental conduction properties.

- **Differential site pacing**: This pacing maneuver is especially useful when an inferior/paraseptal pathway is suspected. It involves pacing from the apex and then comparing VA conduction patterns and timing with pacing occurring close to the base of the right ventricle. If retrograde conduction is via the AV node, then, when pacing from the base, conduction needs to proceed down toward the apex before entering the right bundle, His, and then the AV node and atrium. Thus, the VA time is long.

![Figure 2.1.9](image)

**Figure 2.1.9** Para-Hisian pacing. Pictured from top to bottom on the left panel are surface leads II and V1 followed by EGMs recorded from the high right atrium (HRA), His bundle proximal (His 4) to distal (His 1), RV apex, and coronary sinus proximal (CS 9/10) to distal (CS 1/2). Top to bottom on the right panel are surface leads II and V1 followed by EGMs recorded from the RV apex (RVA), high right atrium (HRA), His bundle proximal (His 4) to distal (His 1), and coronary sinus proximal (CS 19/20) to distal (CS 1/2). Pacing is occurring from the distal His bundle electrode at high and low output. High-output pacing results in direct capture of the His bundle resulting in a narrow QRS complex (black arrows). As output is reduced direct capture of the His bundle fails and only local ventricular myocardium is captured resulting in a wide QRS complex (red arrows). In the left panel, the VA timing remains the same with both direct capture of the His bundle and with capture of the local myocardium only indicating presence of a retrograde conducting accessory pathway. In the right panel, during capture of only local ventricular myocardium, the VA interval lengthens as a result of increased V to His conduction time. Note the visible His marked by the blue arrow. All conduction in this case proceeds in a retrograde fashion via the AV node. See the text for more details.
The retrograde activation sequence will look similar to when pacing from the apex. At the apex, because the pacing site is close to the entrance of the infra-Hisian conduction system, the VH and VA times will be shorter.

**Electrophysiology dictum #4:** When pacing at the same cycle length closer to the apex results in a shorter VA interval than when pacing close to the base, retrograde conduction is likely through the AV node. Care should be used, however, since some accessory pathways may be long and insert away from the base. Further, left-sided accessory pathways may be equidistant to the AV node when pacing at the right ventricular (RV) base.

- **Combination maneuvers:** Atypical AVNRT can sometimes be difficult to distinguish from AVRT involving a right or left posteroseptal accessory pathway, with atrial activation being very similar to retrograde slow pathway activation. In both cases, the earliest recorded electrograms occur in the region of the CS ostium. A maneuver that can be utilized to discriminate between these two tachycardias involves pacing at the RV apex during tachycardia at a cycle length approximately 20 ms shorter than the tachycardia cycle length. During pacing, entrainment of the tachycardia (retrograde VA conduction and speeding up of the atrial rate to the paced cycle length) should be verified. Upon cessation of pacing, tachycardia continues. Measurement from the last paced beat to the first return electrogram on the RV catheter (postpacing interval) is obtained. The tachycardia cycle length is subtracted from the postpacing interval, and this interval is found to be shorter during orthodromic reciprocating tachycardia (ORT) (because the ventricle is part of the circuit) than during atypical AVNRT. A difference between the postpacing interval and tachycardia cycle length of greater than 115 ms is highly suggestive of atypical AVNRT. Care should be taken, however, since if pacing occurs close to the right bundle, then relatively early penetration into the AV node and entrainment of the tachycardia may occur. Conversely, if the pathway is very decremental and poorly conducting, the maneuver cannot be successfully performed.

**Noninvasive Management of AVNRT**

Management of patients with AVNRT involves careful patient education and different approaches for terminating arrhythmia as well as preventing occurrence. For acute episodes, particularly when the arrhythmia is well tolerated, vagal maneuvers such as invoking a gag reflex, Valsalva maneuver, or carotid sinus massage can be safely and easily performed. All of these maneuvers increase vagal tone abruptly and result in termination of tachycardia either because of prolongation of AV nodal or slow pathway refractoriness (Lee et al. 2008a,b). Carotid sinus massage should not be performed bilaterally, simultaneously, and is better avoided in the elderly and contraindicated in those patients with carotid bruits.

The most commonly used drug for acute conversion of AVNRT in a stable patient is adenosine. Adenosine has a very short half-life and very quick onset of action. Usually, 6 mg is given intravenously followed by a fluid bolus, and the dose can be repeated or doubled. Adenosine produces block in the AV node and/or the slow pathway and terminates the tachycardia. The likelihood of termination is close to 100%, with those rare instances of failure to terminate often related to inadequate dosing or infiltration of the IV line. Although the arrhythmia always terminates, it may reinitiate, giving rise to the need for other pharmacological options to prevent recurrence (see below).

A small percentage of patients may develop atrial fibrillation (Blomstrom-Lundqvist et al. 2003). Common symptoms as a result of adenosine effect (not toxicity) following administration include chest pain, flushing, dizziness,
headache, or nausea. Symptoms, however, are short lived because of the very short drug half-life and go away in a matter of seconds.

To prevent recurrence, longer activating AV nodal blocking agents are used. Intravenous or oral beta blockers or calcium channel blockers are frequently utilized. In an acute setting, esmolol is preferred because of its very short half-life and can be quickly discontinued should hypotension or other side effects occur. Other frequently used beta blockers to manage patients with AVNRT are propranolol, metoprolol, and atenolol. Calcium channel blockers, including diltiazem and verapamil, are widely used. Hypotension may be seen with any of these agents, and thus when diagnosis of the arrhythmia is not clear (e.g., if the clinician is unsure whether ventricular tachycardia or AVNRT with aberrancy is occurring), longer acting antihypertensive agents, such as verapamil, are contraindicated. Hemodynamic collapse has been documented with both verapamil and beta blocker therapy, particularly in the elderly and patients with ventricular dysfunction. Although digoxin can be used and does affect AV nodal conduction, as soon as sympathetic tone is elevated, for example with ambulation, digoxin is generally ineffective. For patients with frequent recurring episodes, long-term therapy with daily AV nodal blocking agents is preferred. These agents primarily target the antegrade slow pathway. In patients without structural heart disease, another option is to use membrane active antiarrhythmic agents. Class IC drugs, such as flecainide and propafenone, have been demonstrated to reduce the frequency of AVNRT episodes. These agents depress retrograde fast pathway conduction. In some cases, class III agents such as amiodarone, sotalol, or dofetilide have also been effectively used. However, since these agents carry significant side effects including the risk of proarrhythmia, they are rarely employed. Radiofrequency ablation has fast emerged as a treatment of choice for patients with AVNRT who do not respond to the simplest therapeutic options.

**Invasive Electrophysiological Management of AVNRT**

Radiofrequency ablation has become the mainstay in the treatment of patients with symptomatic documented AVNRT. The groundbreaking demonstration by Jackman et al. (1992) that the slow pathway region could be safely and effectively ablated with high success rates (99%) without significant risk of AV block revolutionized the approach to this arrhythmia.

It should be noted that unlike many other ablations in electrophysiology (accessory pathway, atrial tachycardia, RV outflow tract tachycardia, etc.) ablation is not targeted to the site of earliest atrial activation in typical AVNRT. One would recollect that the earliest electrograms in typical AVNRT are near the fast pathway region very close to the compact AV node and should not be targeted for ablation. It is the antegrade limb of the circuit—the slow pathway—that is ablated. However, as stated above, the antegrade slow pathway cannot be mapped; thus, this is an example of anatomic ablation, that is, ablation at the usual anatomical occurrence location of a particular structure, and in this case the slow pathway.

The relative anatomy of the compact AV node and slow pathway should be completely understood to avoid inadvertent damage to the AV node. The compact AV node is a midseptal structure and is located in the atrium. Thus, keeping ablation energy delivery as posterior (low) as possible and ablating at relatively ventricular locations minimizes the risk of AV nodal damage. Since the slow pathway fibers climb up to the compact AV node on the interatrial septum from the CS and septal sub-Eustachian isthmus, a preferred approach is to ablate in a linear fashion at the level of the CS floor from the ventricle to the CS ostium at its
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During ablation of the slow pathway, a slow steady junctional rhythm, typically <80 beats per minute, will be noted. The operator or an assistant should constantly watch that the junctional rhythm always conducts retrograde to the atrium. If irregular junctional beats, fast junctional beats, or any junctional beat with block to the atrium occurs, energy delivery must be immediately terminated as inadvertent AV nodal or fast pathway damage may be occurring.

Some ablationists have directed energy delivery to sites of so-called slow pathway potentials.
be found at sites close to the AV node where ablation is not desirable given the risk of AV block. Conversely, ablation of some posteriorly located slow pathway potentials will not be enough. Linear ablation may be required to transect these fibers at safe but low sites (Hayashi et al. 2001; Nakagawa and Jackman 2007).

A similar anatomical approach can also be done using cryoablation. An important differ-

Figure 2.1.11 Location of the slow pathway in right anterior oblique (RAO) and left anterior oblique (LAO) view. In the corresponding fluoroscopic inserts, the ablation catheter positioned in the slow pathway region is marked with a yellow arrow. See text for details. (With permission from the Mayo Foundation for Medical Education and Research. All rights reserved.)

These are signals where in a far-field electrogram is succeeded by a sharp near-field electrogram. These are seen in sinus rhythm and sometimes will be reversed in tachycardia. It should be noted, however, that these signals are found at most sites anterior to the Eustachian ridge reflecting the complex interplay between myocardial fibers and transitional fibers to the AV node. Thus, a slow pathway potential may
ence appears to be that junctional rhythm is not consistently seen with cryoablation as is with radiofrequency energy delivery at appropriate locations.

Summary

Typical AV node reentry is the most common SVT and accounts for 50% of patients presenting with palpitation from SVT. Management should be individualized, taking into account the frequency and severity of episodes and coexisting heart disease. The electrocardiographic pattern is characteristic, and electrophysiology diagnosis has been well worked out with very high success rates both for diagnosis and ablative therapy. Medical management and catheter ablation can both be considered choices for first-line management of AVNRT that is symptomatic. Linear slow pathway ablation with thorough understanding of the anatomy of the AV node and circuit, along with careful online monitoring for junctional rhythm, allows the operator to carry out the procedure with low risk of complication while maintaining very high success rates. The ablation approach is primarily anatomical, given the anatomical discreteness of the slow and fast pathways as described above.

AVRT

AVRT is distinguished from AVNRT by the fact that both the atrium and ventricle are required for completion of the AVRT circuit. Two connections between the atria and ventricle must exist for AVRT to occur, one of which is an extra nodal accessory pathway.

An accessory pathway is a muscular connection between the atrial and ventricular myocardium capable of conducting electrical impulses in the antegrade, retrograde, or both directions. These connections are thought to be embryological remnants of the fibrous annular rings typically surrounding the mitral and tricuspid valves during fetal development (Jongbloed et al. 2008). Occasionally, there is a genetic predisposition for developing accessory pathways. Nonseptal muscular connections bridging the atrium and ventricle were first described by Dr. Stanley Kent in 1893, although he thought these to be part of the normal AV conduction system. In 1930, Drs. Wolff, Parkinson, and White described a group of patients with short PR interval (a bundle branch block-like pattern) and SVT. Later, Mahaim, Wood, and Ohnell each described clear evidence of extranodal muscular accessory connections completely independent of the AV nodal conduction access. By the early 1950s, the term pre-excitation (rather than bundle branch-like activation) was used to describe the phenomenon of early extranodal ventricular activation during sinus rhythm with conduction from the atrium to the ventricle (Scheinman 2005). Later, the important work of Drs. Durrer and Wellens (Wellens et al. 1971) using electrophysiological maneuvers confirmed the accessory pathway connection to atrial and ventricular myocardium and the requirement of both cardiac chambers to complete the reentry circuit.

Anatomy and Physiology

AVRT typically involves a macroreentrant circuit utilizing both atrium and ventricle, in addition to the accessory pathway and the AV node.

Terminology

- Pre-excitation: Early activation of ventricular myocardium via an accessory pathway, thus occurring earlier than activation through the AV node. A characteristic slurred upstroke or the QRS complex (delta wave) along with a short PR interval is seen.
- Orthodromic AVRT: When the macroreentrant circuit occurs such that conduction from the atrium to the ventricle is via the AV node and the normal conduction access but
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wide QRS ORT will occur if bundle branch block or a bystander additional accessory pathway is present (Fig. 2.1.12).

Antidromic tachycardia: When the macro-reentrant AVRT involves antegrade conduc-

Figure 2.1.12 Circuit of ORT and ART. The upper panel illustrates orthodromic tachycardia during which antegrade conduction occurs via the AV node and retrograde through an accessory pathway. During tachycardia, the QRS complex is regular and narrow unless bundle branch block is present. The bottom panel illustrates antidromic tachycardia. Antegrade conduction occurs via an accessory pathway with retrograde conduction through the AV node. During tachycardia, a regular wide QRS complex is observed. (With permission from the Mayo Foundation for Medical Education and Research. All rights reserved.)

retrograde conduction back to the atrium is through an accessory pathway, orthodromic AVRT is diagnosed. ORT typically results in a narrow complex tachycardia since the conduction system is being employed; however,
tion down the accessory pathway and retrograde conduction via the AV node back to the atrium to complete the circuit, antidromic tachycardia is diagnosed. Antidromic tachycardia always results in a regular wide complex tachycardia.

- **Pathway-to-pathway tachycardia**: In this arrhythmia, accessory pathways are used for both antegrade and retrograde conduction with the AV node not forming a part of the circuit. Because antegrade conduction is through an accessory pathway, pathway-to-pathway tachycardia also is always a regular wide complex tachycardia.

- **Pre-excited tachycardia**: When the tachycardia mechanism involves some other SVT (not AVRT) such as AVNRT, atrial tachycardia, sinus tachycardia, or atrial fibrillation but a wide QRS rhythm results because of antegrade conduction to the ventricle via an accessory pathway, then a pre-excited tachycardia is said to exist. In pre-excited tachycardias, unlike AVRT, if the pathway is ablated, tachycardia will still continue, except that a wide QRS morphology will no longer be seen.

- **Concealed pathways**: Accessory pathways which cannot be diagnosed from the 12-lead EKG are called concealed pathways. Most of these are a result of pathways that conduct only in the retrograde direction, and thus there is no early activation of the ventricle. Sometimes, antegrade conducting pathways may also be concealed if they conduct poorly or AV node conduction is very brisk or the pathway is located far from the site of origin of the atrial rhythm (e.g., left lateral pathways in sinus rhythm).

- **Wolff–Parkinson–White syndrome (WPW)**: WPW is present when there is evidence of pre-excitation on the 12-lead EKG along with documented atrioventricular arrhythmias. Pre-excited atrial fibrillation is a subtype of pre-excited tachycardias where atrial fibrillation can potentially give rise to a lethal arrhythmia such as ventricular fibrillation as a result of a regular and possibly rapid conduction through an accessory pathway to the ventricle. Pre-excited atrial fibrillation is considered an emergency (Fig. 2.1.13).

Accessory pathways typically occur along the AV valve rings because of the proximity of atrial and ventricular myocardium to each other. These pathways can exist anywhere along the valve continuity but typically involve free walls of the mitral and tricuspid valves. Because of the presence of the aortic valve separating atrial and ventricular myocardium in the region of the anteroseptal mitral annulus, pathways at these locations are rare but sometimes occur and appear to traverse the aortic valve (Suleiman et al. 2008) (Fig. 2.1.14).

There are important anatomical differences between the mitral annulus, the tricuspid annulus, and the common posteroseptal regions. The mitral valve annulus is a distinct structure with well-formed atrial and ventricular insertion sites. On the other hand, the tricuspid annulus shows significant overlap between atrial and ventricular myocardium. Finally, the posteroseptal region is technically an extracardiac, extraseptal structure where the cardiac veins may form the myocardial continuity between atrium and ventricle.

The nomenclature for septal pathways has led to some confusion. Original descriptions of pathway location were derived from viewing the heart in a single plane. Unfortunately, these descriptors do not have much anatomical credibility. For example, the anteroseptal region is a part of the true septum but may involve continuity via the aortic valve, whereas the postero-septal region is not part of the true septum but rather a continuation of the fibro-fatty AV groove. Thus, the term paraseptal rather than septal may be a better descriptor of pathways in this region. The most common site for accessory pathways to occur is on the left free wall (posterior) of the heart in relation to the mitral annulus. The second most frequent site is the
Figure 2.1.13  Pre-excited atrial fibrillation. The 12-lead ECG shows a rapid irregular wide QRS complex tachycardia characteristic of pre-excited atrial fibrillation. Note the irregularity of the QRS complexes in addition to the variation in QRS width caused by differing degrees of pre-excitation. See text for further details.

Figure 2.1.14  Locations of accessory pathways. Accessory pathways typically traverse the free walls of the tricuspid and mitral valves where atrial and ventricular myocardial tissues are in close proximity with one another. While rare, they can also traverse the aortomitral continuity. Other rare locations have been reported and are noted in the chapter text. (With permission from the Mayo Foundation for Medical Education and Research. All rights reserved.)
in inferior paraseptal region of both the right and left sides. Pathways on the right free wall and the anteroseptal region account for the majority of the remaining locations. Rare accessory pathways involve direct connections between the atrial appendage to the RV outflow tract, penetrate through the anterior fibrous trigone, and may involve diverticula or anomalies of the CS.

Certain pathways are so unique that a separate nomenclature has evolved for them.

- **Atriofascicular pathways (Mahaim fibers):** Although Mahaim initially described muscular accessory pathways, his name has become linked to an important subset of accessory pathways called atriofascicular fibers. In many ways, these structures are like AV node tissue located on the tricuspid annulus at the right side free wall. These structures exhibit decremental conduction, never exhibit retrograde conduction (always manifest), and often insert into the His bundle or infra-Hisian conduction system rather than directly to the ventricle. These pathways may be difficult to recognize as the decremental nature of the AV nodalike structure results in very subtle pre-excitation. Further, during tachycardia, a typical left bundle branch block pattern (by virtue of its insertion to the right bundle) will occur, often giving the impression of AVNRT with left bundle branch block.

- **Fasciculoventricular tracts:** These are not true accessory bypass tracts since they do not conduct from atrium to ventricle or vice versa. With the normal AV conduction system, the His bundle and proximal right bundle are insulated such that the earliest site of ventricular activation via the AV node conduction access is about two-thirds the distance to the apex at the site of right bundle breakthrough to the ventricle. In some patients, this insulation is incomplete with early breakthrough from the His or proximal right bundle directly to the ventricle. These are termed fasciculoventricular tracts. Importantly, there is no clinically relevant arrhythmia associated with this type of tract, and its presence results only as a benign mimic of pre-excitation.

### Clinical Diagnosis

The clinical features of AVRT are very similar to that observed with AVNRT. Tachycardia onset is sudden, and offset is sudden as well. Patients also sometimes described an urge to micturate following arrhythmia termination and have some residual fatigue. One difference from AVNRT is patients sometimes will recognize deterioration of their arrhythmia to an irregular “different” rhythm disturbance associated less with palpitation and more with fatigue. This is because of the more significant association of atrial fibrillation with AVRT compared with AVNRT.

### Electrocardiographic Diagnosis

The reader must be familiar with accessory pathway related electrocardiography in three contexts. With antegrade conducting pathways and manifest pre-excitation, the first precept is recognition that pathway activation is present. Once verified, the delta wave vector can provide clues as to pathway location. Second, during ORT, analysis of the P wave at times helps localize a retrograde only conducting pathway. Finally, recognition of pre-excited atrial fibrillation is critical.

The 12-lead EKG is characteristic of pre-excitation when the PR interval is short (<120 ms duration) and a delta wave or slurring of the initial deflection of the QRS complex is observed. The QRS duration itself is usually lengthened. The delta wave is a result of early activation of ventricular myocardium, which is not ideally suited for conduction and results in an abnormal slurred deflection when compared with the sharp reflection characteristic of
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the AV node, pre-excitation is obvious and marked. Conversely, if a pathway conducts poorly and the AV node conducts well, little or no pre-excitation despite the presence of an antegrade conducting pathway is noted (Fig. 2.1.15).

The presence of pre-excitation on an EKG does not necessarily imply that AVRT is possible. Some patients, for example, will have only antegrade conduction, and the circuit time or lengths are such that antidromic tachycardia does not occur in the patient’s lifetime. Similarly, a retrograde conducting pathway may be present at EP study, but occasionally, orthodromic AVRT cannot be induced and in those instances represents an incidental finding.

A number of algorithms have been developed to predict the location of an accessory pathway (Arruda et al. 1998).

Figure 2.1.15 A 12-lead EKG showing SR with pre-excitation and corresponding intracardiac electrograms. The left panel shows the 12 lead EKG. A delta wave is present. Note the initial slurring of the QRS complex as it leaves baseline, the short PR interval, and slightly widened QRS complex. The right panel shows the corresponding IEGMs. Pictured from top to bottom are surface leads II, V1, and V6 followed by EGMs recorded from the high right atrium (HRA), His bundle proximal (His 4) to distal (His 1), RV apex, and coronary sinus proximal (CS 19/20) to distal (CS 1/2). The broken line shows that the ventricular myocardium is activated preceding or near simultaneous to His bundle activation (HV interval = 0). The ventricular EGM recorded on the coronary sinus catheter is early and is bracketed (black arrow).
Electrophysiological Findings

If a PVC placed during tachycardia does pre-excite the atrium but with a different atrial activation sequence (PVC placed during His bundle refractoriness), this is also diagnostic of a retrograde conducting accessory pathway. However, the pathway is not responsible for the tachycardia and the atrial activation sequence seen during tachycardia. Similarly, if a PVC pre-excites the atrium when placed at a time of His bundle refractoriness and there is no change in the atrial activation sequence yet the tachycardia is not reset, once again, a pathway is present but does not participate in tachycardia, and a second mechanism of arrhythmia (AVNRT or atrial tachycardia, etc.) is the primary problem and should be targeted for therapy as well.

Electrophysiology dictum #5: A PVC placed at a time of His bundle refractoriness that pre-excites the atrium but either changes the atrial activation sequence or, when not changing the atrial activation sequence, fails to reset the tachycardia indicates that a bystander accessory pathway is present.

An equally significant finding of PVCs placed during tachycardia is the phenomenon of post-excitation. Here, the PVC again placed when the His bundle is refractory causes a delay without a change in the atrial activation sequence. This is diagnostic of ORT with a retrograde accessory pathway. Reset does not have to be demonstrated in this situation when delay is the observed phenomenon. As an extension of this phenomenon, either spontaneously or with PVCs, if there is termination of tachycardias without VA conduction and the PVC is placed at the time of His bundle refractoriness, ORT using a retrograde accessory pathway is diagnosed (Fig. 2.1.16).

Figure 2.1.16  PVC terminates tachycardia without return to the atrium. Pictured from top to bottom are surface leads II and V1 followed by EGMs recorded from the high right atrium (HRA), His bundle proximal (His 4) to distal (His 1), RV apex, and coronary sinus proximal (CS 17/18) to distal (CS 1/2). A PVC is inserted into tachycardia at a time when the His bundle is refractory (HH interval remains unchanged at 428 ms). Tachycardia terminates in the ventricle without return to the atrium. This is diagnostic of ORT.
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Dynamic Maneuvers in ART

The reader should be familiar with a peculiar phenomenon that occurs in ORT when aberrancy from bundle branch block occurs. When a left-sided accessory pathway is present and used during ORT, and a left bundle branch block appears, then there is a lengthening of the VA interval since now the circuit must exit through the right bundle, cross the septum, and then utilize the accessory pathway for retrograde conduction (Fig. 2.1.17). In the same patient, the appearance of right bundle branch block will not affect at the circuit length. Conversely, in a right free wall accessory pathway participating in ORT, if right bundle branch block were to occur, there would be a lengthening in the VA interval. It is important that the VA interval be measured from the earliest ventricular activation to the earliest atrial EGM. Lengthening of the VA interval does not necessarily translate into slowing of the tachycardia since there may be compensatory increased conduction velocities through the AV conduction axis as a result of the longer retrograde VA time.

Dynamic Maneuvers in ART

Just as PVCs placed during narrow complex tachycardia are extremely useful in defining the mechanism of arrhythmia, sensed premature atrial contractions (PACs) placed during wide complex tachycardia are revealing. The maneuver, however, is more complex, and details are beyond the scope of this work. In general, however, a PAC that is placed from the lateral right or left atrium that can pre-excite the ventricle without advancing the atrial electrgrams in the vicinity of the AV node diagnoses the presence of an antegrade conducting accessory pathway. With analogous reasoning to that
Adenosine also is highly effective and safe to use in both ORT and ART but is contraindicated as are any other AV nodal slowing agents in the presence of pre-excited atrial fibrillation. Thus, either a regular narrow or wide QRS tachycardia will terminate with an AV nodal blocking agent, but an irregular wide complex tachycardia, when recognized, should serve as a contraindication for any maneuver or agent to block the AV node.

Symptomatic patients with recurrent arrhythmias should be considered for radiofrequency ablation, but medical management is an option for minimally symptomatic individuals. Long-term AV nodal blocking agents or the use of membrane active drugs are effective, but particularly with membrane active drug use in older patients, the potential for proarrhythmia with coexisting structural heart disease becomes a concern.

Radiofrequency Ablation of Accessory Pathways

Catheter ablation is very effective in eliminating accessory pathway conduction. The success rate routinely reported is >90%. However, to achieve these success rates and to do so with minimal complications, a thorough knowledge of the methods to recognize an appropriate target for ablation is required. Further, not all pathways require the same approach, and some specific situations are complex, with the techniques required for success being quite different from others. Here we will outline the selection of targets for ablation, specific situations requiring different approaches, and the results that can be expected to be obtained with such approaches.

Selecting a Target for Accessory Pathway Ablation

In general, the primary methods used in practice to identify a target for pathway ablation...
are earliest ventricular activation during atrial pacing with antegrade pathways, earliest atrial sites during ventricular pacing or tachy- 
cardia with retrograde conducting pathways, sites of AV fusion, and recognizing the pathway potential.

- **Earliest ventricular activation:** Utilized with pathways that show manifest pre-excita-
tion on the 12-lead EKG or when pre-
excitation is present during EP study when 
pacing the atrium at more rapid rates. One 
approach that is frequently employed is to 
select the sites of earliest ventricular activa-
tion. Theoretically, this can be done during sinus rhythm, atrial pacing from any site in the 
atrion, or with an induced antidromic tachycardia. However, at times, only some approaches are practical. Antidromic tachycardia is rarely seen and not easy to induce. In sinus rhythm, there can be significant fusion with AV nodal conduction so that rec-
ognizing earlier ventricular sites is complex 
because of the contaminating AV node con-
duction. When pacing the atrium at rapid 
rates, it is not always certain that the AV 
node will block before the accessory pathway, 
and depending on the pacing sites, AV node 
conduction may be favored over that of 
pathways. Therefore, ideally, an atrial pacing 
site should be selected at its flow to the 
accessory pathway (which may have been 
proximately mapped with basic EP study 
and EKG analysis) at a rate where maximal 
pre-excitation is observed.

Once the pacing site and rates in the 
atrion have been determined, point-to-
point mapping using the ablation catheter 
all along the annulus is done. Because some 
pathways may exit further into the ventricle 
and exactly at the annulus, care should be 
taken to ascertain that there is no earlier ven-
tricular activation site in all directions (along 
the annulus and apex to base). Typically, at 
the true ventricular insertion site, any cath-
eter movement away from that location will 
result in later activation, and when com-
pared with the earliest deflection of the delta 
wave on the surface EKG, the local electro-
gram will be 30–40 ms earlier. If unipolar 
recordings are used at the true ventricular 
insertion site, a reversal of polarity will be 
noted. At the best site, a completely negative 
unipolar electrogram will be noted. If a posi-
tive deflection occurs earliest, then the oper-
ator is not at the true insertion site, which 
may be nearby on the endocardial surface or 
possibly epicardially located.

Mapping of earliest ventricular activation 
is a viable method to ablate pathways but 
has serious limitations including the inabil-
ity to produce maximal pre-excitation with 
atrial pacing and the fact that the best target 
is the pathway itself (on the annulus) rather 
than the ventricular insertion site. With any 
approach used, catheter contact is important 
to enable appropriate energy delivery at the 
targeted site. This can be difficult in the ven-
tricle as compared with in the atrium or on 
the annulus.

- **Earliest atrial activation:** With retrograde 
conducting pathways, mapping the site of 
earliest atrial activation during ventricular 
pacing or ORT is a frequent method used. 
With ventricular pacing, care should be used 
since fusion via the AV node commonly 
occurs, especially with septal or paraseptal 
pathways. It can be difficult to know whether 
a given early site is activated via the AV 
node or an accessory pathway, and inadvert-
ent ablation close to the conduction system 
may occur. This problem is much less likely 
when mapping is performed during ORT 
since conduction cannot be retrograde 
through the AV node. ORT mapping, 
however, can be problematic because of 
catheter instability from the rapid rates and 
the occasional case where more than one 
accessory pathway is present producing 
fusion. The preferred approach for catheter
stability is ventricular pacing at a stable rate and from a location close to the pathway so as to produce sole pathway conduction (no AV node conduction).

When this cannot be done, ORT can be induced and the site of earliest activation mapped with pacing the ventricle close to the rate of tachycardia performed simultaneously. This maneuver prevents a major change in catheter position during ablation when tachycardia breaks.

As described above with early ventricular site mapping, careful analysis of the local electrograms (including unipolar when available) and being sure in all directions that there is no earlier atrial activation site is important. Unlike mapping of the earliest ventricular signal where the delta wave can serve as a reference, mapping early atrial electrograms completely relies on being sure that all neighboring sites are activated late. Finally, the atrial insertion is only one end of the pathway, and the ideal targeted site is the pathway itself on the anulus.

* Fusion between atrial ventricular electrograms: Of all the methods available to define an appropriate site for ablation, fusion between atrial and ventricular electrograms is the least reliable and should never be the sole criteria for selection. The principle is that at the site of the accessory pathway, since ventricular activation will occur very soon after atrial activation (with antegrade conduction and vice versa when retrograde conduction is present), a site where both electrograms are fused represents the pathway sites. The major limitation with this approach is that even when a pathway is present at a very different location or there is no pathway at all, there may happen to be locations where the atrial and ventricular wavefront arrives simultaneously. For example, during sinus rhythm with AV nodal conduction, the sinus wavefront will reach the left atrium late. If AV node conduc-

tion is fairly quick and the left bundle is conducting well, ventricular activation may reach the annulus at a site where atrial activation is getting there at the same time. This will produce fusion when in fact there is no accessory pathway there at all. Some of the most egregious examples of mistakes with ablation are related to relying on AV fusion. Similarly, with ventricular pacing even when AV nodal conduction is present, there may be sites that the atrial wavefront reaches the annulus at the same times as the ventricular wavefront, creating the same confusion.

Once the operator understands this very important limitation with AV fusion match, then attempts to make a mistake less likely can be resorted to. For example, if ventricular pacing has been done from very close to the pathway site (or atrial pacing in the case of antegrade conduction) or when mapping during a reentrant tachycardia that uses that pathway, fusion of electrograms is a relatively more reliable finding. Even here, however, the issue of pathway slant should be considered.

* Pathway potential: Conduction across an accessory pathway produces a discrete signal different from the neighboring atrial and ventricular electrograms. The pathway potential is located on the anulus with most pathways and is the ideal target for ablation. Experienced operators often will have trouble recollecting an instance in their entire career where catheter contact at the site of a recognized pathway potential failed to permanently eliminate accessory pathway conduction. This potential, however, can be difficult to recognize, and care must be taken with appropriate maneuvers to be sure that a fragmented atrial or ventricular electrogram is not being mistaken for a pathway potential. Further, with epicardial pathway, the pathway potential will not be seen endocardially and require venous or epicardial mapping.
Technique of Ablation

Most pathways require positioning along the mitral or tricuspid valve annulus. Commonly used methods to access the mitral valve annulus include transseptal or retrograde aortic approaches. Various sheaths with curve will help positioning preferentially along the anterior, lateral, or posterior annulus and are often used. The tricuspid valve is usually approached from femoral access and the inferior vena cava. However, the superior approach from a subclavian or internal jugular access can sometimes be preferred for catheter stability.

Once catheter movement is stable, mapping is based on the principles outlined above. A common oversight with mapping is the failure to distinguish accurately between AV pathway (and in some instances CS) signals seen on the mapping catheter. For a concealed pathway (retrograde only conduction), a simple maneuver is to insert an early coupled PVC, which results in retrograde pathway block. This allows one to immediately distinguish which part of a complex signal arises from the ventricle and which is atrial. When VA block occurs, any signal that disappears is atrial. Similarly, when pathway block occurs, the atrial electrogram will be seen to occur later depending on retrograde AV nodal conduction. In a similar manner, early coupled atrial premature beats that result in antegrade block through the pathway or complete AV block can be used to discriminate the early or fragmented portion of the ventricular electrogram. A combination of these maneuvers will help identify the pathway potentials since on the annulus, any electrogram that can be dissociated from the ventricle (with atrial pacing and from the atrium with ventricular pacing) is the pathway potential (Nakagawa and Jackman 2007).

Electrophysiology dictum #8: The ideal ablation for accessory pathway involves stable catheter contact on the annulus targeting the accessory pathway potential.

Although annular ablation is ideal, there are some exceptions. One example occurs when a retrograde aortic approach is utilized. The catheter can, in a stable fashion, be tucked under the valve leaflet. Here, a very small atrial electrogram will be seen since the catheter is relatively ventricular. Nevertheless, because of excellent contact and the fact that the tip lays on the annulus under the valve, ablation can be successful.

Another common error in pathway mapping involves the failure to appreciate pathway slant. Many pathways cross the valve annulus in an oblique fashion, particularly those at free wall locations. Thus, targeting the site of the shortest AV electrogram intervals will not work. An approach to overcome the limitation imposed by pathway slant involves pacing on either side of the pathway to reverse the direction of wavefront activation (Gonzalez and Rivera 2006). This is accomplished by pacing at a fixed cycle length on either side of the atrial insertion (with antegrade conduction) or ventricular insertion (retrograde conduction) and observing for changes in the AV or VA interval. Generally, when a very short interelectrogram interval is noted, the pacing wavefront is parallel to the direction of the slant. On the other hand, by reversing the wavefront direction at the same pacing cycle length, the atrial and ventricular electrograms will be separated and the pathway potentials will be visualized easier. The reader is referred to two excellent articles that illustrate this principle in detail (Gonzalez and Rivera 2006; Nakagawa and Jackman 2007).

Unique Accessory Pathway Locations

- **Free wall tricuspid annulus**: Ablation on the tricuspid annulus off the septum can be problematic because of catheter contact. This difficulty is exaggerated in patients with Ebstein’s anomaly because of the ambiguity of where the annulus is located. Further, unlike the mitral annulus where the CS
forms a stable surrogate for annular activation, there is no equivalent with the tricuspid annulus.

To overcome these difficulties, placing a multielectrode catheter either on the tricuspid annulus or in the right coronary artery can be considered. The operator should be aware of the various options to improve stability on the annulus, including the use of specific sheaths, a superior approach to tuck the catheter under the tricuspid valve, and magnetic navigation.

- **Septal pathways**: An issue that arises when ablating on the septum for accessory pathways is conduction tissue damage. Anterior septal pathways can often be very close to the bundle of His. The His bundle itself is rarely damaged during ablation because of the fibrous extensions and encased sheath structure. The danger site is the compact AV node, which is located in a midseptal and slightly atrial location. Thus, when ablating on the septum, making sure that ablation energy is in a slightly ventricular location and not on the midseptal location is ideal. When midseptal ablation or annular/atrial locations are a must for energy delivery, cryoablation can be considered (Kimman et al. 2003). Perhaps at no other location should a definite effort be made to identify the pathway potential and not rely on inexact methods of mapping to avoid the undesirable scenario of AV conduction injury without pathway success.

Cryomapping and ablation is an option that allows cooling of the tissue at the site without causing permanent damage. If, during test mapping (cooling to $-35^\circ C$), no untoward effects on conduction are observed but pathway block is seen, then cooling further to $-75^\circ C$ can result in permanent ablation of the pathway. Occasionally, difficult to ablate anterior septal pathways actually represent activation using the musculature above the aortic valve, and ablation from one of the coronary cusps can be successful (Asirvatham 2007a,b; Balasundaram et al. 2008; Suleiman et al. 2008).

- **Ablation of Mahaim (atriofascicular) fibers**: The reader will recall that this type of pathway found on the right side free wall has AV nodelike properties, conducts in an antegrade fashion only, and the ventricular insertion site is related to the right bundle exit. Thus, ventricular mapping for the earliest site during atrial pacing is of absolutely no value since it is exactly the same site that is activated early with right bundle activation. Since these pathways do not conduct retrograde (by definition), mapping during ventricular pacing is ineffective. Thus, the only target that can be used is identification of the pathway potentials.

- **Venous pathways**: In some cases, the myocardium of the vein branches of the CS give rise to accessory pathways. The most common occurrence for this finding is middle cardiac vein pathways. The myocardium of the vein is similar to atrial myocardium as it sheaths the CS and some of its branches in all normal hearts. While in normal hearts the connection of these fibers with the atrium is universal, occasional patients will show connection of these fibers to the ventricular myocardium as well. When this happens, a type of epicardial pathway with the AV conduction using the venous musculature occurs. The venous musculature itself will be targeted for ablation. This, however, can be difficult because of the neighboring coronary arteries. Using cryoablation or intravascular ultrasound to monitor the arteries during ablation can result in success without arterial damage.

**Summary**

Accessory pathway ablation can be among the most satisfying in terms of results for both the electrophysiology personnel and the patients. However, a careful understanding of what constitutes a target for ablation and the technical
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know-how of methods to obtain optimal catheter stability is required to obtain these expected excellent results.

ATRIAL TACHYCARDIA

Atrial tachycardia is an inclusive term for a variety of arrhythmias and involves the mechanisms grouped together based on the requirement of atrial myocardium and the lack of requirement of the AV node or ventricle for arrhythmogenesis.

Macroeentrant atrial tachycardias or atrial flutters and atrial tachycardias exhibiting fibrillatory conduction through the atrial myocardium are discussed in detail elsewhere. Here we review automatic atrial tachycardia in the context of SVT diagnosis and management.

Automatic atrial tachycardia is less common than AVNRT (Chen et al. 1998). The arrhythmia occurs in all age groups. The automatic and more persistent form is seen more frequently in children. The true incidence of automatic atrial tachycardia may be underappreciated. It has been increasingly recognized that atrial fibrillation, as well as some forms of atrial flutter, have as their initiating and sometimes maintaining arrhythmia an automatic tachycardia often in one of the thoracic veins. For the purpose of our discussion, we will limit ourselves to automatic atrial tachycardia as a stand-alone arrhythmia.

Anatomy and Physiology

The primary mechanism of focal atrial tachycardia is abnormal automaticity, although triggered automaticity in microreentry has also been described (Lee 2006; Roberts-Thomson et al. 2006a,b). For those patients who seek medical attention, the characteristic warm up rate after a relatively sudden onset is consistent with enhanced automaticity as the underlying mechanism.

Most patients do not have an anatomical or congenital anomaly in the atrium to explain the reason for the tachycardia. However, certain anatomical sites are far more common in housing the substrate generating the abnormal automaticity and tachycardia origin.

In the right atrium, the crista terminalis (Roberts-Thomson et al. 2006a,b), superior vena cava (SVC), ostium of the CS, and Bachmann’s bundle region are relatively frequent sites of origin. In the left atrium, the pulmonary veins, left atrial appendage, and mitral annulus appear to be sites with a proclivity for atrial tachycardia. Recently, atrial myocardium related to or within the aortic valve cusps has been reported (Gami et al. 2008) (Fig. 2.1.18).

Clinical Presentation

Unlike AVNRT and AVRT, patients sometimes will report a relatively gradual onset of symptoms with atrial tachycardia. Often, a history of exercise, abrupt change in position, swallowing cold fluids, and so on will be obtained when questioning the patient regarding the onset of palpitations. Very commonly, patients will report varying rates, but the palpitations will remain regular (compare with atrial fibrillation with varying rates but irregular). Syncope and presyncope are rare except when the patients are dehydrated or have coexisting structural heart disease. Most patients who present for management of atrial tachycardia have structurally normal hearts; however, a subset of patients—usually young—will have persistent slower variants of atrial tachycardia that may give rise to a tachycardia-related cardiomyopathy and resultant heart failure.

Electrocardiographic Diagnosis

The hallmark of atrial tachycardia is P wave morphology distinct from sinus rhythm noted on 12-lead electrocardiography, rhythm strips, or Holter monitor tracings. A careful analysis of P wave morphology will not only help identify
the site of tachycardia origin but sometimes also help distinguish atrial tachycardia from AVNRT and AVRT. Tang et al. (1995) reviewed P wave morphology during atrial tachycardia in 31 patients. A positive P wave in lead V1 predicted a left atrial focus of 93% sensitivity and 88% specificity. A positive P wave in lead AVL indicated a right atrial focus with sensitivity of 88% and specificity of 79%. A notable exception to this rule was found with atrial tachycardias originating from the supraventricular vein because of the superior exit with such arrhythmia (Tang et al. 1995). The P waves in septal tachycardias are typically narrow because of near simultaneous depolarization of the left and right atrium. A wider P wave, however, can occur not only from the free walls of the atrium but also from the posteroseptal atrium since this structure lies outside the true atrium and is part of the perimeter space.

In the right atrium, the most common site of automatic atrial tachycardia is the crista terminalis. Because the sinus node is located close to the superior end of the structure, the differences between tachycardia P waves and sinus rhythm can be subtle, and these patients are sometimes misdiagnosed as having inappropriate sinus tachycardia. Conversely, during inappropriate sinus tachycardia, the sinus exit site may exhibit morphology changes at different rates. This gives the impression of atrial tachycardia based on P wave change when the diagnosis actually is inappropriate sinus tachycardia.

**Electrophysiology Diagnosis and Ablation**

During electrophysiology study, atrial tachycardia is usually readily diagnosed by an activation pattern that is earlier at locations away from the annulus (high to low activation) differing from accessory pathways and the AV
node-related pathways that exit close to the annulus (low to high activation).

The primary difficulty arises when atrial tachycardia with low to high activation needs to be excluded since the focus may be on the annulus similar to AVRT and AVNRT. Atrial tachycardia often can be demonstrated to be dissociated from the ventricle and AV conduction system. While this excludes AVRT, AVNRT remains a possibility and can be difficult to distinguish from atrial tachycardia.

PVCs placed during tachycardia as described for AVNRT and AVRT diagnosis can also help exclude atrial tachycardia. When PVCs that are placed during tachycardia are demonstrated to advance the retrograde His by >50 ms but do not have effect on the atrium, then the diagnosis of atrial tachycardia is highly likely.

Another valuable maneuver to ascertain diagnosis is that of ventricular pacing during atrial tachycardia. This maneuver validated by Knight et al. (2000) requires pacing of the ventricle at a rate slightly faster than the tachycardia cycle length (approximately 20 ms). During pacing, the atrium should be “entrained” (the atrial cycle length should be the same as the ventricular paced cycle length), and importantly, tachycardia should not terminate. Observation of the atrial activation during the period of ventricular pacing is important since the activation sequence clearly changes when VA connections are not operative in arrhythmia mechanism. After the sequence has been analyzed, the response when pacing stops is examined. Upon cessation of pacing, if a VAAV sequence of electrograms is observed, this strongly suggests atrial tachycardia. The explanation for this response is as follows: the last ventricular paced beat conducts through the AV node to the atrium. There is no way for the VA conduction to result in a subsequent ventricular activation (no pathway or dual AV nodal physiology). The atrial tachycardia focus beats again, which then conducts to the ventricle giving rise to the VAAV response. With AVNRT and AVRT, a VAV response occurs since there is an alternative route other than that which was used for retrograde activation of the atrium to return back to the ventricle (Knight et al. 2000) (Fig. 2.1.19).

Another simple observation that strongly helps differentiate atrial tachycardia from AVNRT or AVRT is to look and see how arrhythmia terminates. If, with tachycardia, termination is with an atrial electrogram (P wave) and this finding is consistently observed, atrial tachycardia can virtually be excluded. This is because with atrial tachycardia when the automatic arrhythmia stops, the last beat would be expected to conduct down to the ventricle. Only the serendipitous simultaneous occurrence of cessation of an atrial tachycardia focus with AV block could possibly result in this observation.

Pharmacological management of atrial tachycardia is typically unrewarding. Success rates with class IC agents such as flecainide or propafenone are in the 40–50% range. Although amiodarone may be more effective, its long-term risks of organ toxicity greatly limit its use.

Catheter ablation for focal atrial tachycardia is generally a straightforward procedure with either multielectrode or three-dimensional mapping focusing on the site of earliest atrial activation. The most common strategy involves comparison of intracardiac sites of interest to the onset of the surface P wave during tachycardia. The typical target sites precede the surface P wave by at least 25 ms. The placement of catheters with multiple poles (e.g., closely spaced 20 pole catheters on the crista terminalis) can assist the point-to-point mapping procedure. Another complimentary strategy involves pacing from the site of interest at the tachycardia cycle length. If the P wave morphology is identical to that seen during tachycardia, the origin site is probably close to where pacing occurs.

Three-dimensional mapping tools serve as an “aid to memory,” clearly defining with color coding the site of earliest activation
Chapter 2.1 AVNRT, AVRT, and Atrial Tachycardia

Focal atrial tachycardia is not as common as AVNRT or AVRT, but it is an increasingly recognized arrhythmia, particularly when it coexists either as a cause or a concomitant arrhythmia with atrial fibrillation and typical atrial flutter.

Suggested Algorithm for Approach to SVT

See Figure 2.1.20 for a suggested algorithm for approach to SVT.

Possible pitfalls with mapping and ablation of atrial tachycardia include failure to consider the possibility of an epicardial focus, a thoracic vein focus, or multiple sites of atrial tachycardia arising while mapping.

Typical radiofrequency ablation involves the delivery of 25–50 W of energy to the earliest defined site of atrial activation. Acceleration of tachycardia prior to termination is often seen typically occurring 3–10 ms after energy delivery is initiated. With crista terminalis or SVC tachycardia, care should be taken to avoid phrenic nerve damage (Dib et al. 2008). Cryoablation can be useful in these circumstances as well as when tachycardia focus is near the conduction system.

Summary

While focal atrial tachycardia is not as common as AVNRT or AVRT, it is an increasingly recognized arrhythmia, particularly when it coexists either as cause or a concomitant arrhythmia with atrial fibrillation and typical atrial flutter.

Suggested Algorithm for Approach to SVT

See Figure 2.1.20 for a suggested algorithm for approach to SVT.
Cardiac Arrhythmia Management

Figure 2.1.20 SVT algorithm.

<table>
<thead>
<tr>
<th>Approach to SVT</th>
<th>AVNRT</th>
<th>AVRT</th>
<th>ATACH</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clinical features: sudden onset, sudden offset tachypalpitations</td>
<td></td>
<td></td>
<td>Features: gradual onset or offset palpitations, fatigue or cardiomyopathy</td>
</tr>
<tr>
<td>AVNRT</td>
<td>AVRT</td>
<td>ATACH</td>
<td></td>
</tr>
<tr>
<td>EKG</td>
<td>Regular narrow complex tachycardia usually short R-P interval</td>
<td>Regular narrow complex tachycardia in ORT, R-P interval never &lt;80 ms</td>
<td>Narrow complex tachycardia typically with long R-P interval</td>
</tr>
<tr>
<td>Pseudo r’ in lead V1</td>
<td>Pseudoexcited atrial fibrillation</td>
<td>EKG</td>
<td></td>
</tr>
<tr>
<td>EP findings</td>
<td>V-A interval may be &lt;60-70 ms</td>
<td>Regular wide complex tachycardia in ART</td>
<td>EKG</td>
</tr>
<tr>
<td>Dual-AV node physiology</td>
<td>Para-Hisian pacing excludes accessory pathway</td>
<td>Preexcited atrial fibrillation</td>
<td></td>
</tr>
<tr>
<td>Early atrial activation in region of fast or slow pathway</td>
<td>EP findings</td>
<td></td>
<td></td>
</tr>
<tr>
<td>EP maneuvers during tachycardia</td>
<td>Short H-V interval becoming negative with decremental atrial pacing</td>
<td>No evidence of accessory pathway</td>
<td></td>
</tr>
<tr>
<td>Sensed PVCs during tachycardia preexcites the atrium only by preexciting the retrograde His without a change in activation sequence and resets the tachycardia</td>
<td>Para-Hisian pacing demonstrates retrograde activation when present</td>
<td>Activation sequence during tachycardia distinct from activation sequence with ventriculoatrial conduction</td>
<td></td>
</tr>
<tr>
<td>V-A-V or V-A-H-V response seen with the Morady maneuver</td>
<td>Usually no evidence of decrement with rapid pacing</td>
<td>EP maneuvers during tachycardia</td>
<td></td>
</tr>
<tr>
<td>Ablation</td>
<td>PVCs placed during tachycardia preexcites the atrium only by preexciting the retrograde His without a change in activation sequence and resets the tachycardia</td>
<td>PVCs do not preexcite the A with a similar activation sequence</td>
<td></td>
</tr>
<tr>
<td>Anatomic ablation in the region of the slow pathway</td>
<td>PVCs placed at a time of His bundle refractoriness may delay the next A without a change in activation sequence or terminates the tachycardia without reaching the atrium</td>
<td>V-A-A-V sequence with the Morady maneuver</td>
<td></td>
</tr>
<tr>
<td>Slow junctional rhythm with intact retrograde conduction predicts positive outcome</td>
<td>V-A-V response with Morady maneuver</td>
<td>Ablation</td>
<td></td>
</tr>
<tr>
<td>Site of pathway potential localization</td>
<td>Earliest site of activation in the atrium during tachycardia</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Case 2.1.1

A 65-year-old male was seen in consultation for evaluation of documented tachycardia. The patient reported a 3-year history of rapid palpitations. He described episodes as sudden in onset. He was frequently able to resolve these episodes with Valsalva maneuver. While performing physical labor, he experienced significant palpitations that did not readily terminate and resulted in slight chest pressure. This prompted a visit to the local emergency room. His past medical history included the diagnoses of coronary calcification, hypertension, hyperlipidemia, and diabetes. A recent treadmill stress echo was negative for ischemic changes and demonstrated preserved left ventricular function. Laboratory studies were unremarkable. Current medications included daily use of a beta blocker.

The 12-lead EKG obtained in the emergency room is shown in Figure 2.1.21.

What treatment options are available for this patient?

Case Discussion

The 12-lead EKG reveals a regular narrow complex tachycardia with a rate of 165 bpm. The RP interval
is short (<80 ms), excluding an accessory pathway. A small positive deflection is observed at the end of the QRS complex in lead V1 (pseudo-R'). The EKG is most consistent with typical AVNRT. In the emergency room, 6 mg of IV adenosine was administered, which terminated tachycardia.

The treatment options discussed with the patient included increasing beta blocker dose, use of an antiarrhythmic agent, or catheter ablation therapy. Antiarrhythmic agents, particularly IC agents such as flecainide, are contraindicated in patients with known coronary disease. Flecainide would be effective in this patient; however, in this case, it may be difficult to use as the patient grows older and his risk for coronary disease becomes more significant. During past episodes of tachycardia, an increased dose of beta blockers resulted in fatigue, making this option undesirable as well.

Catheter ablation is considered a class I, level B indication for the treatment of recurrent symptomatic AVNRT. The patient underwent electrophysiological study. Dual AV node physiology was observed. With programmed atrial stimulation, typical AV node reentry tachycardia was induced (Fig. 2.1.22). Introduction of PVCs during tachycardia pre-excited the retrograde His and the retrograde A without changing the activation sequence and reset the tachycardia.

Slow pathway ablation was performed. During ablation, junctional beats with retrograde conduction to the atrium were noted. Following ablation, repeat electrophysiology study was performed. AVNRT was unable to be induced. Recovery occurred without incident. The patient has been arrhythmia free in follow-up visits.
Case 2.1.2

An otherwise healthy active 12-year-old female was referred for the evaluation of palpitations. She described episodes of rapid heart rate occurring over a period of a year with exertion and at rest. An episode lasting 2 hours in duration resulted in a visit to a local emergency room. A regular narrow complex tachycardia with a rate of 225 bpm was documented. Following doses of 6, 6, and 12 mg of adenosine, tachycardia terminated with resumption of sinus rhythm. A 12-lead EKG obtained during sinus rhythm is shown in Figure 2.1.23.

What is the likely diagnosis?

Case Discussion

The 12-lead EKG reveals normal sinus rhythm with evidence of pre-excitation. The PR interval is short and a delta wave or slurring of the upstroke of the QRS complex is clearly visible. The QRS duration is slightly widened. Given this finding, the diagnosis most likely accounting for the documented tachycardia is AVRT. Rhythm strips obtained during tachycardia also demonstrated P waves in the ST segment. Orthodromic AVRT presents as a regular narrow complex tachycardia unless preexisting bundle branch block or bundle branch aberrancy exists. The P wave can be observed in the T wave because retrograde atrial activation follows ventricular depolarization.

The patient was counseled regarding treatment options including medical therapy and catheter ablation. The patient and family opted to proceed with electrophysiology study.

Orthodromic AVRT was induced with a right anterolateral accessory pathway participating as the retrograde limb of the circuit. PVCs introduced into tachycardia at a time when the His bundle was refractory resulted in retrograde atrial pre-excitation without change in the eccentric sequence of retrograde atrial activation and reset tachycardia.
Figure 2.1.23  A 12-lead EKG showing normal sinus rhythm with evidence of pre-excitation. A delta wave (red arrow) is clearly visible in multiple leads indicating the presence of an accessory pathway. Using the algorithm developed by Arruda et al. (1998), examination of the initial delta wave polarity clearly excludes a left-sided pathway (delta wave positive in lead I, R < S in lead V1).

Figure 2.1.24  Intracardiac electrograms obtained during ORT. Right bundle branch block resolves and the QRS configuration is normal. The VA interval is longer during right bundle branch block confirming the participation of a right-sided pathway in tachycardia. (Black arrows denote onset of the V as measured in the QRS complex. Red arrows mark onset of A as measured on the His bundle EGM).

addition, with the development of right bundle branch block ipsilateral to the pathway, a 30-ms prolongation of the VA interval was observed (Fig. 2.1.24).

Successful radiofrequency catheter ablation was performed at the right anterolateral tricuspid valve annulus using a long sheath to stabilize catheter position.
Case 2.1.3
A 72-year-old male is seen by his family physician for his annual wellness exam. His history is significant for hyperlipidemia, benign prostatic hypertrophy (BPH), and degenerative joint disease. A routine 12-lead EKG is obtained (Fig. 2.1.25). The EKG shows a narrow complex tachycardia with a ventricular rate of 110 bpm. The patient is entirely asymptomatic. A 24-hour monitor is ordered and the patient is referred for electrophysiology consultation. The monitor displays frequent runs of SVT (the longest being 2,962 beats in duration), with a maximum heart rate of 133 bpm. Medical record review during consultation reveals a history of ablation for a concealed left lateral accessory pathway 10 years earlier. An echocardiogram and stress test are obtained. The echocardiogram demonstrates abnormal left ventricular systolic function with an estimated ejection fraction of 40% and global hypokinesis. The stress test does not show evidence of ischemic heart disease.

What differential diagnoses exist? What management options are available? Can tachycardia affect left ventricular function?

Case Discussion
The 12-lead EKG shows a regular narrow complex tachycardia. P waves are present and precede each QRS. The P wave morphology is somewhat difficult to discern. The RP interval is long (RP > PR). The differential diagnosis for a long RP tachycardia includes atrial tachycardia, atypical AVNRT, and paroxysmal junctional reciprocating tachycardia (PJRT).

The test results are consistent with a diagnosis of tachycardia-induced cardiomyopathy. This phenomenon has been well reported in the literature. Treatment options are discussed with the patient. An angiotensin-converting enzyme (ACE) inhibitor is initiated due to depressed LV function. Proposed strategies to manage the tachycardia included trial of antiarrhythmic agents such as sotalol or catheter ablation therapy. Given his prior positive experience, the patient opts for ablation therapy.

Electrophysiology study is carried out. Baseline study discloses no evidence of the accessory pathway that had been ablated in the past. Episodes of a narrow complex tachycardia are induced with paired atrial premature contractions (APCs) and occur spontaneously. Findings include VA dissociation during ventricular pacing without termination of tachycardia (Fig. 2.1.26). A VAAV response is observed following cessation of ventricular pacing, which entrains the atrium. During entrainment, the atrial activation differs from that of tachycardia, providing affirmation to the diagnosis of atrial tachycardia.

Three-dimensional mapping is carried out and an area of early activation identified adjacent to the tricuspid valve annulus (Fig. 2.1.27). Local electrogram precedes the surface P wave by 25 ms. Application of radiofrequency energy results in tachycardia acceleration.

Following ablation and a 60-minute waiting period, repeat electrophysiological testing both on and off Isuprel infusion fails to induce tachycardia.

Figure 2.1.25 A 12-lead EKG showing narrow complex tachycardia with a long RP interval.
Intracardiac electrograms obtained during tachycardia. VA dissociation is observed when pacing the ventricle at a rate faster than tachycardia. Tachycardia does not terminate but continues at the same rate. Poles 1–10 on the CS catheter lie in the coronary sinus with 9/10 at the CS ostium. A 60 mm space exists between pole 10 and pole 11. Poles 11–20 lie in the posterior lateral right atrium along the tricuspid annulus. Early atrial activation is observed on CS poles 11/12.

A three-dimensional activation map of the right atrium showing in the RAO view is displayed. The site of early activation is denoted in red.
REFERENCES


RESOURCES

INTRODUCTION

Atrial fibrillation (AF) is a challenging arrhythmia affecting more than 3.5 million people in the Untied States alone. Besides its significant impact on patient well-being and the decreased quality of life associated with it, AF has a significant economical impact on the patient and on the health care system. This chapter will review AF incidence, risks, and medical management in general but will concentrate on the emerging and latest trends in catheter-based AF therapies. Since this chapter is written for the allied health professional, it will address many of the issues dealing with managing those patients during the pre-, intra- and postprocedural periods, with emphasis on reducing potential complications and improving outcomes. We will also touch on the classification and management of atrial flutter.

AF EPIDEMIOLOGY AND PREVALENCE

AF is the most common cardiac arrhythmia seen in clinical practice. It is estimated that AF affects more than 3.5 millions in the United States, and it is projected that this number may increase to 5 million by the year 2050. The reason for the increased prevalence of AF is not yet completely understood; however, it is mainly attributed to aging population with higher prevalence of cardiovascular diseases such as hypertension. AF affects 10% of individuals who are 80 years old or older (Braunwald 1997; U.S. Census Bureau 2000).
Atrial fibrillation has, over the years, been classified to distinguish between the three types of AF:

1. Paroxysmal AF, defined as recurrent (>2 episodes), terminating spontaneously within 7 days
2. Persistent AF, defined as AF that is sustained beyond 7 days, or lasting less than 7 days but necessitating pharmacological cardioversion (PCV) or electrical cardioversion (ECV)
3. Permanent AF (often referred to as “chronic AF”); this classification has been used for AF that is resistant to attempts at cardioversion

In 2007, the Heart Rhythm Society (HRS) has recommended that the classification of AF be modified to exclude the term “permanent AF.” The HRS Consensus Statement (Calkins et al. 2007), recommended that AF be classified into the two classifications mentioned above as paroxysmal and persistent, and to modify what was previously classified as permanent to be now labelled as “long-standing persistent AF,” which is now defined as continuous AF of greater than 1-year duration.

### The Cost of AF

Although AF is generally not life threatening, it is associated with several health risks. These risks include thromboembolic strokes, heart failure, and most of all, decline in the quality of life due to the symptoms associated with AF (Wolf et al. 1991). Since AF is the most common arrhythmia among hospitalized patients in the United States admitted with a primary diagnosis of arrhythmia, the economic consequences are considerable (Weintraub and Krumholz 2004). Although the cost of AF associated with missed days of work, and decreased productivity and quality of life cannot be quantified, it is estimated that the cost of AF related to cardiovascular accidents (CVAs) alone in the United States is around $12 billion (Rosamond et al. 2007).

### Etiology of AF

AF can occur in patients with or without heart disease, and is associated with advancing age. It is believed that there are many etiologies for this arrhythmia (Table 2.2.1). AF is associated with many underlying heart conditions that can

<table>
<thead>
<tr>
<th>Table 2.2.1 Most common etiology of atrial fibrillation.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Conditions which cases changes in the atrial myocardium</td>
</tr>
<tr>
<td>Coronary artery disease</td>
</tr>
<tr>
<td>Ventricular hypertrophy due to valvular diseases</td>
</tr>
<tr>
<td>Intracardiac thrombus or tumor</td>
</tr>
<tr>
<td>Pulmonary embolism and pulmonary hypertension</td>
</tr>
<tr>
<td>Cardiomyopathy and heart failure</td>
</tr>
<tr>
<td>Inflammatory conditions</td>
</tr>
<tr>
<td>Myocarditis</td>
</tr>
<tr>
<td>Pericarditis</td>
</tr>
<tr>
<td>Amyloidosis</td>
</tr>
<tr>
<td>Systemic lupus</td>
</tr>
<tr>
<td>Conditions that increase sympathetic nervous system activity</td>
</tr>
<tr>
<td>Hyperthyroidism</td>
</tr>
<tr>
<td>Caffeine and alcohol consumption</td>
</tr>
<tr>
<td>Medications (e.g., epinephrine)</td>
</tr>
<tr>
<td>States of anxiety and agitation</td>
</tr>
<tr>
<td>Intense exercises</td>
</tr>
<tr>
<td>Conditions that increase parasympathetic nervous system activity</td>
</tr>
<tr>
<td>Sleep</td>
</tr>
<tr>
<td>Full stomach after a meal</td>
</tr>
<tr>
<td>Congenital heart conditions</td>
</tr>
<tr>
<td>Patent ductus arteriosis</td>
</tr>
<tr>
<td>Atrial septal defect</td>
</tr>
<tr>
<td>After corrective surgery for tetralogy of Fallot or transposition of great vessels</td>
</tr>
<tr>
<td>Genetic factors as in lone AF</td>
</tr>
</tbody>
</table>
lead to changes of the atrial myocardium. These changes can include fibrosis, hypertrophy, and ischemia, among others. In addition, atrial fibrosis can increase the prevalence of AF and seems to increase with age. Other factors contributing to AF may also include pulmonary diseases, thyroid disease, systemic infections, and environmental or chemical toxins.

**MEDICAL MANAGEMENT OF AF**

The goals in managing AF include eliminating or reducing the risk of stroke with anticoagulation while maintaining sinus rhythm or rate control to reduce the risk of heart failure.

**Anticoagulation**

The strong association between AF and stroke has been known for many decades. Due to the incomplete emptying of the left atrium (LA) during AF, pooling or stasis of the blood occurs, leading to formation of left atrial thrombi. The Framingham Study showed that patients with nonvalvular AF have up to 5% risk of stroke per year, which is significantly higher than the general population (Wolf et al. 1978). This risk can increase to 17% in patients with underlying valvular or structural heart disease. Many other studies have been published since then, emphasizing the importance of anticoagulation. In 2001, the American College of Cardiology (ACC)/American Heart Association (AHA) in conjunction with the HRS published guidelines for the management of AF and made specific recommendations for anticoagulation (Albers et al. 2001). The risk of ischemic stroke with AF varies widely depending on the patient individual risk factors (Tables 2.2.3 and 2.2.4).

**Table 2.2.2** AF symptoms.

| Palpitations | Shortness of breath with exertion | Inability to exercise | Dizziness | Fainting or near fainting | Vague sensation in the chest or throat | Excessive urination | Anxiety | Tingling in one arm or hand | Chest pressure | Excessive sweating | Cold hands and/or feet |

**Warfarin or Aspirin or Other?**

Several studies have confirmed the superiority of warfarin in the primary and secondary prevention of embolic strokes in AF (Hart et al. 1999). The anticoagulation guidelines using the CHADS$_2$VASc score are recommended (Table 2.2.5).

In those patients with AF who are considered at low risk of developing embolic events, the decision whether to use aspirin (ASA) versus warfarin, should be made with the patient after he or she has fully understood the risks and benefits of each option. Low risk of
Cardiac Arrhythmia Management

Table 2.2.3  Risk factors for embolic events in AF.

<table>
<thead>
<tr>
<th>Risk Factor</th>
<th>CHADS score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Previous stroke, TIA</td>
<td>0</td>
</tr>
<tr>
<td>Age &gt;65</td>
<td>1</td>
</tr>
<tr>
<td>Hypertension</td>
<td>2</td>
</tr>
<tr>
<td>Diabetes mellitus</td>
<td>3</td>
</tr>
<tr>
<td>Heart failure</td>
<td>4</td>
</tr>
<tr>
<td>Prosthetic valve</td>
<td>5</td>
</tr>
<tr>
<td>Coronary artery disease</td>
<td>6</td>
</tr>
<tr>
<td>Mitral stenosis</td>
<td></td>
</tr>
<tr>
<td>Left ventricular dysfunction</td>
<td></td>
</tr>
<tr>
<td>Enlarged left atrium</td>
<td></td>
</tr>
<tr>
<td>Left atrial thrombus</td>
<td></td>
</tr>
<tr>
<td>Severe left atrial mechanical dysf.</td>
<td></td>
</tr>
</tbody>
</table>

Several risk stratification methods are used to assess these risks; however, the most widely used is the CHADS score, where every risk factor is assigned a value (score). The score ranges from 0 to 6, with 0 meaning low risk and 6 representing highest risk.

1. Congestive heart failure (1 point)
2. Hypertension (1 point)
3. Age over 75 years (1 point)
4. Diabetes mellitus (1 point)
5. Stroke or TIA history (2 points)

Table 2.2.4  Assessing yearly risk of CVA using CHADS score.

<table>
<thead>
<tr>
<th>CHADS score</th>
<th>Yearly risk of stroke (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>1.9</td>
</tr>
<tr>
<td>1</td>
<td>2.8</td>
</tr>
<tr>
<td>2</td>
<td>4.0</td>
</tr>
<tr>
<td>3</td>
<td>5.9</td>
</tr>
<tr>
<td>4</td>
<td>8.5</td>
</tr>
<tr>
<td>5</td>
<td>12.5</td>
</tr>
<tr>
<td>6</td>
<td>18.2</td>
</tr>
</tbody>
</table>

CVA does not mean the absence of risk. Similarly, warfarin, although superior in preventing embolic events, may not be suitable for certain patient populations, such as the elderly, due to the increased risk of bleeding. Other anticoagulation options have been used to lower the risk of stroke; these include low-molecular-weight heparin as enoxaparin and antiplatelet drugs as clopidogrel alone or in combination with ASA.

RHYTHM CONTROL

Patients who are symptomatic with AF episodes usually feel better when converted to normal sinus rhythm (NSR). This may need to be done urgently in cases where the patient is hemodynamically unstable, hypotensive, or having active cardiac ischemia. Rhythm control can be achieved by either pharmacological or ECV, or both. In addition, procedures such as radiofrequency (RF) ablation or the surgical MAZE procedure (not discussed) can be used to maintain NSR.

ECV

ECV to NSR can be achieved by applying direct current (DC) to the chest through either paddles or patches. The amount of current used depends on the kind of external defibrillator used and the size of the patient. Using a biphasic external defibrillator, successful cardioversion can be achieved with 50–100 J. More energy (up to 200 J in one defibrillator and 360 J in another model) can be used if necessary as in cases of long-standing AF or patients with a large body habitus.

Table 2.2.5  New anticoagulation guidelines using the CHADS score.

<table>
<thead>
<tr>
<th>Treatment recommendations based on CHADS score</th>
</tr>
</thead>
<tbody>
<tr>
<td>CHADS score</td>
</tr>
<tr>
<td>0</td>
</tr>
<tr>
<td>1</td>
</tr>
<tr>
<td>2+</td>
</tr>
</tbody>
</table>
Chapter 2.2 Atrial Fibrillation and Flutter

Cardioversion should be used after appropriate anticoagulation if the duration of the AF is unknown. Any patient presenting with AF lasting more than 48 hours, who has not been anticoagulated, should not be cardioverted before ruling out intra-atrial thrombus with transesophageal echo (TEE). These patients with AF lasting longer than 48 hours will also need anticoagulation after the cardioversion regardless of the CHADS\textsuperscript{2} risk score.

Immediate success of cardioversion varies between 70 and 94% (Lown 1967). This wide variation depends on many factors and clinical characteristics of the patient. In general, ECV seems to be more successful with the use of a concomitant antiarrhythmic drug (AAD). Other factors that can affect success include duration of AF, size of LA, and the patient’s size, especially the chest.

**PCV and Maintenance of Sinus Rhythm**

PCV is mainly used to convert persistent AF, of relatively short duration, to NSR. This is achieved by using a number of available AADs.

The most widely used AADs are the Class IC drugs flecainide and propafenone, or the class III drugs ibutilide, dofetilide, amiodarone, and dronedarone. The use of other agents such as digoxin, beta blockers, or calcium channel blockers is usually not effective. Before choosing any of the AADs mentioned above to pharmacologically cardiovert a patient, it is important to rule out any underlying structural heart disease. Class I drugs are not suitable for patients with ischemic heart disease or congestive heart failure. The class III agents dofetilide and sotalol can increase the QT interval and are initiated as inpatients. Both these drugs are also excreted by the kidneys and the dosage should be adjusted (or even discontinued) for renal dysfunction. Amiodarone, while a very effective medication, is also one that is associated with many potential side effects or adverse effects. Amiodarone can affect pulmonary function, liver function, the skin, the eyes, and the thyroid. Baseline tests of pulmonary function, liver function, and thyroid function should be performed and followed in patients on this medication.

In general, the concomitant use of suitable AADs with ECV can achieve more successful cardioversion than using either one of these alone. In addition, the patient must be adequately anticoagulated or evaluated for a left atrial clot prior to initiating drugs for the purpose of cardioversion.

Antiarrhythmic medications can also be used to maintain NSR after cardioversion in patients with persistent AF and can be used in the long-term management of these patients. In addition, many patients with paroxysmal AF may be treated with antiarrhythmic medications in order to lessen the frequency and duration of episodes of AF. In many cases, the use of antiarrhythmic medications can result in the successful suppression of AF for many months.

**CATHETER-BASED MANAGEMENT OF AF**

Catheter ablation for AF is a well-established management strategy for AF. Over the last decade, catheter ablation has emerged as the only promising means for the long-term treatment, and potentially the cure, of AF.

Recent technological advances in imaging and mapping have helped electrophysiologists gain better understanding of cardiac anatomy, specifically the LA and the pulmonary veins (PVs) in relation to surrounding structures, thus improving the safety and efficacy of the procedure (Fig. 2.2.1).

**AF Ablation, PV Isolation (PVI)**

Based on the recent increased understanding of the pathogenesis of AF, catheter ablation tech-
needed to make this procedure available to more patients with paroxysmal and persistent AF. Most of the published data available on outcomes of AF ablation have been in relation to paroxysmal AF, where success rate is reported to be as high as 94% (Bhargava et al. 2004; Chen et al. 2004). Recurrence rate in patients with persistent and long-standing AF remains high, compared with the recurrence rate with paroxysmal AF. In general, PVI is offered to symptomatic patients who have failed at least one AAD, regardless of the duration of AF.

**Preprocedure Care and Preparation**

It is important that the patient and the family gain thorough understanding of the procedure, what to expect, and the treatment plan. The PVI procedure requires thorough preparation to ensure its safety. Many centers across the world have established multidisciplinary AF programs where the patient interacts with several health care providers at different stages of the pre-, peri- and postprocedural periods. This
team approach guarantees that the patient has access to professionals who can educate and guide them through the process. The team is usually made up of nurse practitioners (NPs), registered nurses (RNs), and others who follow written protocols for coordinating the patient care with the electrophysiologist. Another very important function of the AF center is to track long-term outcomes of the procedure. Table 2.2.6 shows an example of data reporting tracking outcomes.

Prior to the ablation procedure, several tests are performed. An echocardiogram may be done to rule out valvular or structural disease and to assess LA size and volume index. This data may shed light on the long-term success of the procedure.

Three-dimensional (3-D) multislice computed tomography (CT) may be required if 3-D mapping will be utilized. The CT scan may also serve as baseline data to be compared with a postprocedure CT scan in case pulmonary stenosis is suspected as a complication of the procedure (Figs. 2.2.3 and 2.2.4).

Other tests that may be performed prior to the procedure may also include a Holter monitor, event recorder, and 12-lead electrocardiogram (ECG) to document AF burden along with heart rates during AF or the transition from AF to NSR.

Table 2.2.6 2006 AF ablation program quality data.

<table>
<thead>
<tr>
<th>Number of cases</th>
<th>Success after one procedure</th>
<th>Success after two procedures</th>
<th>Partial success with AAD</th>
<th>Major complication</th>
<th>Minor complication</th>
</tr>
</thead>
<tbody>
<tr>
<td>455</td>
<td>77%</td>
<td>93%</td>
<td>3%</td>
<td>0.6%</td>
<td>1.2%</td>
</tr>
</tbody>
</table>

Success = no atrial arrhythmias off AAD after 3 months blanking period; major complications—stroke, tamponade, pulmonary vein stenosis, death; minor complications—hematoma, vascular injury.

Figure 2.2.3 This image is a CT scan of the left atrium demonstrating a large common left-sided pulmonary vein trunk. This anatomical variation may have some implications on the approach used for ablation in this individual. PV, pulmonary vein.

Figure 2.2.4 This image is a CT scan of the left atrium demonstrating a mid right-sided vein, another anatomical variation. The colored dots represent areas of ablation.
AADs and rate control medications may be discontinued for a few days prior to the procedure.

Patients undergoing AF ablation should be anticoagulated for 4–6 weeks prior to the procedure, maintaining an international normalized ratio (INR) of 2–3. This needs to be closely monitored. A TEE may be required on the day of the procedure to rule out intracardiac thrombus if anticoagulation is believed to have been subtherapeutic at any time during the last 4–6 weeks prior to PVI. Anticoagulation protocols may vary from one institution to another. While most of the protocols necessitate that warfarin must be discontinued 2–3 days prior to the procedure, others continue Coumadin through the procedure to maintain therapeutic INR. A recent study of 3,000 patients who underwent PVI while maintaining therapeutic INR (2–3) recently published (Oussama et al. 2007) has shown that there were no significant complications with this approach.

When Coumadin is stopped prior to the procedure, Lovenox is usually used to maintain anticoagulation until 12 hours prior to the procedure, then resumed again immediately after the procedure until INR is again at a therapeutic level.

**Periprocedural Anticoagulation**

Thromboemboli events during left atrial ablations are a well-known risk. Appropriate periprocedural anticoagulation is the most important factor to ensure safety during the procedure.

Protocols for periprocedural anticoagulation vary widely. However, several studies have been published indicating that an activated clotting time (ACT) of 350–400 seconds should be maintained as long as left atrial instrumentation is in progress. To minimize the formation of clots at the tip of the transseptal sheaths, a heparin bolus can be given immediately prior to the first transseptal puncture. The heparin bolus is calculated at 150 IU/kg. Heparin intravenous (IV) infusion is also started at 150 IU/kg/h. Immediately after the second transseptal puncture, a second heparin bolus is administered, 50–70 IU/kg. ACT is initially checked in 15–20 minutes and dosage is adjusted to maintain the target ACT. The ACT needs to be monitored continuously throughout the procedure at regular defined intervals and can vary by institution, 20–30 minutes, with dosage adjustments made to retain the therapeutic ACT value.

**Delivery of RF Energy**

To gain access to the LA, double transseptal access is obtained as shown in Figure 2.2.5.

![Figure 2.2.5](image) This image is a rendering of the heart showing an intracardiac ultrasound catheter in the right atrium, and an ablation catheter and a multipolar circular mapping catheter passed into the left atrium via the septum.
Intracardiac echo is used in many instances and can facilitate both the transseptal access in real time, as well as help in the localization of the catheters (Figs. 2.2.5 and 2.2.6).

Figure 2.2.6 Intracardiac ultrasound image of the left atrium and the septum, as well as the pulmonary vein. Note that the tubular portion of the pulmonary vein can easily be distinguished from the body of the left atrium using ultrasound.

Guided by EGM, RF energy is most commonly delivered using an open irrigated-tip catheter, although some centers may use large-tip or closed-loop irrigation catheters (Figs. 2.2.7 and 2.2.8). At our center, RF energy is started at 30 W and gradually titrated to 40–50 W to reach a temperature of 40°C, except when RF energy is delivered in the proximity of the esophagus, in this case energy is dropped to 30 W with maximum temperature of 30°C. Lesions are typically 30–60 seconds in duration depending on the location of the catheter tip in relation to the different landmarks in the LA. The use of intracardiac echocardiogram (ICE) during this stage can be very useful in determining the power and temperature used in the various anatomical structures of the LA. There is no existing guidelines or consensus regarding these parameters for energy and temperature limits; however, careful attention to the EGM, power, temperature, and impedance rise is prudent and essential to prevent complications.

There should be one dedicated staff member watching these parameters at all times, alerting

Figure 2.2.7 This is a typical recording during a pulmonary vein ablation procedure. Note the pacing artifact as the patient is being paced from the coronary sinus catheter. Also note the pulmonary vein potentials that appear sharp, and with high frequency, prior to ablation.
the operator and titrating the power according to preset protocols.

**Potential Complications during PVI Ablation**

Major complications during PVI have been declining in the last few years due to increasing experience with the procedure and with the advent of new available technologies. In a study published in 2005, which included 8,745 worldwide patients who underwent PVI, the investigators reported the most common complications and their incidence (Table 2.2.7).

A rare but serious complication of PVI is atrioesophageal fistula (AEF). It is difficult to estimate its incidence because it is probably underreported especially when it occurs several days or even weeks after the procedure. It is difficult to diagnose AEF as the presentation can be nonspecific and can be attributed to various different causes.

Various approaches have been employed to accurately locate the esophagus during ablation in the LA. The most common approach is positioning a temperature probe within the esophagus to continuously monitor the temperature within the intramural esophageal. Reducing the use of maximum power and reduced total duration time can greatly reduce the risk of this devastating complication.

Another potentially devastating complication is the development of a pericardial effusion and tamponade. Therefore, access to a pericardial drainage kit is of vital importance, as is the rapid attention to significant drops in systolic blood pressure.

**Table 2.2.7** Common potential complications of PVI.

<table>
<thead>
<tr>
<th>Complication</th>
<th>Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thromboembolic events</td>
<td>0.94%</td>
</tr>
<tr>
<td>Pulmonary vein stenosis</td>
<td>1.63%</td>
</tr>
<tr>
<td>Cardiac tamponade</td>
<td>1.22%</td>
</tr>
<tr>
<td>Death</td>
<td>0.05%</td>
</tr>
<tr>
<td>Permanent diaphragmatic paralysis</td>
<td>0.11%</td>
</tr>
<tr>
<td>Atrioventricular fistula</td>
<td>0.95%</td>
</tr>
</tbody>
</table>

**Figure 2.2.8** This is a typical recording during a pulmonary vein ablation procedure. This recording is after some ablation has occurred. Note the pacing artifact as the patient is being paced from the coronary sinus catheter. Also note that some of the pulmonary vein potentials that were evident in the prior figure are now gone.
Chapter 2.2 Atrial Fibrillation and Flutter

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and the opposite is true as well. Risk factors for atrial flutter and associated conditions are also similar to those related to AF.

There are several types of atrial flutter: typical atrial flutter (sometimes referred to as common flutter), reverse typical atrial flutter, and atypical flutter. Both typical and reverse typical atrial flutter utilizes the same circuit in the right atrium around the tricuspid valve. The wavefront with typical atrial flutter is counterclockwise when looking at the heart fluoroscopically in the left anterior oblique (LAO) view, whereas in the reverse typical type, flutter is clockwise (Fig. 2.2.9). The pattern on the 12-lead ECG is also different; while typical atrial flutter has a negative “sawtooth”

Postprocedural Care and Follow-Up

Follow-up protocols vary widely among practitioners. It is crucial that anticoagulation is therapeutically maintained in the few weeks following the procedure. Patients usually resume Coumadin the evening of the procedure or the day after if they had to stop prior to the procedure. Antiarrhythmic medications may be resumed after PVI and continued for 6-8 weeks thereafter.

Patients are usually discharged from the hospital the day following the PVI procedure. However, some physicians may hospitalize the patient for a longer time for observation or until INR is back to therapeutic level. Patient’s rhythm is monitored closely for the 6-8 weeks post PVI by an event recorder. The patient is asked to send random rhythm transmission two to three times a week and whenever the patient is symptomatic so recurrence can be documented. It is not uncommon for patients to experience early recurrence of AF in the first few weeks after PVI. This does not mean failure of the procedure and patients need to be given reassurance of that during this time period. Late recurrence (after 8–12 weeks) of AF or other atrial tachyarrhythmias may indicate that the complete isolation of the PVs was not successfully completed and another procedure may be warranted to assure arrhythmia elimination. Successful PVI is defined as freedom from AF or atrial arrhythmias at 1 year post procedure. Patient should be in NSR without the need for AADs.

ATRIAL FLUTTER

Atrial flutter is considered a macroreentrant arrhythmia, typically with an organized rapid atrial rate.

The exact incidence of atrial flutter is not known; however, it is less common that AF. Patients with AF may also have atrial flutter, and the opposite is true as well. Risk factors for atrial flutter and associated conditions are also similar to those related to AF.

Figure 2.2.9 An electroanatomical map of typical counterclockwise atrial flutter. The right atrium is represented in a modified LAO view. The white arrow shows the direction of activation of this atrial flutter. IVC, inferior vena cava; TV, tricuspid valve; CS, coronary sinus. (Modified from Al-Ahmad A, Callans DJ, Hsia HH, Natale A (eds.). 2008. Electroanatomical Mapping: An Atlas for Clinicians. New York: Wiley-Blackwell.)
Cardiac Arrhythmia Management

Respect to warfarin use based on the CHADS₂ score are reasonable guidelines to follow.

Antiarrhythmic medications and cardioversion are also important strategies to maintaining NSR in patients with atrial flutter. Cardioversion and the use of these medications have been discussed earlier in this chapter.

RF ablation for typical and reverse typical atrial flutter is highly effective, with a success rate in the range of 95%. Ablation using either a large-tip catheter or an irrigated-tip catheter starting from the tricuspid valve and ending at the inferior vena cava interrupts the flutter circuit and terminates the arrhythmia (Fig. 2.2.12). For atypical flutter, the challenge is to identify the narrowest essential part of the atrial flutter circuit and deliver ablation energy in that location (Fig. 2.2.13). Care must be taken to ensure that the location where energy is

Appearance of the flutter waves that can best be appreciate in the leads II, III, and aVF, reverse typical flutter will have a positive “sawtooth” appearance in these same leads (Fig. 2.2.10). Atypical atrial flutter may utilize other circuits in the right or the left atrium typically related to areas of scar or slow conduction that may have been caused by surgery or prior catheter ablation for AF (Fig. 2.2.11).

Treatment of Atrial Flutter

The treatment of atrial flutter is similar to that of AF. Medications that are used for heart rate control with AF can also be effective in controlling the ventricular rate in patients with atrial flutter. In addition, the risk of stroke with atrial flutter is felt to be similar to that of AF, and therefore the same recommendations with respect to warfarin use based on the CHADS₂ score are reasonable guidelines to follow.

Antiarrhythmic medications and cardioversion are also important strategies to maintaining NSR in patients with atrial flutter. Cardioversion and the use of these medications have been discussed earlier in this chapter.

RF ablation for typical and reverse typical atrial flutter is highly effective, with a success rate in the range of 95%. Ablation using either a large-tip catheter or an irrigated-tip catheter starting from the tricuspid valve and ending at the inferior vena cava interrupts the flutter circuit and terminates the arrhythmia (Fig. 2.2.12). For atypical flutter, the challenge is to identify the narrowest essential part of the atrial flutter circuit and deliver ablation energy in that location (Fig. 2.2.13). Care must be taken to ensure that the location where energy is

Figure 2.2.10 ECG of atrial flutter. (A) Typical atrial flutter; note the negative “sawtooth” pattern of the flutter waves seen in the leads II, III, and aVF. (B) Reverse typical atrial flutter, with a positive “sawtooth” pattern on the flutter waves in leads II, III, and aVF.
**Figure 2.2.11** ECG of atypical atrial flutter. This ECG shows an atypical right atrial flutter that utilized prior scarring in the right atrium with the atrial wavefront going around the scar. This patient had a scar in the right atrium as a result of cardiac surgery. Note that the atrial flutter waves are less “sawtooth” in appearance and are more rounded.

**Figure 2.2.12** An electroanatomical map of typical counterclockwise atrial flutter. The right atrium is represented in a modified LAO view. The red dots represent the ablation lesions that span the area from the tricuspid annulus to the IVC. IVC, inferior vena cava; TV, tricuspid valve; CS, coronary sinus. (Modified from Al-Ahmad A, Callans DJ, Hsia HH, Natale A (eds.). 2008. Electroanatomical Mapping: An Atlas for Clinicians. New York: Wiley-Blackwell.)
delivered is safe in order to minimize collateral damage from the ablation (example damage to the phrenic nerve). Postablation care and follow-up is similar but less intensive as compared with AF ablation follow-up. Care should be taken to monitor patients after the ablation for AF as these rhythm disturbances are often seen in the same patient.

CONCLUSION

AF is the most common arrhythmia in adults and is increasing in incidence. AF remains a challenging arrhythmia to treat as it involves many different medications as well as potential procedures. PVI is safe and effective as an approach for the treatment and even potential cure of AF. The ideal therapy for each individual patient can vary depending on the overall health of the patient, the symptoms of the patient, and the desires of the patient. Patient education is paramount in making these types of therapeutic decisions. Atrial flutter shares many of the same treatment options as AF, and again, individualizing care is important. Patients who have atrial flutter should also be screened for AF.

The allied health professional role is essential for patient safety and for supporting the patient throughout the process. A comprehensive team approach has proven to be the most successful in making this process seamless and providing adequate support for the patients and their families.

REFERENCES


RESOURCES


INTRODUCTION

Cardiovascular disease (CVD) remains the leading cause of death since 1900; with one in eight death certificates mentioning heart failure. Heart failure is most often a consequence of coronary heart disease, hypertension, cardiomyopathy, valvular disease, or diabetes. Although overall cardiovascular mortality is decreasing, 5.7 million people have cardiomyopathy and heart failure and experience sudden death six to nine times more often than the general population (Lloyd-Jones et al. 2009). Patients with cardiomyopathy commonly have frequent ventricular arrhythmias, ranging from benign premature ventricular contraction (PVC) to lethal ventricular tachycardia (VT) or ventricular fibrillation (VF), causing sudden death. VT or VF is the first recorded rhythm in 20–38% of out-of-hospital cardiac arrests (Lloyd-Jones et al. 2009). Recent guidelines for management of ventricular arrhythmias indicate that the major substrate for ventricular arrhythmias is coronary artery disease (Zipes et al. 2006).

Presence of ventricular arrhythmias increases the overall mortality; it is not necessarily predictive of sudden death. For optimal patient management, one must take into consideration the symptoms and potential mechanism of ventricular tachyarrhythmias, in addition to the types and severity of the underlying structural heart disease (Stevenson et al. 2002). It is important to understand the mechanisms of ventricular arrhythmias and to utilize evidence-based approaches for classifications of complex cardiomyopathies. This chapter focuses on VT associated with cardiomyopathies, its diagnosis, and current treatment modalities.
DEFINITIONS AND CLASSIFICATIONS OF VENTRICULAR ARRHYTHMIAS

Ventricular arrhythmias have different classifications and definitions (Table 2.3.1). PVC often refers to a single ectopic ventricular beat that originates from the ventricle and occurs before the normally conducted ventricular activation. If PVCs originate from the same focus in the ventricle, they are characterized as unifocal and identical in morphology (monomorphic) during ventricular depolarization. Ventricular depolarization is noted on the electrocardiogram (ECG) as the QRS complex (Fig. 2.3.1). Multifocal PVCs are from different ventricular foci and not identical in morphology. PVCs can appear in pairs, every other beat, every third, or fourth beat between normal beats, and are described as paired (couplet), bigeminal, trigeminal, and quadrigeminal, respectively (Fig. 2.3.2). The causes of PVCs are many and often are multifactorial, including ischemia, electrolyte imbalance such as hypokalemia or hypercalcemia, fever/stress, hypovolemia, myocardial stretch from volume overload, or ventricular scarring from prior myocardial infarction (MI).

PVCs >10 per hour is a prognostic indicator of increased risk of overall mortality and sudden death in patients with recent MIs or history of nonsustained VT (NSVT). VT is defined as a ventricular arrhythmia with three or more consecutive complexes in duration at a rate greater than 100 bpm (Zipes et al. 2006). NSVT is defined as an episode of VT that lasts <30 seconds in duration, whereas sustained VT represents episodes of VT that lasts >30 seconds in duration, or VT associated with hemodynamic instability that requires intervention for termination (Fig. 2.3.3).

Morphologically, VT presents as either monomorphic or polymorphic. Monomorphic VT consists of a single, stable QRS complex that are multiform or variable, suggesting that ventricular activation originates in different locations in the ventricles. Torsades de pointes (TdP) is a French term meaning “twisting of the points” and represents a specific variant of polymorphic VTs with characteristic electrocardiographic features (Fig. 2.3.4). Torsade is commonly associated with a prolonged interval between ventricular depolarization and repolarization, called the QT interval, with a “twisting” QRS pointing upward and downward (Cranefield and Aronson 1988). It may be related to congenital ion channel defects, metabolic derangement, bradycardia, or drug toxicities.

Etiology of VT, either monomorphic or polymorphic, can result from a variety of disease entities. Coronary artery disease is the most common cause of VT, followed by other structural heart diseases with ventricular dysfunction. Genetic abnormalities such as long QT syndromes or Brugada syndrome with abnormal cellular or molecular substrates can also play a role.

It is important to distinguish a monomorphic from a polymorphic ventricular arrhythmia. Monomorphic QRS implies a relatively fixed early ventricular activation and impulse formation. This is most commonly associated with a scar-based reentrant circuit with a “fixed” substrate or a focal idiopathic VT. Polymorphic VT, on the other hand, suggests variable patterns of ventricular activation and impulse propagation, most commonly caused by metabolic abnormalities, acute ischemia, drug toxicity, and congenital or acquired long QT syndromes.

BASIC CELLULAR ELECTROPHYSIOLOGY

Knowledge of the cardiac action potential (AP) is essential to understanding normal conduction process and provides insight to abnormal
Table 2.3.1  Classification of ventricular arrhythmias.

<table>
<thead>
<tr>
<th>Hemodynamically stable</th>
<th>Asymptomatic</th>
<th>The absence of symptoms that could result from an arrhythmia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Minimal symptoms</td>
<td></td>
<td>(e.g., palpitations)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Patient reports palpitations felt in either the chest, threat, or neck as described by the following:</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Heartbeat sensations that feel like pounding or racing</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• An unpleasant awareness of heartbeat</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Feeling skipped beats or a pause</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Hemodynamically unstable</th>
<th>Presyncope</th>
<th>Patient reports presyncope as described by the following:</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>• Dizziness</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Light-headedness</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Feeling faint</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• &quot;Graying cut&quot;</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Syncope</th>
<th>Sudden loss of consciousness with loss of postural tone, not related to anesthesia, with spontaneous recovery as reported by the patient or observer; patient may experience syncope when supine</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>Sudden cardiac death</th>
<th>Death from an unexpected circulatory arrest, usually due to a cardiac arrhythmia occurring within an hour of the onset of symptoms</th>
</tr>
</thead>
</table>

| Sudden cardiac arrest    | Death from an unexpected circulatory arrest, usually due to a cardiac arrhythmia occurring within an hour of the onset of symptoms, in whom medical intervention (e.g., defibrillation) reverses the event |

**Classification by electrocardiography**

<table>
<thead>
<tr>
<th>Nonsustained VT</th>
<th>Three or more beats in duration, terminating spontaneously in less than 30 seconds; VT is a cardiac arrhythmia of three or more consecutive complexes in duration emanating from the ventricles at a rate of greater than 100 bpm (cycle length less than 600 ms)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Monomorphic</td>
<td>Nonsustained VT with a single QRS morphology</td>
</tr>
<tr>
<td>Polymorphic</td>
<td>Nonsustained VT with a changing QRS morphology at cycle length between 600 and 180 ms</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Sustained VT</th>
<th>VT greater than 30 seconds in duration and/or requiring termination due to hemodynamic compromise in less than 30 seconds</th>
</tr>
</thead>
<tbody>
<tr>
<td>Monomorphic</td>
<td>Sustained VT with a stable single QRS morphology</td>
</tr>
<tr>
<td>Polymorphic</td>
<td>Sustained VT with a changing or multiform QRS morphology at cycle length between 600 and 180 ms</td>
</tr>
</tbody>
</table>

| Bundle branch reentrant tachycardia | VT due to reentry involving the His–Purkinje system, usually with LBBB morphology; this usually occurs in the setting of cardiomyopathy |
Classification by electrocardiography

Bidirectional VT        VT with a beat-to-beat alternans in the QRS frontal plane
axis, often associated with digitalis toxicity

Torsades de pointes        Characterized by VT associated with a long QT or QTc, and electrocardiographically characterized by twisting of the peaks of the QRS complexes around the isoelectric line during the arrhythmia:
• “Typical,” initiated following “short-long-short” coupling intervals
• Short-coupled variant initiated by normal-short coupling

Ventricular flutter        A regular (cycle length variability 30 ms or less) ventricular arrhythmia approximately 300 bpm (cycle length: 200 ms) with a monomorphic appearance, no isoelectric interval between successive QRS complexes

Ventricular fibrillation        Rapid, usually more than 300 bpm/200 ms (cycle length 180 ms or less), grossly irregular ventricular rhythm with marked variability in QRS cycle length, morphology, and amplitude

Classification by disease entity

Chronic coronary heart disease
Heart failure
Congenital heart disease
Neurological disorders
Structurally normal hearts
Sudden infant death syndrome
Cardiomyopathies
Dilated cardiomyopathy
Hypertrophic cardiomyopathy
Arrhythmogenic right ventricular cardiomyopathy


LBBB, left bundle-branch block; VT, ventricular tachycardia.

Table 2.3.1 (Continued)

Text not available in the electronic edition
Figure 2.3.1  An example of sinus rhythm with unifocal PVC and nonsustained VT.

Figure 2.3.2  An example of sinus rhythm with monomorphic PVCs in a bigeminal pattern.

Figure 2.3.3  An example of sustained monomorphic VT.
Cardiac cells are excitable and capable of creating and propagating electrical impulses. Cardiac AP is a complex process of multiple, coordinated ionic influx and efflux across cell membranes, resulting in impulse formation and propagation. Normal myocytes have a resting negative membrane potential of \(-85\) to \(-90\) mV and remain negatively polarized until they are electrically stimulated. The AP is depicted in five phases (Fig. 2.3.5). The five phases represent the depolarization, repolarization, and resting state of the cardiac cells. Cellular depolarization occurs with the initial

Figure 2.3.4 Torsades de pointes (TdP), the “twisting of the points,” typically describes a polymorphic ventricular tachycardia associated with prolonged QT intervals. The underlying rhythm is sinus bradycardia with QT prolongation. Arrow indicates PVC with “R on T” that induced TdP.
Chapter 2.3 Ventricular Tachycardia Associated with Cardiomyopathies

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in the regulatory protein receptors, or electrolyte abnormalities are all contributing factors to arrhythmogenesis.

MECHANISMS OF VT

The mechanism of VT is categorized into abnormal impulse formation or abnormal impulse conduction; specifically automaticity, triggered activity, and reentry (Fig. 2.3.6).

Figure 2.3.5 Example of cardiac action potential depicting the five phases (0–4) of cellular depolarization and repolarization. The corresponding transmembrane electrolyte changes and ionic current flow are also represented.

Extracellular

intracellular

Na+

Na+

Na+

Na+

Ca++

K+

K+

Na+

K+

K+

Na+

Figure 2.3.6 Flowchart depicting mechanisms of ventricular tachycardia.

Mechanisms of Arrhythmias

Abnormal Impulse

Formation

Abnormal Impulse

Conduction

Automaticity

Trigged

Activity

Reentry

Enhanced

Abnormal

Anatomical

Functional

EADs

Early Afterdepolarizations

DADs

Delayed Afterdepolarizations

rapid influx of positively charged sodium ions into the myocytes (phase 0) and a small efflux of K+ (phase 1), yielding a positive cell membrane potential. This is followed by Ca++ influx (phase 2), resulting in the plateau phase of the AP and cardiac contraction. The next phase (phase 3) consists of efflux of potassium (K+), resulting in repolarization and back to the resting state (phase 4). Functional alterations or structural changes of the ionic channels, defects in the regulatory protein receptors, or electrolyte abnormalities are all contributing factors to arrhythmogenesis.
Reentry, abnormal automaticity, and triggered mechanisms may be anatomically or functionally based (Josephson et al. 1978a; Stahmer and Cowan 2006; Das et al. 2008). The mechanisms and etiology of VT have been studied extensively in animal models, intraoperative surgical mapping, and electrophysiology laboratories (Josephson et al. 1978a,b; Richardson et al. 1999; Zei and Stevenson 2006; Antzelevitch 2007). Clinically, most ventricular arrhythmias due to automaticity are seen in the setting of acute ischemia (such as MI), hypoxemia, metabolic disturbances (acid-base and/or electrolyte), increased adrenergic tone, and drug toxicities. In the chronic setting, reentrant arrhythmias are more common due to scar-related formation that initiates VT and can occur years after the initial insult.

**Abnormal Impulse Formation**

**Abnormal Automaticity**

*Automaticity* is the ability of a cell to spontaneously depolarize without any extracellular stimulation. Abnormal automaticity occurs when the cardiac cell fires independently, creating an impulse that is not from the normal sinus node source. This creates an interruption in the normal conduction in the heart and is characterized by a premature atrial or ventricular contraction, and junctional or ventricular heart rhythm.

**Triggered Activity**

Triggered activity occurs when there are abnormalities of APs that trigger another electrical event by way of abnormal depolarization. Two types of triggered activity are early and late afterdepolarizations (Fig. 2.3.7).

- **Early afterdepolarization (EAD):** EADs occur due to abnormal phase 2 and 3 depolarization, causing early AP to occur before the cellular membrane potential returns back to baseline. EADs may occur when the heart rate is markedly slowed, reducing the outward potassium current. EADs is usually associated with bradycardia or during pauses and is thought to initiate TdP.

- **Delayed afterdepolarization (DAD):** DADs occur due to elevated levels of calcium influx during phase 4 depolarization when the AP is almost fully repolarized. The elevated cellular membrane potential reaches the threshold, causing the cell to refire, initiating ventricular ectopy. DADs are typically associated with certain digitalis toxic arrhythmias as well as catecholamine-dependent atrial and ventricular tachycardias.

![Figure 2.3.7](image-url) Triggered activities are caused by abnormal cell membrane voltage oscillations, induced by preceding action potentials. Early afterdepolarizations (EADs) occur in late phase 2 or phase 3 of the action potential, before complete cellular repolarization (A). When such membrane voltage oscillation reaches the threshold, a second membrane depolarization was induced (B). Repetitive EADs may occur with bradycardia or reperfusion injury (C). Delayed afterdepolarizations (DADs) occurs in late phase 3 or early phase 4 when the membrane potential is fully repolarized (D). DADs are thought to be responsible for certain digitalis toxic arrhythmias as well as catecholamine-dependent atrial and ventricular tachycardias (E).
Chapter 2.3 Ventricular Tachycardia Associated with Cardiomyopathies

Abnormal Impulse Conduction

Reentry

Reentry is the most common mechanism for the majority of clinically relevant arrhythmias, both atrial and ventricular tachycardias. A reentrant rhythm requires an anatomical/functional circuit with multiple potential “pathways” of different electrophysiological properties (Fig. 2.3.8). The hallmark of reentrant arrhythmias is slow conduction, most commonly due to structural heart disease with abnormal scar-based nonuniform conduction delays. The prerequisites of reentry include (1) unidirectional block within the circuit, often due to a premature impulse, (2) impulse propagation down an alternative “pathway”, and (3) sufficiently slow conduction within the circuit that allows recovery of refractoriness, with electrical impulse reentering at the site of initial block.

The circuit for reentry may be anatomical, functional, or both. The most common cause of VT is macroreentry based on anatomical scar substrate in patients with structural heart disease, such as scar formation from a previous MI (Fig. 2.3.9). Coronary artery disease, ischemic in nature, causes myocardial death, fibrotic scarring, cellular hypertrophy, and chamber dilatation that contribute to arrhythmogenesis. Other types of cardiomyopathies can also have macro- or microscar formation and provide an anatomical substrate for VT.

Functional reentry is not associated with a fixed anatomical location and is not necessarily related to a physical obstacle. The circulating wavefronts can change location, size, and rotational directions in cardiac tissue, propagating variable shifting reentrant circuits. This mechanism is important in polymorphic VT or ventricular/atrial fibrillation (Kleber and Rudy 2004).

DEFINITIONS AND CLASSIFICATIONS OF CARDIOMYOPATHY

Cardiomyopathy is described as a heart muscle disorder, be it mechanical or electrical, that results in dysfunctional myocardium with structural and functional abnormalities, often with depressed ventricular contractions. Causes of cardiomyopathy include MI, hypertension, valvular disease, viral infection, familial and/or genetic inheritance, or idiopathic, indicating
Figure 2.3.9 A diagrammatic illustration of a potential VT circuit. The VT substrate has complex geometry and dimension for scar-based reentry. Multiple sites within the circuit are depicted: entrance, central common pathway (isthmus), exit, and outer loop sites. In addition, multiple “bystander” sites or “blind loops” are also present.

The underlying cause is undetermined (Fig. 2.3.10). The etiology and classification of cardiomyopathy helps distinguish underlying pathology, subsequent risk of ventricular arrhythmias, and appropriate treatment. The World Health Organization (WHO) classified cardiomyopathy into categories in 1995 but has not updated it since, despite an explosion of molecular genetics and discovery. The four categories are dilated cardiomyopathy (DCM), hypertrophic cardiomyopathy (HCM), restrictive cardiomyopathy (RCM), and arrhythmogenic right ventricular dysplasia/cardiomypathy (ARVD/C). In addition, a subset of specific cardiomyopathies associated with a specific cardiac disease or systemic disorders have been recognized (Richardson et al. 1996). The American Heart Association (AHA) and European Society of Cardiology Working Group on Myocardial and Pericardial Diseases have recently published position and consensus statements to update the decade-old WHO classification. The authors propose updated but conflicting cardiomyopathy definitions of the heterogeneous disease group. The AHA uses “primary cardiomyopathy,” which confines the malady to the heart itself, and “secondary cardiomyopathy” as a systemic disorder affecting the heart muscle (Elliott et al. 2008; Maron et al. 2006). The European group disagrees and offers an alternative classification. The WHO classification of cardiomyopathies is distinguished by those with and without the causative genetic defect (Elliott et al. 2008). Ischemic cardiomyopathy, coined in the 1970s, is used as a definition in research and is a common cause of cardiomyopathy (Burch 1973; Felker et al. 2002) (Fig. 2.3.11). Both documents note the exclusion of the term cardiomyopathy resulting from hypertension, coronary artery, congenital, and valvular heart disease.

DCM

DCM is the most common form of cardiomyopathy, expressed as left ventricular or biventricular dilatation or impaired systolic function, and accounts for 55% of cases (Richardson et al. 1996). The ventricular walls are thin, with a dilated ventricular cavity and a low ejection fraction (EF). Causes can be ischemic or nonischemic, familial, viral, metabolic, drug or alcohol
Figure 2.3.10  Flowchart depicting classifications of cardiomyopathy. HCM, hypertrophic cardiomyopathy; DCM, dilated cardiomyopathy; ARVD/C, arrhythmogenic right ventricular dysplasia/cardiomyopathy; LVNC, left ventricular noncompaction; LQTS, long QT syndrome; SQTS, short QT syndrome; CPVT, catecholaminergic polymorphic VT; SUNDS, sudden unexplained nocturnal death syndrome.

Figure 2.3.11  An echocardiogram image in a patient with normal left ventricular wall thickness and size (A). An echocardiogram image in a patient with a diagnosis of dilated cardiomyopathy. The left ventricle is much enlarged with wall thinning (B).
induced, or derived from an autoimmune response. Prior MI results in myocardial scar or fibrosis formation. The healthy myocytes stretch and thicken, causing hypertrophy and eventual heart failure. Mitral ring stretching occurs, so mitral regurgitation is common. Patients complain of dyspnea, exercise intolerance, and an elevated resting heart rate, and congestive heart failure (CHF) is a familiar presentation. Local conduction slowing and abnormalities constitute the substrate for scar-based reentry. Nonischemic cardiomyopathy reflects all the other forms of ventricular dysfunction and accounts for 20% of sudden death (Dec and Fuster 1994). Patients with nonischemic cardiomyopathy may present with diffuse myocardial fibrosis, with triggers for focal VT. VT utilizing the cardiac conduction system is also prevalent in patients with nonischemic cardiomyopathy and is often associated with underlying His–Purkinje system conduction disease. However, myocardial reentry is the most common mechanism of sustained monomorphic VT (Hsia et al. 2003; Soejima et al. 2004). Familial cardiomyopathy is seen in families, and 40% are considered autosomal dominant.

**HCM**

HCM is characterized by a thickened myocardium, usually located in the intraventricular septum, and is a leading cause of sudden cardiac death (SCD) in young people (Maron et al. 1994, 1996). Left ventricular outflow obstruction can occur, with a normal or elevated EF and left ventricular hypertrophy (LVH) (Fig. 2.3.12). Sarcomere protein mutations result in impaired contractility, hypertrophy, and for some, is familial and considered an autosomal (nonsex chromosome) dominant genetically inherited disorder. Some patients are asymptomatic, and sudden death after vigorous exercise may be the first clinical manifestation of the disease. Additionally, patients can present with chest pain, dyspnea, or syncope. Risk factors for sudden death in this disease appear to be related to (1) a family history of sudden death, (2) hypotensive response during exercise stress test, (3) a history of unexplained syncope, (4) severe septal thickness (>3 cm), and (5) presence of spontaneous NSVT.

**RCM**

RCM is differentiated by a restrictive pathology, with reduced diastolic volume, ventricular filling, atrial enlargement, impaired relaxation, and subsequent heart failure. It is characterized by myocardial stiffness that causes a precipitous rise in pressure with small increases in volume (Kushwaha et al. 1997). Systolic function is usually normal and cardiomegaly is absent. It is the least common of the cardiomyopathies (Abelmann 1984). Amyloidosis and sarcoidosis are common causes of RCM, as well as cardiac tumors, hemochromatosis, radiation, and scleroderma. Myocardial biopsy is utilized to aid with diagnosis. Amyloidosis carries a poor prognosis; at least 50% of the patients die of an arrhythmia or heart failure (Shah et al. 2006). Cardiac sarcoidosis is a rare disease of granulomatous inflammation, causing a myo-
PATIENT MANAGEMENT

A thorough history and physical examination is essential in the management of patients with VT associated with cardiomyopathies. Risk stratification is critically dependent on the type and extent of the underlying structural heart disease. A depressed EF remains the most consistent predictor of SCD in patients who have structural heart disease, irrespective of etiology. A patient’s functional capacity can also be determined using the New York Heart Association (NYHA) classification, which portends prognostic significance in patients with cardiomyopathies. It is important to note that although the mortality is highest in patients with severe cardiomyopathy in advanced stage of heart failure, sudden death accounts for only a modest proportion of the total mortality. Progressive hemodynamic deterioration and pump failure is the major cause of death in this population. On the contrary, sudden presumably arrhythmic death accounts for a significant proportion of total mortality in patients with mild to moderate ventricular dysfunction. The relative risk of sudden death due to ventricular tachyarhythmias is higher in patients who are generally minimally symptomatic from their cardiomyopathies (Fig. 2.3.13).

It is important to obtain an accurate patient history, particularly focusing on the underlying ventricular dysfunction, as well as the type and extent of cardiomyopathies. Symptom presentations, syncope or near syncope, or family history of SCD are important factors to consider in the risk stratification process. Physical examination includes evaluation of vital signs, heart and lung auscultations, and assessment of jugular vein distention. Augmentation of a crescendo-decrescendo systolic murmur along the left sternal border during a Valsalva maneuver is suggestive of systolic outflow obstruction in HCM patients. Lower extremity edema may indicate fluid overload, right, or left ventricular failure.
Cardiac Arrhythmia Management

After obtaining the patient history and physical examination, noninvasive tests to consider include a 12-lead ECG, ambulatory Holter monitor, event monitor, and most importantly, an echocardiogram to assess the type and degree of structural heart disease. Laboratory studies are considered to rule out metabolic, electrolyte, hematologic, and infectious causes of cardiomyopathies.

12-Lead ECG

The 12-lead ECG is an important diagnostic test to evaluate a patient for VT and cardiomyopathy. It provides baseline information on the intrinsic rhythm (sinus, atrial fibrillation/flutter, paced rhythms), the underlying structural heart disease (conduction disturbances, atrial/ventricular hypertrophy, chamber dilation, and the presence and location of myocardial scar). During spontaneous ventricular arrhythmia, the ECG helps identify the morphology and possible location of early ventricular activation of the ectopic arrhythmia. Various algorithms have been developed to "regionalize" the probable "site of origin" of PVCs or VT based on the locations of prior MIs and the morphological characteristics of the QRS (Miller et al. 1988; Kuchar et al. 1989). A negative QRS morphology in lead V1, designated as the LBBB pattern, suggests the VT is originating from either the RV or the left ventricular septum. If the QRS in V1 is predominantly positive, designated as the right bundle branch block (RBBB) pattern, the VT is originating from the left ventricle (LV). The QRS axis in the frontal plane dictates superior or inferior VT site of origin, and the patterns of precordial transition (V1 to V6) indicates apical versus basal locations (Fig. 2.3.14). These ECG characteristics allow the clinician to ascertain the location of the VT, which is helpful if VT ablation is considered.

Echocardiogram

In patients with cardiomyopathies, transthoracic echocardiogram is a valuable tool in defining the underlying arrhythmia substrate. Assessment of the EF carries both diagnostic and prognostic information, with a lower EF predicting greater total mortality. In patients with ischemic heart disease and prior MI, locations of the myocardial scar may be confirmed by regional wall motion abnormalities, local thinning of the infarcted area, with aneurysm or thrombus formation. In patients with DCM, diffusely enlarged left ventricle with impaired contractility is usually seen. HCM is marked by

Figure 2.3.13  Annual mortality in patients with cardiomyopathy and heart failure. Prevalence of sudden and non-sudden death and 1-year mortality are categorized by the New York Heart Association (NYHA) functional class.
Figure 2.3.14  A. A 12-lead ECG of sustained monomorphic VT in a patient with a remote myocardial infarction and severe ventricular dysfunction. The VT has a left bundle branch block (LBBB) QRS pattern in lead V1, with a predominantly negative QRS in lead I (rightward axis), with an inferiorly directed axis. The LBBB pattern indicated that the VT is originating from either the right ventricle (RV) or the left ventricular septum. The right inferior frontal axis regionalized the site of early activation to a superior location, off the septum. The precordial transition of this LBBB right inferior (LBRI) VT showed significant R waves, except for lead V6, which suggested a basal site-of-origin. B. The corresponding endocardial three-dimensional electroanatomical voltage map of the patient showed a large anterior septal myocardial scar. Purple-colored areas represent normal endocardium (amplitude ≥1.5 mV) with dense scar depicted as red (amplitude <0.5 mV). The border zone (amplitude 0.5–1.5 mV) is defined as areas with the intermediate color gradient.
a thickened septum with dynamic LV outflow tract (LVOT) obstruction. RCM has abnormal contractility and relaxation indices. ARVD/C is marked by regional RV wall motion abnormalities, often localized at the RV apex, RV outflow tract (RVOT), and the lateral basal RV near the tricuspid annulus. Biventricular involvement may also be present with an LV cardiomyopathy and multiple VTs originating from both chambers (Fig. 2.3.15).

**Holter Monitor**

A Holter monitor is utilized to evaluate the overall heart rate profile, the incidence and frequency of ectopy, as well as the morphology of arrhythmias. Patients document their symptoms in a diary that can be correlated to the time stamp on the Holter recording. In patients with frequent symptomatic episodes and a relatively high arrhythmia “burden,” the clinicians

![Figure 2.3.15](image_url)
can use the multichannel Holter monitor to evaluate the frequency of NSVT and VT, and VT morphology.

**Event Monitor**

If a patient experiences symptoms that are infrequent or too short to obtain on a 12-lead ECG, an event monitor is beneficial to correlate symptoms with heart rate and rhythm.

**Cardiac MRI**

Advances in MRI have provided unique abilities to identify morphological changes in the cardiac soft tissues in various cardiomyopathies. Gadolinium-enhanced imaging identifies areas of fibrosis and provides detailed characterization of the potential electroanatomical substrate. The extent and heterogeneity of the myocardial scar increase susceptibility to ventricular arrhythmias in patients with prior MI and LV dysfunction. Cardiac MRI is also used to confirm the diagnosis of ARVD/C, to identify myocardial fibrosis/scar in patients with ischemic and nonischemic DCMs, and to locate cardiac tumors.

**T Wave Alterans (TWA)**

The TWA test measures the microscopic variations of the cardiac repolarization T wave amplitude on a beat-to-beat basis. The standard ECG cannot see the variations or “alterations” in the T wave. The results are computer analyzed, and a positive test indicates the patient has a higher risk for VT and VF. The test measures vulnerability to functional conduction block, which accounts for the breakup of propagating wavefronts and can cause wavelets to disperse and lead to VT or VF.

**Signal-Averaged ECG (SAECG)**

The SAECG uses signal averaging technique to reduce noise of surface ECG to assess ventricular conduction delay and/or slow conduction that manifest as microscopic “late potentials” (LPs) at the end of the QRS. It is often positive in patients with ARVC/D and prior MI. The slow conduction constitutes the substrate for reentry and VT.

**Underlying Coronary Artery Disease**

It is important to assess the possibility of obstructive coronary artery disease. Myocardial ischemia may predispose the patient to subsequent infarction, as well as electrical instability and incessant ventricular tachyarrhythmias (VT storm). Either a noninvasive stress test or coronary artery angiogram is recommended for routine assessment in patients with VT and cardiomyopathy. Ventricular irritability and arrhythmia inducibility can be determined by the electrophysiology study with programmed stimulation, especially for VT based on a reentry mechanism.

After careful analysis of the objective data, diagnostic testing, and laboratory results, the clinician can formulate a strategy and management plan.

**TREATMENT OPTIONS FOR VT ASSOCIATED WITH CARDIOMYOPATHIES**

**Medical Management**

Medical therapy in patients with cardiomyopathy and VT should focus on the underlying structural heart disease and ventricular dysfunction. Treatment for myocardial ischemia and electrolyte abnormalities are paramount, and optimization of heart failure therapy is essential. Evidence-based practice recommendations from the AHA and the American College of Cardiology (ACC) for the routine management of heart failure should include
afterload reduction therapy (angiotensin-converting enzyme inhibitors [ACEIs] or angiotensin II receptor blockers [ARBs]), diuretic, and beta blocker (or a combination). Symptomatic patients may benefit from the addition of digoxin for its contribution to neurohormonal systems and increased heart contractility (Hunt et al. 2009). In addition, the Eplerenone Post-AMI Heart Failure Efficacy and Survival Study (EPHESUS) trial postulated a reduction of sudden death with the use of aldosterone receptor blockers (Pitt et al. 2001). The use of antiarrhythmic drugs (AADs) in patients with ventricular arrhythmias has to be highly individualized. The most commonly used AADs include the Vaughn Williams classification class II and III agents such as beta blockers, amiodarone, sotalol, and dofetilide (Fig. 2.3.16). Beta blockers have shown a statistically significant reduction in mortality by suppressing ventricular ectopy (Teo et al. 1993) (Fig. 2.3.17). Although amiodarone showed benefit in early trials (Doval et al. 1994; Singh et al. 1995) to reduce ventricular ectopy in patients with CHF, its pharmacological side effects were substantial. Subsequent trials demonstrated that amiodarone did not reduce sudden death in patients with ischemic heart disease (Cairns et al. 1997). In the Sudden Cardiac Death in Heart Failure (SCD-HeFT) trial, amiodarone, optimal medical therapy, and a single-lead implantable cardioverter defibrillator (ICD) were compared. Amiodarone showed no survival benefit as compared with placebo; however, ICD therapy reduced the overall mortality by 23% (Bardy et al. 2005).

**Figure 2.3.16** Antiarrhythmic drug actions on cardiac cellular action potential. Cardiac action potential consists of phase 0 rapid upstroke of cellular depolarization, mostly due to rapid sodium (Na⁺) influx. Repolarization is divided into three phases. Phase 1 represents early and rapid repolarization. Phase 2 is known as the plateau phase. Phase 3 represents the final repolarization of the cell. Calcium (Ca²⁺) influx is essential in maintain the plateau membrane potential and potassium (K⁺) efflux is predominately responsible for cellular repolarization. Phase 4 represents spontaneous depolarization with cellular automaticity. Also depicted are the effective (absolute) and relative refractory periods. Antiarrhythmic drug actions: Class I drugs (sodium channel blockers) block the fast Na⁺ channels of the rapid phase 0 depolarization. Class IV drugs (Ca²⁺ channel blockers) inhibit calcium influx of phase 2. Class III drugs (potassium channel blockers) inhibit K⁺ efflux and prevent phase 3 repolarization. The action potential duration and refractory period is prolonged. Class II drugs are beta blockers, which act mostly at phase 4, as well as elevating the ventricular fibrillation.
The decision to use AAD therapy depends on the goals of therapeutic end points, arrhythmia burden, patients’ symptomology, left ventricular function, long-term compliance, and the side effect profiles. If a patient fails antiarrhythmic therapy, nonpharmacological therapy such as defibrillator or VT ablation should be considered.

**ICD**

Multiple clinical trials have demonstrated the benefit of ICDs in primary or secondary prevention of SCD in patients with coronary artery disease. The Multicenter Unsustained Tachycardia Trial (MUSTT) in 1999, Multicenter Automatic Defibrillator Implantation Trial (MADIT-II) in 2002 and the SCD-HeFT in 2004 demonstrated the superiority of ICDs over pharmacological therapy in primary prevention of sudden death in patients with reduced EF. The Defibrillators in Nonischemic Cardiomyopathy Treatment Evaluation (DEFINITE) trial has also demonstrated the benefit of ICDs for primary prevention in non-ischemic cardiomyopathy patients. ICDs have also proven to be effective in preventing sudden death in HCM patients with at least a single risk factor (Maron and Spirito 2008). ARVD/C patients are at risk for SCD and may opt for ICD, but risk stratification and indications are not as well defined (Fontaine & Prost-Squarcioni 2004). Cardiac resynchronization therapy (CRT), also known as biventricular pacing, has reduced mortality in patients with progressive or chronic heart failure. Patients enrolled in clinical trials included NYHA class II–IV heart failure, widened QRS (120–140 ms), and reduced EF of 35% or less. The addition of an ICD to CRT provided additional survival benefit (Abraham et al. 2002; Bristow et al. 2004; Cleland et al. 2005). Despite its life-saving potential, some patients report depression, anxiety, and decreased quality of life after ICD implantation and/or discharge (Thomas et al. 2006). ICD development has progressed to provide
antitachycardia pacing (ATP) for VT safely and to minimize shock therapy by 70% as evidenced by the PainFree Rx I and II (Wathen et al. 2004). Further research is indicated to address the adverse psychological consequences of inappropriate ICD shocks.

**Ablation**

VT ablation is a viable option for patients with VT, especially those refractory to drug therapy. Developed in the past 20 years, VT ablation was initially surgically performed. Although very effective in experienced centers, the operative mortality rate may be as high as 15%. Catheter-based VT ablation is a complex procedure that requires expertise to locate, map, and then successfully ablate the VT using catheters placed in the heart. Sophisticated mapping systems that couple the electrophysiological information with anatomy are utilized. Different techniques are deployed to identify and “map” the origins of arrhythmogenic foci and locations of the reentrant circuits. These include substrate mapping, activation mapping, entrainment mapping, and pace mapping. Radiofrequency heat energy or cool cryoenergy is deployed via catheter application to kill the cells and circuits responsible for initiating the VT (Fig. 2.3.18). Such catheter VT ablation has been shown to be

![Figure 2.3.18](image_url)
effective in eliminating or controlling recurrent VTs. Endocardial ablation involves placing catheters on the inside of the myocardial tissues. Epicardial ablation is defined as having a VT on the outside layer of the heart, and a subxiphoid access for percutaneous puncture is often utilized.

**SUMMARY**

CVD remains the leading cause of death in the United States, with cardiomyopathy prevalence of 1:2,500. There are different forms of cardiomyopathies, and etiology is important for appropriate treatment. Advances in molecular genetics and research continue to grow to identify cellular impairment and treatment possibilities. VT is the most common arrhythmia recorded and a frequent cause of SCD. VT is caused by abnormal impulse formation, abnormal impulse conduction, or a combination of both. Patient management includes AAD therapy and ICD implantation for prevention of SCD. Catheter ablation is available for treatment of VT refractory to medical management and/or in patients with frequent ICD therapies. With the advent of technological advances in catheters, mapping systems, and research, skilled and well-trained electrophysiologists are able to provide endocardial and epicardial ablation as a viable option to patients for modification or elimination of VT.

**Case 2.3.1**

Mr. A is a 64-year-old male who obtained a second opinion for his cardiomyopathy after complaining of increasing shortness of breath. Prior medical history included diagnosed nonischemic cardiomyopathy with an EF of 27%. He underwent a biventricular ICD implantation after documenting VT on a stress test and was treated with amiodarone. Subsequent nuclear imaging and coronary angiogram indicated a 90% stenosis of the distal left circumflex (LCX) and a 50% stenosis of the right coronary artery (RCA) with a fixed inferior basal wall defect. He has a diagnosis of mixed ischemic-nonischemic cardiomyopathy.

He presented to clinic for a routine ICD check that demonstrated multiple episodes of VT due to a fractured RV lead. The patient was admitted for RV lead revision and generator change. The patient returned 1 month later with chest pressure, multiple episodes of VT treated successfully with ATP, and one shock. Restenosis of his RCA was noted, and percutaneous intervention was successfully implemented. Despite revascularization, the patient continued to have VT. Analysis of the 12-lead ECG noted the VT was not from the suspected fixed defect (scar) at the inferior wall (12-lead ECG). He was placed on antiarrhythmic drug therapy and continued with sustained VT at a slower cycle length while hospitalized.

Mr. A was taken to the Catheterization Lab for endocardial VT ablation. Mapping confirmed that the location was suggestive of trigger-mediated idiopathic VT originating near the RVOT and suspected epicardial focus. An epicardial ablation was then attempted, and the focal VT was noted at earliest activation in the anterior septal right ventricular outflow tract region. Postoperatively, the rate of the patient’s VT recurrence decreased significantly, and he has been able to resume his activities of daily living.

**Case 2.3.2**

Mr. J is a 52-year-old male with a history of hyperlipidemia, myocarditis, and subsequent VT. He received a dual-chamber ICD at an outside hospital and presented to the emergency room with VT storm after receiving four shocks from his ICD and needing pacing intervention for five additional VT episodes.
The echocardiogram was normal, previous angiogram noted clean coronary arteries, and the patient was on sotalol.

Mr. J did well for 2 years and then experienced another episode of VT storm, ICD shocks, and hospitalization. He was started on amiodarone. Echocardiography findings revealed a mildly dilated LV, and worsening LV systolic function with apical hypokinesis. The patient returned for a VT ablation in hope of obliterating the ventricular focus and ICD shocks.

Pace and entrainment mapping indicated that the VT had a RBBB morphology left superior (RBLS) axis with a basal site of origin. There was easily inducible monomorphic VT consistent with a scar-based reentry circuit in the basal LV segment, and ablation was performed; however, a suspected epicardial circuit existed. The patient returned for an epicardial ablation. Voltage mapping demonstrated a large posterior lateral and posterior septal scar near the mitral annulus. Two VTs were induced, primarily a RBBI left inferior (RBLI) QRS axis VT at 410 ms cycle length and another RBLS at 350 ms cycle length that degenerated into VF, which required direct current cardioversion (DCCV). Catheter ablation was performed for successful substrate modification of VT.

Case 2.3.3

Mr. R is a 67-year-old male with previous history of coronary artery disease, status post large MI in 1984, without subsequent revascularization procedures. The patient developed ischemic cardiomyopathy, with a left ventricular ejection fraction (LVEF) of approximately 25–35%. He had a single-chamber ICD placement in 1997 for VT. The patient had multiple ICD firings and presented for evaluation of his VT and possible ablation.

His other risk factors include diabetes mellitus type 2, hypertension, dyslipidemia, and former smoker. Myocardial ischemia was ruled out and the patient is currently on sotalol, digoxin, lisinopril, ezetimibe, and a statin for his ischemic cardiomyopathy. The patient underwent endocardial ablation and induced four sustained monomorphic VTs with different cycle lengths. Electroanatomical mapping identified potential reentry circuits within the region of large anterior-apical-septal scar, and linear ablation lines were performed to transect the potential circuits. The patient’s medications were changed to include carvedilol for his cardiomyopathy and coronary artery disease (CAD) and discontinuation of the sotalol.

The patient did well for over 1 year and presented with VT at 150 bpm. He had a successful cardioversion and was referred back for repeat ablation. His angiogram noted a 50% stenosis in his LCX and an 80% stenosis of his distal LAD. His NYHA classification was II and EF remained at 25%. A sustained monomorphic VT with RBBI right inferior (RBRI) QRS axis was induced, consistent with his clinical tachycardia and was successfully ablated. No other VTs were inducible. The patient has subsequently remained free of VT.

REFERENCES

Chapter 2.3 Ventricular Tachycardia Associated with Cardiomyopathies


INTRODUCTION

Although most ventricular tachycardias (VT) occur in the setting of identifiable heart disease, 10% of patients who present with VT have no significant structural heart disease (Lerman et al. 2000). The more common causes of VT in the absence of structural heart disease include right ventricular outflow tract (RVOT) VT, left ventricular outflow tract (LVOT) VT, idiopathic left ventricular tachycardia (ILVT), long QT syndrome (LQTS), and Brugada syndrome.

These arrhythmias can be further classified according to prognosis as either benign or malignant (Table 2.4.1). The VT syndromes that are frequently referred to as “idiopathic,” including the outflow tract VTs and ILVT, carry a more benign prognosis, whereas the inheritable arrhythmia syndromes of long QT and Brugada are considered more malignant, with a strong association with sudden cardiac death (SCD). In this chapter, we will review the general features of VT in structurally normal hearts, and then discuss the management of the common causes in more detail.

GENERAL FEATURES

Clinical Presentation and Diagnosis

Patients that present with VT in the setting of structurally normal hearts tend to be younger than those affected by VT and concomitant heart disease. Although clinical presentation does vary, typical presenting symptoms include palpitations, presyncope, and syncope. In general, cardiac arrest is rare in this population, but can be the primary presentation in patients with a genetic arrhythmia syndrome.

The diagnosis of VT is first suspected when a wide complex tachycardia is detected on any form of electrocardiogram (ECG). Although
Once the diagnosis of VT is made, a thorough evaluation for underlying heart disease follows. Patients with VT in the setting of coronary artery disease, dilated cardiomyopathy, hypertrophic cardiomyopathy, and other forms of structural heart disease have a much higher risk of SCD and have a worse prognosis. The initial evaluation includes echocardiography and stress testing, and can be extended to include cardiac angiography and cardiac magnetic resonance imaging (MRI) if necessary. It is only after structural heart disease has been excluded that the diagnosis of “idiopathic VT” is finally made.

Table 2.4.1 Causes of VT in structurally normal hearts.

<table>
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<tr>
<th>Causes of VT in structurally normal hearts</th>
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<tr>
<td>Idiopathic VTs (benign prognosis)</td>
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<tr>
<td>Outflow tract VTs</td>
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<tr>
<td>RV outflow tract VT</td>
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<tr>
<td>LV outflow tract VT</td>
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<tr>
<td>Idiopathic LV tachycardia*</td>
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<tr>
<td>Genetic arrhythmia syndromes (increased risk of SCD)</td>
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<tr>
<td>Long QT syndrome</td>
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<tr>
<td>Brugada syndrome</td>
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<tr>
<td>Catecholaminergic polymorphic VT (rare)</td>
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<td>Short-coupled torsades de pointes (rare)</td>
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* Also known as verapamil-sensitive fascicular VT.

VT, ventricular tachycardia; RV, right ventricular; LV, left ventricular; SCD, sudden cardiac death.

Wide complex tachycardia, defined as a rhythm with a heart rate >100 bpm and a QRS duration >120 ms, has a number of potential etiologies, VT is the diagnosis 80% of the time (Steinman et al. 1989). A full 12-lead ECG should be immediately obtained before the tachycardia terminates or is treated because the diagnosis of VT frequently hinges on morphological features observed on multiple ECG leads. It is too often the case that only a single-lead ECG strip of the wide complex tachycardia is captured, and the diagnosis of VT cannot be made definitively.

Features on 12-lead ECG not only help distinguish VT from other causes of wide complex tachycardia such as supraventricular tachycardia with aberration (Fig. 2.4.1) but can also help identify the origin of the VT (Brugada et al. 1991). Typically, VT originating from the right ventricle (RV) will produce a left bundle branch block (LBBB) pattern (a predominantly negative complex in lead V1 of the ECG), and conversely VT originating from the left ventricle (LV) will produce a right bundle branch block (RBBB) pattern (a predominantly positive complex in V1).

Figure 2.4.1 Brugada algorithm for diagnosing ventricular tachycardia (VT). At each step, if a criterion is fulfilled, ventricular tachycardia (VT) is diagnosed. If none of the criteria are met, supraventricular tachycardia (SVT) with aberrancy is diagnosed. * Morphologic criteria for VT in RBBB-like tachycardia are monophasic R, QR, or RS in V1, and R/S < 1, monophasic R, QS, or QR in V6; in LBBB-like tachycardia, criteria are R > 30 ms, >60 ms to nadir S, or notched S in V1, and any Q wave, QR, or QS in V6. (Figure based on Brugada et al. 1991.)

Once the diagnosis of VT is made, a thorough evaluation for underlying heart disease follows. Patients with VT in the setting of coronary artery disease, dilated cardiomyopathy, hypertrophic cardiomyopathy, and other forms of structural heart disease have a much higher risk of SCD and have a worse prognosis. The initial evaluation includes echocardiography and stress testing, and can be extended to include cardiac angiography and cardiac magnetic resonance imaging (MRI) if necessary. It is only after structural heart disease has been excluded that the diagnosis of “idiopathic VT” is finally made.
Mechanisms of Idiopathic Ventricular Tachycardia

An understanding of the underlying mechanisms of these arrhythmias is helpful in making a diagnosis and in guiding appropriate therapy. Certain mechanisms are only responsive to particular medications, and the observed response to a medication often points to the etiology. The two major mechanisms of VT in structurally normal hearts are triggered activity and reentry.

Triggered Activity

Triggered activity is an abnormal cell depolarization (electrical activation) that is triggered by oscillations in membrane potential during, or immediately following, normal cell repolarization (electrical resetting). Triggered activity that occurs during the last phase of repolarization is called early afterdepolarizations (EADs), contrasting it to those that occur immediately following repolarization called delayed afterdepolarizations (DADs) (Fig. 2.4.2).

DADs are the cause of the outflow tract VTs. An increase in beta adrenergic activity initiates an increase in cyclic adenosine monophosphate (c-AMP). This in turn leads to an influx of intracellular calcium that triggers the afterdepolarization. Adenosine and beta blockers lead to a decrease in c-AMP, and the nondihydropyridine calcium channel blockers (diltiazem, verapamil) lead to a decrease in intracellular calcium concentration, thereby effectively terminating and treating tachycardias due to this form of triggered activity.

EADs are the cause of VT that occurs in LQTS. In this syndrome, the long QT interval is the result of a genetically determined prolonged repolarization phase. Oscillation in membrane potential can occur during this abnormal phase, resulting in triggered activity. Because an increase in heart rate shortens the QT interval, rapid pacing at heart rates >80–90 bpm usually prevents recurrence of VT in this setting until further treatment can be instituted.

Reentry

Reentry is a repetitive loop of electrical activation that utilizes an abnormal circuit within the heart. Reentry is most encountered when there is structural heart disease leading to scarred regions of myocardium that become the basis for these abnormal circuits. Reentry, however,
Cardiac Arrhythmia Management

Modality, EPS also offers therapeutic benefit through catheter ablation. To evaluate the VT, the arrhythmia is first induced using specific pacing sequences from within the RV. Isoproterenol infusion is sometimes needed to facilitate arrhythmia induction, in fact the outflow tract VTs can spontaneously initiate during isoproterenol infusion. With the patient in a sustained tachycardia, the origin of the tachycardia is located using mapping techniques and technologies. Radiofrequency energy is then delivered from the tip of an ablation catheter, and by heating the tissue to 50–60°C the VT focus or reentry circuit is eliminated.

The ultimate success of radiofrequency ablation in eliminating VT can exceed 95% in RVOT VT and LVOT VT and 80% in ILVT (Stevenson and Soejima 2007). Serious complications associated with VT ablation are rare, but include heart block (<1% occurrence), perforation of the myocardium resulting in pericardial effusion (<1%), and damage to the coronary arteries (<1%). The current American College of Cardiology (ACC)/American Heart

Electrophysiology Study (EPS) and Catheter Ablation

An EPS offers information that is not provided by 12-lead ECG and plays an important role in refining the diagnosis of VT. The site of origin and underlying mechanism of VT are confirmed during EPS by the response of the tachycardia to pacing maneuvers and pharmacological therapy. In addition to being a diagnostic modality, EPS also offers therapeutic benefit through catheter ablation.

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Association (AHA)/European Society of Cardiology (ESC) guidelines for management of ventricular arrhythmias recommend catheter ablation (class I indication) in patients with structurally normal hearts and symptomatic drug-refractory VT arising from the RV or LV, as well as in patients in whom drugs are not tolerated (Zipes et al. 2006).

**OUTFLOW TRACT VT**

**Right Outflow Tract Ventricular Tachycardia**

RVOT VT is the most common cause of VT in the setting of no significant structural heart disease and accounts for up to 80% of all such diagnoses (Lerman et al. 2000). This arrhythmia often affects young, otherwise healthy patients and occurs in women more frequently than in men (Nakagawa et al. 2002). Many patients report that their symptoms are precipitated by exercise or emotional stress. The prognosis is favorable and the risk of sudden death is extremely low.

The characteristic morphology of RVOT VT on a 12-lead ECG is an LBBB pattern in lead V1 with an inferior axis (markedly positive in II, III, and aVF) (Fig. 2.4.4). The mechanism of RVOT VT is c-AMP-mediated triggered activity, which explains the association with exercise and stress. Adenosine decreases c-AMP production and reliably terminates this arrhythmia when administered intravenously.

Treatment options for RVOT VT include medical therapy and catheter ablation. Beta blocker therapy is typically the initial pharmacological treatment of choice, given its relatively high level of efficacy and generally low side effect profile. Beta blocker therapy can be coupled with class IC or class III antiarrhythmic medications for increased efficacy. If medical therapy leads to undesired side effects or if the arrhythmia proves to be refractory to medical therapy, catheter ablation is the treatment of choice in both RVOT and LVOT VT. Ablation is also appropriate as initial therapy if patients do not wish to take medications or if symptoms are debilitating and include presyncope or syncope.

It is important to note that the relatively benign diagnosis of RVOT VT must be differentiated from arrhythmogenic right ventricular dysplasia (ARVD), a condition that is characterized by progressive fibro-fatty displacement of RV free wall and associated with SCD. When ARVD affects the outflow tract, the VT can have morphological characteristics similar to RVOT VT; however, it is not responsive to adenosine. Cardiac MRI should be considered in patients presenting with presumable RVOT VT that is nonresponsive to adenosine, or that demonstrated RV abnormalities on echocardiography, to help evaluate for ARVD.

**Left Outflow Tract Ventricular Tachycardia**

Although RVOT VT accounts for the majority of outflow tract tachycardias, the LVOT can be the site of origin in 10% of cases (Lerman et al. 1997). LVOT VT has morphological characteristics similar to RVOT VT on ECG with an LBBB pattern in V1 and an inferior axis. Whereas in RVOT VT the precordial lead transition from a predominantly negative QRS complex to a positive complex happens between V3 and V4, the “R wave transition” in LVOT VT occurs earlier in lead V2 (Lerman et al. 2000).

The mechanism of LVOT VT is also c-AMP-mediated triggered activity and this arrhythmia therefore responds to intravenous adenosine. Given the similar underlying mechanism, the treatment options for LVOT VT mirror that of RVOT VT, including catheter ablation.

**ILVT**

In contrast to outflow tract tachycardias, ILVT is more common in men than in women and
Figure 2.4.4 A 12-lead electrocardiogram (ECG) of right ventricular outflow tract (RVOT) ventricular tachycardia. Beats 2–4 represent a three-beat salvo of RVOT ventricular tachycardia. The LBBB pattern is evident in V1–V3, with an R wave transition at V4. The prominent R-waves in II, III, and aVF represent an inferior axis.

tends to occur slightly earlier in life, between the ages 15 and 40 (Nakagawa et al. 2002). It was initially described as a VT that occurred during rest but has since been found to also occur with exercise and emotional stress. Similar to RVOT VT and LVOT VT, ILVT carries a favorable prognosis, with a minimal risk of sudden death.

The morphology that is most characteristic of ILVT on 12-lead ECG is an RBBB pattern in lead V1 with a left superior axis; VT from the left posterior fascicle is the most common form of ILVT (Nogami 2002). The underlying mechanism of ILVT has been shown to be reentry, utilizing fascicular and perifascicular tissue as portions of the abnormal circuit (Nakagawa
et al. 1993) (see Fig. 2.4.3). Given the ability of verapamil to slow conduction within perifascicular tissue, this tachycardia can terminate with intravenous verapamil. The treatment options for ILVT include long-term oral verapamil, or catheter ablation, which offers a high probability of definitive cure. During catheter ablation, the left posterior fascicle is targeted for ablation, and by transecting conduction through the fascicle the reentry circuit is eliminated.

**LQTS**

Congenital LQTS is another inherited disorder in patients with structurally normal hearts that is associated with a high incidence of malignant ventricular arrhythmias and SCD. The underlying mechanism is a genetic defect in cardiac ion channels (primarily potassium or sodium channels) that results in an EAD, a premature depolarization before the completion of repolarization, that initiates a characteristic type of polymorphic VT known as torsades de pointes (Fig. 2.4.5). Although numerous genotypes have been identified, LQT1, LQT2, and LQT3 constitute more than 90% of all congenital LQTS diagnosed (Shimizu 2008).

**Diagnosis and Genotypes**

Because the corrected QT (QTc) interval serves as the foundation in diagnosing LQTS, the interval must be measured and calculated carefully. The QT interval is measured from the onset of the QRS complex to the end of the T wave in leads II and V5 or V6, with the longest value serving as the baseline QT interval (Fig. 2.4.6). This interval is then corrected for the shortening effect of faster heart rates on the QT interval by employing the Bazzett’s formula ($\text{QTc} = \frac{\text{QT}}{\sqrt{\text{RR(seconds)}}}$). In general,

![Figure 2.4.5](image1)

**Figure 2.4.5** Episode of torsades de pointes during initiation of dofetilide therapy. Torsades de pointes, or “twisting of the points” in French, is a characteristic polymorphic ventricular tachycardia that occurs in the setting of QT prolongation that has the appearance of a tracing that twists around on a point. In this particular example, excessive QT prolongation occurred during the initiation of dofetilide (class III antiarrhythmic agent) that resulted in sustained tachyarrhythmia.

![Figure 2.4.6](image2)

**Figure 2.4.6** Measurement and calculation of the corrected QT interval. The QT interval is first measured from the onset of the QRS complex to the end of the T wave. The longest value in leads II, V5, or V6 serves as the baseline QT interval. This interval is then entered into the Bazzett’s formula shown at the bottom of the figure. The RR interval is measured (units in seconds) between two consecutive R wave peaks.
a QTc interval length >500 ms constitutes a diagnosis of LQTS, although QTc intervals >440 ms can lead to a diagnosis of LQTS depending on genotype, gender, and presenting symptom.

The three primary genotypes of LQTS are associated with variable arrhythmia triggers. Exercise-induced events, especially during swimming, are common in patients with LQT1. Exercise restrictions are therefore recommended to these patients. In patients with LQT2, startling auditory stimuli can trigger arrhythmias. These patients are advised to avoid certain acoustic stimuli, such as loud telephone ring tones and alarm clocks. Patients with LQT3 tend to have cardiac events during rest or sleep, and behavior modification plays very little role (Goldenberg and Moss 2008).

Many pharmacological agents can cause prolongation in the QT interval leading to “acquired” LQTS. Other causes of acquired LQTS include myocardial ischemia, electrolyte imbalances, and hypothermia. Although patients that present with acquired LQTS may in fact have an underlying genetic abnormality that predisposes them to pronounced prolongation in the QT interval with provocation, the acquired and congenital forms of LQTS are considered distinct. The treatment of acquired LQTS is the removal of the offending agent or reversal of the underlying cause of QT prolongation.

**Management**

Beta blockers have been the mainstay of pharmacological therapy for LQTS, although the effectiveness appears to be genotype specific. Beta blockers are effective in decreasing the sympathetic input to the myocardium and blunting the maximal heart rate achieved during exercise, thus are most effective in preventing cardiac events in patients with LQT1, a genotype where the QT interval does not shorten normally with increasing heart rate. Patients with LQT2 genotypes also benefit from beta blockers, although it does not appear to be as protective as in LQT1. Beta blockers have not been shown to benefit patients with LQT3. Mexilitine, a sodium channel blocker, has demonstrated shortening of the QT interval in patients with LQT3; however, its effectiveness in preventing SCD has not been established (Goldenberg and Moss 2008).

Internal cardioverter defibrillator (ICD) therapy has been shown to be effective in preventing SCD in patients with LQTS. The 2006 ACC/AHA/ESC guidelines for management of ventricular arrhythmias recommend ICD therapy for patients with LQTS who have survived a cardiac arrest (class I indication), and for patients with LQTS who experience syncope or VT while receiving beta blocker therapy (class IIa indication) (Zipes et al. 2006).

Relatives of a patient with LQTS should all be screened for LQTS, starting with ECGs and a detailed survey of symptoms. If genetic testing is able to identify a known defect in the patient with LQTS, family members can be definitively screened by looking for the same defect. There are still genetic defects that lead to LQTS that have not been isolated, but fortunately this number is growing smaller.

**BRUGADA SYNDROME**

Brugada syndrome, first described in 1992 by Joseph and Pedro Brugada (Brugada and Brugada 1992), is a genetic disorder that is associated with a high incidence of SCD. It is currently estimated that the Brugada syndrome is responsible for 4% of all sudden death, and in certain populations, up to 20% of sudden death in patients with no structural heart disease (Antzelevitch 2006). It is more common in men compared with women and is typically diagnosed in adulthood. Brugada syndrome is inherited through autosomal dominant transmission.

Mutations in the gene SCN5A, a gene that encodes for the cardiac sodium channel, have been linked with Brugada syndrome. The defect
in myocardial sodium channels caused by these mutations results in a reduction of sodium inflow currents, which causes a disruption in the usual balance between differing currents that maintain the normal action potentials, especially in the region of the RVOT. The prevailing theory is that this imbalance can generate a reentrant ventricular extrasystole that initiates polymorphic ventricular tachyarrhythmias.

**Diagnosis**

The diagnosis of Brugada syndrome is considered when one of three characteristic ECG patterns (types 1–3) is found (Fig. 2.4.7). The type 1 pattern is the most specific and consists of a coved ST segment elevation ≥2 mm followed by a negative T wave in more than one of leads V1–V3. In the type 2 pattern, the ST segment elevation has a saddleback appearance with an initial elevation of ≥2 mm followed by a trough with ≥1 mm elevation. The type 3 pattern can have either a coved or saddleback ST segment elevation that is <2 mm (Antzelevitch et al. 2005).

It is important to note that these ECG patterns are not always manifest and sometimes only appear when precipitated by provoking factors such as a febrile state, electrolyte imbalances, alcohol and cocaine toxicity, and the introduction of various pharmacological agents including sodium channel blockers and tricyclic antidepressants (Antzelevitch et al. 2005).

Once a Brugada ECG pattern is recognized, the syndrome is diagnosed in conjunction with associated specific conditions. The type 1 pattern coupled with any of the following establishes a diagnosis of Brugada syndrome: syncope, inducible VT on EPS, spontaneous polymorphic VT, ventricular fibrillation, nocturnal agonal respiration, or family history of early SCD prior to the age of 45 years (Antzelevitch et al. 2005). The type 2 and 3 patterns are less specific. Pharmacological testing with drugs that interfere with the cardiac sodium channel, such as flecainide and propranolol, can convert type 2 and 3 patterns to the more diagnostic type 1 pattern.

**Therapy**

Management of Brugada syndrome is guided largely by symptoms. According to the 2006 ACC/AHA/ESC guidelines for management of ventricular arrhythmias, ICD therapy is recommended for Brugada syndrome patients who have survived a cardiac arrest (class I indication) and for Brugada patients who have had a syncopal episode or VT (class IIa indication) (Zipes et al. 2006).
Pharmacological therapy is generally ineffective in treating Brugada syndrome. Quinidine is a class IA antiarrhythmic agent that has significant transient outward current blocking properties that helps reverse the imbalance caused by the SCN5A sodium channel defect, thus normalizing the ST segment and suppressing the initiation of arrhythmia. There is growing clinical evidence that quinidine is effective in preventing cardiac arrest (Antzelevitch 2006). In situations where ICD therapy is not feasible, quinidine can be considered.

Asymptomatic patients with type 1 Brugada pattern on ECG with no family history of SCD should undergo close follow-up and be counseled to report symptoms that may lead to a change in the treatment plan. First-degree relatives of patients with Brugada syndrome should be evaluated with ECGs and survey of symptoms.

CONCLUSION

Ventricular arrhythmias in the absence of significant structural heart disease constitute approximately 10% of all VT occurrences. The majority of these tachyarrhythmias originates from a focus within either the right or left outflow tracts, and has a benign prognosis. They are effectively treated either by antiarrhythmic medications or by catheter ablation. Genetic arrhythmia syndromes such as long QT and Brugada are the cause of a smaller subset of these ventricular tachyarrhythmias. Because of a high risk of sudden death in these syndromes, ICD therapy becomes the treatment of choice in most cases. It is vital that patients with genetic arrhythmia syndromes are recognized early on by their symptoms and characteristic ECG patterns, so that life-saving treatment can be initiated before an episode of cardiac arrest.

Case 2.4.1

A 32-year-old athletic woman with a long-standing history of intermittent palpitations presented with a 2-week history of recurring episodes of dizziness and occasional presyncope. She reported that the episodes were more common when she was working out at the gym. By 24-hour Holter monitoring, she was found to have >12,000 uniform PVCs and repeating salvos of monomorphic tachycardia. A 12-lead ECG of the tachycardia (see Fig. 2.4.4) demonstrated an LBBB pattern with inferior axis, consistent with RVOT VT. She was unable to tolerate metoprolol (extended release 12.5 mg once a day) because of fatigue and severe limitations to her exercise routine. She underwent EPS with catheter ablation. The tachycardia was induced with isoproterenol infusion and the focus was localized to the RVOT utilizing a three-dimensional (3-D) electroanatomical mapping system (see Fig. 2.4.8). A second PVC focus within the RVOT was localized just anterior but separate from the VT focus. Radiofrequency ablation successfully eliminated both foci.

This case illustrates a common presentation of RVOT VT. Because it is a c-AMP-mediated tachycardia, it is frequently associated with exercise and...
easily induced with isoproterenol infusion. Although many antiarrhythmic agents including beta blockers, calcium channel blockers, and sodium channel blockers are effective in treating this tachyarrhythmia, catheter ablation can be offered as first-line therapy. Computerized 3-D anatomical mapping systems utilized during EPS enables precise localization and effective elimination of these foci.

Case 2.4.2
A 47-year-old woman presented with progressive exertional dyspnea over a 1-month period and was found to have congestive heart failure. Echocardiography found her to have newly discovered global LV dysfunction with an ejection fraction of 40%. Coronary angiography found no significant coronary artery disease. On 24-hour Holter monitoring, she was found to have >24,000 uniform PVCs. By 12-lead ECG, these PVCs demonstrated an LBBB pattern with an inferior axis. The R wave transition, however, was at V2, suggesting an LVOT focus. EPS was performed and the PVC focus was localized with 3-D electroanatomical mapping to the LVOT just below the aortic valve. The focus was effectively eliminated with radiofrequency ablation. At follow-up, repeat 24-hour Holter monitor discovered <100 PVCs, and echocardiography confirmed an improvement in the LV ejection fraction to 50–55%.

Outflow tract triggered foci do not always present as ventricular tachyarrhythmias. They often present as frequent PVCs associated with intermittent palpitations. When the density of PVCs exceeds several thousand in a 24-hour period, the dyssynchrony from the PVCs is thought to result in cardiomyopathy. This was a case in which there was no identifiable cause of cardiomyopathy except for her very high density of PVCs. Successful elimination of the PVCs with catheter ablation coincided with a dramatic improvement in LV function.

Case 2.4.3
A 53-year-old man was admitted to the hospital with fevers and productive cough and was diagnosed with pneumonia. His admission ECG demonstrated a type 1 Brugada pattern. As his fever subsided, the ECG normalized. Because he had no prior history of syncope or nocturnal agonal breathing, and no family history of sudden death, no further evaluation or treatment was recommended at this time. He was counseled to report any symptoms suggestive of tachyarrhythmia. Approximately a year later, he presented with unexplained syncope that occurred suddenly without warning. An extensive cardiac evaluation demonstrated no structural heart disease and no evidence of coronary disease. He underwent insertion of an ICD at this point.

This case is typical of how patients are discovered to have Brugada syndrome through a routine ECG. Fever is known to accentuate the ECG abnormality, and was what uncovered the genetic abnormality in this patient. Because he had no symptoms suggestive of tachyarrhythmias and no family history of cardiac arrest, he was appropriately monitored without further therapy at this stage. The subsequent unexplained syncopal event appropriately prompted ICD therapy.

Section 3
Implantable Device Management
Today, there exist cardiac devices that can restore the rate, rhythm, and synchrony of the heart. In 1958, Rune Elmquist, an engineer, and Dr. Åke Senning made history by implanting the first pacemaker in Arne Larsson. It delivered a constant rate of 70–80 bpm (Heart Rhythm Society [HRS] online; www.hrsonline.org/News/ep-history/notable-figures/akesenning.cfm). Made of nickel cadmium and extremely large, approximately 55 mm in diameter and 16 mm thick, the battery could only hold a charge for a short period of time. Once weekly, the batteries were recharged by beaming radio energy through the skin to the pacemaker antenna (Larsson et al. 2003). Made of nickel cadmium and extremely large, approximately 55 mm in diameter and 16 mm thick, the battery could only hold a charge for a short period of time. Once weekly, the batteries were recharged by beaming radio energy through the skin to the pacemaker antenna (Larsson et al. 2003). Pacemakers of today not only provide a heart rate necessary to meet the metabolic demands of the body but also provide diagnostic information, last 7 and 10 years or more, and can detect and pace terminate atrial arrhythmias (Moses et al. 1995; Lloyd et al. 2000; Kenny 2008). Figure 3.1.1 shows how pacemakers have changed through the years.

Implantable cardioverter defibrillators (ICDs) were first developed and implanted in 1980 by Dr. Michel Mirowski (Mirowski 1985). Implantation of these devices took place in the operating room where the lead systems were placed epicardially via thoracotomy. Due to the large size, they were placed in the abdomen. Once the heart rate exceeded a set limit, a high-energy shock was delivered. Significant technological advances in lead systems, defibrillation waveforms, and implantation techniques have allowed for almost all of these devices to now be implanted in the Electrophysiology Laboratory below and parallel to the clavicle without the need for thoracotomy (Hayes 2000; Hesselson 2005; Kenny 2006). Not only do current ICDs terminate ventricular tachycardia (VT) or ventricular fibrillation (VF) with high-energy shocks, but multiple tachycardia zones...
Cardiac resynchronization therapy (CRT) devices were approved in the United States in 2001. Restoration of ventricular synchrony in the presence of heart failure is accomplished by pacing both the right ventricle (RV) and left ventricle (LV). Whereas standard pacemakers typically use a single lead for pacing of the RV, a CRT device uses two leads in the ventricles; one lead in the RV and a second lead placed via the coronary sinus (CS) through a coronary vein to the LV. Although a lateral vein is the ideal site for the CS lead, placement is dependent on the patient’s venous anatomy (Ellenbogen 2004). CRT devices can also provide all the specialized features of pacemakers and ICDs; however, three leads must be taken into account, making them the most complicated devices on the market.

Antibradycardia devices (pacemakers and CRT pacemakers [CRT-P]) and antitachycardia devices (ICDs and CRT-defibrillators [CRT-DS]) are implanted for a variety of reasons (Hayes and Friedman 2000a) (Tables 3.1.1–3.1.3). The American College of Cardiology (ACC) and the American Heart Association (AHA) Task Force on Practice Guidelines in collaboration with the American Association for Thoracic Surgery and Society of Thoracic Surgeons recently revised the ACC/AHA/North American Society of Pacing and Electrophysiology (NASPE) 2002 Guidelines for the Implantation of Cardiac Pacemakers and Antiarrhythmia Devices. Decisions to implant these devices are based on patient symptoms, etiology of rhythm disturbance, comorbidities, and optimal pharmacological therapies. A classification system was devised to help aid physicians in the implantation decision process (Table 3.1.4).

This chapter will provide a basic overview of the common components of pacemakers, ICDs, and CRTs; the fundamental electrical principles for pacing and defibrillation; and the principal means for communicating device functionality.

Artificial cardiac pacing is the result of a combination of electrical, physical, and...
Table 3.1.1  Indications for pacemaker placement.

<table>
<thead>
<tr>
<th>Bradyarrhythmias (intrinsic rate less than 60 beats per minute)</th>
<th>Sick sinus syndrome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Symptomatic bradycardia</td>
<td></td>
</tr>
<tr>
<td>Sinus arrest due to failure of sinus node to produce an electrical impulse</td>
<td></td>
</tr>
<tr>
<td>Sinoatrial node exit block due to the failure of the electrical impulse to conduct normally</td>
<td></td>
</tr>
<tr>
<td>Atrial fibrillation with slow ventricular response (&lt;60 bpm)</td>
<td></td>
</tr>
<tr>
<td>Tachycardia-bradycardia syndrome</td>
<td></td>
</tr>
<tr>
<td>Atrioventricular heart block</td>
<td>First-, second-, or third-degree block</td>
</tr>
</tbody>
</table>

Carotid sinus hypersensitivity

Indications for pacemaker placement usually require the presence of symptoms and documented evidence of arrhythmia. Symptoms include irritability, fatigue, forgetfulness, palpitations, chest pain, dyspnea, weakness, dizziness, presyncope, and syncope.


Table 3.1.2  Indications for internal cardioverter defibrillator placement.

**Primary prevention:**
Patient is at high risk for event due to:

- Ischemic heart disease that are least 40 days post MI or nonischemic cardiomyopathy >3 months
- And NYHA functional class II or III symptoms (NYHA class I = class Ila indication)
- And have reasonable expectation of survival with a good functional status for more than 1 year.
- With a LVEF less than or equal to 40%,
- And are on optimal medical therapy
- Long QT syndrome

**Secondary prevention:**
Patient has a history of:

- Sustained ventricular tachycardia/ventricular fibrillation
- Cardiac arrest
- Unexplained syncope
- Arrhythmogenic RV cardiomyopathy with documented VT/VF
- Brugada syndrome with documented VT, or cardiac arrest, or syncope

*Must have all conditions to qualify.
**Must have any of the indications to qualify.

Five main pathologies for ICD implant: LV dysfunction relating to a prior MI; congenital heart disease; certain metabolic and inflammatory conditions, including myocarditis, rheumatic disease, endocarditis, diabetes, end-stage renal failure; pericardial disease; or valvular heart disease.

MI, myocardial infarction.

biochemical processes. An implantable pulse generator (IPG) is a closed-loop electrical circuit in which an electrical stimulus is generated from the battery and travels to electrodes at the ends of conducting wires (leads) to the muscle of the heart. If this electrical stimulus is successful, the myocardium is “captured,” causing the heart to contract and beat. Electrical information is sent back to the battery, closing the circuit (Lloyd et al. 2000; Barold et al. 2004; Kay and Shepard 2007).

### Table 3.1.3  Indications for cardiac resynchronization therapy.

- Patients with LVEF less than or equal to 35%
- NYHA functional class III or ambulatory class IV symptoms despite recommended, optimal medical therapy
- Have cardiac dyssynchrony, which is currently defined as a QRS duration greater than 120 ms
- Patients with standard ICD indication will have CRT-D placed


### Table 3.1.4  ACC/AHA/NASPE classifications for device placement.

<table>
<thead>
<tr>
<th>Class</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Class I</td>
<td>Evidence and/or/general agreement that device therapy is beneficial, useful, and effective</td>
</tr>
<tr>
<td>Class II</td>
<td>Conflicting evidence and/or divergence of opinion as to the necessity of device therapy</td>
</tr>
<tr>
<td>Class IIa</td>
<td>Evidence is weighed in favor of device therapy</td>
</tr>
<tr>
<td>Class IIb</td>
<td>Usefulness less well established by evidence</td>
</tr>
<tr>
<td>Class III</td>
<td>Agreement that device therapy is unnecessary</td>
</tr>
</tbody>
</table>


### COMPONENTS OF PACEMAKERS, ICDs, AND CRTs

All pacemakers and internal ICDs are composed of four main components: the pulse generator (PG), the leads, the programmer, and the patient (Kenny 2008).

**PG**

The PG can be considered the powerhouse and brain of the pacing system (Moses et al. 1995). Contained within a titanium metal case (also called the can) are the connector(s), the battery, and the circuitry (Fig. 3.1.3). Early pacemakers were very large, had limited functionality, and only lasted a few months (Orestes 1988). Today’s pacemakers are extremely complicated devices capable of storing diagnostic information, retrieving real-time information, and a multiplicity of programming options. Despite the complexity of these components, today’s pacemakers are very small in size and weight, yet they can sometimes last more than 10 years.

Titanium is the most common metal used by various pacemaker companies to make the can that encases the components of the PG (www.medtronic.com/mdlConnectPortal; www.

### Figure 3.1.3  Pulse generator basic components.

(Permission to reproduce this image granted by St. Jude Medical, Inc.)
Chapter 3.1 Basics of Pacing and Defibrillation

For simplicity, Ohm’s law is used to discuss resistance and impedance.

Battery

Many different power sources have been used for pacemakers and defibrillators. Today, most pacemaker batteries are made of lithium iodine (LiI), while most defibrillator batteries are made from lithium vanadium silver pentoxide or silver lithium vanadium oxide (Kenny 2006; Untereker et al. 2007;).

In basic terms, a battery can be thought of as an electron delivery system. All batteries have three basic components: an anode, a cathode, and an electrolyte that helps the conduction of ions and prevents the transfer of electrons. The definitions of anode and cathode are different when referring to the flow of energy in a pacemaker/ICD battery than when referring to the flow of energy through external sources such as the leads and heart tissue. In a battery, electrons flow from the negative pole of the battery (anode), through the leads and the heart tissue, back to the positive end of the battery (cathode) (see Fig. 3.1.4). In a conventional electronic circuit, the portion of the electrode that receives electrons is also called the cathode (negative end), and the portion of the electrode that gives electrons back is the anode (positive end). Electrons flow from negative to positive in the

Electrical Principles

Before one can understand the functioning of pacemaker and ICD systems, an understanding of electrical principles is necessary (Moses et al. 1995; Lloyd et al. 2000; Kay and Shepard 2007). Current (I) is the rate of flow through a conductor and is measured in amperes. When talking about pacing, current is expressed in milliamperes (1 A = 1,000 mA). Voltage (V) is the unit of electrical pressure or force that causes current to flow and is measured in volts. In pacing terms, voltage is often referred to as the amplitude or output used to stimulate the heart muscle. Resistance (R) is the opposition to flow of electrical current and is measured in ohms (Ω). Ohm’s law states that voltage (V) is equal to current (I) times resistance (R), mathematically expressed as $V = IR$. The ohm is the unit of measurement. In order for the voltage to remain constant, as the current increases, the resistance must decrease.

When talking about pacing circuits, impedance and resistance are used interchangeably; however, the two are in fact different. Impedance not only includes resistance but also takes into account all factors that impede the flow of current including lead wire resistance and electrode-tissue resistance (polarization impedance will be discussed later in this chapter under the “Leads” section). The calculations for impedance are detailed and will not be discussed in this chapter.
battery circuit, and from positive to negative in a conventional circuit, but always from anode to cathode (Moses et al. 1995; Barold et al. 2004; Hesselson 2005). Examples of cathodes include the electrode tip. Anodes include the ring electrode on a bipolar lead and the can of a PG when a unipolar lead is used (Fig. 3.1.5). (Bipolar and unipolar leads are discussed later in the “Leads” section.) An electrolyte is a chemical compound that can conduct electric current with ions and not electrons. Examples include intracellular and extracellular fluids in the heart and around the pacemaker and defibrillator leads, as well as the chemical compounds within a battery.

Using the example of a LiI battery, the electrolyte is the solid LiI. The anode is formed from the oxidation reaction in which the lithium (Li) atoms lose electrons (e−) and become Li+ ions (2Li → 2Li+ + 2e−). The electrons are pushed out of the electrolyte of the battery and are sent to the cathode of the electrode, through the heart, to the anode end of the electrode, and back to the battery cathode through a reduction reaction. Iodine combines with the two electrons to yield two iodine ions (I2 + 2e− → 2I−). LiI is continuously formed by the reaction of Li and I (2Li + I2 = 2LiI). With each breakdown and reformulation of LiI in the battery, a barrier (resistance) is built up that makes it more difficult for the Li and the I to combine. This increasing resistance causes a decrease in the battery voltage, Ohm’s law.

The more electrons a battery has to give off, the longer the device will last. The battery in pacemakers and ICDs is not designed to last forever. In fact, pacemaker batteries last, on average, 7 years, and ICD/CRT batteries last an average of 5 years (www.medtronic.com/mdtConnectPortal; www.sjmprofessional.com; www.bostonscientific.com). Longevity is an important characteristic of a device battery. As a general rule, larger devices have a larger battery, which translates to longer longevity. A trend in the device industry has been to make smaller and smaller devices; however, it has been shown that patients actually prefer longevity over size (Wild et al. 2004). Longevity is dependent on three factors (Untereker et al. 2007).

1. The amount of electrical energy it takes to provide the needed therapy to the patient (e.g., pacing, sensing, antitachycardia pacing, and defibrillation).
2. The amount of energy consumed by the electronic circuitry to perform the functions of the device (e.g., operating the microprocessor, electrocardiogram [EGM] storage, telemetry, and charging).
3. The energy capacity of the battery.

Batteries must be able to generate enough voltage to stimulate the myocardium, must not lose power when not in use, should drain at a predictable rate, and be hermetically sealed, meaning that gases from the chemical compounds in the battery cannot leak out (Kenny 2006, 2008).
**Leads**

While pacemaker and ICD batteries today are not designed to last forever or be rechargeable, other components of these devices, such as lead wires, are designed to outlast the PGs. The components of a lead (Fig. 3.1.6) include the lead body, the electrode, fixation mechanism, and the connector (Russo and Marchlinski 2007).

Pacemaker leads have been described as the “weak link” in the pacemaker system (Hayes 2000). The heart beats approximately 35–40 million times per year. Because pacemaker leads must function in the unforgiving environment of the heart, they must be strong, flexible, noncorrosive, highly conductive and resistive, able to maintain good contact with the myocardium, and survive the natural defenses of the body to foreign substances. There are several device factors that contribute to their success or failure: lead design and polarity, electrode design, the electrode tissue interface, fixation mechanisms, and connectors. Patient factors also contribute to the success or failure of leads including the condition of the myocardium (cardiomyopathy, myocardial infarction, fibrosis, etc.), medications, and electrolyte imbalances (Moses et al. 1995; Hayes and Friedman 2000b; Kenny 2006, 2007, 2008; www.medtronic.com/mdtConnectPortal).

**Lead Body**

Essentially, *leads* are insulated conductor wires that deliver the electrical impulses from the battery in the PG to the myocardium. *Conductors* are wires in the middle of the lead that can pass electric current well due to their large numbers of free electrons. *Insulators*, on the other hand, are materials with few free electrons. They pass electric current poorly and cover the conductor wires. Conductors are the wires in the middle of the lead that allows for the passage of electrical current (Moses et al. 1995). Conductors and insulators are imperative for the proper functioning of cardiac pacing and defibrillation leads.

In a pacing lead, the conductor facilitates the flow of electrical current from the PG to the electrode tip. Conductors must be strong with low resistance. There are three basic varieties of conductor design: coils, either unifilar or multifilar; cables; or a combination of coil and cable. Cables allow for a smaller lead body, while coils aid in the passage of the stylet for lead implantation. Design varies from company to company and includes coaxial and multilumen construction (Fig. 3.1.7). In a coaxial design, there is an inner coiled conductor wire that is separated by a layer of insulation, an outer coiled conductor wire, and another layer of insulation. Multilumen designs have coiled and straight conductors running parallel through a single insulator. In addition, each conductor is covered by its own layer of insulation. While most conductors are made from a nickel alloy, the drawn brazen strand (nickel alloy wires brought together with heated silver) is also used (Kenny 2008). Silicone and polyurethane are the two most common insulators used today. Both have advantages and disadvantages. Both have shown to be reliable, biocompatible, and biostable. Silicone can tear easily, but can be repaired with medical adhesive. It has also been around for about 40 years and is very flexible. Polyurethane, on the other hand, is a newer material that has been shown to be
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Additionally, bipolar leads can be programmed to unipolar pace and/or sense. On the other hand, bipolar leads are larger, stiffer, and have a smaller spike on a surface EGM (Barold et al. 2004).

Electrode

An electrode is a conductor located at the distal end of the lead that is in direct contact with the myocardium. This electrode–tissue interface is stronger, tears less easily, but is less flexible (Russo and Marchlinski 2007; Kenny 2008).

Unipolar leads have one internal conductor wire (Fig. 3.1.8), and bipolar leads have two internal conductors (Fig. 3.1.9). Current flows from the PG can (anode) to the tip of the lead (cathode). In a bipolar lead, current flows from the lead tip to the lead ring. Bipolar leads are the most commonly implanted leads for many reasons. There is less influence of extracardiac signals on sensing, pacing, and defibrillation, more lead design options, and there is less electromagnetic interference (Hayes and Friedman 2000b). Additionally, bipolar leads can be programmed to unipolar pace and/or sense. On the other hand, bipolar leads are larger, stiffer, and have a smaller spike on a surface EGM (Barold et al. 2004).
very important for suitable delivery of the electrical impulse that will stimulate contraction of the myocardium (Russo and Marchlinski 2007). The electrode is also responsible for sensing the electrical signals from the heart tissue and relaying this information through the lead to the PG circuitry (Moses et al. 1995). Selection of the metal used in the electrode is important for the long-term function of the lead. Many different materials have been used for electrodes: platinum, platinum-iridium, elgiloy (an alloy of cobalt, chromium, molybdenum, nickel, and manganese), and carbon. An important feature of electrode design is to provide optimal pacing and sensing thresholds. This is best accomplished with an electrode that has a small radius and large surface area (Hynes et al. 1981). Low-impedance electrodes have a large surface area; however, the current flow through the myocardium is not concentrated; whereas high-impedance electrodes have a smaller surface area and current through the myocardium is more concentrated (Barold et al. 2004). These electrodes greatly reduce the current drain from the battery, thus improving longevity. A porous layer on the surface of the electrode allows for increased contact with the myocardium and has helped reduce lead dislodgement, decrease pacing thresholds, and reduce polarization (Russo and Marchlinski 2007).

Polarization is the phenomenon that occurs due to the accumulation of charges of opposite polarity in the myocardium at the electrode-tissue interface. This is created during the pacing stimulus. During each pacing stimulus, ions are produced that accumulate on or near the electrode. For instance, in a bipolar lead, the electrode tip is negatively charged and the electrode ring is positively charged. The positive ions in the electrolyte move to the negatively charged tip, while the negative ions move to the positively charged ring (Barold et al. 2004). Figure 3.1.10 illustrates polarization.

This polarization effect causes impedance to the movement of charge from the electrode to the myocardium (Ohm’s law). With time, the impedance increases, making it necessary for higher voltages to stimulate the myocardium. In between pacing pulses, these polarized (separated) ions stay apart for a brief time and are called afterpotentials. These are important because they can be sensed as intrinsic depolarizations and lead to inhibition of pacing. Smaller

![Figure 3.1.10 Polarization](https://example.com/polarization.png)
leads may be a better choice to prevent lead dislodgment. Active fixation, as the name implies, means that the lead is “actively” physically embedded in the myocardium by a screw that is either fixed or retractable. Figure 3.1.12 shows an active fixation screw in lead. These leads can be positioned anywhere in the atrium or ventricle and are easier to remove if a problem occurs than passive fixation leads.

Both active and passive fixation leads can have steroid elution to minimize the damage to the heart that can occur as the lead is passed through the heart and secured into the myocardium (Ceviz et al. 2000). The steroid slowly dissolves from the tip of the lead, reducing the inflammatory response and resulting in less scar formation. Less inflammation means lower pacing thresholds and longer battery life. Leads that do not have steroid-eluting tips have higher pacing thresholds due to the initial swelling in the myocardium and secondary chronic formation of a fibrous capsule at the lead tip (Ceviz et al. 2000).
Note on Implantation of Leads

Pacemaker, ICD, and CRT leads can be implanted using two approaches: epicardial or transvenous. Epicardial leads are surgically placed directly on the outside of the heart using sutures or other active fixation mechanisms via thoracotomy (Fig. 3.1.13). Transvenous leads are passed through a vein, usually the cephalic or subclavian, into the heart. The passive and active fixation leads in Figures 3.1.11 and 3.1.12 are examples of transvenous leads. Most leads today are placed transvenously in the Electrophysiology Laboratory. Patches are also sometimes used for defibrillation. The high-voltage epicardial patch is secured directly on the outside of the heart (Fig. 3.1.14). A subcutaneous patch or array is implanted under the skin, not the epicardium of the heart, and is an added electrode to a transvenous lead system (www.medtronic.com/mdtConnectPortal).

Differences between Pacing and ICD Leads

Pacing and defibrillation leads share many of the same features. Unlike pacemaker leads, ICD leads are always bipolar to minimize inappropriate oversensing of intracardiac and extracardiac signals, which could lead to inappropriate therapies such as ATP, cardioversion, or high-energy shocks. An ICD lead consists of many more conductor wires separated by an insulation layer. These extra conductors allow the lead to function as it does in a pacemaker system, with the added ability to deliver shocks through a separate pathway or lead coil; however, this also increases their complexity and complication rates. These leads can have a
The most common lead pace/sense lead configuration in CRT devices is LV lead tip to RV ring/coil (Ellenbogen 2004). Some research has shown an increase in anodal stimulation (capture of the RV when only LV capture is intended) with true bipolar leads than with IBP leads (Freedman et al. 2009).

**CS Leads**

CRT devices have three leads, one in the RA, one in the RV, and one in the LV. The LV lead is placed via the CS into a coronary venous branch. Unlike right atrial or ventricular pacing leads, a CS LV lead has no fixation mechanism. Even if a lead with tines is used, there is no trabeculation in these veins, meaning there is no fibrous mesh for the tines to embed themselves in. Innovations in lead size and shape have helped increase the stability of LV lead placement. LV leads, more than RV leads, can cause a problem with diaphragmatic stimulation, especially with more lateral placement of the lead, due to the proximity of the LV lead to the phrenic nerve. Early leads were only unipolar. Newer leads are bipolar and have steroid elution.
Chapter 3.1 Basics of Pacing and Defibrillation

Connector
A connector on the proximal end of the lead attaches to the header of the PG.

Capacitors and Transformers
Capacitors are present in both pacemakers and ICDs. They contain two conductors separated by an insulator and are used to store energy (Hesselson 2005). Similar to pacemakers, PGs in ICDs contain a battery to produce electrical energy and a capacitor to store it, as well as a specialized electronic circuit called a transformer. The transformer in the ICD has voltage multiplying capabilities that allow the low-voltage energy needed for pacing to be changed into the high-voltage energy needed for shock delivery. The voltage multiplying circuit can produce up to 800 V for storage of a large amount of energy in the high-voltage capacitors. High amounts of energy and high voltage are delivered rapidly to the heart at a precise time when a shock is needed to terminate tachyarrhythmias.

The output voltage of the ICD is regulated to supply the amplitude selected (via the programmer) even as the battery energy depletes (up to its end of life). The energy used for both pacing and defibrillation comes from the same battery (Hesselson 2005).

Circuitry
Sophisticated circuitry is what controls the operations of pacemakers and defibrillators (Fig. 3.1.16). These complicated circuits are powered by the battery. Circuit designs may vary by make, model, and manufacturer; however, there are several features that are common to all devices: feedthrough, reed switch, telemetry circuit, oscillator, capacitors, diodes, resistors, and microprocessors (www.medtronic.com/mdtConnectPortal; www.sjmprofessional.com; www.bostonscientific.com).

Since the circuitry is packaged inside the PG and is hermetically sealed, there needs to be a means to convey the signal between the electronics within the case and the lead system. The feedthrough is a small flat metal wire that enables this to happen. Another thin flat metal wire inside the circuit is the reed switch, which is activated when a magnetic field is applied over the device (Kenny 2008). Magnet application causes the reed switch to bend, creating its own electric circuit. Pacemakers will revert to asynchronous pacing at a rate specific to the device manufacturer or the programming. On the other hand, the pacemaker function of an ICD is not affected by a magnet application. Instead, magnet application suspends tachycardia detection and therapy delivery (Hesselson 2005; Kenny 2006).

An antenna inside the PG allows communication from the various circuits to the industry-specific programmer. This telemetry circuit sends a radiofrequency signal that allows diagnostic information (arrhythmic events, therapies, etc.); real-time measured data including battery status, lead impedance, battery impedance, current, pulse amplitude and duration thresholds, and EGMs with markers; and programmed data to be obtained (Moses et al. 1995).
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These features are highly complex and will only be discussed basically in this chapter. Generation of a pacing output or stimulus is an important function of pacing circuitry. The output capacitor charges and discharges to the cathode and anode of the pacing leads. To review Ohm’s law, in a basic pacemaker circuit, the heart provides impedance ($R$) or resistance to this current flow when connected to a battery source. The flow of current causes the heart to be stimulated. The discharged current must be of sufficient strength to cause the myocardium in contact with the electrode to depolarize. The lowest amount of energy needed to “capture” the heart causing depolarization is called the stimulation threshold. A pacing stimulus is derived from two sources: the pulse amplitude and the pulse duration. These are important values for proper programming of the device to deliver the needed energy to pace the heart and for conservation of battery life. Programming is not discussed in this chapter.

The heart can also act as a voltage source causing current to flow. This intrinsic electrical signal is registered by the sensing circuit. If the sensing circuit “sees” intrinsic myocardial electrical activity, it relays this information back to the output circuit. Based on this, the stimulus is either inhibited or delivered. In an ICD, the

![ICD components](image-url)

**Figure 3.1.17** ICD components. (Reproduced with permission from Medtronic, Inc.)
Chapter 3.1 Basics of Pacing and Defibrillation

The components of the ICD are very similar to those of a pacemaker. In fact, both contain a battery to provide energy for pacing, an antenna and reed switch for communication with the programmer, and electronic circuits for control of decision making, therapy delivery, and information storage. These components are housed inside a hermetically sealed case to keep body fluid out. Leads connect to the PG case via the header to deliver electrical impulses to the heart and to relay information back to the electronic circuits. Additional design components in ICD leads and circuitry allow for the storage and delivery of high-voltage shocks needed for defibrillation as well as the low energy needed for pacing.

The decision to implant an antibradycardia device (pacemaker or CRT-P) or an antitachycardia device (ICD or CRT-D) is based on the patient’s symptoms and condition as well as published guidelines. It is a daunting and impractical task to try and communicate the programming of an individual patient’s pacemaker, ICD, CRT-P, or CRT-D. However, the NASPE, now known as the HRS, and the British Pacing and Electrophysiology Group (BPEG) in 1987 developed a universal code that allowed clinicians to convey the basic programming features of antibradycardia and antitachycardia devices (Bernstein et al. 1987). In 1993, the NASPE/BPEG group developed a separate defibrillator code that described in more detail the features of an ICD (Bernstein et al. 1993). In 2002, the group released an updated version of the original 1987 code (Bernstein et al. 2002). Whereas the previous NASPE/BPEG code from 1987 described antibradycardia as well as antitachycardia devices, the 2002 code only describes antibradycardia functions. Like the 1987 code, the 2002 code still contains five positions describing pacemaker behavior. Positions I–III all describe the chamber(s) paced (I), chamber(s) sensed (II), and response to sensing (III). These have not changed since 1987. The last two positions describe rate modulation (IV) and multisite pacing (V). This newly established table is described in Table 3.1.5 and only describes antibradycardia devices, including adaptive rate

sensing circuit also provides input to the defibrillator circuit. This seems like a relatively simple process; however, in reality, this is very complicated (Tse and Lau 2007). The intrinsic signal is sent from the pacing electrodes to the sensing circuit where it is amplified and filtered. This amplified and filtered signal is then compared with the sensitivity that is programmed into the device. Signals that are below the sensitivity setting may be classified as noise, extracardiac myopotentials, or T waves. If the amplitude, measured in millivolts (mV), is greater than or equal to the programmed value, this information is then sent to the timing and logic circuits. A crystal oscillator regulates the components of the timing cycle including the pacing cycle length, refractory periods, blanking periods, and atrioventricular intervals (Kroll and Levine 2007). These signals are also sent to the logic control circuit that operates all the internal clocks, which in turn regulate all the timing cycles of the PG.

Other features that may be included in pacing circuitry are activity sensors, minute ventilation sensors, and alarms. Activity sensors adapt pacing rates on the detection of body movement and muscle motion. The piezoelectric crystal and the accelerometer are two types of activity sensors available today (www.medtronic.com/mdtConnectPortal; www.sjmprofessional.com). The first works by detection of body vibrations. The drawback with this type of sensor is that all vibrations can be detected, not just those within the body. The second detects body motion rather than vibration (www.bostonscientific.com). Almost all pacemakers with rate responsiveness use one of these activity sensors. Minute volume is another type of sensor less commonly used in devices. The cyclical differences in transthoracic impedance caused by respiration are used to estimate minute ventilation. Alarms can also be programmed in to the devices to alert the patient to changes in lead impedance, pacing outputs, battery life, and so on.
Table 3.1.5  The revised NASPE/PBEG generic code for antibradycardia pacing.

<table>
<thead>
<tr>
<th>Position:</th>
<th>I</th>
<th>II</th>
<th>III</th>
<th>IV</th>
<th>V</th>
</tr>
</thead>
<tbody>
<tr>
<td>Category:</td>
<td>Chamber(s) paced</td>
<td>Chamber(s) sensed</td>
<td>Response to sensing</td>
<td>Rate modulation</td>
<td>Multisite pacing</td>
</tr>
<tr>
<td>O = None</td>
<td>O = None</td>
<td>O = None</td>
<td>O = None</td>
<td>O = None</td>
<td>O = None</td>
</tr>
<tr>
<td>A = Atrium</td>
<td>A = Atrium</td>
<td>T = Triggered</td>
<td>R = Rate modulation</td>
<td>A = Atrium</td>
<td>V = Ventricle</td>
</tr>
<tr>
<td>V = Ventricle</td>
<td>V = Ventricle</td>
<td>I = Inhibited</td>
<td></td>
<td></td>
<td>V = Ventricle</td>
</tr>
<tr>
<td>D = Dual (A + V)</td>
<td>D = Dual (A + V)</td>
<td>D = Dual (A + V)</td>
<td></td>
<td></td>
<td>D = Dual (A + V)</td>
</tr>
<tr>
<td>Manufacturers’ designation only:</td>
<td>S = Single (A or V)</td>
<td>S = Single (A or V)</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

and multisite pacing. In 1993, the NASPE put out a policy statement on codes for defibrillation in response to the increased capabilities of ICDs. This code has four positions. Position I describes the chamber(s) where shocking occurs. Position II describes the chamber(s) in which antitachycardia pacing occurs. Position III describes how tachycardia detection occurs either from an EGM alone or with the addition of other features such as transthoracic impedance and blood pressure. The last position, position IV, describes the chamber(s) in which antibradycardia pacing occurs. Both codes would need to be used together to completely describe the basic functioning of a device. For example, a CRT-D device programmed to shock in the RV, antitachycardia pace in the RA and RV; detect tachycardia events from the EGM, with the following antibradycardia features; pace and sense in the RA and RV; respond to sensing in these chambers with triggering and inhibition; with rate modulation on; an additional pacing lead in the LV would be coded DDE-DDDRD. Further examples of coding are described in the 1993 and 2002 versions of the NASPE/BPEG policy statements. While most clinicians do not choose to use the coding for the four positions of the 1993 ICD policy, most do use the first four positions of the 2002 antibradycardia coding.

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INTRODUCTION

Implantation of pacemakers and implantable cardioverter defibrillators (ICDs) have come a long way from the initial pacemaker implant in 1958 by Senning and the first ICD implant by Mirowski in 1980 (Mirowski et al. 1980). The volume of pacemaker and defibrillator implantation has expanded over the last few decades. The increase in pacemaker implantation is largely due to an increase in implantation for sinus node dysfunction, whereas expanding indications for sudden cardiac death prophylaxis account for the expansion in ICD implants (Josephson and Wellens 2004 and Birnie et al. 2006). Furthermore, the introduction of cardiac resynchronization therapy in patients with symptomatic heart failure who have a wide QRS complex has led to a further increase in the use of implantable devices in cardiovascular disease (McAlister et al. 2004). It is estimated that there are 4.5 million individuals worldwide with cardiac rhythm devices, 1.8 million in the United States alone (Hauser 2005 and Borek and Wilkoff 2008).

The implanting team, physicians, nurses, and technicians alike, should be familiar with current guidelines for implanting pacemakers and ICDs. These guidelines are promulgated by the leading professional associations and incorporate the findings of major clinical studies pertaining to the field and the experience of some of the world’s leading experts in this field (Epstein et al. 2008).

THE FACILITY

The critical need for aseptic technique in implanting devices mandates that the implant
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entail long procedure time and high radiation exposure. The high frame rates used in cineangiography exceed the requirements of simple device implantation. The resolution obtained at a frame rate of 7.5 per minute is adequate for purposes of implanting devices. The added capability of the imaging system to store and display contrast venograms of the coronary sinus or of a subclavian vein would assist in guiding a coronary sinus lead into the desired branch, in the case of resynchronization therapy, or assist in accessing an axillary or subclavian vein percutaneously for lead introduction.

In addition to the implanting physician(s), the laboratory should be staffed by nurses with extensive experience managing patients with advanced cardiovascular problems. The section on perioperative complications will demonstrate the need for an experienced nurse for the duration of the implant. Recipients of pacemakers are more likely to be advanced in age, and recipients of ICDs, in general, have extensive cardiovascular disease and may be at risk for serious complications. A technician should be

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**Figure 3.2.1** The operator is scrubbed in the same fashion as one would scrub in the operating room. The patient is draped in a single, fenestrated sterile drape extending from the head to the foot.
available in the room with experience in operating the fluoroscopy equipment, testing the implanted leads, and troubleshooting the electrophysiological monitors.

Once a decision has been made to implant a pacemaker or an ICD, every effort should be directed toward the safe and comfortable implantation of the device. A thorough review of the history, the physical examination, labs, and other pertinent investigations allows the operator and the staff to anticipate the particular needs of the patient scheduled for the implant. From a nursing perspective, the two most important considerations are sedation and the potential development of complications. At the outset, the patient’s identity, the site of the proposed implantation, and the intended device (manufacturer, single-chamber vs. dual-chamber device, pacemaker vs. ICD, and with or without resynchronization therapy) should be confirmed with the operating physician before any sedation is administered.

**DOCUMENTATION OF HARDWARE**

As patients live longer, more patients are coming for device replacement (Hauser 2005). At the time of the device implant, the need to have detailed and accurate records of the leads used at the time of the implant is of paramount importance. Lead/device mismatch would preclude a successful implant and may require a second procedure, raising the risk of developing a pocket infection. For this reason, the implanting team should document the model and serial number of the leads and device being implanted and any additional hardware incorporated such as a subcutaneous patch or an adapter. Documentation should be stored in the patient’s record and device chart, with copies mailed to the manufacturing company to store in their archives with the patient’s name and contact. The manufacturing company should have records of all patients who are recipients of their hardware should the need arise in the future to contact the patients, for example, in the event of a recall or the need for heightened surveillance (Mark et al. 2006).

The patient’s record and device chart should document the access site, pacing and sensing thresholds, as well as pacing impedances. This information will prove invaluable for the duration of the lead’s operation pacing and sensing. A patient who presents for an ICD replacement of a “pectoral” device may have had the device implanted in the subpectoral area. A cursory examination of the ICD implant site may not establish the true location of the ICD and unless the implant report contains that information, accessing the device using the standard surgical approach for a prepectoral device change will lead to a difficult and traumatic explant. In the case of ICDs, the defibrillation threshold (DFT) and the lead polarity at which that threshold was established should be documented. Before ICD defibrillation testing of a replaced device is undertaken, the implant report should be reviewed to anticipate problematic DFT testing. If the proposed implant site is not a standard implant site, for example, subpectoral implantation, the physician should assist in positioning the patient to help identify the anatomical landmarks that invariably become blurred once the patient is positioned and draped.

**INTRAOPERATIVE MONITORING**

Continuous pulse oximetry and noninvasive blood pressure monitoring are mandatory equipment required for patient safety throughout the procedure (see Fig. 3.2.2). Occasionally, patients with severe left ventricular dysfunction undergoing ICD or resynchronization device implantation will require invasive blood pressure monitoring with an arterial line. This would enhance the nurse’s ability to safely administer sedation and analgesia without
Patients who have had a history of venous instrumentation on the side of the proposed implant (preexisting leads in the case of a device upgrade, history of an ipsilateral central line) should have the patency of the ipsilateral vein confirmed in advance of the site’s surgical preparation to determine its patency or lack of. The safety of using iodinated IV contrast should be determined at the outset of the procedure. True anaphylactic reaction to iodinated contrast agent is an absolute contraindication to its use. Patients who describe a variety of cutaneous reactions should be premedicated with corticosteroids, diphenhydramine, and an H2 receptor blocker. In addition, the use of iodinated IV contrast should be weighed against its potential nephrotoxicity in patients with impaired renal function or patients with heart failure and diabetes mellitus who are particularly prone to the nephrotoxicity from these agents (McCullough 2008). N-acetylcysteine and simple IV hydration have been found to help reduce the nephrotoxicity precipitously dropping the patient’s blood pressure. It would also allow the nurse to detect changes in blood pressure early in the event a complication has occurred. The decision to use invasive monitoring should be made in conjunction with the implanting physician before the patient is draped.

**PREPARING THE PATIENT**

All patients should have radiolucent defibrillator patches placed on their thorax and connected to a defibrillator, with a second “backup” defibrillator available in the laboratory in the event one fails. The best location for the posterior patch is over the spine at the level one would expect the upper cardiac border, and anteriorly, at the apex of the heart.

The patient should have two intravenous (IV) lines, one ipsilateral to the implant site to allow the administration of IV contrast if there is difficulty accessing the subclavian vein or if the plan is to use the axillary vein for access.

![Standard equipment next to where the nurse stands at the head of the operating table. These include continuous electrocardiographic monitoring, pulse oximetry, and noninvasive blood pressure monitoring. The defibrillator attached to the chest pads is immediately accessible to the nurse. Given the occasional need for general anesthesia, the equipment they need is immediately available for them.](image)
of iodinated IV contrast. In addition, using the nonionic, iso-osmolar agent iodixanol (Visipaque®; GE Healthcare Inc., Princeton, NJ) helps further reduce the risk of contrast nephropathy (McCullough 2008). The contrast volume used throughout the procedure should be documented to allow the medical team to anticipate the likelihood of developing contrast nephropathy.

Each laboratory should follow the adopted protocol in the sterilization of the surgical field (see Figs. 3.2.3 and 3.2.4). If the standard protocol employs povidone iodine preparation, a substitute such as chlorhexidine (Hibiclens®; Mölnlycke Health Care, Norcross, GA) should be available if the patient describes cutaneous allergy to iodine.

**SEDATION AND ANALGESIA**

In general, device implantation requires sedation and analgesia coupled with local anesthesia. If subpectoral implantation is planned, one should expect the need for higher doses of IV analgesics. The two most commonly used local anesthetics are short-acting lidocaine (Xylocaine®; Hospira, Lake Forest, IL) and long-acting bupivacaine (Marcaine®; Hospira). Care must be taken to determine the total dose administered. Overdose of lidocaine may lead to mental status change and seizures. The total amount of lidocaine administered should not exceed 60 mg (60 mL of the 1% solution).

The systemic agents most commonly used in our laboratory for sedation and analgesia are midazolam and fentanyl; morphine is rarely used. Midazolam (Versed®; Hospira) is a benzodiazepine and is administered slowly, intravenously. While a dose of 1 mg may induce adequate sedation, the dose should not exceed 2.5 mg in individuals younger than 60 years of age and 1.5 mg in individuals older than 60 years. Midazolam should be given over 2 minutes and its effect should be assessed 2 minutes later. The nurse administering midazolam should monitor respiratory parameters and blood pressure, including pulse oximetry. If the patient becomes excessively somnolent and hypopneic...
with hypoxia, the effect of midazolam, or any other benzodiazepine, can be reversed with flumazenil (Romazicon®, Genentech, Inc., South San Francisco, CA). The initial dose of flumazenil is 0.2 mg (2 mL). If by 45 seconds there is no improvement, a further 0.2-mg boluses can be administered for up to a total of 1 mg.

Fentanyl (Sublimaze®; Taylor Pharmaceuticals, Decatur, IL) is a short-acting opioid analgesic that is given at a dose of 0.05–0.1 mg over 1–2 minutes. It can cause respiratory depression and apnea. Respiratory parameters should be monitored closely and resuscitative equipment available to provide manual ventilation. The opioid antagonist naloxone (Narcan®; Endo Pharmaceuticals Inc., Chadds Ford, PA) is used to reverse the effect of fentanyl; the starting dose is 0.4–2 mg given in 2–3-minute intervals to a maximum of 10 mg.

Dexmedetomidine (Precedex®; Hospira) is a powerful sedative. It is a relatively selective alpha-2 adrenoreceptor agonist, which explains its most serious side effects: hypotension and bradycardia. Patients also require intensive observation of respiratory status. National accreditation organizations such as the Joint Commission have set up training standards that need to be fulfilled by the staff administering conscious sedation to promote the safe administration of these agents. Electrophysiology laboratories involved in the implantation of these devices with the administration of conscious sedation should adopt written training protocols using the Joint Commission criteria.

**PERIOPERATIVE ANTIBIOTICS**

One of the most feared complications of device implantation is device infection, with an overall risk of 0.8–1.5% (Lai and Fontecchio 1998; Wilkoff 2007). Observing strict aseptic and meticulous surgical technique is the best and most effective intervention to minimize the risk of device infections (Borer et al. 2004). Some studies have shown that preoperative fever, the presence of a temporary pacing wire, and reinsertion raise the risk of device infection,
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The most common site for implanting a device, pacemaker or ICD, is the infraclavicular area anterior to the pectoralis major muscle. In our practice, we select the site contralateral to the dominant arm and after discussing the merits of one site over the other with the patient. In individuals with sparse chest wall soft tissue (muscle and fat), patients with recurrent device erosion, individuals with small body frame where there is a greater risk of future device erosion, or for cosmetic reasons, the implanting physician may elect to implant the device, usually an ICD, in the subpectoral area. The nurse and technician staffing the laboratory should know in advance since this approach may require different patient positioning. In addition, pain management requirements both during and after the procedure are greater when the subpectoral area is selected rather than the standard prepectoral implants. Since the axilla will be exposed to the surgical field, identifying a subset of patients at higher risk of developing this dreaded complication (Klug et al. 2007). The use of preoperative antibiotics has been demonstrated to reduce the risk of device infection (Da Costa et al. 1998). The preoperative administration of prophylactic antibiotics is directed toward reducing the risk of infection and targets the most common responsible pathogens: *Staphylococcus aureus* and *Staphylococcus epidermidis* (Borek and Wilkoff 2008). The most frequently used agent is cefazolin 1–2g intravenously 30 minutes to 1 hour prior to the procedure and may be continued for 3–5 days postoperatively (Bertaglia et al. 2006). In patients allergic to penicillin or cephalosporins, or if there is concern that prevailing skin organisms have become methicillin resistant, such as patients with extended hospital stay, vancomycin can be used. Antibiotic irrigation is an additional adjunctive measure used to further help reduce the risk of an infection. While widely adopted, the efficacy of this measure remains unproven.

**POCKET SITE**

The most common site for implanting a device, pacemaker or ICD, is the infraclavicular area anterior to the pectoralis major muscle. In our practice, we select the site contralateral to the dominant arm and after discussing the merits of one site over the other with the patient. In individuals with sparse chest wall soft tissue (muscle and fat), patients with recurrent device erosion, individuals with small body frame where there is a greater risk of future device erosion, or for cosmetic reasons, the implanting physician may elect to implant the device, usually an ICD, in the subpectoral area. The nurse and technician staffing the laboratory should know in advance since this approach may require different patient positioning. In addition, pain management requirements both during and after the procedure are greater when the subpectoral area is selected rather than the standard prepectoral implants. Since the axilla will be exposed to the surgical field,
particular care should be directed toward its preoperative surgical preparation.

When the pocket is being fashioned, at the onset of the case or after the leads have been implanted, the nurse should be notified. This step may cause discomfort, and the patient should be alerted and a supplemental dose of local anesthesia as well as systemic sedation/analgesia may need to be administered.

**VENOUS ACCESS**

Pacemaker implantation requires the introduction of pacing lead(s), and until subcutaneous ICDs become available, ICD implantation requires the introduction of a high-voltage lead into the right ventricular cavity via a venous structure. The cephalic vein courses between the deltoid muscle and pectoralis major muscle in the deltopectoral groove. Accessing the cephalic vein requires some expertise in dissection and performing the venous cut down but offers the opportunity of introducing lead(s) without having to access a more central vein with its attendant risks—pneumothorax, hemothorax, and lead crush in the event the subclavian vein is used. If the cephalic vein cannot be identified, iodinated contrast injection from the ipsilateral arm may help identify this vein. Otherwise, the subclavian or axillary vein can be used for venous access.

Once the subclavian or axillary vein has been accessed, the implanting physician should alert the staff. There is a higher risk of inadvertently puncturing the ipsilateral lung or the axillary artery, resulting in a pneumothorax or a hemothorax, respectively. A sudden drop in pulse oximetry or in blood pressure while attempting to access the vein or immediately thereafter should alert the implanting team to the possibility that the patient may have developed a tension pneumothorax, an emergency that requires the immediate decompression of the pleural space. Fluoroscopy of the ipsilateral lung field would help exclude this possibility.

Most instances of pneumothorax are small in volume but may increase in size with time. If a central venous access is used, the patient should have a portable chest X-ray (CXR) immediately after the procedure and a more formal posteroanterior (PA) and lateral view on the following day looking for a pneumothorax and confirming lead location.

Any unsuccessful attempt at accessing a central vein should be followed by adequate time to determine if a slow-leaking pneumothorax had developed before any attempt is undertaken to access a central line on the contralateral side. The development of a pneumothorax on both sides would have catastrophic consequences. Many operators prefer to abandon the procedure in favor of bringing the patient back the next day. The subclavian venous approach carries the added risk of compressing the leads between the clavicle and first rib, a condition known as subclavian crush syndrome (Belott and Reynolds 2007; Borek and Wilkoff 2008).

**LEAD INTRODUCTION AND POSITIONING**

Once a vein has been accessed, a retained wire is introduced and secured. A poorly secured wire may dislodge and migrate into the vascular space. The retained wire is used to introduce a peel-away sheath through which the lead is inserted unless the operator is familiar with direct cannulation of the lead into the cephalic vein. The insertion of a nonhemostatic sheath carries the risk of introducing air into the vascular space during respiratory inspiration when the intrathoracic pressure becomes negative. Air embolism may lead to hypoxia, hypotension, and even loss of consciousness. The air bolus can be seen fluoroscopically wedged below to the pulmonary vein. If hemodynamically significant, the air bolus may be aspirated using a fluoroscopically guided Swan-Ganz catheter. The patient who has been kept fasting
overnight would be more prone to this complication, and maintaining adequate IV hydration before and during device implantation may help reduce the risk of this complication. Steps that can be taken, in addition to IV hydration, to help reduce this complication include placing the patient in Trendelenberg after the sheath is introduced, placing a wedge beneath the patient’s legs to increase thoracic venous return, asking the patient to take shallow respirations during lead insertion, and using sheaths with hemostatic valves.

The ventricular lead is then introduced under fluoroscopic guidance using the softest stylet to support the lead. Once the lead enters the cavity of the right atrium, the stylet is withdrawn a few inches and the lead is prolapsed across the tricuspid valve. The operator should be aware that if the patient has a preexisting left bundle branch block, advancing the lead across the valve might result in trauma to the right bundle branch, leading to complete heart block. The preoperative review of the data by the physician, nurse, and technician staffing the laboratory would alert the team to this potential risk at the outset of the case. The nurse and technician in the operating room should be notified when the operator plans to prolapse the lead across the tricuspid valve. The operating physician may elect to connect the pacing wires to the lead, usually in a unipolar mode, before advancing the lead, while the technician is prepared to initiate pacing in an asynchronous mode should the need arise.

Lead entry into the right atrium and ventricle is often associated with some premature atrial and ventricular contractions, respectively. Rarely, in patients with depressed ventricular function, nonsustained or even sustained ventricular tachyarrhythmias may ensue from the manipulation of the lead in the right ventricle. The absence of premature ventricular contractions with advancement of the right ventricular lead should raise suspicion that the lead may have entered into the coronary sinus. This may not be appreciated on a standard anteroposterior (AP) fluoroscopic view. Using the left anterior oblique (LAO) view should help confirm or exclude this possibility. The ventricular lead may inadvertently cross a patent foramen ovale or an unrecognized atrial septal defect. If the lead crosses either one of these two defects, it will end up in the left ventricle with catastrophic future consequences (systemic thromboembolism). This may be missed if the operator relies solely on the fluoroscopic AP projection or is unaware of this risk. Fluoroscopy in the LAO or lateral projection would help by confirming the anterior location of the ventricular lead (retrosternal) when the lead is in the right ventricular cavity. Coronary sinus or a left ventricular location of the lead would place it posteriorly in the lateral projection. When the ventricular lead is in the right ventricular cavity, the paced ventricular complex (and the premature ventricular beats) should have a left bundle-branch block-like morphology on the physiological monitor (precordial lead V1). Another way for the operator to reduce the risk of inadvertent left ventricular insertion is by advancing the lead across the tricuspid valve and into the right ventricular outflow, thus excluding inadvertent left heart entry, before slowly sliding the lead back until the lead tip reaches the desired location against the right ventricular endocardium.

Lead tips are secured in the desired chamber once a location with favorable pacing and sensing properties is found. There are two types of fixation mechanisms: passive and active. Passive fixation employs a set of tines attached to the lead tip that hook onto surrounding trabeculae. An active fixation mechanism employs a distal screw that is deployed once the desired location is found. Leads with passive fixation mechanisms are less amenable to manipulation and are only stable in the apex of the right ventricle. The fibrous reaction surrounding the tines over time makes the lead tip more stable but makes any future attempt to extract the lead more difficult than in leads with an active fixation system. Active fixation mechanisms...
leads are easier to manipulate in the right ventricle and offer a wider choice of lead tip positioning particularly if there is a preference to implant the lead tip at the base of the septum distal to the bundle of His. Active fixation leads are easier to extract in the future if such a need were to arise. When using an active fixation mechanism, deployment in the apex may increase the likelihood of perforation or inadvertent diaphragmatic stimulation (Aizawa et al. 2001 and Geyfman et al. 2007). In our practice, once the lead is secured at the site where pacing and sensing properties are favorable; pacing is initiated at maximum output to confirm the lack of diaphragmatic pacing.

Once the lead tip has been secured (actively or passively), the pacing and sensing properties are redetermined before the proximal portion of the lead is secured to the underlying pectoral muscle/fascia using the securing sleeves and nonabsorbable suture. A pacing system analyzer is used to determine the pacing and sensing thresholds during lead mapping. Once the active lead tip is deployed, impedances are measured. These indices are measured again after the stylet is withdrawn and the lead is anchored to the underlying muscle using the anchoring sleeve.

Given the anatomy of the right atrium, right atrial leads are usually active fixation leads. Preformed stylets allow adequate lead tip localization, usually in the right atrial appendage. Particular care should be observed in making sure the lead is secure as dislodgement of the lead may result in lead prolapse into the right ventricle, precipitating ventricular arrhythmias. The unexplained emergence of premature atrial or ventricular contractions after the right atrial lead has been “secured” should alert the operator to the possible development of lead dislodgement. The deployment of the distal fixation screw in the right ventricle or right atrium should be observed fluoroscopically using a magnified view. The operator should be familiar with the fluoroscopic appearance of a deployed lead tip screw.

Once the leads are secured, and lead testing confirms favorable pacing and sensing properties, the device is handed to the implanting physician. The lead pins are inserted into the device header, carefully making sure that the pins are clean of any blood. The operator should confirm that the lead pins are beyond the set screws before the screws are tightened. The device is interrogated and if there is any change in pacing properties, the connections between the lead and the header should be reevaluated.

Improperly set connections can be a cause of failure to pace or sense early after implantation. In ICD implantation, the inadvertent insertion of one high-voltage pin into the contralateral receptacle is one of the causes of high DFTs. Loose set screws can lead to inappropriate detection and shock.

DFT TESTING

The defibrillator is designed to terminate lethal arrhythmias. Ventricular fibrillation (VF) is terminated by the delivery of a shock, and ventricular tachycardia by either delivering a shock or a burst of antitachycardia pacing (ATP). The energy required by the ICD to successfully terminate VF can vary according to the prevailing sympathetic tone, degree of cardiac compensation, serum electrolytes, and circulating drug levels. Hence, to safely implant an ICD, the operator must verify that an adequate safety margin exists between the minimum shock that would reliably terminate VF, the DFT, and the maximum energy the device is capable of delivering. In the current age of pectoral, biphasic ICDs, the need for DFT testing has come into question (Birnie et al. 2008; Blatt et al. 2008, and Viskin and Rosso 2008). In our practice, we require a 10-J safety margin between the DFT and the first programmed shock. Determining the DFT at the time of the implant allows the operator to make changes in the lead positioning to improve the reliability of the shock delivered or change the shock lead polarity, if that
feature is not available noninvasively. Lowering the DFT not only enhances the reliability of the ICD but also allows the programming of lower energy shocks, which, through shorter charge times, expedites the delivery of therapy. This would reduce the likelihood the patient will lose consciousness from the ventricular tachyarhythmia, especially as the battery begins to age and the charge time begins to increase.

Different protocols exist to help determine the DFT; the most common methods entail the induction of VF. There is some risk inherent in the induction of VF, especially if done repetitively. Another method of determining the DFT is by determining the upper limit of vulnerability (ULV) (Swedlow et al. 1996, 2007). A commonly employed method of inducing VF is by delivering a low-energy shock on the T wave. As higher shocks are delivered, VF ceases to be induced. This defines the ULV, which correlates with the DFT and does not require the induction of VF (Swedlow et al. 1996, 2007).

DFT testing requires deeper anesthesia. The staff should be prepared for the possibility that the induced VF may not terminate with the shocks delivered by the ICD, thereby requiring external defibrillation. In our laboratory, the implanting physician will discuss the sequence of steps to be undertaken in the event a delivered shock fails. This may entail having the ICD deliver its first two or three shocks, at increasing levels of strength, before delivering an external “bailout” shock at the device’s maximum output. The polarity of the last programmed ICD shock is reversed to enhance efficacy in the event of failure of earlier shocks. The nurse will charge the external defibrillator after the first internal shock fails to terminate VF to reduce the time until rescue external defibrillation is delivered. Failure to terminate VF with internal or even external shocks should alert the operator to exclude a pneumothorax as cause of the high DFT (Kroll and Tchou 2007). Each laboratory should have a protocol for the management of VF that has been induced but cannot be terminated.

**IMPLANTATION OF RESYNCHRONIZATION DEVICES**

Approximately a third of patients in the New York Heart Association (NYHA) class III or IV heart failure have a wide QRS (>120 ms) (Hunt et al. 2001). These patients have more pronounced heart failure symptoms than similar patients with narrow QRS width. This has been attributed to interventricular and intraventricular dyssynchrony. Most of these patients have left bundle branch block, and the introduction of a “left ventricular” lead into a lateral branch of the coronary sinus has been found to improve their subjective symptoms, objective performance, reduce hospitalization, and reverse remodeling of the left ventricle (McAlister et al. 2004). These leads are most frequently implanted in conjunction with an ICD.

A coronary sinus lead is introduced via an additional venous access site, increasing the likelihood of complications such as pneumothorax. The introduction of a lead into the coronary sinus and finding an appropriate branch in the lateral wall of the left ventricle can be challenging at multiple levels. The coronary sinus and its branches are subject to multiple anatomical variations. Left ventricular leads do not have the reliable anchoring mechanisms available to endocardial leads. The team must have a plan in place in advance in the event accessing the coronary sinus, using the device manufacturer’s sheaths, proves challenging. This may entail using multipolar, preformed, or steerable catheters, rarely requiring femoral venous access to identify the os of the coronary sinus. Identifying and navigating the coronary sinus and its branches often requires the use of IV radiographic contrast agents. The patient’s renal function and the absence of allergy to iodinated contrast material should be determined in advance, and a record of the total amount of contrast
delivered should be kept to help anticipate any nephrotoxicity.

Once a favorable spot for pacing in a branch of the coronary sinus is found, inadvertent diaphragmatic pacing should be excluded both at baseline and at high pacing output. The proximity of the phrenic nerve to the epicardial branches of the coronary sinus may lead to its inadvertent stimulation leading to diaphragmatic contractions. The operator can see diaphragmatic stimulation fluoroscopically. Compared with implanting a pacemaker or an ICD, implanting a resynchronization device requires more time and entails slightly greater blood loss, and the average patient has more advanced cardiac disease with a greater risk of a complication.

**DEVICE REPLACEMENT**

As the indications for device implantation increase and the mortality of patients with cardiovascular disease decrease, the number of patients with cardiac rhythm devices who need device replacement will increase (Hauser 2005; Birnie et al. 2006). A less common reason for device replacement is device recalls due to component failure. The replacement of a device entails more preparation in advance of the procedure than the procedure itself or the preparation for a fresh implant. Determining the indications for the original implantation, the types of lead(s) used, DFTs at the time of the original implant in the case of an ICD, and the need for a new lead should all be determined in advance. If there is a problem with one or more preexisting lead, this is the time to try to fix it or replace it. If a new lead implantation is anticipated, the patency of the ipsilateral vein should be determined in advance of the patient being prepped and draped. The most serious long-term consequence of device replacement is the higher risk of a pocket infection (Wilkoff 2007). No effort should be spared in reducing the likelihood of an infection.

**POSTOPERATIVE CARE**

The introduction of active fixation leads has loosened the once strict bed confinement recipients of devices were once subjected to. However, given that coronary sinus leads used in resynchronization therapy have no mechanism of securing the lead tip, recipients of coronary sinus leads and recipients of pacemakers for severe bradycardia and syncope are subjected to the 24-hour bed rest rule. Otherwise, patients are on strict bed rest for 6 hours and may use the bathroom after that but only with assistance. Confining patients to bed rest for periods longer than 6 hours would require using a Foley catheter, particularly in male patients who cannot use bedpans, or compel them to surreptitiously go to the bathroom and risk falling. A Foley catheter may promote a nosocomial urinary tract infection, and a fall may expose the lead to dislodgement, the risk of either event outweighs any potential benefit bed confinement may offer.

The use of an arm sling is controversial. While routinely used in our institution until the day following the procedure, arguments have been made against their routine use. Artificially restricting arm movement to reduce the likelihood of dislodging a lead will give the implanting physician the false sense of lead stability (Belott and Reynolds 2007). Had the patient had the opportunity to fully deploy his or her shoulder, a poorly anchored lead tip would have dislodged, uncovering lack of lead stability and prompting early intervention. There is also fear that a sling may promote a syndrome of “frozen shoulder,” which can be painful (Belott and Reynolds 2007).

On the following day, patients will have an upright, PA, and lateral CXR along with a baseline electrocardiogram, and in the case of a pacemaker, after the application of a magnet. These maneuvers help confirm adequate lead sensing and pacing. ICDs are deactivated by magnets and a magnet electrocardiogram is not performed. At our institution, we conduct a
more formal, bedside evaluation of the implanted leads.

Once discharged, daily wound inspection and dressing is carried out either by the patient, a visiting nurse, or a family member. One week later, the wound is assessed in the pacemaker clinic. If the wound is healing well, the patient is allowed to shower. Patients are allowed to drive 2 weeks after the implantation if there is no other reason barring them from driving. In the case of a left pectoral implant, the seatbelt will come across the device, patients are advised to fold a towel and place it between the seatbelt and the implant site for the first 6 weeks. Oral antibiotics are continued for 3 days post implant. The amount of time before showering, driving, and the total antibiotic time can vary between institutions.

**COMPLICATIONS**

Device-related complications could be divided into perioperative, immediate postoperative, and late complications. It has been established that physicians and implant centers with higher volumes have lower complication rates (Eberhardt et al. 2005).

**Perioperative Complications**

Hypotension is not uncommonly encountered during device implantation and causes can vary from causes as simple as dehydration or excessive sedation to causes as dangerous as cardiac tamponade. These are complications that are real and every laboratory should have a contingency plan available to manage these complications. Failure to do so can convert a potentially reversible complication into a lethal one.

Cardiac tamponade, the rapid accumulation of blood in the pericardial space during lead manipulation, would have lethal consequences if not addressed immediately. Risk factors for perforation include the presence of a temporary pacing wire, the use of active fixation leads, and the concomitant use of steroids, while a right ventricular (RV) systolic pressure of ≥35 mm Hg have been found to be associated with a lower incidence of this complication (Mahapatra et al. 2005; Geyfman et al. 2007). The first clue to a tamponade is a sudden drop in blood pressure; the drop does not have to be dramatic. It is usually one that does not respond or responds transiently to the administration of IV fluids or IV inotropic agents such as phenylephrine (Neo-Synephrine®; Hospira) 0.1–0.5 mg. Other clues include the preceding development of chest discomfort that may be pleuritic in character. Significant changes in pacing or sensing characteristics of an implanted lead may further raise suspicion of cardiac perforation. A sudden and rapid drop in blood pressure should be considered cardiac tamponade until proven otherwise. Every lab should be equipped with several pericardiocentesis kits to avert any delay in relieving the raised intrapericardial pressure preventing right ventricular filling. The diagnosis can only be confirmed by echocardiography and some implant laboratories carry portable echocardiography machines that can be used to urgently confirm the presence of fluid collection around the heart and can also help safely guide the insertion of a pericardial drain. While most tamponades are managed by percutaneous pericardial catheter drainage, if bleeding does not stop, some patients will need to have the perforation corrected with heart surgery.

More commonly, hypotension is due to the combined effect of dehydration and sedation. The temporal relationship with the administration of sedation or IV analgesia establishes the cause of hypotension and usually responds to the administration of IV fluids, IV phylephrine, and if sedation/analgesia are suspected to be the cause; flumazenil 0.2–3 mg for benzodiazepines, naloxone 0.4–2 mg for opioid analgesics. Dexmedetomidine (Precedex) is more likely to cause hypotension and bradycardia than other agents used for sedation or analgesia; there are no antidotes for this agent. In the event hypotension or bradycardia occur and are attributed to this agent, IV fluids and pressor agents are used.
for hypotension, and atropine and glycopyrrole can be used for bradyarrhythmias.

The advantage of the cephalic vein cut down for venous access is the elimination of the risk of pneumothorax. The percutaneous subclavian vein approach is associated with a higher risk of pneumothorax and hemothorax. Pneumothorax is seen in up to 2% of patients in whom the subclavian approach is used. Advanced age and lower body weight appear to portend a higher risk for this complication (Birnie et al. 2006). In our laboratory, we will often employ a long, thin “finder” needle to access the subclavian vein before using the smallest gauge needle that would allow the introduction of a retaining wire (Micro Puncture®; Bard Access Systems, Inc., Salt Lake City, UT) to help minimize the likelihood of an accidental pulmonary perforation or minimize its consequences if it does occur. Patients whose implant required a subclavian approach should have a portable CXR to exclude having developed a pneumothorax shortly after the patient leaves the laboratory. This should be followed by a formal PA and lateral CXR the following day. A slow-growing pneumothorax may only be detected the following day. Tension pneumothorax is a true emergency and is fortunately very rare. The cardinal findings are loss of lung markings in the ipsilateral hemithorax, in addition to a mediastinal shift to the contralateral side and drop in blood pressure. The pleural space should immediately be decompressed percutaneously. To avoid developing a bilateral pneumothorax, the failure to access a subclavian vein on one side should only be followed by an attempt to access the contralateral vein after a sufficient amount of time has passed to confirm that a slow-growing pneumothorax had not developed in the interim.

Immediately Postoperative
Pocket hematomas occur in up to 5% of patients undergoing device implant. The use of postoperative heparin, and therapy with combined aspirin and clopidogrel appear to portend a higher risk of this complication (Wiegand et al. 2004). While some studies have not associated the development of hematomas to pocket infections, other studies have found that early re-intervention carries a higher incidence of late (12 months) pocket infection (Klug et al. 2007).

The incidence of lead dislodgement has decreased significantly since the introduction and the widespread use of active fixation leads. Nonetheless, a CXR the day following the procedure should be done to confirm proper lead placement and would also help in the future to determine if any changes in lead location had taken place. It also helps exclude inadvertent coronary sinus or left ventricular lead placement if the operator relies only on an AP view during implants. The application of a magnet to a pacemaker converts the pacemaker from a sensed to a nonsensed mode. This helps confirm appropriate chamber pacing in nonpacemaker-dependent patients. An electrocardiogram without and with the application of a magnet helps confirm adequate sensing and pacing, functions that would be altered by lead dislodgement or perforation. If the implanted device is an ICD, the application of a magnet would suspend therapy for tachyarrhythmias and is not done as part of the routine, postimplant follow-up. A more formal interrogation with a programmer is necessary to confirm adequate device pacing and sensing. The need to re-intervene to reposition the lead results in a higher risk of pocket infection (Klug et al. 2007).

The complications of device implantation are not limited to the above. Venous occlusion/thrombosis, device/lead infection, lead/device erosion, lead fracture, and loss of insulation, while they may occur soon after the implant, are usually long-term complications. The discussion of these complications is beyond the scope of this chapter.

CONCLUSIONS
The successful implantation of a cardiac rhythm device—a pacemaker, an ICD, or a resynchro-
nization therapy device—entails multiple steps. The success of the procedure is dependent on anticipating these steps, enhancing the likelihood of a successful implant while minimizing any potential complication associated with each one of these steps. The process begins with understanding the indication for the implantation and planning the procedure with the patient’s unique clinical characteristics in mind. The procedure can only succeed if all members of the implant team understand what the procedure entails and what the expectation of each member of the team is.

**Case 3.2.1**

A 78-year-old male patient is admitted to the hospital after a witnessed syncopal episode while having dinner with his family. He was seen to suddenly slump, his head striking the table. His daughter, an internist, immediately tried to feel his pulse but was unable to palpate any pulse. He regained consciousness within seconds and, apart from the pain of striking his head, denied any symptoms. This was the third such episode in 3 months. His electrocardiogram showed sinus rhythm, a borderline prolonged PR interval, and a complete left bundle branch block with a QRS width of 160 ms. His echocardiogram was unremarkable, and an electrophysiological study was performed; there was evidence of delayed infranodal conduction with an HV interval of 70 ms (normal 35–55 ms). AV Wenckebach was demonstrated at 140 beats per minute. No ventricular tachyarrhythmias could be induced.

**Case Discussion**

Given that the most likely cause of his loss of consciousness was a bradyarrhythmia secondary to intermittent infranodal atrioventricular block, a dual-chamber pacemaker was implanted (Epstein et al. 2008).

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**Case 3.2.2**

An 82-year-old female patient with a history of hypertension is brought to the emergency room after losing consciousness. As a result, she fell and, upon regaining consciousness, suffered severe left hip pain and was unable to get up. Using her Lifeline® (Philips Healthcare, Andover, MA), she called 911. In the emergency room, she was found to have suffered an intertrochanteric fracture of the left femur. An electrocardiogram in the emergency room showed sinus bradycardia with a heart rate of 52 beats per minute. A bedside echocardiogram showed a hyperdynamic left ventricle with mild to moderate concentric left ventricular hypertrophy. The left atrium was moderately dilated. Mitral annular calcification and thickening of the aortic valve cusps were seen. She was transferred to the operating room for internal fixation of the fracture. While in the operating room, her heart rate suddenly shot up to 150 beats per minute, and the monitor showed an irregularly irregular rhythm consistent with atrial fibrillation. There was no associated hemodynamic compromise; she received metoprolol 2.5 mg IV, slowing her heart rate down to 100 beats per minute. However, she suddenly became asystolic and after an 8-s pause, a slow junctional rhythm was seen before sinus activity began to emerge.

**Case Discussion**

She was diagnosed with the tachy-brady syndrome, a form of sinus node dysfunction. Two days later, a permanent pacemaker was implanted (Epstein et al. 2008).
Case 3.2.3
A 27-year-old male patient loses consciousness at work after running up a flight of stairs. This was preceded by sudden palpitations and light-headedness. There is no family history of recurrent syncope or of sudden cardiac death. A 12-lead electrocardiogram showed T wave inversion in leads V1–V3. A transthoracic echocardiogram showed a moderately enlarged right ventricle with mild impairment in systolic function. Excessive trabeculation of the right ventricular apex was noted. The left ventricle was normal in size, systolic function, and wall thickness. A signal-averaged electrocardiogram (SAECG) was positive for late potentials using 2/3 criteria.

Case Discussion
The patient was diagnosed with arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC/D) and was advised to have an ICD (McKenna et al. 1994; Epstein et al. 2008).

Case 3.2.4
A 72-year-old male patient who is known to suffer from an ischemic cardiomyopathy and diabetes mellitus with mild impairment of renal function presented with class III congestive heart failure. His electrocardiogram showed normal sinus rhythm and left bundle branch block with a QRS width measuring 156 ms. An echocardiogram showed a dilated left ventricle, anteroseptal and apical akinesis, and moderate to severe mitral regurgitation, left ventricular ejection fraction (LVEF) 30%. Cardiac catheterization showed multivessel coronary artery disease. He underwent coronary artery bypass graft surgery and mitral valve repair with a Carpentier annular ring. His postoperative course was complicated by heart failure and atrial fibrillation requiring IV amiodarone and cardioversion.

Three months later, he returns still complaining of exertional dyspnea and NYHA class III symptoms. He is on a diuretic, a beta blocker, and an angiotensin-converting enzyme inhibitor. His echocardiogram now shows his LVEF at 20% with trace mitral regurgitation (MR); the left ventricle remains dilated. The patient is advised to have a prophylactic ICD with resynchronization therapy (Hunt et al. 2001; Epstein et al. 2008).

Given his renal insufficiency, the patient is admitted the day before the procedure to begin gentle IV hydration with isotonic saline 1 mL/kg/h. Acetylcysteine 600 mg twice daily was started the day before the procedure and continued on the day of the procedure, both to help reduce the risk of contrast nephropathy. A bipolar lead is successfully implanted in a posterolateral branch of the coronary sinus and, after implanting the right ventricular and right atrial leads, an ICD is connected to the three leads. No diaphragmatic pacing was detected at maximum output. A total of 25 mL of ioxithalamate (iso-osmolar nonionic contrast agent) is administered throughout the case. Six weeks later, the patient returns for follow-up, and his functional status is now closer to NYHA class II.

Case 3.2.5
Transtelephonic monitoring detects that the pacemaker battery of a 78-year-old female patient with complete heart block and a 7-year-old pacemaker, has crossed the elective replacement interval (ERI). The patient is contacted and advised to come in for an evaluation in advance of her generator replacement. Records show that the leads, implanted 7 years ago, have IS-1 pins and have displayed good sensing and pacing properties without any major changes in impedance. This was confirmed on her
preoperative visit. In contacting the manufacturing company, no alerts or recalls have been issued relative to the lead model she has. She is on warfarin for permanent atrial fibrillation, and she has not had a history of a stroke. No recent attempt had been undertaken to determine the presence of an escape rhythm.

The patient is advised to hold her warfarin 3 days before the procedure to allow the insertion of a temporary pacemaker wire given the lack of a ventricular escape. Her preoperative white blood cell (WBC) count is normal and she has been afebrile. Her urinalysis is unremarkable. On the day of the procedure, a temporary wire is inserted after confirming the absence of an escape rhythm. The patient is prepped and draped with meticulous care for sterility. After receiving a dose of cefazolin, the old generator is retrieved and after confirming adequate pacing from the external pacemaker, the generator is disconnected from the leads. The leads are tested to confirm adequacy of pacing and sensing and the absence of changes in impedance that may have resulted from lead manipulation. The new generator is inserted in the pocket and the pocket is closed in three layers.

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RESOURCES
Pacemaker Timing Cycles, Programming, and Troubleshooting

Christine C. Chiu-Man, Anders Nygren, and Laura De Souza

PACEMAKER TIMING CYCLES

Pacemaker timing cycles are essential to our understanding of how the pacemaker responds to paced and sensed events. A timing cycle is defined in milliseconds (ms) and regulates the delivery of the pacing stimuli to achieve the programmed rate in beats per minute (bpm). Depending on the pacemaker mode, programmed rates refer to low rate interval (LRI), maximum tracking rate (MTR), and/or maximum sensor rate (MSR). Each timing cycle is initiated by a paced or sensed event that will complete its cycle (timer), that is, the programmed LRI, MTR or MSR. In the demand mode, the timing cycle is reset by an intervening sensed event.

A number of pacemaker intervals are initiated with each event. This chapter explains how pacemaker intervals contribute to the timing cycle, pacemaker electrocardiography (ECG) interpretation, and how to troubleshoot events arising from pacemaker function.

WHAT IS CAPTURE AND SENSING?

Pacemaker capture occurs when electrical energy delivered from the pacemaker to the lead electrode tissue interface stimulates the heart muscle to cause a depolarization or heartbeat. In the case of atrial capture, depending on the location of lead in the atrium, the P wave that immediately follows the pacing spike may be upright, inverted, or isoelectric. In the case of right ventricular capture, the paced morphology is usually a wide QRS complex that looks different than the intrinsic QRS (Fig. 3.3.1). A fusion beat occurs if the paced beat captures at the same time with an intrinsic beat such that the fused morphology is
Sensing is the pacemaker’s ability to recognize and respond to the patient’s intrinsic rhythm. Sensing is critical to preventing the pacemaker from pacing to compete with the patient’s intrinsic heart rhythm as this may induce arrhythmias. The pacemaker’s sense amplifiers will detect the intrinsic deflection within the local electrogram (EGM) if the deflection is sufficiently large as compared with the programmed sensed signal. Depending on the lead position, sensing may occur at the onset, mid-

Figure 3.3.1 Ventricular pacing is at 70 bpm. The fifth QRS complex from the left is an intrinsic beat, while the 6th QRS is a paced fusion beat. The rest of the wider QRS complexes are the fully paced morphology.

Figure 3.3.2 Top panel shows atrial pacing at 70 bpm. The bottom panel shows pseudo-pseudofusion beats in complexes 1, 4, and 5 from the left. This is due to accelerated junctional rhythm similar to the atrial pacing rate.

Partially between or a hybrid of the fully paced and the intrinsic morphology. When there is a pacing spike delivered to the tissue that has been depolarized completely by an intrinsic beat, the morphology is called pseudofusion since the pacing spike did not capture because the tissue has already depolarized and is refractory. When an atrial pacing spike is seen in front of an already depolarized spontaneous ventricular beat, it is called pseudo-pseudofusion beat (Fig. 3.3.2).
portion, or offset of the intrinsic P wave or QRS (Fig. 3.3.3). Far-field sensing refers to the situation when there is sensing of remote EGM from another chamber other than the local EGM in which the lead is placed. An atrial lead maybe positioned close to the atrioventricular (AV) junction and records both the local atrial EGM along with a far-field ventricular EGM. Consequently, the atrial channel may sense both EGMs (Fig. 3.3.4). This is known as far-field R wave sensing. Less common is the case of far-field P wave sensing on the ventricular EGM.

**PACEMAKER INTERVAL Timers**

These intervals determine the delivery of pacing stimuli or resetting of the programmed rate limits, and thus are considered as part of the timing intervals (Fig. 3.3.5).

**Low Rate Interval (LRI)**

This is the minimum allowed pacing rate (bpm) or the longest pacing interval (ms) that is allowed by the pacemaker between consecutive paced or sensed events. The purpose of the LRI is to make sure that the patient’s heart rate does not fall below this limit.

**Figure 3.3.3** Ventricular pacing is at 60 bpm (equivalent to 1,000 ms). The third QRS complex from the left is an intrinsic QRS. When we march backward from the fourth QRS that is paced, we see that the pacemaker is sensing from this midportion of the third intrinsic QRS complex. Depending on the ventricular lead location, sensing may occur at the onset, mid, or offset of the QRS complex.

**Figure 3.3.4** The recording was obtained during sensing test in a patient with complete heart block and a dual-chamber pacemaker. The atrial electrogram shown on the bottom channel shows a large atrial signal with a smaller far-field QRS.
Cardiac Arrhythmia Management

It may be possible to program a secondary low or base rate interval that is related to the patient’s inactivity such as during sleep or rest. The purpose is to reduce the amount of pacing during rest when the patient does not need a faster pacing rate. This may be achieved by programming the clock time when the patient is usually in bed or it may be tied in to the activity sensor’s detection of nonactivity over a 20–30-minute period. The purpose of this rate interval is to help conserve battery life by minimizing unnecessary pacing, and occasionally, for patient comfort during rest or sleep.

**Hysteresis**

For patients who are not pacemaker dependent, one can program another lower rate limit called the hysteresis rate (bpm), which is the longest sensing interval (ms) allowed by the pacemaker (e.g., hysteresis 40 bpm). This is helpful to promote the patient’s intrinsic rhythm, and yet when the pacemaker needs to pace, it will pace at the LRI (e.g., low rate 60 bpm). Thus, on the ECG, one would see a longer interval after an intrinsic beat than after a paced beat.

**AV Interval**

This is initiated after an atrial event (paced or sensed) and is the duration during which the ventricular channel is alert for any ventricular sensed event while the atrial channel is refractory to any atrial sensed events. If no ventricular sensed event occurs, then a ventricular pacing stimulus is delivered at the end of the AV interval. This interval simulates the PR interval during sinus rhythm. In most pacemakers, the paced AV interval (AP) is programmed about 20–50 ms longer than the sensed AV interval (AS) to account for the lag time between delivery of the pacing stimulus and local atrial depolarization. For example, AP may be programmed 150 ms, while AS is programmed at 120 ms. This is known as the

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**Figure 3.3.5** This shows the relation of the sensing intervals and timing intervals in a dual-chamber pacemaker. See discussion in the text.
differential AV interval. The AV interval can be programmed to be “rate adaptive,” with gradual shortening up to a programmed minimum AV interval as the heart rate accelerates (e.g., one might allow rate-adaptive AV interval to no shorter than 80 ms). This is also known as rate-responsive AV interval and is to simulate the shortening of the PR interval during sinus tachycardia.

**Postventricular Atrial Escape (VA) Interval**

Since it is expected that an atrial event should follow a ventricular event, the VA interval is thus initiated after a ventricular event (sensed or paced). It is the duration during which the atrial channel is alert for any atrial sensed event. If no atrial sensed event occurs, then an atrial pacing stimulus is delivered at the end of the VA interval.

**Maximum Tracking Interval (MTR)**

This is the maximum allowable ventricular pacing rate to sensed P waves (or tracking P waves) in DDD or VDD modes. The purpose is to make sure that ventricular pacing is not any faster than this interval. There is maintenance of 1:1 AV synchrony by the pacemaker up to this rate limit; if atrial rate exceeds this maximum tracking interval, the pacemaker will ignore some of the P waves so that there is intermittent loss of AV synchrony and pause in the ventricular tracked rhythm. For patients who may be symptomatic from this loss of AV synchrony, one should program a higher MTR to permit maintaining 1:1 AV synchrony.

**Maximum Sensor Rate (MSR)**

This is the maximum allowable pacing rate in response to a rate-responsive sensor. It is used for patients with chronotropic incompetence whereby their intrinsic heart rate does not increase appropriately with physical activity. For pacemakers that are programmed to DDD with mode switch on, the sensor rate becomes important during mode switch to DDIR in the event of atrial tachycardia (AT). For patients who are symptomatic from pacemaker upper rate behavior (URB), for example, 2:1 block during exercise, turning on the rate-responsive sensor may help prevent the abrupt rate changes in response to sinus tachycardia related to exercise.

**PACEMAKER SENSING INTERVALS**

These intervals do not reset the programmed rate timers but do impact the ability of the pacemaker to sense.

**Postventricular Atrial Refractory Period (PVARP)**

In dual-chamber pacemaker programming, the PVARP is the duration of time after a ventricular event (sensed or paced) during which the atrial channel does not respond to any atrial sensed events. The purpose of PVARP is to prevent atrial sensing of ventricular depolarization, retrograde-conducted P waves, or premature atrial complex (PAC). In general, it is usually programmed around 250 ms. Having a long PVARP may place limits on how fast the pacemaker can track the sinus rate, that is, the MTR. Rate-responsive PVARP will allow longer PVARP at a slower heart rate, with gradual shortening of this interval in association with rate increase. This is useful when there is a need to program a longer PVARP at the lower heart rates such as to avoid tracking PAC, but have a shorter PVARP at the faster sinus rates to allow a higher programmable MTR.
Total Atrial Refractory Period (TARP)

In dual-chamber pacemaker programming, the TARP is the duration of time when the atrial channel is insensitive to any atrial sensed events. It is the sum of the AV interval plus the PVARP. This interval is an important consideration as it imposes how fast the pacemaker can track atrial rates. In patients with complete heart block and dual-chamber pacemaker, TARP defines the interval during which 2:1 P wave tracking occurs (simulating the appearance of 2:1 block) as the atrial rate increases above the MTR. In the example below, it will not be possible for the pacemaker to achieve 1:1 tracking of atrial rate 180bpm as TARP will result in 2:1 P wave tracking when the atrial rate reaches 150bpm or above (Table 3.3.1).

Table 3.3.1  Example of a dual-chamber pacemaker setting.

<table>
<thead>
<tr>
<th>Interval (ms)</th>
<th>Rate (bpm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sensed AV interval</td>
<td>150</td>
</tr>
<tr>
<td>PVARP</td>
<td>250</td>
</tr>
<tr>
<td>TARP</td>
<td>400</td>
</tr>
<tr>
<td>MTR</td>
<td>333</td>
</tr>
</tbody>
</table>

The pacemaker will not be able to reach the 1:1 atrial MTR based on these settings.

Refractory Period

The ventricular refractory period is the duration of time after a ventricular event (sensed or paced) during which the ventricular channel will not respond to any sensed events. The purpose of this interval is to prevent resetting of timing cycles from sensing of ventricular depolarization and of T waves. In pacemakers with tachycardia detection algorithms such as mode switch or high rate counter, events sensed within the refractory period may include them in the diagnostic counter and its determination of the presence of tachycardia. In general, a refractory period is usually programmed around 250–300 ms. It is worthwhile to keep this short in the ventricular channel (e.g., 200–220 ms), particularly in patients with fast intrinsic heart rates, such as children or young adults, to ensure that premature ventricular complex (PVC) are sensed outside of the refractory period to allow reset of the timing cycle and avoid pacemaker competition. Likewise, the atrial refractory period in a single-chamber atrial pacemaker can be programmed around 250–300 ms or longer if there is need to avoid sensing far-field R waves.

Blanking Period

The purpose of blanking periods is to completely switch off the pacemaker sensing amplifiers so that nothing is detected. For example, the most common purpose is to avoid detection of the pacing stimulus as a sensed event. There are three types of blanking periods:

- Blanking within the refractory period after a paced or sensed event
- Ventricular blanking after an atrial pace event within the AV interval
- Postventricular atrial blanking within the PVARP

At the beginning of each refractory period, the duration of the short blanking period or absolute refractory may be nonprogrammable and differs between pacemakers and programmed modes, but is usually in the range of 60–100 ms to avoid detection of the pacing stimulus in the chamber that it is paced from. The ventricular blanking after atrial pacing is programmable and is usually set short at 12–20 ms. Postventricular atrial blanking is programmable, and the nominal setting is usually around 150–180 ms.
Chapter 3.3 Pacemaker Timing Cycles

PACEMAKER MODES

Pacemaker modes are determined by the clinical indication for pacing. They will dictate which timing intervals are operational and how they interact.

Single-Chamber Pacing

Single-chamber timing cycles are the easiest to understand and function in a similar way regardless of whether it is atrial or ventricular pacing.

Demand Mode: AAI, VVI

This is the simplest pacing modality; a single lead is placed into either the atrium (for AAI) or the ventricle (for VVI). In the demand mode, a sensed event will inhibit the subsequent paced event and reset the timing cycle. The cycle begins with a paced or sensed event; this initiates the timer for the LRI, the refractory period along with the blanking period. Any events that fall within the refractory period will be ignored by the pacemaker timer, and thus will not reset the timing cycle. The pacemaker’s alert period is the time from the end of the refractory period to the end of the programmed low rate limit. This is the time when pacemaker timing can be reset by sensed events. Any events falling outside the refractory period will reset the pacemaker timer and initiate a new timing cycle. If no events are sensed during the alert time, then at the end of the programmed low rate limit, the timer expires and a pacing spike is delivered, thus restarting the next timing cycle.

Asynchronous Mode: AOO, VOO

The pacemaker will pace continuously at the programmed low rate regardless of any events. The LRI could not be reset by any intrinsic events since there is no sensing in this mode. Since there is no sensing, there is no pacemaker refractory period either. This mode is rarely used clinically as a permanent programmed setting. In fact, it is potentially dangerous since asynchronous pacing can compete with the patient’s own intrinsic rhythm or pace into the vulnerable period, the tissue’s repolarization phase, and induced arrhythmias. However, asynchronous pacing is of use as emergency or temporary management to avoid oversensing such as intraoperative electrocautery-induced inhibition of pacing. One must ensure that the patient is monitored with ECG in case the patient’s intrinsic heart rate accelerates above the asynchronous pacing rate or if arrhythmia occurs, in which case, the pacemaker should be reprogrammed to the demand mode.

Triggered Mode: AAT, VVT

This mode is rarely used clinically but the initial design was intended to address oversensing problems. In addition to initiation of the low rate timer, the pacemaker will deliver a pacing spike immediately upon a sensed event outside of the refractory period. On ECG, this spike will distort the native signal (P wave in AAT or QRS in VVT). From this perspective, the pacing energy is wasted since there is intrinsic rhythm. However, in the presence of oversensing due to noise, triggered pacing may result in pacing above the low rate, up to the factory-determined maximum rate or the runaway limit of the pacemaker. This may induce competition with the patient’s underlying rhythm. Historically, the triggered mode can be used as a marker for the point of sensing on the intrinsic signal or for chest wall stimulation to perform noninvasive electrophysiological study (NIPS). However, current-day pacemaker programmers can accomplish these two tasks via telemetered marker channel data and availability of electrophysiological stimulation menu to perform NIPS via the
permanent pacemaker. Thus, triggered mode has limited clinical value or utility.

**Rate-Responsive Mode: AAIR, VVIR**

Rate-responsive pacing is important for those patients whose heart rate cannot increase with exercise. Increasing the heart rate will have a direct effect of increasing cardiac output. The most common rate-responsive sensor is the body motion activity piezoelectric accelerometer. The sensor is mounted onto the circuit board inside the pacemaker. When the rate-responsive feature is turned on, the accelerometer detects the back-and-forth motion such as when the patient is running. The motion will deflect the piezoelectric sensor. The amount of mechanical deflection of the piezoelectric crystal is translated proportionally to increase the pacemaker’s pacing rate. This will result in a quick increment of the pacing rate at the onset of exercise and gradual decrement at the offset of exercise; the duration of the onset and offset is programmable. The variability in pacing rate will depend on the amount of activity detected by the sensor. In most manufacturers, sensor-driven pacing will occur for two levels of activity. There is usually a walking or daily activity rate for moderate activities, usually about 20–30 bpm faster than the programmed low rate. From this walking or daily activity rate, pacing can further increase up to the programmed MSR depending on the amount of vigorous activity or exercise that the patient is doing.

**Dual-Chamber Pacing**

In dual-chamber pacemakers, the complexity of the timing cycle is related to the simultaneous interplay of the pacemaker intervals on both atrial and ventricular channels as well as the basis of the low rate timing (ventricular-based, atrial-based, or modified atrial-based timing). These pacemaker intervals are shown in Figure 3.3.5. Conceptually, the purpose of dual-chamber pacing is the restoration of AV synchrony or 1:1 relationship of atrium and ventricle in patients with heart block.

Historically, dual-chamber pacemakers were designed as ventricular-based timing. This means that the timers work solely on the ventricular rates. The low rate limit is the longest interval between consecutive ventricular events (V-V timing). The hallmark of ventricular-based timing is the initiation of a constant VA interval following each ventricular event. The VA interval is determined as the difference between the LRI minus the AV interval. It is an atrial alert period whereby if an atrial sensed event does not take place prior to the end of the VA interval, an atrial pacing spike is delivered.

There are two problems associated with ventricular-based timing. One is the potential for acceleration of pacing rate when the patient has intrinsic conduction that occurs prior to the end of the AV interval. In this case, the constant VA interval necessitates delivery of atrial pacing earlier than the low rate limit; the amount of rate acceleration is equal to the difference between the sensed AV interval and the paced AV interval. During sensor-driven pacing (e.g., DDDR), the amount of rate acceleration may be significantly higher than that dictated by the sensor. The second problem is that the low rate may oscillate slightly faster than or equal to the programmed low rate limit if the patient is alternating between ventricular paced and ventricular sensed (from conducted beats) events.

Pacemaker manufacturers developed atrial-based timing in an effort to abolish the rate fluctuations seen in ventricular-based timing devices. In atrial-based timing systems, the timers are reset based on the atrial rates only. The low rate limit is the longest consecutive interval between atrial events (A-A timing). Ventricular sensed events prior to the end of the AV delay interval inhibits ventricular pacing but does not reset the low rate timer. Thus, the atrial pacing rate remains constant at the programmed low rate limit. However, if one measures the V-V intervals during these ventricular
sensed events within the AV interval, they will be slower than the programmed low rate limit. If there is alternation between ventricular sensed and ventricular paced events, the V-V intervals will alternate between slower and faster rates than the programmed low rate limit but the A-A intervals will remain constant at the programmed low rate limit. In the event of a sensed PVC, the PVC will reset the low rate timer, simulating a compensatory pause.

Most pacemakers used a modified atrial-based timing system. The VA interval is used after a sensed PVC to reduce the amount of the pause that would be seen in a pure atrial-based timing system. Otherwise, the low rate timing is based on consecutive fixed A-A intervals. Due to variations of the timing systems used by the different pacemaker manufacturers, reference to their technical manuals may be necessary.

**Tracking Modes: DDD/VDD**

In the tracking mode, the pacemaker is pacing in the ventricle in response to sensed P waves at rates above the LRI and below the MTR. The VDD pacing may be achieved by a single-pass ventricular lead that has four electrodes: a proximal pair of “floating” bipolar electrodes in the atrium and a distal pair of bipolar electrodes in the ventricle. Since the proximal bipoles are not attached to the atrial tissue, atrial pacing is not possible. In VDD mode, AV synchrony is maintained by tracking within the programmed rate limits (low rate and MTR); however, when the atrial rate falls below the LRI, ventricular pacing at the low rate limit is maintained and gives the ECG appearance of VVI pacing, resulting in loss of AV synchrony (Fig. 3.3.6). In DDD mode, when the atrial rate falls below the LRI, AV sequential pacing will take place, and thus AV synchrony will be maintained. In the case of a patient with intermittent AV conduction and a DDD pacemaker, in addition to tracking and AV sequential pacing, it is possible to see intrinsic sensed P wave and conducted QRS (giving appearance of sinus rhythm with no pacing) or atrial pace with ventricular conduction (simulating ECG appearance of AAI pacing).

A sensed P wave will inhibit atrial pacing and initiates the sensed AV interval, the LRI, and MTR. At the end of the AV interval, ventricular pacing will be delivered if there is no

![Figure 3.3.6](image-url)  
**Figure 3.3.6**  Pacemaker setting is VDD 60/180 bpm (low rate/maximum tracking rate). During a brief period in the midportion of the recording, the sinus rate (P waves) falls below the low rate limit of 60 bpm. The pacemaker will maintain ventricular pacing at the low rate. In this portion of the recording, it gives the appearance of VVI pacing.
ventricular sensed event. A ventricular paced event will initiate the VA interval, PVARP, and ventricular refractory period. At the end of the VA interval, if no P wave is sensed, atrial pacing will be delivered. This will initiate the paced AV interval, the LRI, and MTR.

**Rate-Responsive Modes: DDDR, VDDR, DDIR**

For patients with chronotropic incompetence, rate-responsive pacing may be needed in addition to dual-chamber pacing. Rate-responsive sensors are built into most present-day pacemakers. In DDDR, there are two programmable upper rate intervals: MTR and MSR. These two rates may be programmed the same or different depending on the patient needs. Rate-responsive PVARP and AV interval are useful features to ensure that at the upper rate intervals, an adequate atrial sensing window is preserved. In differential rate programming, if the MTR is programmed lower than the MSR, it is possible for atrial events to be sensed above the MTR that will inhibit atrial pacing during sensor-driven pacing. This gives the ECG appearance of atrial tracking above MTR, but in fact, the sensor is driving the rate at this point.

**VDDR** is somewhat a misnomer as there is only ventricular pacing at the sensor-indicated rate, so functionally, it is VVIR during sensor-driven pacing. In this case, AV synchrony is maintained at the atrial tracking rate (between the LRI and MTR); loss of AV synchrony may result during sensor-driven pacing if atrial rate is below the sensor-indicated rate.

**DDIR** is an option for those patients who are prone to chronic atrial arrhythmias that we do not want the pacemaker to track, yet the rate-responsive sensor allows rate increases in proportion to exercise activities. In the presence of atrial arrhythmias, sensor-driven pacing gives the appearance of VVIR pacing when the atrial events are sensed and inhibits sensor-driven atrial pacing. In DDD pacemakers with mode switch feature, the mode switch is commonly to DDIR.

**MISCELLANEOUS PACEMAKER BEHAVIORS**

There are times when these complex timing cycles can result in pacemaker behavior that
may be due to the limitations of the programmed parameters, rather than true malfunction. The following are examples.

**Upper Rate Behavior (URB)**

In dual-chamber programming, the pacemaker is not allowed to ventricularly tracked atrial rates above the MTR. When the sensed atrial rate is faster than the MTR, the pacemaker must postpone the delivery of the ventricular pacing until the MTR times out. This delay in delivery of ventricular pacing is achieved through a forced AV interval extension beyond what is programmed. The delay in ventricular pacing will have the effect of eventually having a P wave falling inside the PVARP, causing it to be untracked. The degree of AV interval extension (ms) is determined as the difference between MTR and TARP intervals. The Wenckebach window is the difference between MTR and TARP in terms of rate (bpm).

Depending on the atrial rate and the programmed MTR and TARP, there may be varying cycles of P waves not being tracked (e.g., every second or third P wave) when the atrial rate exceeds the MTR. When this happens, the ECG appearance is similar to that of varying degrees of “block” (such as group beating in Wenckebach block, 2:1, or 3:1 block), but the “block” is really the single cycle pause created by the pacemaker withholding from ventricular pacing.

It is ideal in programming to have a Wenckebach window of about 20–40 bpm before 2:1 block occurs to minimize the patient becoming symptomatic from the sudden rate change. This is particularly important when the patient does not have intrinsic ventricular conduction (i.e., complete AV block) and the ventricular rate is controlled solely by the pacemaker. Depending on how the intervals are programmed (AV interval, PVARP, and MTR), there may be sudden shifts in pacing rate during URB (e.g., the ventricular rate may change abruptly in half from 1:1 tracking to 2:1 block). It may be possible to minimize sudden rate changes by maximizing the MTR, increasing the difference between the MTR and TARP. If there is still URB despite optimal programmed settings in which the patient is symptomatic, then one can consider using programming features such as turning on rate response to use the sensor to help control the maximum pacing rate. Consideration may also be given to turn on rate regularizing features such as rate smoothing, fallback, or rate stabilization that operates on the premise of reducing pauses or sudden rate changes by controlling the pacing rate changes at gradual decrements.

**Cross Talk**

Cross talk occurs when there is cross-chamber sensing of the pacing stimulus. It is a real concern, particularly in heart block patients when ventricular sensing of the atrial pacing stimulus inhibited the delivery of ventricular pacing. This may happen in the situation of programmed high atrial output setting coupled with very sensitive ventricular sensitivity setting. This is detrimental in a pacemaker-dependent patient as it can result in an asystolic situation.

In most pacemakers, there is also a nonprogrammable shortened paced AV interval that is manufacturer specific, known as safety pacing or the ventricular safety interval (Medtronic, Minneapolis, MN; 110 ms) or nonphysiological AV interval (St. Jude Medical, Minneapolis, MN; 120 ms). Boston Scientific Guidant pacemakers (Boston Scientific, Natick, MA) do not have this feature. It is designed to intervene when cross talk occurs. The AV interval is made of the initial short blanking period followed by a noise sampling window (usually about 60 ms) and then the rest of the AV interval (Fig. 3.3.7). Ventricular sensing during the noise sampling window will initiate safety pacing, characterized by shortened AV delay. Ventricular sensed events in the remainder of the AV interval will inhibit ventricular pacing.
Blanking Noise sampling window

**Figure 3.3.7** This illustrates the blanking period and noise sampling window within the AV interval. Ventricular sensed events inside the noise sampling window will initiate ventricular safety pacing at the shortened AV interval. The noise sampling window occurs immediately after the blanking period and is the short duration of time when sensed events might be due to detection of pacing stimulus (such as the polarization effect from the stimulus), or it might be intrinsic QRS. It is a window of uncertainty for the pacemaker; therefore, rather than pace at the programmed AV interval (which might be set at longer duration) or inhibit from pacing, it will deliver a pacing spike at the shortened interval. Pacing at the shortened interval is unlikely to pace onto the T wave of an intrinsic QRS. Note that any ventricular sensed event beyond the noise sampling window will inhibit ventricular pacing.

**Pacemaker-Mediated Tachycardia (PMT)**

A form of pacemaker-related tachycardia deserves special mention as prompt recognition is critical to patient management. It is most commonly called pacemaker-mediated tachycardia as the pacemaker pacing is an integral part of the tachycardia circuit; sometimes it may be known as endless-loop tachycardia. This form of tachycardia is seen only in dual-chamber pacemakers with tracking modes DDD or VDD, and if the patient has intact retrograde VA conduction. The onset is sudden, and the tachycardia rate is usually at or near the upper tracking rate (Fig. 3.3.8).

PMT is initiated by an event that uncoupled AV synchrony for one cycle during normal DDD or VDD pacing. The potential initiating events include PVC with retrograde conduction, loss of atrial capture, or tracked PAC near the MTR.

The ventricular event (PVC or paced) has the opportunity to conduct retrograde reactivating the atrium as a retrograde P wave that is tracked by the pacemaker. The cycle of tracking retrograde P waves perpetuates. One way to get around this is to test and find out what the retrograde conduction time is (from ventricular paced to sensed P wave) and program the PVARP about 20–50 ms longer so that the retrograde P wave falls inside PVARP and is not tracked. This may necessitate programming a long PVARP and impose limitation on programming a higher MTR. The other alternative is to turn on the rate-responsive PVARP, PMT algorithm as well as PVC response. These algorithms and features differ depending on pacemaker manufacturer, but the general aim is to prevent PMT by PVARP extension for one cycle. If PMT is in progress, this PVARP extension will prevent the retrograde P wave from being tracked and break the PMT cycle. This will be followed by resumption of ventricular tracking of sinus rhythm.

**Intrinsic Conduction Search**

Some of the new pacemaker models allow a search algorithm for intrinsic conduction during the AV interval in dual-chamber pacemakers. This is helpful consideration for patients with some intrinsic AV conduction since minimizing ventricular pacing can help save battery life. Furthermore, research findings have indicated that ventricular pacing may be deleterious, particularly in patients with impaired ventricular function. Programmable features that promote intrinsic conduction include AV search hysteresis, minimal ventricular pacing, or ventricular intrinsic preference. Occasionally, one may observe AV conduction prolongation if the patient is on cardiac medications such as beta blockers or calcium channel blockers and exhibits varying degrees of AV block until the pacemaker has completed its search criteria before reverting back to 1:1 tracking mode. For these patients, it may be
**Figure 3.3.8** The recording is obtained from a patient with DDD pacemaker, showing ECG lead II, marker channel, and atrial electrogram. The top panel shows atrial pacing threshold test is being conducted. The surface P wave is not clear, but the local atrial electrogram helps determine the presence or absence of atrial capture. On the fifth atrial pacing spike from the left, atrial capture is lost and a retrograde atrial signal is seen after the ventricular paced event. This atrial activity is tracked by the pacemaker, and pacemaker-mediated tachycardia (PMT) is initiated. PMT continues, and at the bottom panel, PMT breaks when retrograde conduction was lost (eighth QRS complex from the left). AV sequential pacing resumes after this.
worthwhile to consider disabling this feature and program a sufficiently long AV delay.

SYSTEMATIC APPROACH TO PACEMAKER ECG INTERPRETATION

Assessment of pacemaker ECG includes rhythm strip, 12-lead ECG, ambulatory Holter, event recorder, or stress exercise testing. Although it is often possible to recognize the pacing mode from a pacemaker ECG, it is important to know some of the key programmed settings to allow for accurate interpretation of the ECG. These settings include the programmed mode, rate limits such as low rate, upper rate (MTR or MSR), hysteresis rate, rest or sleep rate, refractory periods, AV interval if applicable, and any relevant advanced programming features. It is extremely helpful to have a set of calipers to help measure the intervals accurately. Occasionally, there may be a need to convert the programmed rates from bpm to ms. This is accomplished by using the formula of \( \frac{60000}{\text{bpm}} = \text{ms} \).

The purpose of pacemaker ECG assessment is to determine if the pacemaker is sensing well, capturing, and if the programmed settings are appropriate for the patient’s needs. Pacemaker ECG is often complex due to the interplay of advanced programming features, the need to understand the timing intervals, and how they work. The best way to learn and be good at this is practice.

In some ways, ECG interpretation involves a degree of initial pattern recognition. This must always be followed by having in your mind a list of possible differential diagnosis of the pattern. At this point, you will need to work out the timing cycles to find out which diagnosis best fits the ECG pattern shown. We will follow through this approach with some examples to make this easier to understand. There are four basic rules that will help make pacemaker ECG interpretation less intimidating.

1. Observe what the paced and intrinsic morphology looks like.
2. Look for two consecutive paced events during the same period of time when a sensed event occurs or at the LRI. This helps determine what the prevailing pacing rate should be at that time.
3. March the calipers forward from the sensed event to determine when the next pacing spike should be delivered.
4. When possible, march the calipers backward from a paced event to determine where sensing is timed from.

The first step is identification of the rhythm. Is it the patient’s intrinsic, is it paced, or is it a combination of both pacing and intrinsic? If pacing is present, is it pacing in the atria or ventricle or both? Similar to regular ECG interpretation, we want to identify if it is sinus rhythm, if the atrial rhythm is associated with the ventricular rhythm, or if the rhythm is consistent with AV sequential pacing, atrial, or ventricular pacing.

The next step is to determine if the heart rate is consistent with the programmed rate limits. If the heart rate is faster than the programmed MTR or MSR, then it should be the patient’s own intrinsic rhythm. If the heart rate is slower than the programmed low rate, then the possibility exists that the pacemaker is not functioning or other special features such as hysteresis or rest/sleep rate, AV search hysteresis are turned on (Table 3.3.2). This information will need to be confirmed from the printed programmed parameters. Advanced programming features such as automatic threshold or sensing determination, rate regularizing features such as rate smoothing or stabilization, or specific search algorithms such as blanked flutter search, AV search hysteresis may alter the usual pattern of pacemaker behavior. In these instances, it is hard to differentiate pacemaker problems from normal functioning of these advanced features.
Chapter 3.3 Pacemaker Timing Cycles

by programming the sensitivity setting to a more sensitive setting, that is, a smaller number, to ensure that the pacemaker will “see” events that have a lower amplitude.

The caveat is that one must confirm the intrinsic beat that was not sensed was outside the pacemaker refractory period. If in fact the intrinsic beat was in the pacemaker’s refractory period and thus did not reset the timer, then this is normal function and this intrinsic beat is noted as “functional undersense.” For example, if this was a PAC that is within PVARP, and we do not want the pacemaker to track PAC, then this is normal and we would not do anything further. If this was a PVC that fell inside the ventricular refractory period, then it will not reset the pacemaker timer even though the pacemaker can sense it. To correct this problem, we should shorten the refractory period to allow the PVC to reset the timer rather than adjust ventricular sensitivity.

Table 3.3.2 Pacemaker features that affect pacemaker timing.

<table>
<thead>
<tr>
<th>Feature</th>
<th>Description</th>
<th>Consequence</th>
</tr>
</thead>
<tbody>
<tr>
<td>PVC response</td>
<td>Extend PVARP longer from the sensed PVC to avoid atrial sensing of retrograde P wave</td>
<td>First P wave after PVC may not be tracked</td>
</tr>
<tr>
<td>PMT algorithm</td>
<td>Regularly check for PMT by extension of PVARP for one cycle to try to ignore one P wave; if PMT is going on, this will break the tachycardia cycle</td>
<td>May see single P wave that is not tracked in regular cycles</td>
</tr>
<tr>
<td>Hysteresis</td>
<td>Allows a separate programmed lower rate (hysteresis rate) for sensing that is different from the programmed low rate</td>
<td>Spontaneous intrinsic rhythm or ectopic beats will activate the hysteresis rate, thus allowing a lower intrinsic rate than the programmed low rate (Fig. 3.3.12)</td>
</tr>
<tr>
<td>AV search hysteresis</td>
<td>Allows prolongation of AV interval to search for intrinsic conduction; after completing the search criteria, the AV interval reverts to the programmed interval</td>
<td>Lower rate intervals may be violated for brief periods during search algorithm</td>
</tr>
<tr>
<td>Ventricular safety pace</td>
<td>AV pace occurs at a shorter fixed AV interval due to ventricular sensed event in the noise sampling window of the AV interval</td>
<td>Shortened paced AV delay (Fig. 3.3.13)</td>
</tr>
</tbody>
</table>

Finally, we should determine if sensing and capture are appropriate. This is perhaps the most important step in the interpretation as it may lead us to further assessment of the patient or indicate to us specifically if we need to make certain pacemaker programming adjustments to correct any problems.

ECG Recognition of Sensing Problems

Sensing problems may manifest as undersensing or oversensing. Undersensing occurs when the patient’s intrinsic heartbeat is not seen by the pacemaker and a pacing spike is delivered in close proximation to the intrinsic beat (Fig. 3.3.9). On ECG, undersensing appears to show “too much pacing” or overpacing. If one marches backward from the paced event, it becomes clear that the preceding intrinsic beat was not sensed. This problem can be corrected by programming the sensitivity setting to a more sensitive setting, that is, a smaller number, to ensure that the pacemaker will “see” events that have a lower amplitude.

The caveat is that one must confirm the intrinsic beat that was not sensed was outside the pacemaker refractory period. If in fact the intrinsic beat was in the pacemaker’s refractory period and thus did not reset the timer, then this is normal function and this intrinsic beat is noted as “functional undersense.” For example, if this was a PAC that is within PVARP, and we do not want the pacemaker to track PAC, then this is normal and we would not do anything further. If this was a PVC that fell inside the ventricular refractory period, then it will not reset the pacemaker timer even though the pacemaker can sense it. To correct this problem, we should shorten the refractory period to allow the PVC to reset the timer rather than adjust ventricular sensitivity.
Oversensing occurs when the pacemaker sees signals other than the patient’s intrinsic rhythm and inhibits from pacing (Fig. 3.3.10). The pacemaker is resetting the timing from noise (electrical noise, interference, myopotential) or signals such as T wave, other than the P wave or QRS complex. This may result in pauses if patient is pacemaker dependent or in heart rate slower than the programmed low rate if the patient has a slower underlying rhythm. On ECG, oversensing appears to show “too little pacing” or not pacing when it should have paced. In the case of noise oversensing in a dual-chamber pacemaker in tracking mode, ventricular tracking of noise may lead to inappropriately fast pacing rates. It may be possible to reduce oversensing by reducing the sensitivity of the pacemaker, that is, programmed to a larger number on the sensitivity setting.

Occasionally, oversensing can lead to undersensing. In such a case, when one marches backward with calipers from the paced event, it may confirm to us the point of oversensing. If there is an intrinsic beat that followed the oversensed event, this beat may fall within the

**Figure 3.3.9** Pacemaker is programmed to backup rate of 60 bpm VVI. Patient has intrinsic sinus rhythm with occasional AV block, followed by a paced escape at 60 bpm. However, the PVC (sixth QRS complex from the left) was not sensed and pacemaker paced inappropriately onto the T wave. One can appreciate the potential danger of initiating ventricular fibrillation or arrhythmia by pacing competition with the intrinsic.

**Figure 3.3.10** Pacemaker is programmed VVI 60 bpm. There is inappropriate inhibition from pacing after the third QRS complex: Myopotential noise is seen on the ECG without any intrinsic QRS. This is oversensing.

**Differential diagnosis of undersensing:**
- Functional undersense
- Oversensing leading to undersense
- Magnet-induced asynchronous pacing
pacemaker refractory period and was not sensed. In this case, the primary problem is oversensing rather than under sensing, thus one should adjust the pacemaker sensitivity to a less sensitive setting to correct the oversensing problem.

### Differential diagnosis of oversensing:
- Secondary low rate (rest, sleep)
- Hysteresis

**ECG Recognition of Capture Problems**

Pacemaker capture is identified by seeing pacing spikes followed by atrial (in atrial pacing) or ventricular (in ventricular pacing) depolarization. Loss of capture occurs when there is no depolarization after the pacing spike (Fig. 3.3.11). This is corrected by rest-testing the stimulation threshold and increasing the pacemaker output. The pacing spikes of bipolar pacemaker are often not very visible on the ECG. It may be helpful in distinguishing between a paced versus an intrinsic morphology to look at the morphology of the atrial or ventricular complex at the programmed low rate as this may be likely the paced morphology.

If the pacing spike was delivered close to a preceding spontaneous beat such as PVC or P wave, it is possible that pacing will not be able to cause depolarization as the tissue is still in repolarization. In this case, this is known as “functional loss of capture” and is unrelated to pacemaker output.

However, sensing and capture problems may be related to lead hardware or, in rare cases, pacemaker hardware circuit malfunctions. In these instances, pacemaker programming adjustments may or may not alleviate the problems.

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**Figure 3.3.11** Pacemaker is programmed DDI 60 bpm in a patient with intermittent AV block and RBBB. There is loss of ventricular capture by the pacing spike after the second QRS complex, followed by a spontaneous junctional escape beat of RBBB morphology. This was followed by AV sequential pacing at the low rate of 60 bpm, and the QRS morphology is similar to the intrinsic junctional escape beat, hence, these may be pseudofusion beats or absence of ventricular capture in the presence of intrinsic conduction.
Programming Features That May Alter the Basic Pacemaker Timing Cycle

In pacemaker ECG interpretation, one should be aware that some programmed features may alter the basic pacemaker timing cycle, and thus should not be interpreted as a problem (Figs. 3.3.12 and 3.3.13).

PACEMAKER FOLLOW-UP

With our knowledge of pacemaker timing cycles and intervals in mind, it is time to take care of our patient with a pacemaker. Present-day pacemakers can do more than just pace and sense. They provide powerful diagnostics, measurements, and may have algorithms that can be turned on to prevent arrhythmia devel-

**Figure 3.3.12** Pacemaker is programmed VVI 60 bpm, hysteresis 40 bpm. The recording shows a longer pause (more than 1,000 ms) after the first two intrinsic QRS complex, followed by ventricular pacing at 60 bpm. The pause is due to the hysteresis function. This is normal pacemaker function.

**Figure 3.3.13** Pacemaker is programmed DDD 60/180 bpm, paced AV interval 150 ms. The fifth QRS complex is an intrinsic beat in between the atrial and ventricular pacing spikes (best seen on the second channel). The measure paced AV interval is 120 ms, shorter than the programmed AV interval. This is due to sensing of the QRS within the noise sampling window. This is not undersensing.
opment. Thus, one of the impetus in pacemaker follow-up is to ensure that problems are recognized and programming is tailored to individual patient needs. Pacemaker check can be divided in four parts.

1. The patient
2. Pacemaker interrogation and testing
3. Programming considerations
4. Additional testing as required

It is important to always have a systematic approach and take the time as needed to achieve the best results possible for the patient.

**Before Seeing the Patient**

A person performing follow-up should have some basic information readily available (see Table 3.3.3).

It is helpful to have a dedicated chart or database system with the essential information available on the patient as well as to track pacemaker follow-up visit issues, measurements, and programming changes in the form of tables. These systems can be tailored to the needs of the clinic (some examples are seen in Tables 3.3.4–3.3.6). Keeping these kinds of records is influenced by legislations and policies about documentation and paper charts. In most hospitals, there is electronic documentation, and patient records should always incorporate appropriate information from pacemaker follow-ups.

**Before Starting the Pacemaker Interrogation**

One of the most important parts of the pacemaker check is to take an adequate history from the patient. There is a difference between a patient who says that everything is fine and a patient who had two episodes of syncope or who has increased signs of heart failure. Some of the important symptoms to be aware of and what to look for during follow-up are shown in Table 3.3.7.

Sometimes, the symptoms can be directly related to the behavior of the pacemaker and can be easily resolved by the person performing the check. More often, however, information found during interrogation has to be seen in the context of both the patient’s symptoms and the underlying condition. These need to be reviewed with the responsible physician. It is therefore essential to have good communication between the pacemaker clinic and the physician if they are separate teams. It is a good idea to inspect the pacemaker implant site in case of any changes in appearance such as device erosion.
Table 3.3.4  Example of a short table to track performance of battery and leads.

<table>
<thead>
<tr>
<th>Date</th>
<th>Battery</th>
<th>Lead impedance</th>
<th>Threshold</th>
<th>Sensitivity</th>
<th>Misc</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Atrial Ω</td>
<td>Ventricular Ω</td>
<td>Atrial V</td>
<td>Ventricular V</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Atrial Ω Pol</td>
<td>Ventricular Ω Pol</td>
<td>Atrial Dur Pol</td>
<td>Ventricular Dur Pol</td>
</tr>
<tr>
<td>14-Dec-08</td>
<td>2.76</td>
<td>&lt;1 kΩ Bi 542 Pol</td>
<td>1.25 0.4 Bi</td>
<td>1.0 0.4 Bi</td>
<td>2 mV Bi 7.6 mV Bi</td>
</tr>
</tbody>
</table>

V, voltage; Ω, impedance; Pol, polarity; Dur, pulse duration; Bi, bipolar; Vent, ventricle.
Table 3.3.5 Example of a short table to track diagnostic information.

<table>
<thead>
<tr>
<th>Date</th>
<th>Events</th>
<th>Mode-switch</th>
</tr>
</thead>
<tbody>
<tr>
<td>14-Dec-08</td>
<td>AS-VP</td>
<td>Number</td>
</tr>
<tr>
<td></td>
<td>AS-VS</td>
<td>95%</td>
</tr>
<tr>
<td></td>
<td>AP-VP</td>
<td>&lt;1%</td>
</tr>
<tr>
<td></td>
<td>AP-VS</td>
<td>4%</td>
</tr>
<tr>
<td></td>
<td></td>
<td>&lt;1%</td>
</tr>
<tr>
<td></td>
<td></td>
<td>6</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Flutter</td>
</tr>
<tr>
<td></td>
<td></td>
<td>4%</td>
</tr>
</tbody>
</table>

AS, atrial sense; AP, atrial pace; VS, ventricular sense; VP, ventricular pace.

**Pacemaker Interrogation and Testing**

Most pacemaker manufacturers have interfaces on the pacemaker programmer, such as summary screen or quick checklist routine, designed to facilitate and speed up the pacemaker check. These should be regarded as a good supplement and a useful tool but can never replace a true understanding of the principles of reviewing diagnostic information and performing testing systematically. We will try to cover the most common programming features in general terms, but the operation of these features are very manufacturer specific and one needs to refer to the technical manuals of the pacemakers to truly appreciate how they work.

**Diagnostic Information**

**Baseline ECG**

Record an ECG strip before placing the interrogation wand above the pacemaker. This can usually be done through the programmer and if not, a surface ECG with rhythm strip is sufficient. Occasionally, one may encounter patients who are not willing to cooperate (such as small children); it is then a good idea to place the ECG leads on the patient’s back, out of reach, and have them sit on the parent’s lap. The baseline ECG is mainly a documentation of the pacemaker function and provides a snapshot of possible issues (such as AF or loss of capture) to focus on during pacemaker check.

**Diagnostics: Paced and Sensed Events**

From the first overview page, all programmers will have a diagnostic display. The first thing that must be kept in mind when interpreting diagnostics from the pacemaker is that the pacemaker shows “what it thinks it saw” and this may not correlate with what is truly going on with the patient’s rhythm. The diagnostics are helpful but can never truly replace the value and utility of ambulatory Holter recording or exercise test or basic ECG recording.

Different pacemaker companies will have different nomenclature. They will provide a summary of the different events in the pacemaker including the amount of paced or sensed events. In DDD mode, four different events will be shown by combination of sensed and paced events in the atrial and ventricular channels. Examples of expected and unexpected numbers or proportions of paced and sensed events depending on the patient’s indication for pacing are shown in Table 3.3.8. The counter may also include information about isolated ectopy or runs of ectopy. This will provide useful information in the management of the patient. A percentage of cardiac cycles in AV synchrony may also be given. For example, less than 100% AV synchrony in DDD mode may be caused by atrial ectopy/arrhythmias resulting in mode
Table 3.3.6  Example of a table with final settings.

<table>
<thead>
<tr>
<th>Date</th>
<th>Mode</th>
<th>Base</th>
<th>Sleep</th>
<th>MTR</th>
<th>MSR</th>
<th>AV/PV</th>
<th>PVARP</th>
<th>Output</th>
<th>Sensitivity</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Atrial</td>
</tr>
<tr>
<td>14-Dec-08</td>
<td>DDDR</td>
<td>60</td>
<td>50</td>
<td>140</td>
<td>130</td>
<td>150/120</td>
<td>225</td>
<td>2.5/0.4</td>
<td>Bi</td>
</tr>
</tbody>
</table>

Pol, polarity; Vent, ventricle.
Table 3.3.7 Patient symptoms and possible causes to be aware of during interrogation.

<table>
<thead>
<tr>
<th>Patient symptoms</th>
<th>Possible causes</th>
<th>Possible findings in clinic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Feeling well with episodes of syncope/presyncope</td>
<td>Intermittent loss of capture</td>
<td>Pacing threshold approaching output, unstable impedance, loss of capture on provocation</td>
</tr>
<tr>
<td></td>
<td>Atrial arrhythmias</td>
<td>Seen on diagnostic counter, log</td>
</tr>
<tr>
<td></td>
<td>Ventricular arrhythmias</td>
<td>Seen on diagnostic counter, log</td>
</tr>
<tr>
<td></td>
<td>Oversensing</td>
<td>Inappropriate inhibition of pacing</td>
</tr>
<tr>
<td>Sudden onset of fatigue</td>
<td>Lead failure of different etiologies</td>
<td>Sudden change in lead impedance, change in threshold</td>
</tr>
<tr>
<td></td>
<td>Automatic switch to VVI when approaching elective replacement</td>
<td>seen on interrogation</td>
</tr>
<tr>
<td></td>
<td>Oversensing</td>
<td>Inappropriate inhibition of pacing</td>
</tr>
<tr>
<td></td>
<td>Pacemaker dysfunction</td>
<td>Settings differ from previously set (e.g., electrical reset)</td>
</tr>
<tr>
<td>Gradual onset of fatigue, signs of heart failure, or change in mental status</td>
<td>Pacemaker syndrome</td>
<td>See separate section</td>
</tr>
<tr>
<td></td>
<td>Atrial arrhythmias</td>
<td>Usually seen on interrogation depending on pacemaker</td>
</tr>
<tr>
<td>Exercise intolerance</td>
<td>Inadequate rate response</td>
<td>Low peak sensed heart rate</td>
</tr>
<tr>
<td>Intermittent palpitations</td>
<td>Atrial arrhythmias</td>
<td>Low response of rate sensor</td>
</tr>
<tr>
<td></td>
<td>Ventricular arrhythmias</td>
<td>Seen on diagnostic counter, log</td>
</tr>
</tbody>
</table>

Table 3.3.8 Examples of different event counters in DDD mode.

<table>
<thead>
<tr>
<th></th>
<th>CHB</th>
<th>SSS AV1</th>
<th>SSS AV1*</th>
</tr>
</thead>
<tbody>
<tr>
<td>AS-VS</td>
<td>&lt;1%</td>
<td>20%</td>
<td>3%</td>
</tr>
<tr>
<td>AS-VP</td>
<td>92%</td>
<td>3%</td>
<td>20%</td>
</tr>
<tr>
<td>AP-VS</td>
<td>&lt;1%</td>
<td>70%</td>
<td>7%</td>
</tr>
<tr>
<td>AP-VP</td>
<td>7%</td>
<td>7%</td>
<td>70%</td>
</tr>
</tbody>
</table>

* SSS AV1: sick sinus syndrome and first-degree heart block—unexpected values with increased amount of ventricular pacing, consider prolongation of AV delay and activation of AV hysteresis/ventricular intrinsic preference; can also be caused by progression of heart block.

AP-VP, atrial pacing—ventricular pacing; AS-VP, atrial sensing—ventricular pacing; AP-VS, atrial pacing—ventricular sensing; AS-VS, atrial sensing—ventricular sensing; CHB, complete heart block; SSS AV1, sick sinus syndrome and first-degree heart block—expected values.

Heart Rate Counter/High Rate Counter

This provides more details in the form of a rate histogram, a counter with numbers for different heart rate bins, or as a log list that has date and time of arrhythmia occurrence. The data that is collected and the format of the data display vary somewhat between manufacturers and pacemaker models. Commonly, both ventricular and atrial counters/histograms are given, and these are expected to be the same if the AV synchrony is 100%. Examples of normal atrial and ventricular histograms are found in
Cardiac Arrhythmia Management

Cardiac rate. Adjustment: Depends on the patient’s underlying condition and presence of symptoms. If higher ventricular tracking rates are desired, then one should increase the MTR to 170 or 180 bpm.

- In the atrial histogram, there is a cluster of high atrial rates; consider atrial arrhythmias (Fig. 3.3.16). In pacemaker check, one should determine if patient has symptoms consistent with atrial arrhythmias, but would also need to rule out presence of oversensing with the sensed events filling the higher atrial rate bins in the histogram.

Sensor Rate Counter

The activity sensor is used in patients with decreased chronotrophic competence. Today, almost all pacemakers have a built-in programmable activity sensor. The same kind of counter or histogram as provided for atrial and ventricular heart rates is usually provided for the activity sensor rates. If the pacemaker is in a rate response mode, the sensor will adjust pacing rate in response to physical activity. In some patients with some intrinsic response to exercise, the intrinsic rhythm may be able to exceed the sensor rate. A blunted peak heart rate on the sensor in a VVIR pacemaker has to be correlated with the patient’s activity level or

Figures 3.3.14 and 3.3.15. Usually, the pacing and sensing events are labeled in the counters/histograms as well as at what rates pacemaker interpreted as PACs or PVCs occur. In some models, you can activate storing of high atrial and ventricular rates. In some models, a figure of heart rate over time for the last 24 hours is provided. Here are two examples of data you might find in pacemaker check.

- DDD pacemaker, MTR 130 bpm: The atrial histogram shows an expected pattern of cluster around lower rates and tailed toward higher rates, with the peak rate at 170 bpm. The ventricular histogram shows no rates over 130 bpm. Cause: MTR is programmed lower than patient’s potential sinus tachycardia rate. Adjustment: Depends on the patient’s underlying condition and presence of symptoms. If higher ventricular tracking rates are desired, then one should increase the MTR to 170 or 180 bpm.

- In the atrial histogram, there is a cluster of high atrial rates; consider atrial arrhythmias (Fig. 3.3.16). In pacemaker check, one should determine if patient has symptoms consistent with atrial arrhythmias, but would also need to rule out presence of oversensing with the sensed events filling the higher atrial rate bins in the histogram.
symptoms. Programming adjustments should be considered to address both symptoms and the patient’s needs.

**AV and Ventriculoatrial Intervals**

If this information is given, one expects the AV times to be no longer than the programmed AV/PV intervals nor any shorter than your minimal settings if you use rate-adaptive AV/PV intervals. One should be aware of different kinds of programmable algorithms for intrinsic conduction search that may allow longer AV times intermittently in order to conduct its search for intrinsic conduction. Clustering of both shorter and longer times is usually caused by atrial and ventricular arrhythmias. The VA time overview (when provided in some pacemakers) can show interesting information. If one finds a cluster of VA time intervals around 300–400 ms and is fixed, then this is suspicious of the presence of retrograde conduction to the atrium outside the PVARP. This may potentially cause PMT.

**Mode Switch**

In a DDD pacemaker, atrial tachyarrhythmia with high ventricular tracking rates is an obvious concern. All pacemakers have algorithms trying to prevent this. The simplest form is when the pacemaker transiently changes the mode of operation when the atrial rate increases above a programmed rate limit. Usually, the device switches to a nontracking mode (DDI or DDIR), hence the term mode-switch. An example is when we have a DDD pacemaker with a base rate of 60 bpm. The patient then goes into atrial flutter with a rate of 280 bpm. The pacemaker detects the high atrial rate and switches to DDI mode 60 bpm. This means that the device will continue to sense the high atrial rate but maintains ventricular rate no slower and no faster than 60 bpm. When the atrial flutter terminates and returns to sinus rhythm, mode-switch will occur again to DDD, and the ventricular channel will resume tracking sinus rhythm.

To prevent sudden drops in ventricular rate during mode switch, the mode may switch to DDIR where the ventricular rate will be dependent on the activity sensor. Another possibility is that a pacemaker has programmable rate regularizing features (e.g., rate smoothing, rate stabilization) to make gradual adjustments in the ventricular rate to minimize sudden rate changes. Young people who normally reach higher atrial rates in their day-to-day living are a separate challenge in this regard. The limit for detection of high atrial rates will need to be set higher than the expected sinus rate ranges to avoid inappropriate mode switch from sinus tachycardia.

From the summary information page, most pacemaker models will give a message alert if any high atrial or ventricular rates have been detected in the counter. Sometimes, EGM details are stored that can be reviewed together with the markers. This is particularly helpful as it will help decide if there is true tachyarrhythmia or if detection was due to oversensing. The specific time of the event will be given together with the duration, the atrial rate, and/or the ventricular rate during the event. You should always ask if the patient can remember what he or she was doing or experiencing during that time and if he or she had any symptoms.

**AT/AF Log**

AF and atrial flutter is a significant problem that is not uncommon in the pacemaker patient population. In some pacemakers, this log provides additional information about the arrhythmia rates, duration, total, and relative time spent in AT/AF. This is a useful tool for the clinician in evaluating different treatment modalities and the burden of disease. In some instances, when there is problem with far-field ventricular oversensing on the atrial lead, some
of the oversensing episodes may be included in the AT/AF log.

**Leads**

Lead impedance should always be monitored as it reflects the integrity of the pacing lead. Even though leads have different normal impedance ranges, values under 200 ohms may suggest that there is insulation break. This may result in loss of capture, sensing problems, and stimulation of skeletal muscle if the break is within the pacemaker pocket. Impedance values over 2,000 ohms may be consistent with lead conductor fracture. This may result in the inability to capture, sensing problems, or if there is a complete break of the conductor, pacing output failure due to the stimulation current not reaching the tissue. In general, we monitor for relative changes in lead impedance value trends between follow-ups. If an abrupt change occurs, then further investigation is needed to determine the integrity of the lead. In some cases, if the impedance changes were associated with normal pacing threshold or sensing threshold, and the patient is not pacemaker dependent, then one may elect to continue with close monitoring rather than surgical replacement of the lead. The management options clearly must be individualized for each patient.

Some pacemakers perform daily impedance measurements and present this data in a trend. These trends are helpful in pacemaker follow-up. Impedance trending upward together with increased thresholds and decreased sensing is an indicator that the lead integrity is suboptimal and should warrant closer follow-up or replacement. Chest X-ray can also be useful in this situation to determine if there is any lead tension that might be the cause of the problem.

The advantage of using a bipolar lead is that when there are issues with thresholds or changes in impedance, one should consider assessing the measurements in unipolar configuration. Sometimes, the lead conductor to the distal pole (cathode) may be intact but the lead to the proximal pole (anode) is disrupted. If unipolar measurements are acceptable and within normal, then reprogramming the polarity to unipolar can be made and emergent lead replacement surgery is avoided. In most pacemakers, there is a programmable option to automatically switch polarity from bipolar to unipolar when out-of-range impedances are detected. Before turning this feature on, one should perform threshold measurements in unipolar to ensure that its performance is acceptable, that is, not worst than bipolar. Although changing to unipolar is a reasonable option, the patient should be monitored closely since partial damage to one conductor may indicate potential overall lead problem.

**Battery**

Battery voltage and/or battery impedance are standard information from present-day pacemakers. Most pacemakers will give you an estimation of remaining longevity in time. Factors affecting battery life are pacing output, lead impedance, amount of pacing in respective chamber, heart rate, and internal pacemaker current consumption. The status of the battery may be described as good, elective replacement near (ERN); time to intensify follow-up, elective replacement time (ERT), elective replacement indicator (ERI), or end of life (EOL).

The pacemaker battery depletes in a very predictable manner. As the battery voltage depletes closer to ERI, the battery status should be monitored on a more frequent basis, either monthly or every 2 months. Telephone transmission of the pacemaker’s magnet rate may be useful when monthly monitoring is used to determine when the pacemaker reaches ERI. However, the magnet response is manufacturer specific. For example, the magnet rate may start off at 97.5 or 100 bpm at beginning of life, and then gradually decrease to 85 or 65 bpm at ERI. Figure 3.3.17 shows how the pacemaker battery voltage depletes over time.
Chapter 3.3 Pacemaker Timing Cycles

The basic tests that should be performed are pacing threshold and sensing threshold. Provocative maneuvers may be performed if there are concerns about the lead integrity or oversensing problems. Provocative test may be performed by asking the patient to hold her hands together and pull. If a lot of activity is seen in the EGM and it is sensed by the pacemaker, we call it oversensing of myopotentials. Additional tests such as ambulatory Holter monitoring and exercise testing may provide validation of pacemaker function.

Every clinic’s strategy and data collection method may be different. For example, if our patient has a DDD pacemaker and is sensing the atrium and pacing in the ventricle, it makes sense to start with testing atrial sensing. Many patients with complete heart block experiences discomfort when we are testing for the intrinsic R wave sensing because the base rate sometimes needs to be decreased very low for the intrinsic rhythm to emerge. This part of the test should therefore be the last test to do. The idea is to perform follow-up testing as efficiently as one can with the least amount of programming steps.

**Intrinsic Atrial Rhythm**

This requires the intrinsic atrial rate to be higher than the programmed low rate in the pacemaker. It is helpful to turn rate response off to eliminate sensor-driven pacing (from pushing the programmer wand against the pacemaker area) from confusing the sensing test process. The lower rate should be reduced until intrinsic P waves are seen. When the test starts, the pacemaker either measures the amplitude of the P wave directly or adjusts the sensitivity level until sensing is lost. Depending on which method is used, either a range of signal amplitudes or a single value will be given. Some manufacturer test sensing up to a certain sensitivity value over the sensitivity value which you start the test. If you start your test at 1 mV, you may get >4 mV as a result. If you perform
the same test but starting at 2.8 mV, you will get 5.6–8.0 mV as result (if this is the case). The same principle applies to testing of R waves. After testing, one needs to ensure that the programmed sensitivity is at least twofold more sensitive. In the case of bipolar atrial sensing, one may program it to 0.5-mV sensitivity if there is no evidence of oversensing concerns. The rationale is to ensure the pacemaker can sense atrial arrhythmias whose P waves may have different sensing threshold from sinus rhythm.

Some of the challenges one may find are as follows. Bipolar sensing is often, but not always, better than unipolar. Patients with single-pass lead for VDD pacing may have marginal atrial sensing that is also dependent on body position. This is due to the floating atrial electrodes that are at a fixed distance from the tip of the lead that may be in a position that does not have large atrial signals (e.g., at the superior vena cava [SVC] junction). Patients with sinus node dysfunction have intrinsic junctional rhythm that may make atrial sensing assessment challenging. The P wave may be absent or after the QRS complex as a retrograde P wave. P waves can also be absent in the setting of a silent atrium. It is possible sometimes to get the patient to perform leg lifts while on the bed as a way to stimulate sinus or heart rate acceleration. Reviewing the atrial EGM is helpful and can be used to compare with the EGM snapshots that are stored with high rate detection.

**Intrinsic Ventricular Rhythm**

Appropriate ventricular sensing is important in patients with pacemakers. If an intrinsic ventricular beat is undersensed, there is a risk of pacing at the peak of the T wave, which might cause ventricular fibrillation. This is one of the reasons why VOO mode should never be used other than in very special circumstances. Oversensing, on the other hand, can lead to the inhibition and resetting of the ventricular timer. This can cause pauses or asystole. It is usually not possible to test ventricular sensing in DDD mode because of ventricular tracking of atrial rate. The exception from this are patients with intermittent AV block. In this case, it might be enough just to prolong the AV/PV delay. Otherwise, in complete heart block patients, VVI mode is used to conduct the test. The sensitivity setting should be set at least twofold more sensitive than the sensing margin or set nominal at 2.5 mV.

Some of the challenges one may find are as follows. When performing the test in patients with ventricular pacing, the phenomenon of overdrive suppression may be present. This phenomenon may be seen in atrial paced patients as well. Overdrive suppression means that if the intrinsic heart rate is 40 bpm and we pace 90 bpm, then abruptly stop pacing or have sustained loss of capture, a significantly longer pause may occur before intrinsic rhythm resumes. This may also cause patient symptom or discomfort. Overdrive suppression is more likely to be seen in patients with wide QRS complex escape rhythm, sick sinus syndrome, or unstable escape rhythm. Because of this, the pacing rate should be decreased gradually rather than abruptly decreased from a high rate to a low rate. This will reduce patient discomfort during testing and increase the chance of successfully obtaining stable intrinsic rhythm to conduct sensing test. Some pacemakers permit you to gradually lower the rate when you are in a temporary test. For example, if you start at VVI 50 bpm in the sensing test screen and no intrinsic rhythm is sensed, the programmer will ask and allow you to decrease the low rate while continuing the test in the temporary test screen. In other pacemakers, you may need to change the permanent programmed rate gradually until intrinsic rhythm is seen and then run the sensing test. The patient can also be asked to move legs or perform other exertion maneuvers to help increase the rate of the escape rhythm. Some patients do not tolerate a very low ventricular rate. If no intrinsic rhythm
is seen at a programmed rate of 30 bpm, the patient is regarded as pacemaker dependent.

**Atrial Threshold**

Threshold testing in both the atrium and ventricle is a key feature of a pacemaker check. The principle is easy. The goal is to ensure adequate pacemaker function. The practice is usually to program a twofold amplitude voltage margin or threefold pulse width margin above the pacing threshold. The pacing rate is increased to overdrive the intrinsic rate. The threshold test routine usually permits testing of either output pulse width threshold (while holding voltage constant) or output amplitude (voltage) threshold (while holding pulse width constant). Output is gradually decreased until capture is lost (Fig. 3.3.18). Threshold is considered as the minimum output setting at which consistent capture is maintained.

In the atrium, it can sometimes be difficult to see the atrial response to pacing. If the programmer displays an atrial EGM, this may be a helpful way to see capture versus noncapture. Keep in mind that the morphology of the local EGM to pacing might change as the amplitude of the pacing stimulus decreases. The surface ECG shown in the programmer can also be used, and sometimes, you may have to select and display the lead that shows the best P wave. If no EGM is available and P wave is not seen on the programmer ECG, you may try an external ECG monitor or use 12-lead ECG. In the DDD pacemaker, increasing the AV delay can sometime make it easier to see the P wave.

In patients with AV conduction, a sudden change in ventricular rate implies loss of atrial capture. You can either start the test at the permanent programmed output setting or a few settings above the previous output setting. Here is a suggestion on how to perform the test:

1. Start by testing the amplitude threshold.
2. Check that the temporary test settings are appropriate.
   - AAI/DDD (AAI can only be used with intact AV conduction)
   - Low rate set 10–20 bpm over intrinsic atrial rate
   - AV delay 200 ms
   - Amplitude 1 V over previous results or as permanent setting
   - Leave the pulse width as per the permanent settings, or adjust if you need to check threshold at a different pulse width setting
   - Number of beats between each decrement: five or adjust as needed
3. Perform test.
   - If you are not sure where you lost capture, you can review the saved test strip and identify the point at which loss of capture occurs
4. If the threshold is higher than the current battery voltage or has increased substantially, you should retest the amplitude with a longer pulse width or different pacing configuration (bipolar or unipolar). One expects the voltage threshold to be less with slightly

![Figure 3.3.18 Loss of atrial capture during threshold testing.](image)
Cardiac Arrhythmia Management

wider pulse width. The rationale and goal here is to try to reduce the permanent output voltage by increasing the pulse width setting.

**Ventricular Threshold**

In patients who are fully paced, there is usually little doubt when capture in the ventricle is lost as evidenced by the pause in rhythm. Ventricular threshold test can be done in DDD mode in a dual-chamber system (Fig. 3.3.19) or VVI in a single-chamber system. If intrinsic rhythm is present, the testing rate will need to be increased. The same steps as in testing of atrial threshold can be followed. It is important to watch carefully and stop the test when capture is lost to avoid producing long asystolic periods. Sometimes, threshold rise may occur and the threshold measurement appears to be inconsistent or unstable. Performing the threshold test with a small number of test pulses (e.g., two) may give a false low threshold value. In such cases, repeating the threshold test to allow 10 test pulses per each output decrement may give a more reproducible and realistic measurement. Consideration should also be given to test capture threshold in unipolar pacing if bipolar threshold is high, as sometimes, unipolar threshold may be lower. However, the risk of skeletal muscle stimulation is higher with unipolar leads. High-output testing at the clinic may help assess this possibility and determine if there is a margin of safety in an output setting between absence and presence of muscular stimulation. Patients should be asked to report symptoms of intermittent skeletal muscle stimulation such as pocket stimulation to the clinic staff so that programming adjustments can be made to correct the problem.

**Ventricular Automatic Pacing Threshold Algorithm**

In ventricular automatic pacing threshold determination, the pacemaker must be able to sense the evoked response in the ventricle to a given stimuli to ensure capture. However, during pacing, there is a certain amount of residual lead polarization and the pacemaker has to be able to distinguish this from the evoked response in a safe way. Thus, the setup often includes an assessment of the evoked response followed by automatic threshold test. During this test, loss of capture is not seen since it would be accompanied by delivery of a backup pulse shortly after the loss. Depending on the manufacturer, the automatic testing algorithm is done on a regular basis (e.g., during the day) or on a beat-to-beat basis. It reduces the output until capture is lost and then immediately delivers a backup pace of at least 4V in most devices. The automatic output is then usually set 0.25V or at a programmed margin above the measured threshold. This is to minimize battery consumption.

One nice consequence of this is that you will be provided with a long-term trend of the measured thresholds. At interrogation, you will also be provided with the latest tested threshold. This feature has clinical utility for remote follow-up monitoring of pacemakers. There will still be the option of repeating the test or conducting a “manual” test in clinic, even though this is not mandatory during a follow-
up where everything is unchanged and the trend is stable. If your patient complains of skeletal muscle stimulation at certain intervals of the day, the cause might be the scheduled testing of threshold with subsequent backup pulse delivery (sometimes, this backup pulse is unipolar).

**Atrial Automatic Pacing Threshold Algorithm**

This feature is relatively new. There are two methods of automatic determination of atrial pacing threshold. Direct method measuring evoked response potential (St. Jude Medical) and the indirect method that uses the atrial rhythm response to loss of atrial capture to determine presence of capture (Medtronic). The St. Jude Medical ACAP Confirm operates slightly differently from its counterpart ventricular AutoCapture in that it uses bipolar pacing configuration and does not run on a beat-by-beat basis. ACAP Confirm runs a threshold search every 8 or 24 hours (programmable) and compares the measured evoked response with lead polarization to determine if there is an adequate safety margin to reliably determine capture. If there is an acceptable margin, then the threshold test can run and output setting can be adapted to add 1.0 V above thresholds ≤1.5 V; up to 2.0 V added for thresholds between 2.375 and 3.0 V.

The indirect method uses either the atrial reset method or the AV conduction method to determine atrial capture. Medtronic’s Atrial Capture Management conducts an atrial threshold search once a day only if the atrial rate is not faster than 87 bpm or if the atrial pacing rate is slower than 90 bpm. In the atrial reset method, loss of atrial capture is inferred by refractory sensed atrial event in the AV interval. This method will not work in patients with sick sinus syndrome since the sinus rate is often slower or easily overdrive suppressed by pacing. In the AV conduction method, the patient must have stable 1:1 AV conduction during atrial pacing rate that is no faster than 101 bpm and the AV interval is extended for the duration of the test to achieve atrial pace ventricular sensed rhythm. Loss of atrial capture is inferred when there is absence of ventricular sensed event during the test. This method will not work in patients with complete heart block or second-degree AV block at atrial rates less than 101 bpm.

**Retrograde Conduction Test**

The purpose of performing this test is to determine if the patient has the risk of developing PMT. An automatic test or an abbreviated test can be done manually and with the atrial EGM present. While pacing in the ventricle, the atrial EGM or surface ECG is observed for retrograde atrial conduction. When the VA relationship is stable and clearly associated with the pacing rate, you should measure what the VA time (ms) is. PVARP should be programmed slightly longer than this measured retrograde VA interval to exclude sensing of the retrograde P wave, and the PMT prevention algorithm of the pacemaker may be turned on.

**Rate Response Sensor Test**

If the patient is in need of a rate-responsive mode and there are indications that the current settings are not adequate, a simple activity or exercise test can be performed to determine if the sensor settings are appropriate. The test is simply started by the programmer, and the patient is instructed to do different kinds of physical activity for a couple of minutes such as brisk walking, running, or stair climbing. When the test is done, the programmer will display the results including the actual heart rate response to exercise as well as the predicted heart rates with different settings of the activity sensor.

**Programming**

After the tests are conducted, the decision time comes to determine how best to optimize
programming for the patient. This is where it becomes challenging. Different manufacturers have slightly different nomenclatures and features. Settings must be adjusted according to age, indication, and intercurrent disease. This is also where the problems discovered during diagnostic and testing may be resolved by making programming changes.

**Output**

Early after implant, threshold changes occur due to inflammation and healing as part of the body’s foreign body reaction; a higher output safety margin is maintained for the first 3 months. For acute lead implant thresholds under 1.0 V, we usually leave the pacemaker at the nominal setting of 3.5V and increase to higher outputs if the thresholds are higher.

The general rule for chronic output setting is to always have a two times safety margin for the amplitude or a three times margin for the pulse-width thresholds. In patients who are highly dependent on pacemaker, a greater safety margin may be used. On the basis of the strength duration curve, one rarely programs voltage amplitude below 2.0V, or a pulse width under 0.3ms, since these values approach the steep part of the strength duration curve. In these instances of output settings, slight variations in thresholds may result in outputs that are below the capture threshold curve. If the programmed amplitude setting is more than 2.5 V, a voltage multiplier is used and the battery longevity will be reduced.

**Sensitivity**

The general rule is that the programmed sensitivity for signal detection should be at least half the measured P or R wave amplitude (mV). Bipolar sensing is less prone to oversensing of myopotentials or far-field signals. For bipolar R wave >5mV or P wave >2mV without any issues of oversensing, one can leave the sensitivity setting at the nominal values of about 2.5 or 0.5 mV, respectively. The rationale for this is that ectopic beats or intrinsic rhythm from different foci may have different amplitudes (mV) or slew rates that may not be sensed if the programmed sensing margin is set at the twofold margin.

When we talk about increasing the sensitivity (mV) setting, we mean lowering the actual number, which will decrease the set limit for signal detection. For example, if the programmed sensitivity is 4.0 mV, the sensitivity is increased or made “more sensitive” by changing it to 2.5 mV, as the pacemaker will then be able to sense all signals with an amplitude of 2.5 mV or greater, as opposed to only sensing signals that are larger or at least 4 mV in amplitude.

**Heart Rates**

**Base Rate**

Base rate programming is determined by the patient’s age and intercurrent disease conditions. The nominal setting in most pacemakers is a base rate of 60bpm. In DDD pacemaker in a patient with normal sinus node function, the base rate really is the lowest heart rate that we are comfortable letting the patient experience. Most often, the base rate is left at 60 or 50bpm, and rest rate or sleep rate is set to 50 or 40bpm. In case of sinus node dysfunction or single-chamber pacemaker, the base rate will more often determine the patient’s actual heart rate. In some patients prone to arrhythmias such as atrial flutter, the base rate may be programmed higher than normal in an effort to reduce the potential of premature ectopies that may initiate the arrhythmia. There are many different scenarios and often not much scientific evidence supporting different settings.

**Maximum Tracking Interval (MTR)**

Modern pacemaker allows an MTR to be set as high as 180bpm or even up to 210bpm. The
higher rates are usually applied to the pediatric population or young active persons as needed. The programmed AV interval and PVARP play a big role in determining if the pacemaker can reach the target MTR. Even if a physically active teenager will do perfectly fine with an MTR of 180 bpm, he or she might not appreciate the sudden drop to 95 bpm that happens when the atrial rate reaches 190 bpm (if that is the 2:1 block rate dictated by TARP). If you expect that the peak atrial rate will exceed the 2:1 block rate during activity, you should adjust the settings to push out the 2:1 block rate. You may use the formula of (220 bpm — age) in years to estimate the expected peak heart rate. In some patients, you might want to keep the MTR down such as in ischemic heart disease as they do not tolerate rapid pacing rates.

**Sleep Rate/Rest Rate**

Almost all pacemakers have the option of sleep rate or rest rate not only to mimic to some degree the circadian rhythm but also to help preserve battery longevity by reducing unnecessary pacing during sleep. This feature may function on the basis of a programmed clock time or of the activity sensor’s detection of inactivity over a 20-minutes period. You will also need to define the lower rate that you allow the pacemaker to go to (usually 10 bpm below the daytime base rate).

**Maximum Sensor Rate (MSR)/Activity Sensor**

As always, different manufacturers have somewhat different nomenclature. The general concept for programming the activity sensor is, however, quite straightforward. The following are some considerations in programming.

MSR—this setting depends on the same factors as for the MTR including considerations such as patient’s age, disease condition, and physical life style.

How much activity is needed for the sensor to respond? This is the sensor threshold, the amount of body motion required for the sensor to begin to respond.

How quickly should the rate increase at the start of physical activity? This is the sensor slope and ranges from a very blunt slope (not much increase in sensor pacing rate) to a very steep slope (rapid increase in sensor pacing rate to reach the MSR).

How quickly should the rate increase or decrease to the target range when physical activity has started and stopped? This is the onset or offset and is programmed in time (minutes).

Fortunately, all manufacturers have nominal values or automatic adjustments of them. This leaves you with the MSR to program and tailor the automatic adjustments to the patient’s activity style. For the other parameters, you can almost always start with the standard settings and evaluate it during the next follow-up. Doing an activity/exercise test is a very good way of optimizing sensor responsiveness.

**Rate Hysteresis**

Hysteresis was discussed in previous section. From the point of programming, you just need to turn it on and define the lower hysteresis rate. Some pacemakers require you to define the sensed interval or cycle length (ms) beyond the base rate or cycle length that will be tolerated before the pacemaker starts to pace again at the base rate. An important consideration is that this feature should not be used for patients who are prone to develop bradycardia-induced or pause-dependent arrhythmias and includes patients with long QT syndrome.

**Mode Switch**

To turn mode switch on, you need to program the AT detection rate at the atrial rate at which
you want the mode-switch to occur. One needs to consider that this detection rate is beyond the normal range of sinus rate for the patient. This rate has no impact of the pacemaker timing intervals; it is strictly an atrial rate at which the algorithm begins to function. At follow-up, you should analyze when mode-switch occurred and whether it was appropriate. Oversensing problems may cause inappropriate mode switch to occur because artifact is being sensed and will falsely increase the atrial rate.

**Additional Testing in Pacemaker Follow-Up**

Even though the pacemaker check in the clinic is usually sufficient to identify problems and address them, there are additional tests that may be useful to validate the pacemaker function. These tests are ordered based on clinical indication and some may be performed more routinely.

**Ambulatory Holter Recording/Event Recording**

The Holter is a good adjunctive test to assess pacemaker function in the patient’s activities outside of the follow-up clinic. The indications for performing Holter recording are to detect intermittent problem with sensing and capture, response to activities or work environment, correlation of patient symptoms with pacemaker behavior, or clinical suspicion of pacemaker dysfunction (such as when the thresholds are increasing, diminished sensing threshold). Holter may be performed after implant of new leads, regularly every few years or only based on clinical indications.

Event recording may have some utility by providing an ECG snapshot in pacemaker patients who have intermittent symptoms such as palpititations, presyncope, or dizziness despite normal pacemaker check. Some pacemakers may allow patients to activate storage of EGM strip or marker data by placing a magnet over the pacemaker at the time of symptoms.

**Chest X-ray**

A chest X-ray is always performed after the implant of a new lead to document the lead position as well as to exclude possible implant complications such as pneumothorax or hemothorax. The X-ray can be used to assess a lead fracture, lead dislodgment, lead perforation, lead connections to the pacemaker, and lead tension resulting in a threshold or impedance increase. Lead insulation problems are usually hard to discern from chest X-ray. In growing children, stretching of the lead may be the cause of rising thresholds and warrants close monitoring.

**Exercise Testing**

Besides the usual indications of exercise capacity and ischemia, the exercise test can be used to study the URB of the pacemaker or responsiveness of the activity sensor. It is important to understand that body movement during a bicycle test will not be the same as on a treadmill and may show blunted activity sensor responsiveness.

**Follow-Up Schedule**

There are many factors affecting the frequency and assessments at follow-up such as time from implant, specific monitoring of problems, advisories, and battery reserve. Table 3.3.9 lists suggestions related to time from implant. The general guideline suggests more frequent follow-up in the first 3–6 months post implant, and thereafter every 6 months to once a year depending on pacemaker type, pacemaker dependency, and institutional practice. Follow-up frequency is increased again as the pacemaker battery depletes and approaches ERI.
Table 3.3.9  Suggestion of follow-up intervals and relevant tasks.

<table>
<thead>
<tr>
<th>Time</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>At implant</td>
<td>Test high-output pacing to ensure there is no stimulation of diaphragm or phrenic nerve</td>
</tr>
<tr>
<td>Before discharge</td>
<td>Program output to three times output margin</td>
</tr>
<tr>
<td>1 week</td>
<td>Wound check at local health care facility to ensure absence of infection</td>
</tr>
<tr>
<td>6 weeks</td>
<td>Routine check to ensure lead measurements are within normal</td>
</tr>
<tr>
<td></td>
<td>Optimize other pacemaker settings as needed</td>
</tr>
<tr>
<td>3 months</td>
<td>Routine check to determine chronic threshold; output is adjusted to two times safety margin</td>
</tr>
<tr>
<td>Every 6 months or once a year</td>
<td>Chronic follow-up schedule</td>
</tr>
<tr>
<td>Battery status: ERN</td>
<td>Increase frequency of follow-up to every 2–3 months</td>
</tr>
<tr>
<td></td>
<td>Consider use of telephone transmitter to assess magnet response to monitor ERI status</td>
</tr>
<tr>
<td>Battery status: ERI</td>
<td>Schedule for replacement</td>
</tr>
</tbody>
</table>

**Intercurrent Disease**

**Atrial Fibrillation (AF)/Atrial Flutter**

A patient with chronic AF should not be in DDD mode. Both DDI/R and VVI/R will work the same way and decrease the risk for ventricular tracking of the high atrial rates. If the patient has intermittent AF and good sensing properties in the atrial channel, the DDD mode can be used but special attention needs to be paid to have adequate settings for detection of AF and appropriate mode switch settings.

**Ischemic Heart Disease**

High heart rates increase oxygen consumption and both the MTR and MSR should therefore be limited in patients with ischemic heart disease. There is seldom a need to have the week of life. It then decreases gradually during life. When patient reaches the teenage years, the heart rate range approaches the adult range at the same level of fitness. In a VVI system, it is adequate to start with a base rate of 100bpm in newborns, 90bpm in 1-year-olds, 80bpm from 3 years, 70bpm from age 6, 60bpm in teenagers, and 60 or 50bpm in adults. We usually use the same numbers in a DDD system or lower rates if the sinus node function is preserved to avoid pacing at rates supported by intact sinus node firing. In a DDD pacemaker for children, the MTR is usually set to 180bpm, and AV delay and PVARP should be managed so that the 2:1 block rate is above 195bpm. In a VVI system, the rate response should be turned on once the child moves in a way that activates the sensor. Keeping the MSR around 160bpm is often enough. In adults, depending on the underlying disease condition and patient activity, the MTR of a DDD pacemaker or the MSR of a VVIR pacemaker may be programmed to 120–150bpm.
MTR/MSR rate higher than 130 bpm in these cases.

**Hypertrophic Cardiomyopathy**

As the name implies, the myocardium is thickened and often has a reduced perfusion. These patients may develop symptoms of dyspnea or angina. From this standpoint, the consideration of reduced MTR/MSR should be observed. There is also some evidence that right ventricular pacing may improve hemodynamics in this group of patients. This sometimes requires a short AV delay to be programmed. These patients are also at risk of developing arrhythmias; it is a good idea to track the number of PVCs or nonsustained ventricular tachycardia by the pacemaker diagnostics. This can be helpful in the clinical management of the patient should they require an upgrade to an ICD.

**Complex Congenital Heart Defects**

Heart block after cardiac surgery is the most common indication for permanent pacemaker in children. Some of these have had a complete repair with no residual lesions, and these patients often tolerate VVIR pacing well. Patients with significant residual lesions in which maintenance of AV synchrony is essential for their hemodynamic support require dual-chamber pacing. The largest group are patients with univentricular circulation such as post-Fontan repair. The surgery repair done at a young age is palliative and performed in stages. After the last surgery, they will have their superior and inferior caval veins directly connected to the pulmonary arteries (to bring deoxygenated blood to the lungs), such that the single ventricle will only pump the blood to the body (systemic circulation). If these patients have complete heart block, they will benefit from dual-chamber pacing for two reasons. First, they need the atrial contraction for optimal filling of the ventricle. Second, an increased atrial pressure due to contraction against closed AV valves in ventricular pacing (lack of AV synchrony) will impair the pulmonary blood flow that requires low pulmonary artery pressures and low pulmonary vascular resistance. A slightly higher base rate may be programmed in an effort to prevent atrial arrhythmias. The other consideration for this group of patients is the lack of venous access to the heart. Thus, the pacemaker leads are generally placed on the outside surface of the heart (epicardial).

**Pacemaker Patient’s End of Life Issues**

Medical considerations may arise in situations where it has been deemed necessary to withdraw care in a dying pacemaker patient. The pacemaker is considered a medical treatment that is ongoing and, unlike an ICD, rarely is the cause of the patient’s suffering. The decision to withdraw care becomes a dilemma in terms of what to do with the pacemaker. In a pacemaker-dependent patient, turning off the pacemaker will immediately terminate life or hasten death. There are currently few established guidelines to help clinicians in making these decisions: the ethical conduct, the processes that should be followed to carry out device deactivation, and what the legal implications are.

Clearly, if pacemaker deactivation is deemed necessary, there should be apparent documentations of prior discussion and signed consent with the patient, the family, or their legal personnel. The do-not-resuscitate (DNR) order should clearly specify if the pacemaker should be turned off at the time of carrying out a DNR order.

There are two ways in which a pacemaker can be turned off. For pacemakers in which a nonpacing mode (OVO, OAO, ODO) is avail-
able, this can be programmed. If this is not available, one can reduce the pacing outputs to the minimum settings (e.g., 0.2 V, 0.03 ms) and decrease the rate to the minimum (e.g., 30 bpm). It should be clearly documented in the patient’s chart who gave the order, who carried out the order, who is present, the programmed settings, and a copy of the programmed changes should be filed in the chart.

**TROUBLESHOOTING**

When problems arise in pacemakers, the key to troubleshooting is recognizing the problem and the causes and considering the differentials that should be ruled out. Pacemaker-related problems can be caused by any link in the system: the patient, the lead, the pacemaker, or its settings. Problems that are caused by hardware dysfunction can sometimes be solved by reprogramming, but often, there is a need for lead or pacemaker replacement. Patient-related problems can be the most challenging to address. For example, sometimes, patients may present in the clinic with AF or periods of ventricular tachycardia. The conventional management such as initiation of medications still apply. But further to this, one may optimize the pacemaker settings (AF suppression, PVARP extension on PVC, mode switch) that may intervene or prevent some of these arrhythmias. The findings must be communicated with the responsible physician so decisions about further treatment and examination can be made. In the following section, we will try to address different issues and how they can be resolved.

**Battery Depletion**

Pacemaker battery technology has proven reliability and predictable depletion characteristics. In the present day, it is unusual for a patient to present emergently with unexpected battery depletion. Occasionally, accelerated battery depletion may occur and it is not clear what the cause is. It may be the result of circuit malfunction or related to battery-intensive features (such as storage of EGMs). When there is earlier-than-anticipated battery drain, the device should be returned to the manufacturer for analysis.

Ideally, the pacemaker should be replaced while it is in ERI status. The concern with the battery that is at EOL is the erratic and unpredictable function of the pacemaker that can cause patient compromise. Low battery levels may trip the pacemaker to an electrical reset condition, causing the programmed parameters to change to the factory nominal settings or reset settings that may not be appropriate for the patient. The EOL behavior of pacemakers should be considered, particularly if the plan is to leave the pacemaker in situ (e.g., implanting a new pacemaker system at a different site or if the patient no longer meets pacing indications).

For patients in whom the pacemaker has been deactivated by reprogramming to nonpacing mode, when the battery depletes, it may trip the ERI indicator and revert to the ERI pacing mode, thus paradoxically reactivating the pacemaker.

**Lead Problem**

Lead hardware problems are challenging in terms of recognition and management options. During pacemaker interrogation, there are several clues that alert the clinician that something might be wrong with the leads: increased threshold, decreased sensitivity, change in impedance, loss of capture, no visual output on surface ECG, and stimulation of skeletal muscle or diaphragm. Suspicion of this warrants a chest X-ray. Sometimes, lead problems can be solved by reprogramming; if not, then lead replacement should be performed. The risks and benefits of surgical replacement must be balanced and differs for individual patients.
Lead insulation damage may present with low impedance measurements, oversensing, undersensing, or failure to capture. Likewise, lead fracture may present with similar findings except that the impedance will be abnormally high. Sometimes, the only way to unmask intermittent lead problems is by performing provocative testing. It is helpful during this test to continuously record the telemetered EGM and marker data to prove if lead “noise” or artifact is present.

**Oversensing**

**Far-Field Sensing**

An example of far field is when the atrial channel senses activity in the ventricle. One situation when this might occur is in case of sinus bradycardia with junctional escape rhythm and AAI pacing. If you try to perform a test of P wave amplitude in these patients, you will often find that the atrial channel is actually sensing the R wave. In such case, premature ventricular or junctional beats may reset the atrial timer and pace at a rate lower than the low rate limit. This can be minimized by decreasing the sensitivity (by selecting a bigger sensitivity number) or by increasing the atrial refractory period. On the other hand, in a DDD pacemaker, problems caused by far-field sensing of the R waves are less common due to the ability to program the postventricular atrial blanking (PVAB) to exclude far-field R wave sensing. This problem rarely warrants lead replacement unless it is causing significant pacemaker dysfunction.

**Myopotential**

If skeletal muscle activation (myopotential) is detected, the pacemaker may be inhibited from pacing, resulting in pauses. Unipolar sensing is more prone to myopotential inhibition and can be demonstrated by performing provocative maneuvers and watch the EGM and marker channel for sensing of myopotentials. To resolve this issue, the sensitivity may be programmed to be less sensitive. If this is not possible due to small intrinsic P wave or R wave, and patient has significant symptoms, revision of the pacemaker system to bipolar leads will help.

The occasional pauses from oversensing are usually well tolerated, and lead replacement is rarely indicated.

**Undersensing**

Undersensing is always a serious problem such that recognition is essential and ruling out the possible differentials is important. The concern with undersensing is the risk of competitive pacing-induced arrhythmia. Failure to sense P waves in a DDD system can lead to loss of AV synchrony. If the patient has an AAI system for sinus bradycardia with junctional escape rhythm, this is less of a problem. More concerning is undersensing of R waves. Inappropriate ventricular pacing on the peak of the T wave may risk inducing ventricular tachycardia or fibrillation. True undersensing may be corrected by repeating the sensing threshold test and increasing the sensitivity setting (selecting a smaller sensitivity number). Functional undersensing must be ruled out, for example, due to long programmed refractory period or blanking period, due to specific advance programmed features (e.g., PMT termination algorithm, PVC response), or due to magnet-induced asynchronous pacing. In these instances, adjustment of sensitivity value will not correct the problem.

**Capture Problems**

**Output Failure**

Output failure is present if the pacemaker does not deliver a pacing stimulus when expected. It can also be caused by battery depletion, setscrew disconnection, or lead fracture. In addition to the pacemaker check, an X-ray may
be performed to review the integrity of the lead, its location, and its connection to the pacemaker.

**Failure to Capture**

Causes of failure to capture include threshold rise that exceeded the safety margin of the output of the system, lead dislodgment, lead fracture, or insulation break. With the evolution of steroid-eluting leads, threshold problems are somewhat less common. It is important to remember that immediately after implant, there is often a rise in the thresholds that may eventually decrease to a lower chronic threshold.

**COMPLICATIONS OF LONG-TERM PACING**

Despite new advance programming features, a pacemaker will always be second best to sinus rhythm with normal conduction. This is especially true regarding pacing in the ventricle. Two major issues will be addressed.

**Table 3.3.10**  Signs and symptoms of pacemaker syndrome (adapted from Hurst).

<table>
<thead>
<tr>
<th>Signs</th>
<th>Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cannon A waves</td>
<td>Cough</td>
</tr>
<tr>
<td>Increased jugular venous pressure</td>
<td>Shortness of breath</td>
</tr>
<tr>
<td>Palpable liver pulsation</td>
<td>Neck pulsations</td>
</tr>
<tr>
<td>Peripheral edema</td>
<td>Generalized fatigue, weakness</td>
</tr>
<tr>
<td>S3 gallop on auscultation</td>
<td>Chest fullness</td>
</tr>
<tr>
<td>Pulmonary rales</td>
<td>Confusion</td>
</tr>
<tr>
<td>Decrease in systolic blood</td>
<td>Dizziness, presyncope, rarely</td>
</tr>
<tr>
<td>pressure &gt;20 mm Hg during ventricular pacing</td>
<td>syncope</td>
</tr>
</tbody>
</table>

**Pacemaker Syndrome**

A subgroup of patients who are paced in the ventricle without 1:1 AV synchrony may develop symptoms known as pacemaker syndrome. The signs and symptoms (Table 3.3.10) are a consequence of decreased cardiac output with the loss of AV synchrony. During ventricular pacing, regardless of the presence or absence of retrograde VA conduction, the atrium contracts against a closed AV valve. This increases the pulmonary venous and right atrial pressures leading to a vagal reflex of peripheral vasodilation and decrease in systolic blood pressure. Loss of AV synchrony may also occur in other settings such as loss of atrial capture in a dual-chamber pacemaker, AF, accelerated junctional rhythm, sinus rhythm with marked first-degree AV conduction, or PMT. A patient may develop pacemaker syndrome at any time post pacemaker implant; hence, it is important to recognize the symptoms. Restoration of AV synchrony through optimized programming such as increasing atrial output if loss of atrial capture is the concern or upgrade to dual-chamber pacing via implantation of atrial lead can alleviate the hemodynamic compromise.

**Ventricular Dysfunction**

Traditionally, ventricular leads are positioned in the apex and sometimes in the outflow tract of the right ventricle. This will create an abnormal activation pattern, where the right side will contract a little bit before the left, and thus reduce the effective cardiac output of the left ventricle. There is some evidence that reducing the amount of pacing in the ventricle is beneficial for the patient in the long term. For a patient with intermittent heart block, we can optimize the settings with prolonged AV interval, rate hysteresis, and intrinsic conduction search when available. For a patient with complete heart block, it gets more complicated. Rate hysteresis and intrinsic conduction search are of
Little use. AV/PV intervals can mostly be kept around nominal values of 150/120 ms. The question here is if the patient should have a single- or dual-chamber system and if the MTR should be high or low. One good example of this is the impact of long-term pacing in patients with congenital heart block.

**Postoperative Considerations**

**Lead Dislodgement**

Lead dislodgement is mostly a problem in the early postoperative period, before fibrous tissue has been formed around the lead-myocardium interface and around the actual pacemaker. Since the vast majority of pacemakers are transvenous, and the lead is inserted through the subclavian vein, we recommend that our patient not do excessive arm movement on the side of the pacemaker system, lift heavy weights, or have pressure over the shoulder for 2 months to allow for lead stabilization.

**Bleeding**

Bleeding in the pocket is the most common reason for early reintervention after pacemaker implant, and it also increases the risk for infection. High-dose heparin and aspirin in combination with thienopyridine has been described as risk factors for bleeding in adults. Direct trauma to the pacemaker-pocket can also cause bleeding and possible damage to the pacemaker/lead.

**Infection**

Pacemaker pocket infection is a serious complication to pacemaker insertion. Signs of infection include redness, swelling, pus oozing from the incision site, fever, tenderness, and warmth. Antibiotic prophylaxis may be given post implant. The acute early infection is therefore quite uncommon. More common, but still unusual, is the chronic infection with pain, swelling over the pacemaker, fluctuation caused by subcutaneous fluid collection, rubor, and ultimately, breakthrough with discharge of pus. Oral antibiotics, and sometimes intravenous treatment, have often been attempted, with temporary relief of the infection. If this does not resolve the infection, explantation of the pacemaker system is indicated. How long the patient should be without pacemaker after this depends on the underlying condition. Different institutions have different ways of addressing this. Some implant a new device in another location immediately, while some may use isoprenaline or a temporary pacemaker to increase the ventricular escape rhythm for a period of time until the new system is implanted.

Additional precaution to minimize risk of infection is to avoid getting the incision site wet until a crust is formed (usually 3–5 days) and the site should not be immersed in water until the crust has fallen off and there is a scar (usually an additional 2 weeks).

**Twiddler’s Syndrome**

Twiddler’s syndrome is a description of a condition whereby the patient turns the pacemaker around in its pocket before it has completely healed, consequently causing tangling of the leads in the pacemaker pocket and eventually resulting in lead dislodgment. Sometimes, patients subconsciously manipulate or massage the pacemaker pocket without realization of this potential risk of dislodging the lead. Thus, patient education is critical to prevent this from happening. The diagnosis is usually confirmed by chest X-ray showing the lead and pacemaker position.

**Extracardiac Stimulation**

Extracardiac stimulation is more common in unipolar leads or high-output settings. During lead implant, high-output pacing should be
Chapter 3.3 Pacemaker Timing Cycles

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Pure discomfort or pain without any underlying causes (besides the pacemaker being there) is, however, best treated conservatively. The patient needs to be reassured that everything is working according to plan and there is no serious problem with the system; the patient may also be referred for counseling as appropriate. With this, along with careful follow-up, and support, the symptoms often fade away.

Shoulder or upper arm pain of the affected surgical side may result when patients are overvigilant about restricting their activity to the implant side. The reduced movement of the shoulder can result in a “frozen shoulder,” or a shoulder that is causing pain because the muscles are tight as a result of inactivity. Slow, increased activity will result in pain relief, but sometimes, a physical therapy consultation will be required to provide the patient the necessary muscle retraining.

Electromagnetic Interference

Proximity of electronic devices to the pacemaker area may result in transient interference to the pacemaker function by oversensing of the electrical noise. Transient disturbance of pacemaker function may include inhibition from pacing, triggering pacing, asynchronous pacing, or inappropriate mode switch. As soon as the source of interference is removed, the pacemaker returns to normal function.

Very strong EMI, like electrocautery or defibrillation, can cause complete reset of the pacemaker to VVI or VOO mode. Magnetic resonance imaging is usually not recommended and if it needs to be performed, the pacemaker manufacturer should be contacted for details of compatibility and recommended MRI conditions. If surgery and interventions requiring electrocautery is needed, the pacemaker-dependent patient should be reprogrammed to VOO or DOO to assure pacing during the procedure, despite potential artifact or EMI interfering with pacemaker sensing. This can also be...
accomplished by placing a magnet over the pacemaker. The patient should be monitored throughout the procedure and the pacemaker should be rechecked after surgery if any reprogramming was done.

In general, electronics should be kept at least 6 in. (15 cm) away from the pacemaker area. This applies to cell phones, cordless phones, wireless communication items, electric shavers, and so on. Even the small magnets found in toys, earphones, and household magnets have been shown to interfere with the pacemaker if held immediately over the device. For some electronics or equipment, a larger distance from the pacemaker area may be needed. Depending on the duration and significance of the interference, the patient may or may not be symptomatic. If the diagnostic data suggest problems caused by electrical noise interference or if patient has symptoms, then careful history taking is needed to determine if something in the patient’s environment is the cause.

Patient education is the key to prevent this problem.

**Drug Effects on Thresholds**

Drugs such as flecainide and propafenone have been shown to significantly increase pacing threshold, while drugs such as glucocorticoids, epinephrine, or ephedrine may decrease pacing threshold. Other small studies have suggested that disopyramide, propanolol, verapamil, aldactone, or aldosterone are associated with an increase in pacing threshold, but the number of patients studied were very small. Isoproterenol was shown to cause an initial decrease followed by increase in pacing threshold. When a pacemaker patient is started on antiarrhythmic drugs (especially class I, III, or IV), it is recommended to recheck pacing and sensing thresholds after the drugs have reached steady state, particularly in patients with borderline or suboptimal pacing thresholds.

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### Case 3.3.1

A patient was assessed in the intensive care unit for irregular heart rhythm after dual-chamber pacemaker (St. Jude Medical) insertion. The pacemaker is programmed to DDD 60/180 bpm; the settings are shown in Figure 3.3.20. The rhythm strip of the irregular heart rhythm is shown in Figure 3.3.21. What is the differential diagnosis that one should consider as potential causes of the rhythm? What are the steps that we should go through to rule out the potential but unlikely causes of this irregular rhythm? What programming changes should we make to correct the problem?

**Discussion**

The rhythm showed ventricular tracking of atrial (probable sinus) rhythm in a group beating pattern at a regular atrial rate of 100 bpm; pauses occurred intermittently after a P wave that were not followed by ventricular pacing. Since the atrial rate is above the low rate limit, there is appropriately no evidence of atrial pacing. On the basis of this pattern, potential differential diagnosis include pseudo-Wenckebach URB, PMT algorithm, atrial undersensing, ventricular oversensing, and ventricular loss of capture.

Since the atrial rate is not faster than the upper rate of 180 bpm, this is not consistent with pseudo-Wenckebach URB: 1:1 P wave tracking will occur up to atrial rates of 180 bpm; URB is seen only when atrial rates exceed 180 bpm. PMT may occur at any rate depending on the retrograde conduction time. There is no evidence of any initiating events along with the group beating pattern that may induce loss of AV synchrony to start PMT. The upright P wave in lead II is also not consistent with a retrograde conducted P wave. The variable group beating pattern (3:2, 4:3) is not consistent with the device’s PMT algorithm. Thus, PMT is unlikely the
### Basic Parameters

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<thead>
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<td>Rest Rate</td>
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</tr>
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<tr>
<td>Post Vent. Atrial Blanking (PVAB)</td>
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<tr>
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### Sensor Parameters

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<tr>
<td>Max Sensor Rate</td>
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<td>130 ppm</td>
</tr>
</tbody>
</table>

*Not Applicable

T ⇒ Temporary programmed value

--- Parameter error(s)

---

**Figure 3.3.20** Pacemaker settings.

cause. Whenever ventricular pacing occurs, there is capture preceded by a distinct pacing spike on the rhythm strip. The absence of the pacing spike at the time of the pauses rules out ventricular loss of capture but may be consistent with ventricular oversensing.

This ECG pattern remains consistent with possible intermittent atrial undersensing or ventricular oversensing. From the ECG alone, we cannot rule out these two potential causes. If this occurred near the low rate and with atrial pacing present, we can use calipers to march backward from the atrial pacing spike to determine where ventricular sensing occurs (i.e., the VA interval).

Pacemaker check was performed and the marker channel data revealed that ventricular oversensing of the T wave was present (Fig. 3.3.22). Ventricular sensing testing was performed and sensitivity was decreased from 1.5 mV to 3.0 mV (less sensitive). This resolved the problem and normal DDD pacemaker function was restored.
Figure 3.3.21  Continuous ECG rhythm strip.

Figure 3.3.22  Printout from pacemaker check: surface ECG, marker channel data, and ventricular EGM.
Case 3.3.2

At scheduled follow-up in the pacemaker clinic, a 24-year-old man complained about exercise intolerance. He had just bought a membership to a gym and reported that the first few minutes on the bike or treadmill was fine, but then he “ran into a wall” and became very tired. His underlying diagnosis was complete heart block after repair of a ventricular septal defect at the age of 2. Initially, he had an epicardial right ventricular lead. After one pacemaker generator change, he was upgraded to a transvenous DDD system at the age of 12. This pacemaker was changed at the age of 18 because of battery depletion. The current settings are DDD, low rate 60 bpm, MTR 170 bpm, rate-adaptive AV interval 150/120 ms, paced AV/sensed AV interval 80 ms, PVARP 250 ms, ventricular refractory period 225 ms, and bipolar configuration for sensing and pacing.

On the basis of the clinical history, what are the differential diagnoses of his symptoms at this point? How should we evaluate these?

Discussion

The key features of this young man are as follows:

1. He has been subject to long-term right ventricular pacing. This is now a recognized potential cause of ventricular dysfunction and heart failure.
2. His transvenous leads were implanted at the age of 12. He is now fully grown and the leads may have been stretched, causing intermittent loss of capture or sensing problems.
3. The last pacemaker generator change was performed 6 years ago and the battery might be depleted, causing output failure. If battery depletion is reached, the mode may have switched to VVI 60 (ERI mode), explaining the exercise intolerance.
4. He is 24 years old and has an expected peak heart rate of around 196 bpm. The pacemaker may be exhibiting URB.
5. The symptoms are related to his level of inadequate physical fitness.

How do we address this in a pacemaker clinic?

1. Patient history: Besides the new exercise intolerance, there are no other symptoms to suggest chronic heart failure such as fatigue, shortness of breath, and fluid retention.
2. Baseline ECG at the clinic: Regular P waves 73 bpm with synchronous ventricular pacing without loss of capture. One adequately sensed PVC.
3. Pacemaker check: No changes in thresholds, sensing, and lead impedances to suggest lead dysfunction. Expected battery longevity was 1.5 years and the diagnostics did not suggest any atrial or ventricular arrhythmias; mode switch was not activated.

At this point, battery depletion and lead problem are unlikely. From here, there are several investigations that are appropriate: physical examination, ultrasound of the heart, chest X-ray, and exercise test. In this case, we decided to start with the exercise test since the symptoms are exercise related (Fig. 3.3.2).

What phenomena are occurring on the ECG recording? What changes in settings do you suggest?

The ECG recording of exercise test shows a typical URB. Wenckebach phenomenon starts at the MTR of 170 bpm. When the atrial rate reaches the 2:1 block rate (182 bpm), the ventricular rate drops even more (90 bpm). The 2:1 block rate is decided by the TARP, which equals the PV delay + PVARP (here 80 + 250 ms = 330 ms = 182 bpm). At this point, it is suspicious that this is the cause of the patient’s exercise intolerance.

We need to increase the 2:1 block rate above the patient’s peak atrial rate during exercise. This will expand his pseudo-Wenckebach window (between MTR and TARP) to reduce the sudden changes in ventricular pacing rate. The rate-adaptive AV interval is already turned on, and we should not use a shorter interval than 80 ms as it will be too unphysiological. The PVARP, however, is fixed at 250 ms and by turning on the rate-responsive PVARP here with a lower limit of 200 ms, we will get the 2:1 block rate of 214 bpm, which
Cardiac Arrhythmia Management

should be enough for the patient. We also increased his MTR to promote 1:1 tracking of his sinus at higher atrial rates during his exercise.

At this point, we can choose to let the patient evaluate the new settings in his day-to-day living or in his gym activities. If he still experiences the same symptoms, he should undergo other evaluations to rule out nonpacemaker-related causes that were described.

![Figure 3.3.23 Exercise test recording.](image)

**Case 3.3.3**

A patient was seen by his primary care for evaluation of a 3-week history of episodic dizziness and two episodes of witnessed syncope of up to 30 seconds in duration associated with seizures. This patient has a transvenous VVIR pacemaker implanted 10 months ago and has been functioning normally. Blood work, physical exam, and echocardiography were all normal. ECG and Holter were performed and recordings are as shown. The physician referred the patient back to the pacemaker clinic for assessment. On the basis of the clinical history, what are the possible causes of the patient’s syncope? Based on the recordings, what are the possible diagnoses of the pacemaker function? What other tests should we consider doing to help confirm the diagnosis?
Discussion

Syncope with or without seizures in a pacemaker patient is always a serious concern that one must always first rule out pacemaker malfunction as a possible cause of syncope. Pacemaker malfunction includes lead fracture or insulation damage that can result in episodic symptoms such as dizziness or presyncope. Malfunction related to the pacemaker generator is less common but can include battery depletion resulting in no output, or electrical circuitry reset of the pacemaker parameters that differs from what is programmed and may not be appropriate for the patient. Other causes of syncope may be ventricular tachycardia/ventricular fibrillation, vasovagal syncope, or epilepsy.

In this case, the initial workup by the primary physician was appropriate and did not yield any abnormal findings. ECG and Holter were very helpful as they showed evidence of intermittent pacemaker malfunction resulting in pauses in the paced rhythm, some as long as 1.5 seconds in duration in which the patient did not have any escape rhythm (Figs. 3.3.24 and 3.3.25). Careful inspection of the recordings showed oversensing (no pacing artifacts) and intermittent pacing artifacts without capture were the cause of the pauses in the paced rhythm. Both intermittent oversensing and loss of capture can arise from a number of causes including lead fracture, insulation failure, loose setscrew, and lead dislodgement resulting in false sensing of signals other than the QRS. Other causes of loss of capture may include lead perforation, but the patient would have presented with other severe symptoms such as chest pain, hypotension, and signs such as pericardial effusion or cardiac tamponade.

Pacemaker interrogation showed abnormal lead impedance trend with lead warning occurring 3 months prior. Heart rate histogram and high
Figure 3.3.25  Ambulatory Holter recording showing ventricular pacing with 15 seconds of asystole. Recording speed is 12.5 mm/s.

rate counters showed sensed events that were consistent with intermittent noise oversensing (Fig. 3.3.26).

Pacemaker check was performed and showed stimulation threshold that varied from 3.5 V 0.12 ms to 7.5 V 0.34 ms. Bipolar lead impedance was variable from 818 to >9,999 ohms. During testing, the patient was noted to have overdrive suppression of his intrinsic rhythm when loss of capture occurred. During sensing test of the patient’s intrinsic rhythm at 44 bpm, rare noise artifact was noted on
Figure 3.3.26  Pacemaker diagnostics showing ventricular lead impedance trend on the top and heart rate histogram on the bottom.
the ventricular EGM. Provocative testing with isometric hand squeeze and stretching of the arms elicited the noise artifacts that were sensed by the pacemaker. These noise artifacts were likely due to make-break fracture of the lead. Both the noise artifacts and the variable stimulation threshold were consistent with intermittent lead hardware malfunction. Chest X-ray was performed and a tight turn of the lead was noted near the lead tie down where the lead entered into the left subclavian vein (Fig. 3.3.27). This was suspected as the cause of tension on the lead that may have resulted in the problems that were seen clinically.

The patient underwent immediate pacemaker lead replacement; the old lead retracted easily and was returned back to the manufacturer for analysis. The analysis showed that there was torn insulation adjacent to the partial conductor fracture area due to friction of the lead body with the tie down at the location of the tight bend that was shown on the chest X-ray.

The take-home message from this case is that if a pacemaker-dependent patient has loss of consciousness, pacemaker dysfunction must be ruled out in an emergent manner.

**RESOURCES**

3.4 Cardiac Resynchronization Therapy
Melanie Turco Gura and William R. Lewis

EPIDEMIOLOGY

Heart failure (HF), a progressive and debilitating disease (Pappone et al. 2001), remains a major health problem in western society. In the United States, approximately 4.9 million people have chronic HF, with 400,000–700,000 newly diagnosed cases yearly. (Levy et al. 2002; Roger et al. 2004). As a cardiogeriatric condition (Sweeney 2008), HF is increasing due to the aging population of individuals with coronary artery disease, which is now the principal cause of HF in patients with dilated cardiomyopathy and reduced ventricular function (Kannel and Belanger 1991; American Heart Association 2008; Sweeney 2008). Due to significant changes in the management of patients with progressive HF with dilated cardiomyopathy, mortality has declined. Pharmacological agents, including beta adrenergic blockers, angiotensin-converting enzyme (ACE) inhibitors, angiotensin receptor blockers, and aldosterone antagonists, have yielded significant reductions in mortality due to progression of pump failure (CONSENSUS Trial Study Group 1987; SOLVD Investigators 1991; O’Connell and Bristow 1994; Packer et al. 1996, 1999; MERIT Merit-HF Study Group 1999; Pitt et al. 1999; Bubien and Ching 2004; Sweeney 2008). However, HF still confers a 20–25% risk of premature death in the first 2.5 years after diagnosis (Sweeney 2008). HF is the leading indication for hospitalization in elderly patients (SOLVD Investigators 1991; Packer et al. 1996; MERIT Merit-HF Study Group 1999; Bubien and Ching 2004; American Heart Association 2008), accounting for $40 billion annual expenditures in the United States (O’Connell and Bristow 1994; McMurray and Stewart 2000).
ETIOLOGY

HF may result from diseases of the myocardium, pericardium, endocardium, valve structures, the great vessels, or abnormal rhythms (Gura and Foreman 2004); however, HF usually results from left ventricular (LV) systolic or diastolic dysfunction. Patients with systolic dysfunction, defined as an LV ejection fraction (EF) of $<40\%$, typically present with an enlarged LV and a decreased cardiac output. Patients with diastolic dysfunction, defined as a preserved LVEF $>40\%$, have abnormal relaxation due to a noncompliant LV. Hypertension, restrictive or hypertrophic cardiomyopathy, valvular heart disease, diabetes mellitus, and other conditions commonly cause diastolic dysfunction (Levy et al. 1996; Sharpe and Doughty 1998; Gura and Foreman 2004).

ELECTROMECHANICAL DYSSYNCHRONY

It is common for patients with advanced HF to develop conduction abnormalities, which include prolongation of the PR interval and the QRS complex (Williams et al. 1968; Wilensky et al. 1988; Schoeller et al. 1993; Xiao et al. 1996; Gura and Foreman 2004). Left bundle branch block (LBBB), right bundle branch block and nonspecific intraventricular conduction delays (IVCDs) have been reported in 31–53% of patients with advanced HF (Maguire et al. 1987; Xiao et al. 1996; Aaronson et al. 1997; Shamin et al. 1998). The duration of the QRS complex increases as LV function worsens, with the widest QRS occurring in patients with New York Heart Association (NYHA) class IV HF. Prolongation of the QRS interval has been found to be an independent predictor of mortality in numerous studies of HF patients (Wilensky et al. 1988, Schoeller et al. 1993, Xiao et al. 1996, Shamin et al. 1998, Juliano et al. 2002), with the highest mortality rates occurring in patients with a prolonged QRS duration, and a LVEF $<35\%$ secondary to ischemic and nonischemic cardiomyopathy (Huang et al. 1995; Silverman et al. 1995; Shamin et al. 1998; Juliano et al. 2002; Shenkman et al. 2002; Bade et al. 2004; Freudenberger et al. 2004). These electrical timing abnormalities adversely effect critical mechanical interactions that further impair LV function.

Optimal contraction of the myocardium is a result of the synchronized, sequential activation of the atria and ventricles. Normally, LV activation anticipates right ventricle (RV) activation. However, in patients with dilated cardiomyopathy, five major disturbances in the normal electromechanical events may occur in isolation or in combination. They include atrial decoupling, atrioventricular (AV) decoupling, ventricular decoupling, IVCD, and LV transmural delay (Sweeney 2008).

Atrial Decoupling

In normal sinus rhythm (SR), the right and left atria are activated within 50–80 ms (Takasi et al. 2004). In patients with HF, electrical atrial remodeling results in slow conduction throughout the atria and Bachmann’s bundle, an increase in the atrial effective refractory period, and functional conduction delays at the crista terminalis (Bernheim et al. 2005). This leads to significant interatrial conduction delays (up to $\geq 200\text{ms}$), which may result in a disruption of optimal left-sided AV coupling. In severe cases, left atrial contraction delay can occur simultaneously or after LV contraction, resulting in abnormal hemodynamic and neurohormonal responses (Sweeney 2008).

AV Decoupling

Prolongation of the PR interval, or AV decoupling, has a negative impact on ventricular performance, resulting in suboptimal preload due to the delayed onset of ventricular systole in
Chapter 3.4 Cardiac Resynchronization Therapy

relation to atrial filling (Chevalier et al. 1997; Auricchio et al. 2000). Prolonged AV delay can be measured with Doppler mitral inflow pattern analysis. Atrial contraction can occur immediately after or during the preceding ventricular contraction and leads to early closure of the mitral valve and reduction in diastolic filling times. This delay between the onset of LV pressure rise and atrial filling may have an end result of inversed AV flow and cause diastolic mitral regurgitation (MR) (Fig. 3.4.1) (Breithard 2008; Sweeney 2008).

Figure 3.4.1 A. Intracardiac pressures and mitral inflow during atrioventricular (AV) decoupling (prolonged AV interval). The mitral inflow pattern has two constituents. The initial component (E wave) is due to ventricular filling. The subsequent A wave is due to atrial contraction. The sum of these two constituents comprises the diastolic filling period, which is the interval from the onset of the E velocity to the cessation of the A velocity. B. Effect of left ventricular (LV) conduction delay and prolonged AV conduction on diastolic filling patterns. LV filling is displaced rightward in time, whereas atrial contraction is displaced leftward in time. The result is fusion of filling and atrial contraction (E-A fusion). ic, isovolumic contraction; ir, isovolumic relaxation. (Adapted from Sweeny 2008. Reprinted with permission.)
Cardiac Arrhythmia Management

**IVCD**

IVCD, a conduction delay or the mechanical dispersion of motion within the LV, is responsible for reduced pump function. This LV delayed conduction causes a dyssynchronous contraction, resulting in a redistribution of the mechanical load within the walls of the ventricles, leading to a reduction in stroke volume, increase in wall thickness at the site of earliest activation, ventricular remodeling, and progression of HF. This can be assessed using color tissue Doppler imaging to measure the regional time intervals between the onset of the QRS complex to the peak of the systolic myocardial velocity in four basal segments of the LV (septal, lateral, anteroseptal, posterior) (Bleeker et al. 2004). Intraventricular dyssynchrony is considered by majority of clinicians to be the most important target of cardiac resynchronization therapy (CRT). Intraventricular dyssynchrony can also be present in patients without prolonged QRS (Bax et al. 2004).

**LV Mural Delay**

Intramural (endocardial to epicardial) delays have been noted in detailed studies of the ventricular activation process using noncontact and contact mapping techniques. In patients with LBBB, activation is significantly different in the subendocardial layers compared with the intramyocardial layers. The negative effects of LV mural delay are uncertain (Auricchio et al. 2004).

**CRT**

CRT is the most efficacious nonpharmacological treatment to restore the coordination of contraction and relaxation among cardiac chambers (Leclercq and Kass 2001; Linde et al. 2002; Cleland et al. 2005). A device-based therapy, CRT targets electromechanical ventricular dysynchrony by pacing or sensing the right atrium, (unless the patient has permanent atrial fibrillation [AF]), and simultaneously pacing the RV near the interventricular septum and the LV (using the coronary venous branches), that is, biventricular pacing (Gura and Foreman 2004). The target population (Table 3.4.1) for biventricular pacing (CRT-P) are patients with symptomatic chronic HF, NYHA class III–VI, on optimal medical therapy (OPT), and have an IVCD with a QRS duration >120 ms. A biventricular pacing system with a defibrillator (CRT-D) is indicated in patients with the same comorbidities and an LVEF of ≤35% (Epstein et al. 2008).

Device components include (Figs. 3.4.2 and 3.4.3) the pulse generator (PG) and the pacing or pacing/defibrillation leads. The components of the PG are contained in a titanium case, containing the battery, the microprocessors, the pacing and sensing circuitry, and the capacitors in CRT-D devices.

**CLINICAL TRIALS SUMMARY**

Early observation studies evaluated the acute hemodynamic changes and effects of CRT on
Table 3.4.1  Indications for cardiac resynchronization therapy* in patients with severe systolic heart failure (Epstein et al. 2008).

<table>
<thead>
<tr>
<th>Classification of recommendations and level of evidence</th>
<th>Indications</th>
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<tbody>
<tr>
<td>I A</td>
<td>For patients who have left ventricular ejection fraction (LVEF) less than or equal to 35%, a QRS duration greater than or equal to 0.12 seconds, and sinus rhythm, cardiac resynchronization therapy (CRT) with or without an ICD is indicated for the treatment of New York Heart Association (NYHA) functional class III or ambulatory class IV heart failure symptoms with optimal recommended medical therapy.</td>
</tr>
<tr>
<td>IIa B</td>
<td>For patients who have LVEF less than or equal to 35%, a QRS duration greater than or equal to 0.12 seconds, and AF, CRT with or without an ICD is reasonable for the treatment of NYHA functional class III or ambulatory class IV heart failure symptoms on optimal recommended medical therapy.</td>
</tr>
<tr>
<td>IIa C</td>
<td>For patients with LVEF less than or equal to 35% with NYHA functional class III or ambulatory class IV symptoms who are receiving optimal recommended medical therapy and who have frequent dependence on ventricular pacing, CRT is reasonable.</td>
</tr>
<tr>
<td>IIb C</td>
<td>For patients with LVEF less than or equal to 35% with NYHA functional class I or II symptoms who are receiving optimal recommended medical therapy and who are undergoing implantation of a permanent pacemaker and/or ICD with anticipated frequent ventricular pacing, CRT may be considered.</td>
</tr>
<tr>
<td>III B</td>
<td>CRT is not indicated for asymptomatic patients with reduced LVEF in the absence of other indications for pacing.</td>
</tr>
<tr>
<td>III C</td>
<td>CRT is not indicated for patients whose functional status and life expectancy are limited predominantly by chronic noncardiac conditions.</td>
</tr>
</tbody>
</table>

* All primary SCD prevention ICD recommendations apply only to patients who are receiving optimal medical therapy and have reasonable expectation of survival with good functional capacity for more than 1 year.

Figure 3.4.2  Biventricular pacing system. (Reproduced with permission from Medtronic, Inc.)
**MUSTIC: Multisite Stimulation in Cardiomyopathies Studies**

The MUSTIC study (Cazeau et al. 2000), a single-blind, randomized crossover trial, evaluated 48 patients with NYHA class III/IV HF, a QRS duration of ≥150 ms, LVEF ≤35%, left ventricular end-diastolic dimension (LVEDD) >60 mm, SR, and 6-minute walk test (6MWT) <450 m. Patients were treated for 3 months with CRT and compared with an inactive phase. Patients who were actively paced showed a significant improvement in 6MWT ($P = 0.0001$), peak VO$_2$ ($P = 0.029$), quality of life (QOL) ($P = 0.0001$), and a decrease in QRS duration ($P = 0.0003$). Three-month rehospitalization rates were also statistically reduced in the CRT arm. MUSTIC-AF, a second MUSTIC trial, looked at similar end points in patients with symptomatic HF, AF, and ventricular dyssynchrony (paced QRS of >200 ms) (Linde et al. 2002). Results were similar to MUSTIC in that CRT evoked statistically significant improvement in 6MWT, peak VO$_2$, and hospital readmission rates. MUSTIC-AF conferred that CRT is reasonable for patients with AF with a slow ventricular response, who are pacemaker dependent due to intrinsic conduction disease, or following AV nodal ablation.

**MIRACLE: Multicenter InSync Randomized Clinical Evaluation**

The MIRACLE study (Abraham et al. 2002), a prospective, randomized, double-blind parallel-controlled trial, evaluated 453 patients with NYHA class III–IV, LVEF of ≤35%, LVEDD >55 mm, a QRS duration of ≥130 ms, and a 6MWT ≤450 m. CRT resulted in QRS duration decrease of 20 ms ($P ≤0.01$), an improvement in 6MWT of 39 m ($P = 0.02$), a QOL score of 19 points ($P = 0.017$), a 100-s average increase in Naughton protocol exercise duration ($P = 0.001$), an average decrease in LVEDD of 0.5 cm...
Table 3.4.2  Clinical trials summary of cardiac resynchronization therapy in congestive heart failure.

<table>
<thead>
<tr>
<th>Trial</th>
<th>Purpose</th>
<th>Number of patients</th>
<th>Design</th>
<th>Patient population</th>
<th>Results</th>
<th>Conclusions</th>
</tr>
</thead>
<tbody>
<tr>
<td>MUSTIC (Cazeau et al. 2000)</td>
<td>To evaluate whether multisite BV pacing improves hemodynamics and well-being by reducing ventricular dyssynchrony</td>
<td>58</td>
<td>Prospective, randomized, single-blind, crossover</td>
<td>NYHA class IIIEF &lt;35%QRS ≥150 msNSR</td>
<td>After 3 months of inactive or active pacing, CRT: ↑6-minute distance by 23% (P &lt; 0.001) Improved QOL by 32% (P &lt; 0.001) ↑peak O₂ by 8% (P &lt; 0.03) ↓hospitalizations by 2/3 (P &lt; 0.05)</td>
<td>Atriobiventricular pacing significantly improves exercise tolerance and QOL in patients with ventricular dyssynchrony (IVCD)</td>
</tr>
<tr>
<td>PATH-CHF (Auricchio et al. 1999)</td>
<td>To evaluate the acute benefit of CRT, the chronic benefit with and without an ICD, and the benefit in QRS duration &gt;150 ms compared with those with a QRS of 120–150 ms</td>
<td>41</td>
<td>Crossover, longitudinal study (no pacing vs. LV pacing)</td>
<td>NYHA class III/IVEF ≤30%QRS ≥120 msNSR ≥55 bpmPR ≥150 ms</td>
<td>CRT showed benefit over control in 6-minute walk distance (386 m vs. 342 m; P = 0.001) Peak VO₂ (14.3 ml/kg/min vs. 12.5 ml/kg/min; P = 0.001) NYHA improvement (2.4 vs. 3.3; P = 0.001) QOL (29.5 vs. 48.8; P = 0.001)</td>
<td>CRT improves exercise capacity, NYHA class, and QOLPatients with widest QRS derive greatest benefit</td>
</tr>
<tr>
<td>MIRACLE (Abraham et al. 2002)</td>
<td>To evaluate the clinical benefits of CRT in patients with HF and IVCDs</td>
<td>453</td>
<td>Prospective, randomized double-blind, parallel, controlled</td>
<td>NYHA class III/IVNSRQRS ≥130 msEF ≤35%</td>
<td>Over control CRT showed significant benefit in 6-minute walk distance (39 m vs. 10 ms; P = 0.005) QOL (−19 vs. −10; P = 0.0001) Δ in ≥1 NYHA (52% vs. 32%; P &lt; 0.001)</td>
<td>CRT results in significant clinical improvement in patients with moderate-to-severe HF and an ventricular dyssynchrony (IVCD)</td>
</tr>
<tr>
<td>CONTAK CD (Saxon et al. 1999)</td>
<td>To evaluate the safety and effectiveness in using CRT and ICD therapy to improve functional class and slow the progression of HF</td>
<td>490</td>
<td>Crossover, parallel controlled, double-blind</td>
<td>NYHA class II/IVQRS &gt;120 ms EF ≤35% NSR Indications for ICD therapy</td>
<td>The primary composite end point with CRT showed a reduction in slowing of HF progression, 21% deaths, 23% hospitalization, 13% worsening of HF class, 26% VT/VF incidence, 9%</td>
<td>CRT and ICD combination devices are safe and may provide benefit in a broader patient population</td>
</tr>
</tbody>
</table>

(Continued)
<table>
<thead>
<tr>
<th>Trial</th>
<th>Purpose</th>
<th>Number of patients</th>
<th>Design</th>
<th>Patient population</th>
<th>Results</th>
<th>Conclusions</th>
</tr>
</thead>
<tbody>
<tr>
<td>MIRACLE ICD (Young et al. 2003)</td>
<td>To evaluate safety and efficacy of combined CRT and ICD therapy in patients with moderate to severe HF, ventricular dyssynchrony, and an ICD indication</td>
<td>555</td>
<td>Prospective, randomized, double-blind, parallel-arm, controlled trial</td>
<td>NYHA class III/IVQRS ≥130 msEF ≤35%ICD indication</td>
<td>6-month follow-up: Control versus CRT</td>
<td>CRT + ICD improved: QOL NYHA functional class exercise capacity improvements occurred without compromising ICD functionality</td>
</tr>
<tr>
<td>COMPANION (Bristow et al. 2004)</td>
<td>To evaluate whether OPT therapy + CRT or OPT + CRT-D is superior to OPT alone</td>
<td>1,520</td>
<td>Parallel, randomized 1:2:2, open-label three-arm study</td>
<td>NYHA class II/IVQRS ≥120 msPRI ≥150 msEF ≤35%LVEDD ≥60 mm</td>
<td>Compared with OPT: CRT ↓ combined all-cause morality and all-cause hospitalization by 18.6% (P = 0.015) CRT-D ↓ combined all-cause morality and all-cause hospitalization by 19.3% (P = 0.005) CRT-D ↓ all-cause mortality by 43.4% (P = 0.002)</td>
<td>CRT and CRT-D both significantly reduce the composite of all-cause death and all-cause hospitalizations as well as the composite of all-cause death and HF hospitalizations</td>
</tr>
<tr>
<td>CARE-HF (Cleland et al. 2005)</td>
<td>To evaluate the effect of CRT on long-term morbidity and mortality in patients with advanced HF and dyssynchrony despite OMT</td>
<td>814</td>
<td>Open label, randomized</td>
<td>NYHA class III/IVNSRQRS ≥120 msEcho evidenced dyssynchrony OMT</td>
<td>Overall CRT group showed benefit in ↓ all-cause morbidity (P &lt; 0.001) and mortality (P &lt; 0.002) Improvement (P &lt; 0.001) in NYHA FC QOL LVEF LVESV Neurohormonal measures improved</td>
<td>CRT-P improves symptoms and QOL Reduced complications and risk of death</td>
</tr>
</tbody>
</table>

MUSTIC, Multisite Stimulation in Cardiomyopathies; PATH-CHF, Pacing Therapies for Congestive Heart Failure; MIRACLE, Multicenter InSync Randomized Clinical Evaluation; MIRACLE ICD, Multicenter InSync Randomized Clinical Evaluation ICD; COMPANION, Comparison of Medical Therapy, Pacing, and Defibrillation in Chronic Heart Failure; CARE-HF, Cardiac Resynchronization Heart Failure; BV, biventricular; NYHA = New York Heart Association; EF, ejection fraction; NSR, normal sinus rhythm; CRT, cardiac resynchronization therapy; CRT-P, cardiac resynchronization therapy pacemaker; QOL, quality of life; IVCD, intraventricular conduction delay; LV, left ventricle; HF, heart failure; ICD, implantable cardioverter defibrillator; VO₂, maximum oxygen uptake; VT, ventricular tachycardia; VF, ventricular fibrillation; OPT and OMT, optimal medical therapy; PRI, PR interval; LVESV, left ventricular end-systolic volume; LVEDD, left ventricular end-diastolic diameter; CRT-D, cardiac resynchronization implantable cardioverter defibrillator; FC, functional class.
CARE-HF: Cardiac Resynchronization Heart Failure

CARE-HF (Cleland et al. 2005), a multicenter, open-label, randomized trial, evaluated NYHA class III–IV patients with a LVEF ≤35%, LVEDD ≥30 mm, SR, QRS duration ≥120 ms plus echocardiographic criteria of dyssynchrony, and on stable OPT. Patients (n = 814) were randomized to OPT alone or OPT plus CRT-P. Patients in the CRT arm had a greater relative risk reduction in death, cardiovascular hospitalization (P ≤ 0.002), and exhibited an improvement in LVEF, NYHA class, symptoms, QOL, and other echocardiographic parameters (P ≤ 0.001) than the control arm. Death and hospitalization for deteriorating HF were also reduced in the CRT arm.

CRT, AF, AND HF

There is a 20% prevalence of permanent AF in the HF population (Carson et al. 1993; Heist and Ruskin 2006). AF contributes to worsening HF due to the loss of the atrial contribution to cardiac output, and the irregular and fast heart rates. Patients with AF are not well represented in the landmark trials that established the clinical benefit of CRT; however, small studies support the premise that patients with AF benefit from CRT. Leclercq et al. (2000) studied 37 patients with dilated cardiomyopathy who received CRT and reported greater improvements in exercise capacity, and LVEF in patients with AF as compared with patients in SR.

The Left Ventricular-Based Cardiac Stimulation Post AV Nodal Ablation Evaluation (PAVE) trial (Doshi et al. 2005) was a prospective, randomized trial of the “ablate and pace” strategy combined with CRT. A total of 103 patients with NYHA class III–IV, LVEF 46 ± 16%, and refractory AF requiring AV junction (AVN) ablation were randomized to CRT or RV pacing alone. Patients in the CRT-treated
group experienced significant improvement in 6MWT ($P = 0.04$). At 6 months, LVEF was higher in CRT patients than the RV pacing group (46 ± 13% vs. 41 ± 13%; $P = 0.03$). Ferreira et al. (2008) did a retrospective analysis of 131 consecutive HF patients who underwent CRT implantation. Three groups were considered: SR (n = 78), AF with AVN ablation (n = 26), and AF without AVN ablation (n = 27). The three groups showed a significant improvement in functional class; however, the proportion of responders was significantly lower in AF patients without AVN ablation (52 vs. 79% in SR and 85% in AF with AVN ablation; $P \leq 0.008$). AF without AVN ablation was also independently associated with increased mortality (hazard ratio [HR] 5.22, 95% confidence interval [CI]: 1.60–17.01, $P = 0.006$) and increased hospitalization for HF during the first 12 months (HR 6.23, 95% CI: 2.09–18.54, $P = 0.001$). The outcomes of AF with AVN ablation patients were similar to the outcomes of patients in SR.

A meta-analysis of five prospective studies that determined the effects of CRT in 1,164 patients with HF, IVCD was done by Upadhyay et al. (2008). Ablation of the AVN was performed in 56% of the 367 patients with AF to evaluate how effective CRT was in patient with AF. The meta-analysis validated that although LVEF improves to a similar degree with CRT in patients with SR and AF, SR is associated with significantly greater improvement in functional capacity and QOL. If CRT is used in a patient with AF, it is important to control the rate (either pharmacologically or by AVN ablation), such that there is a high proportion of ventricular pacing, particularly during activity or exertion.

**FORCED RV PACING, HF, AND CRT**

The literature is replete with studies to substantiate that forced RV pacing in patients with pacemakers for sinus node dysfunction, AV block, and ICDs has deleterious effects and causes desynchronization of ventricular electrical activation and contraction, thereby increasing cardiac morbidity and mortality (Anderson et al. 1997; Connolly et al. 2000c; Wilkoff et al. 2002; Sweeney et al. 2003). Forced RV pacing induces a LBBB and results in abnormal depolarization and mechanical dyssynchrony in the ventricle (Lee et al. 1994). Preliminary data suggested that a biventricular pacing upgrade in RV-paced HF patients improves NYHA functional class (Baker et al. 2002). Horwich et al. (2004) studied 15 NYHA class III–IV patients with prolonged QRS of 190 ± 27 ms, and constant forced RV pacing when upgraded to CRT, reported a reduction in QRS duration, LV electromechanical delay, left ventricular end-systolic volume (LVESV), and an improvement in LVEF. However, the limitation of these studies are that the data comes from small observational studies and to date, no prospective randomized clinical trial has been conducted to evaluate the benefit of CRT-in RV-paced HF patients.

**SUDDEN DEATH IN HF PATIENTS**

Initially described in the Framingham Heart Study (Kannel et al. 1988), the association between sudden cardiac death (SCD) and HF has been recognized. HF is associated with a fivefold increase in the risk of SCD. Although remarkable advances in the treatment of HF have occurred, mortality remains high (Uretsky and Sheahan 1997). The implantable cardioverter defibrillator (ICD) has proven to be the therapy of choice for patients (secondary prevention) who have survived an episode of SCD and for those patients (primary prevention) at high risk of SCD. Table 3.4.3 summarizes the clinical trials. A meta-analysis (Connolly et al. 2000b) of AVID (The AVID Investigators 1997), CIDS (Connolly et al. 2000a), and CASH (Kuck et al. 2000) trials support the use of ICDs as...
Table 3.4.3  Clinical trials summary for implantable cardioverter defibrillator therapy for secondary and primary prevention of sudden cardiac death.

<table>
<thead>
<tr>
<th>Trial</th>
<th>Number of patients</th>
<th>Patient population</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>AVID* (The AVID Investigators 1997)</td>
<td>1,016</td>
<td>Survival from VF or sustained VT LVEF 40% Sx of hemodynamic compromise or sustained VT with syncope</td>
<td>ICD ↓ mortality 31% compared with AADs</td>
</tr>
<tr>
<td>CIDS* (Connolly et al. 2000a)</td>
<td>659</td>
<td>Survival of cardiac arrest or documented VF or sustained VT ≥150bpm LVEF ≤35% Sx of hemodynamic compromise Sustained VT with syncope Unmonitored syncope with subsequent documentation of spontaneous &gt;10 seconds or inducible sustained VT</td>
<td>ICD ↓ mortality 20% compared with amiodarone</td>
</tr>
<tr>
<td>CASH* (Kuck et al. 2000)</td>
<td>288</td>
<td>Survival after cardiac arrest due to documented ventricular arrhythmia</td>
<td>ICD ↓ mortality 37% compared with amiodarone or metoprolol; propafenone arm stopped early due to SCD (29.3% vs. 11.5%) risk compared with the ICD group</td>
</tr>
<tr>
<td>MADIT I† (Moss et al. 2002)</td>
<td>196</td>
<td>MI ≥3 weeks prior to entry LVEF ≤35% Spontaneous NSVT (3–30 beats) Inducible VT at EP nonsuppressible with procaninamide NYHA class &lt;IV</td>
<td>ICD ↓ mortality 56%</td>
</tr>
<tr>
<td>MUSTT† (Buxton et al. 1999)</td>
<td>704</td>
<td>CAD NSVT &gt;3 beats LVEF ≤40% Inducible VT at time of EP NYHA class &lt;IV</td>
<td>ICD ↓ mortality 55%</td>
</tr>
<tr>
<td>MADIT II† (Moss et al. 1996)</td>
<td>1,232</td>
<td>Q wave or positive enzyme MI ≥1 month Conventional medical therapy LVEF ≤30% NYHA class &lt;IV</td>
<td>ICD ↓ mortality 31%</td>
</tr>
<tr>
<td>DEFINITE† (Kadish et al. 2006)</td>
<td>458</td>
<td>Nonischemic dilated cardiomyopathy Optimal medical therapy LVEF ≤35% Asymptomatic VTA</td>
<td>ICD ↓ mortality 34%</td>
</tr>
<tr>
<td>SCD-HeFT† (Brady et al. 2005)</td>
<td>2,521</td>
<td>Ischemic or nonischemic dilated cardiomyopathy NYHA II-III ≥3 months Optimal medical therapy LVEF ≤35% Permanent AF if INR ≥2.0 ≥3 weeks</td>
<td>ICD ↓ mortality 23%; amiodarone does not improve survival</td>
</tr>
</tbody>
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Table 3.4.3 (Continued)

<table>
<thead>
<tr>
<th>Trial</th>
<th>Number of patients</th>
<th>Patient population</th>
<th>Results</th>
</tr>
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<tbody>
<tr>
<td>Contak CD (Saxon et al. 1999)</td>
<td>Refer to Table CRT.2</td>
<td></td>
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<tr>
<td>MIRACLE ICD (Gras et al. 1998)</td>
<td></td>
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<tr>
<td>COMPANION (Bristow et al. 2004)</td>
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* Secondary prevention trial; † primary prevention trial.

AVID, Antiarrhythmics Versus Implantable Defibrillators; CIDS, Canadian Implantable Defibrillator Study; CASH, Cardiac Arrhythmia Suppression Study Hamburg; MADIT I, Multicenter Automatic Defibrillator Implantation Trial; MUSTT, Multicenter Unsustained Tachycardia Trial; MADIT II, Multicenter Automatic Defibrillator Implantation Trial II; DEFINITE, Defibrillators in Nonischemic Cardiomyopathy Treatment Evaluation; SCD-HeFT, Sudden Cardiac Death in Heart Failure Trial; VF, ventricular fibrillation; VT, ventricular tachycardia; LVEF, left ventricular ejection fraction; Sx, symptoms; ICD, implantable cardioverter defibrillator; AADs, antiarrhythmic drugs; SCD, sudden cardiac death; MI, myocardial infarction; NSVT, nonsustained ventricular tachycardia; NYHA, New York Heart Association; CAD, coronary artery disease; EP, electrophysiology study; VTA, ventricular tachyarrhythmia; AF, atrial fibrillation.

### IMPLANTATION OF CRT

Implantation of CRT is significantly different compared with implantation of standard ICD or pacemaker therapy. Many of the techniques are borrowed from interventional cardiologists including angiography and the use of intravascular wires to deliver lead systems. In majority of cases, LV leads are delivered to the posterolateral LV wall via the coronary sinus system. Thus, LV leads are thinner and less easy to manipulate compared with standard pacing leads. The technique begins with placement of a long delivery sheath in the right atrium. The coronary sinus is cannulated with this sheath using either a wire or a steerable catheter. At this point, a venogram is performed by injecting contrast into the coronary sinus. This is usually performed using a catheter, with a proximal balloon to occlude the coronary sinus during injection. It is best to advance the balloon catheter out into the coronary sinus over a wire and withdraw it to a proximal position to assure that the balloon is not within a venous branch. Inflation of the balloon or vigorous injection can result in coronary sinus dissection or rupture. The operator then chooses a tributary of the coronary sinus, which lies over the posterolateral wall. Often, this wall is approached via the middle cardiac vein (Fig. 3.4.4), which is a very proximal branch of the coronary sinus. The vein is evaluated for position, ease of access (tortuosity, acute angle ostia, stenosis, etc.), and diameter. There are several types of leads. Leads can be delivered by stylet or over the wire (OTW); leads can be bipolar or unipolar. The lead is advanced into the coronary sinus branch using the appropriate technique (Fig. 3.4.5). Security of the lead position is often difficult. The standard fixation systems used in RV
Figure 3.4.4 A venogram performed in the left anterior oblique position. The balloon (labeled A and outlined with arrows) is positioned distally to see the middle cardiac vein (labeled C and outlined with arrows) adequately. The main coronary sinus is labeled “B.”

Figure 3.4.5 A fluoroscopic image of an LV lead (A) in the right anterior oblique position.

or atrial pacing systems cannot be used on LV leads. Typically, leads are secured by advancing them into the vein as distally as possible. Sensing and stimulation threshold testing are performed using standard methods. There are various fixation techniques that are available (Fig. 3.4.6). Once proper position has been confirmed, temporary pacing at high-output energy (10 V) is performed to assure lack of stimulation of the phrenic nerve, which courses along the lateral wall of the LV. During high-output pacing, the hemidiaphragm is assessed under fluoroscopy for pacing-induced diaphragmatic excursions. Diaphragmatic stimulation can also be felt by hand placement over the hemidiaphragm. If phrenic nerve capture is demonstrated, movement of the lead proximally, distally, or to a different site is required. Once the lead is placed in the posterolateral region, the pacing and sensing thresholds are adequate, and phrenic nerve capture is not present, the sheath must be removed. This usually requires cutting or peeling the sheath while keeping the lead in a steady, fixed position. This part of the procedure may be challenging. Digital image acquisition in standard positions is usually performed to assure that the leads have not dislodged. The leads are then fixed to the pectoralis muscle and attached to the generator in the standard manner. The implantation success rate of CRT therapy is 93.7% (McAlister et al. 2007). The most common reason for an unsuccessful implant is the inability to place or maintain the position of the LV lead due to venous tortuosity and inaccessible target veins. Complications include coronary sinus dissection (0.5 to 4%) (Abraham et al. 2002; Bristow et al. 2004; Cleland et al. 2005), coronary sinus perforation (0.8–2%) (Gras et al. 2005).
Cardiac Arrhythmia Management

1998; Auricchio et al. 1999; Kass et al. 1999; Saxon et al. 1999; Abraham et al. 2002; Young et al. 2003; Bristow et al. 2004), and LV lead dislodgement (5.7–5.9%) (Abraham et al. 2002; Cleland et al. 2005). Additionally, because these patients often have a LBBB, transient mechanical trauma to the right bundle by the coronary sinus sheath may result in complete AV block. To prepare for this, many operators implant the RV lead first during a CRT implant. Overall, this procedure is more challenging and time consuming. The average procedural duration in the MIRACLE study was 2.7 hours (Abraham et al. 2002).

Procedure time has decreased significantly with the advent of advanced delivery systems and increased operator experience. If there is an inability to cannulate the coronary sinus, a surgical approach to LV lead placement can be utilized. The surgical approach will depend on the surgeon’s preference ranging from a minimally invasive surgical approach for lead placement of the epicardial LV pacing lead via a port hole or limited left lateral thoracotomy to a full left lateral thoracotomy, which permits full view of the LV free wall.

OPTIMIZATION OF CRT

In order to achieve maximum CRT efficacy, optimization of the programmed parameters is pivotal. AV synchrony and biventricular pacing >90% should be maintained in order to provide continuous therapy. Initial programming should include mode selection, upper and lower rate limits, and AV delays during both sensed and paced atrial events. Bernheim et al. (2005) revealed that there are acute hemodynamic advantages in CRT patients with intrinsic atrial activation in comparison with paced atrial events. Therefore, programming in the acute phase should include minimizing atrial pacing if the patient is chronotropically competent. Diagnostic heart rate histograms will allow the clinician to evaluate the rate excursion and the appropriateness of the programmed lower rate long term. Device-specific algorithms, such as rate hysteretic to enhance intrinsic atrial events, should be considered to reduce the cumulative incidence of atrial pacing. However, in patients with sinus node dysfunction who maintain high atrial pacing rates, rate modulation should be considered (Tse et al. 2005). Programming of chronotropically incompetent patients should routinely include a dual-chamber rate responsive (DDDR) mode at 60–70 ppm.

However, since most CRT patients have normal sinus node function, upper rate limits should be programmed to maintain maximal tracking above the highest achievable sinus rate. Manufacturer-specific algorithms (ventricular sense response, negative AV delay, rate regularization, conducted AF response, rate fading, triggered mode) should be utilized to maintain complete biventricular capture (Fig. 3.4.7). Other programmed parameters to ensure biventricular pacing include the optimal AV delay and RV and LV pacing outputs. At times, the change in the paced QRS morphology due to loss of biventricular capture may only be a subtle change and evaluation of capture in multiple ECG leads may be necessary (Fig. 3.4.8). In the presence of elevated LV stimulation thresholds or phrenic nerve stimulation, “electrical repositioning” may be an option in some devices. LV pacing configuration may include the use of the LV tip/ring, RV ring/coil, and LV tip to RV coil configuration (Fig. 3.4.9).

AV and V-V Optimization

Most acute and long-term beneficial effects of CRT are due to the improved pumping of the ventricles and independent of the programmed AV delay (Auricchio et al. 1999). However, empiric AV delay programming may not be optimal in CRT patients since studies (Auricchio et al. 1999; Kass et al. 1999) indicated variability from patient to patient. The AV delay interval should be programmed optimally, short enough
Chapter 3.4 Cardiac Resynchronization Therapy

Figure 3.4.7 Ventricular sense response (VSR) (Medtronic, Inc.) operation. The spontaneous conversion to an atrial tachyarrhythmia causes an increase in the intrinsic ventricular rate. The VSR is triggered on the second QRS complex and is evident on complex 3–4 and 9–10. A, atrial; V, ventricular; AS, atrial sensed; AR, atrial sensing in the atrial refractory period; BV, biventricular pacing; VS, ventricular sensing.

Figure 3.4.8 Lead I rhythm strip illustrating a subtle change in QRS morphology with the loss of left ventricular capture.

to assure biventricular pacing, yet allowing adequate time to provide optimal LA contribution to LV filling, maximum stroke volume, shortening of the isovolumic contraction time, and the longest diastolic filling time in the absence of diastolic MR (Pandis et al. 1986; Bax et al. 2005). Noninvasive techniques to optimize the AV delay include automatic device-based algorithms (i.e., St. Jude Medical’s [St. Paul, MN] QUICKOPT™ or Boston Scientific’s [St. Paul, MN]SmartDelay™), echocardiography-guided programming using the Ritter method (Ritter et al. 1995; Kinderman et al. 1997), or aortic Doppler velocity-time integral (VTI) (Kerlan et al. 2006), plethysmography (Butter et al. 2004; Whinnett et al. 2006a,b), and impedance cardiography (Tse et al. 2003; Braun et al. 2005). Optimal AV delay timing will change over time and differ due to physiological influences such as heart rate, activity, and substrate changes with long-term effects of CRT.

Since CRT is used to treat dyssynchrony, it would seem reasonable that the RV and LV pacing stimulus should be delivered simultaneously. This is not always the case. Optimal cardiac synchrony can often be achieved by programming the excitation of the RV or the LV first (V-V optimization). Brignole et al. (2008) demonstrated that IVCD at baseline was 63 ms, and decreased to 44 ms with CRT. Further V-V optimization by programming the RV or LV first resulted in a decrease in the delay to 26 ms. In most cases, the LV is activated before the RV (Brignole et al. 2008). Since it is not clear which
V-V interval will be optimal, repeated testing is required. Bertini et al. (2008) demonstrated that QRS duration was a reasonable marker for V-V optimization. They found a correlation between stroke volume measured by echo Doppler and QRS duration measured by ECG. The optimum V-V interval was measured by 12-lead ECG in this study (Bertini et al. 2008).

Clinically, the optimal V-V delay cannot be identified in the majority of patients, with the range being relatively narrow and most frequently involves LV pre-excitation by 20 ms (Burri et al. 2006). Automatic device-based algorithms may be used to optimize V-V timing, but assessment can also be done with echocardiography using tissue Doppler imaging and strain rate analysis and aortic VTI. Careful evaluation to rule out anodal ring stimulation should be done since this will interfere with a programmed V-V interval. If RV anodal ring stimulation occurs, the V-V interval will become zero (Barold et al. 2007).

**Diagnostic Features**

All clinicians in the management of HF patients should utilize diagnostic data provided by CRT devices. Although manufacturer specific, data includes atrial and ventricular arrhythmias, heart rate trends, battery and lead status, and
the percent of sensing and pacing. The most basic diagnostic feature in CRT therapy is evaluation of the percent of sensed ventricular events. Since the goal is to consistently stimulate the RV and LV as therapy, the percent of ventricular sensing should be minimized. A major reason why ventricular sensed events may occur is AF with increased ventricular rates. Evaluation of atrial tachycardia events and mode switch episodes is important as well. Device-specific algorithms to achieve and maximize ventricular pacing in the event of conducted atrial arrhythmias should be utilized.

Patients with certain comorbidities are at increased risk for decompensation, possibly due to the burden of their diseases and how they interact with the underlying HF. However, the overall predictability of a clinical event in patients with HF, such as a hospitalization for decompensation, continues to be poor. Many CRT devices are equipped with systems to identify when patients are beginning to decompensate. Typically, the patient undergoes a physiological change as they begin to decompensate; that is, the blood pressure rises, pressures within the heart rise, and renal function worsens. There is a window of diagnostic opportunity during this period where the fluid levels within the lung tissue increase (one of the earliest physiological changes) before patients become symptomatic (Whellan 2008). The use of intrathoracic impedance as an indicator for excessive lung fluid content is shown in Figure 3.4.10. Other diagnostic data

![Figure 3.4.10](image_url)

**Figure 3.4.10** OptiVol (Medtronic, Inc., Minneapolis, MN) fluid index monitoring showing a decrease in thoracic impedance, a positive OptiVol crossing and resetting of the OptiVol Fluid index following medical intervention.
will show a series of clinically relevant arrhythmia HF-related measures across multiple dimensions, including arrhythmia episodes, AF burden, heart rate trends, heart rate variability, and patient activity levels, to be used by the caregiver for patient management (Wang 2005).

**RESPONDERS AND NONRESPONDERS**

Not every patient improves after CRT therapy. This has been an area of significant concern and research. A lack of clinical improvement was observed in 20% of patients in the MIRACLE trial (Abraham et al. 2002). The goal of CRT is to improve the synchrony of contraction of the LV, and thus patients are currently selected using QRS duration, which is a crude measure of dyssynchrony. Presumably, the wider the QRS duration, the greater the time it takes to depolarize the septum to the lateral wall. Unfortunately, QRS duration does not adequately predict dyssynchrony. Bleeker et al. (2004) demonstrated in general that QRS duration was predictive of dyssynchrony in patients with depressed LV function. However, as many as 30% of patients with very wide QRS duration (>150 ms) did not have significant dyssynchrony when measured using tissue Doppler echocardiography. Additionally, as many as 27% of patients with narrow QRS duration (<120 ms) had significant delay (Bleeker et al. 2004). In fact, patients with greater degrees of dyssynchrony demonstrate greater benefit of CRT therapy. Bax et al. (2004) showed that patients who improved by at least one NYHA functional class (responders) had at least 65 ms of delay between the septum and the lateral wall of the LV (Bax et al. 2004). In addition, responders improved LVEF, MR, and 6MWT greater than nonresponders (Bax et al. 2004). Thus, significant intraventricular LV delay may be a method of selecting patients most likely to respond to CRT therapy. Additionally, measures of LV delay may be appropriate to select a target for LV lead placement. If the latest point of mechanical contraction of the LV could be identified and if the LV lead could be positioned at that point, it is possible to optimize timing of contraction and minimize dyssynchrony. Ypenburg et al. (2008) evaluated the latest point of LV contraction by speckle tracking radial strain on Doppler echocardiography. They then evaluated the position of the LV lead on chest X-ray. Two important points are made from this article. First, when the LV lead is placed near the area of latest LV contraction, there is a reduction in LVESV and a reduction in events (death and HF hospitalizations). If the lead is not placed in this “sweet spot,” no improvement was observed. Second, the latest site of activation occurred in the lateral and posterior LV walls in 69% of patients (Ypenburg et al. 2008). Thus, the technique of placing the lead in the posterolateral wall region may be the best option if no Doppler guidance is available.

**SUMMARY**

Optimal pharmacological therapy is central in the management of HF patients; however, high rates of morbidity and mortality continue. There is compelling evidence from randomized clinical trials to support CRT in patients with advanced HF and electromechanical dyssynchrony. Electrical-based therapy (CRT-P and CRT-D) has been shown to alleviate symptoms, augment ventricular remodeling, improve NYHA functional class, and decrease mortality. Ongoing randomized clinical trials continue to target refinement of patient selection criteria and defining new subsets of patients that may benefit from CRT.
Case 3.4.1

Case Presentation

A 65-year-old male with diabetes mellitus, idiopathic cardiomyopathy, combined systolic and diastolic HF, LVEF 25%, QRS duration 164 ms, status post implantation of Medtronic (St. Paul, MN) CRT-D system: InSync Sentry 7297, atrial lead 5076 CapSure Fix Novus, RV lead, 6949 Sprint Fidelis, LV lead 4194 Attain OTW. His medications include pioglitazone 15 mg by mouth daily, ramipril 2.5 mg by mouth daily, carvedilol 25 mg by mouth twice daily, and aspirin 81 mg by mouth daily. He recently lost his health insurance and has not been seen in the office for the past 6 months.

He presents to the arrhythmia services with reports of feeling “sluggish and tired” for the last few days. He reports mild dyspnea on exertion (DOE) but denies shortness of breath at rest, orthopnea, edema, weight gain, chest discomfort, presyncope, palpitations, or claudication.

On physical exam, he presents as a well-nourished male in no acute distress. There is no jugular venous distention (JVD) or carotid bruits. Examination of the heart reveals no lifts or heaves, apical impulse palpable at the sixth intercostal space, rate 75 ppm, regular, normal S1, S2, no rubs, murmurs, or gallops. Abdomen is flat with no visible pulsations, lesions, or hernias; bowel sounds present and active in all four quadrants, no bruits. Liver edge at costal margin is smooth and nontender; liver height 9 cm at midclavicular line (MCL).

Lower extremities are warm to touch with no edema, normal hair distribution, and no venous dilation. Radial, brachial, femoral, popliteal, posterior tibial, and dorsalis pedis pulses are equal and of good quality.

Device interrogation data reveals no ventricular arrhythmias or AF. What is the most appropriate next step in evaluating delivery of CRT? What diagnostic features on the cardiac compass will give us clinical insight to consistent delivery of CRT? Does the patient require device or medical intervention?

Case Discussion

The goal of CRT is to consistently stimulate the RV and LV >90%. The percent of ventricular pacing can be evaluated from the cardiac compass, the HF management report (Fig. 3.4.11), the rate histograms, the episode counter report (Fig. 3.4.12), the quick look summary screen, and the ventricular sensing episodes. Data review reveals essentially 100% ventricular pacing. Stimulation thresholds were conducted and verified complete biventricular capture (Fig. 3.4.13). Evaluation of the OptiVol fluid index reveals a decrease in thoracic impedance with a crossing of the OptiVol threshold (Fig. 3.4.14). This crossing and the reports of DOE correlate with early warning of decompensation. Medical treatment consisted of furosemide 40 mg by mouth daily for 3 days, diet counseling, and a basic metabolic panel in 1 week. The patient returned to our services, felt much improved without DOE; laboratory results were within normal limits and the OptiVol fluid index was reset with diuresis (Fig. 3.4.15).
Figure 3.4.11  Cardiac compass and heart failure management report demonstrating the clinical utility of device diagnostics including the percent of atrial and ventricular pacing trends.

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<th>Since Last Cleared</th>
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<td>0.1%</td>
<td>&lt;0.1%</td>
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<tr>
<td>AS-VP</td>
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<td>99.7%</td>
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<td>&lt;0.1%</td>
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Figure 3.4.12  Episode counter report displaying the arrhythmia events and the percentage of atrial and ventricular pacing (arrow).
Figure 3.4.13  Rhythm strips illustrating (A) biventricular capture, (B) right ventricular stimulation threshold, (C) left ventricular stimulation threshold, (D) atrial stimulation threshold, and (E) patient's intrinsic rhythm.
**Observations (1) (Oct 17, 2005 to Apr 05, 2006)**

- Possible fluid accumulation, Apr 04, 2006.

**OptiVol Fluid Trends (May 2005 to Apr 2006)**

OptiVol fluid index is an accumulation of the difference between the daily and reference impedance.

- P = Program
- I = Interrogate

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<th>OptiVol threshold</th>
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<td>40</td>
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<td>80</td>
<td>120</td>
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<td>120</td>
<td>160</td>
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<td>160</td>
<td>200</td>
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Thoracic impedance (ohms)
- Daily
- Reference

**Figure 3.4.14** OptiVol Fluid index monitoring showing the decrease in thoracic impedance and a positive OptiVol crossing which correlated with patient’s symptoms of fatigue and DOE.
Figure 3.4.15 OptiVol Fluid index obtained after administration of loop diuretics, with improvement in impedance and resetting of OptiVol index.
## Case 3.4.2

### Case Presentation

A 74-year-old male status post dual-chamber pacing system for Mobitz II second-degree AV block developed a pacing-induced dilated cardiomyopathy. He presents with an LVEF of 35–40%, no ischemic disease, and NYHA class III HF. An LV lead was added and connected to a CRT-P system: Boston Scientific H125 Contak Renewal TR, atrial and ventricular leads Medtronic 5076 CapSure Fix Novus, and a Boston Scientific LV lead 4525 Easytrak 3. He presents for his 3-week evaluation status post AV delay optimization. He denies SOB, DOE, weight gain, chest discomfort, or palpitations. Physical examination is unremarkable.

From a device interrogation, a review of the HF bradycardia counters (Fig. 3.4.16) reveals 35% atrial pacing with 100% RV and LV pacing. If the goal of CRT is to consistently stimulate the RV and LV >90%, does this data ensure that we have AV synchrony and ventricular synchrony? What is the next appropriate step in the evaluation of this patient?

### Counters - HF/Brady

20-APR-2005 to 13-MAY-2005

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**Figure 3.4.16** Heart failure and Brady counters from a Boston Scientific H125 CRT-P device. The counter shows that the device is pacing 35% in the atrial channel and 100% in the right and left ventricular channels.
Case Discussion

The presenting rhythm strip (Fig. 3.4.17) is consistent with a balanced endless-loop tachycardia since the rate is slower than the programmed upper rate. A comparison of the histogram report before (Fig. 3.4.18A) and after (Fig. 3.4.18B) AV optimization reveals a significant change in the percent of atrial pacing, with frequent atrial sensed rates of >110 ppm. Stimulation thresholds revealed consistent capture with an underlying rhythm of sinus bradycardia (Fig. 3.4.19).

Since there was an inability to induce this arrhythmia with provocative maneuvers, the upper rate limit was decreased to 110 ppm, the premature ventricular contraction (PVC) response was programmed off, and the device was reprogrammed for episode data storage of the electrogram (EGM) for two EGMs at 26 seconds preceding the arrhythmia and 13 seconds following the arrhythmia. A 24-hour Holter was recorded during this period to capture arrhythmia onset and termination.

Figure 3.4.17 Balanced endless loop tachycardia at a rate of 110 ppm due to delayed retrograde VA conduction. The upper rate limit was programmed to 130 ppm.

Figure 3.4.18 Comparison of the pre (A) and post (B) AV delay (AVD) optimization histogram reports collected over a 23-day period showing a significant increase in atrial sensed events greater than 110 ppm.
Figure 3.4.19 Rhythm strips illustrating (A) patient’s intrinsic rhythm, (B) right ventricular stimulation threshold, (C) left ventricular stimulation threshold, and (D) atrial stimulation threshold.
Figure 3.4.20 Atrial and ventricular stored EGMs illustrating the mechanism of the ELT (see text for explanation). AEGM, atrial electrogram; VEGM, ventricular electrogram.

Figure 3.4.21 Three-channel Holter recording illustrating tracking of an atrial premature depolarization as the initiating mechanism of the ELT. The recording system classified this as episodes of supraventricular tachycardia (SVT) consisting of 17 beats in duration.

displayed in Figures 3.4.20 and 3.4.21, the mechanism of the arrhythmia was due to tracking of an atrial premature depolarization and a long ventriculoatrial (VA) conduction time. The post-ventricular refractory period (PVARP) was reprogrammed from 270 ms to 330 ms, and the rate responsive AV delay was programmed on with a 30-ms offset. Since reprogramming, no further pacemaker-mediated tachycardia occurred.
Case 3.4.3

Case Presentation

A 62-year-old thin female underwent implantation of a St. Jude Medical Promote RF CRT-D model number 3213 device in 2009. She had a familial dilated cardiomyopathy with an LVEF of 25%. Coronary arteriograms were normal. The device was implanted for refractory congestive HF (class III) despite OPT with beta blocker, ACE inhibitor, and aldosterone. A 12-lead ECG showed an LBBB with QRS duration of 140 ms. The LV lead (SJM Quicksite 1056 T) was implanted in the posterolateral LV region after demonstration of dyssynchrony.

She returned for follow-up on postoperative day 7 complaining of hiccups when she lies on her left side. Examination demonstrates no evidence of diaphragmatic stimulation in the supine position; however, when the patient moves to the left side, brisk spasm of the left side of the abdomen is demonstrated at a rate of 70 per minute. What is the most appropriate next step?

A chest X-ray confirms that the lead is in a similar position compared to implant. Testing for diaphragmatic stimulation was performed at the time of implant; however, slight changes in posture can change the place of the LV lead in juxtaposition to the phrenic nerve.

The options in this case are several. First, reoperation can be performed to move the lead to another branch or move it to a more proximal or distal location in the current vessel. This is not optimal. Repositioning requires an operation and does not guarantee that the lead will not capture the diaphragm during a posture change again. Second, the output of the LV lead can be programmed to a lower level. This can be successful if the stimulation threshold of the LV is much lower than the stimulation threshold of the diaphragm. The third alternative is electrical repositioning. In this case, the Promote RF device has VectSelect® that allows the pacing configuration to be changed by programming. The following was obtained during testing in the device clinic in the left lateral position (the LV tip to LV ring configuration was programmed during implant) (Table 3.4.4).

In this case, the LV tip to RV coil configuration was chosen. A phone call to the patient 1 month later confirmed resolution of the hiccups.

Table 3.4.4  LV thresholds at different configurations.

<table>
<thead>
<tr>
<th>Configuration</th>
<th>LV threshold</th>
<th>Diaphragm capture threshold</th>
</tr>
</thead>
<tbody>
<tr>
<td>LV ring to RV coil</td>
<td>5.0 V at 0.5 ms</td>
<td>No capture at 7 V at 1 ms</td>
</tr>
<tr>
<td>LV tip to RV coil</td>
<td>1.2 V at 0.5 ms</td>
<td>No capture at 7 V at 1 ms</td>
</tr>
<tr>
<td>LV tip to LV ring</td>
<td>1.8 V at 0.5 ms</td>
<td>5 V at 0.5 ms</td>
</tr>
</tbody>
</table>

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Chapter 3.4 Cardiac Resynchronization Therapy


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INTRODUCTION

Implantable cardioverter defibrillators (ICDs) provide the foundation for contemporary management of patients with lethal ventricular arrhythmias or those at risk. Numerous primary and secondary prevention clinical trials have repeatedly demonstrated the superiority of ICDs in reducing mortality in at-risk patients (Moss et al. 1996, 2002; AVID Investigators 1997; Buxton et al. 1999; Connolly et al. 2000; Kuck et al. 2000; Bardy et al. 2005). It is estimated that in 2007, approximately 235,000 ICDs were implanted in North America and approximately 88,000 ICDs in western and central Europe (Wilkoff et al. 2008). The ever-increasing population of patients with implantable defibrillators creates many unique challenges for health care providers, which include specialized training of personnel, time and space management, resource allocation, as well as economic considerations. The long-term management of patients with implantable defibrillators requires an intricate knowledge of device functionality, device interactions with medications as well as other implantable devices (pacemakers, medication pumps, and neural stimulators), as well as numerous patient comorbidities. The purpose of this chapter is to provide a detailed review of the routine follow-up process as well as to present several commonly seen troubleshooting issues presented in a case study format.

ROUTINE ICD FOLLOW-UP

ICD patients have historically been seen in the office every 3–4 months; however, with increasing device automaticity and the widespread utilization of remote monitoring, the need for routine in-office visits has diminished. Despite this, the importance of in-office visits cannot be underestimated. The visit provides a critical
opportunity to obtain a detailed symptom and arrhythmia history, as well as a focused physical examination of the patient. The primary goals of monitoring ICDs can be summarized in four main categories: patient related, device related, disease related, and communication. Table 3.5.1 summarizes the primary goals of device monitoring (Wilkoff et al. 2008a). The frequency of device follow-up primarily depends on the patient’s clinical status and arrhythmia burden as outlined in Table 3.5.2 (Wilkoff et al. 2008a). Additionally, the Current Procedural Terminology (CPT) codes for cardiac device monitoring were updated in January 2009, which significantly impacted the intervals at which in-office and remote checks are performed. The frequency of visits will also depend on whether or not the physician is providing

Table 3.5.1 Goals of monitoring CIEDs.

<table>
<thead>
<tr>
<th>Patient related</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Optimize the patient’s quality of life</td>
</tr>
<tr>
<td>• Optimize pacemaker/ICD system function to meet the patient’s clinical requirements</td>
</tr>
<tr>
<td>• Identify patients at risk and initiate appropriate follow-up with field safety corrective action/safety alerts</td>
</tr>
<tr>
<td>• Triage non-CIED related health problems and make appropriate referrals</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>CIED related</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Document appropriate CIED function</td>
</tr>
<tr>
<td>• Identify and correct abnormal CIED behavior</td>
</tr>
<tr>
<td>• Maximize pulse generator longevity while maintaining patient safety</td>
</tr>
<tr>
<td>• Identify CIEDs approaching end of battery life, to identify leads at risk of failure, and to organize CIED replacements in a nonemergent manner</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Disease related</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Document the nature and frequency of arrhythmias over time and correlate with patient symptoms and determine the appropriateness of CIED response to these arrhythmias</td>
</tr>
<tr>
<td>• Document (when feasible) hemodynamic status, transthoracic impedance, patient activity and other physiologic parameters over time as part of chronic disease monitoring in heart failure</td>
</tr>
<tr>
<td>• Monitor response to therapy</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Communication</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Maintain a patient database</td>
</tr>
<tr>
<td>• Timely communication to the patient and relevant healthcare providers of CIED- and disease-related information</td>
</tr>
<tr>
<td>• Provide technical expertise and education to colleagues, patients and community</td>
</tr>
</tbody>
</table>

Table 3.5.2 Factors determining the type and frequency of CIED follow-up.

Text not available in the electronic edition
general cardiology care, arrhythmia and/or device management, or a combination of both.

**ICD INTERROGATION**

Vendor-specific programmers are used to interrogate and download device data via bidirectional telemetry to a screen where the patient’s clinical data and programmed settings can be reviewed and the programming of the device parameters can be adjusted as necessary. Depending on the type of device, the interrogation can be performed either with a wand or wirelessly. Of note, most wireless devices regardless of vendor do require a wand or activator to be placed over the device to initiate wireless connectivity. Once activated, communication between the programmer and the device should be seamless. Typically, a paper copy is generated from the initial interrogation for documentation purposes. However, since these data begin in a digital format, the direct transfer to a digital storage system is becoming more feasible and will soon become the norm.

**DEVICE DIAGNOSTICS**

The most important feature of any device follow-up is that the interrogation process is done in a standardized format. Generally, the device diagnostics are reviewed first. The diagnostics are typically displayed in a longitudinal format, which enables the provider to correlate events over a specific time period (typically 14 months) (Fig. 3.5.1).

Table 3.5.3 provides a summary of commonly available device diagnostics, which are summarized below.

**Battery Voltage**

The device will provide a real-time estimate of the battery voltage. Some vendors will provide an elective replacement indicator (ERI) voltage, which provides some guidance as to timing of device replacement.

**Real-Time Electrogram (EGM)**

The real-time intracardiac EGM is obtained during a routine device interrogation or from a remote transmission. A remote transmission provides a 22-second strip of the intracardiac EGM, which typically displays the atrial near-field and shocking EGMs for dual-chamber ICDs (Fig. 3.5.2) and the near-field and shocking EGM for single-chamber ICDs. The stored EGM configuration can be individualized according to provider preference and patient needs. Common practice is to store the atrial near-field and ventricular far-field EGMs, which allows for identification of the A-V relationship. The far-field EGM, which is similar to the surface electrocardiogram (ECG) morphology, is extremely useful for distinguishing ventricular tachycardia (VT) from supraventricular tachycardia (SVT) by comparing the real-time EGM morphology with that of the stored clinical event. The EGM obtained from a remote transmission can be diagnostic, providing the clinician the ability to confirm the presence or absence of atrial fibrillation (AF).

**Episode Counters**

The device will log the total number of detected ventricular events, including nonsustained tachycardias as well as mode switch events for AF. This provides the health care provider with an overall view of the patient’s arrhythmia burden since the last device interrogation (Fig. 3.5.3).

**Treated VT/Ventricular Fibrillation (VF) Episodes**

The device will store an episode log of both treated and nontreated events that meet the specified detection criteria. Each event triggers
Figure 3.5.1  Example of a cardiac compass report (Medtronic, Inc., Minneapolis, MN).
storage of an EGM as well as a detailed episode text and interval plot (Fig. 3.5.4). ICD functionality can be assessed by reviewing the stored events for proper functioning, such as sensing and delivery of therapy (Scher 2003). The log can corroborate the patient’s recall of events and symptoms at the time of the event.

**Table 3.5.3** Available device diagnostics.

- Battery voltage
- Presenting EGM
- Episode counters
- Treated VT/VF episodes
- Non-sustained VT episodes
- Atrial fibrillation (AF) (mode switch, atrial tachycardia response (ATR))
- Ventricular rate during AF
- Percentage of pacing (atrial and ventricular)
- Average ventricular rate
- Heart rate histograms (atrial and ventricular)
- Patient activity (hours/day)
- Heart rate variability
- Lead trending
- Sensing integrity counter *
- Alert notifications

* Vendor specific.

Text not available in the electronic edition

**AF and Ventricular Rate (VR)**

Dual-chamber devices will store a running total of atrial mode switch events or atrial tachycardia response (ATR)—the nomenclature depending on the vendor, thus providing the practitioner with a quantification of the patient’s AF burden since the last interrogation. Additionally, some devices will provide information on the patient’s VR during AF.

**Figure 3.5.2** Example of a real-time electrogram (EGM).
Initial Interrogation: Quick Look

Clinical Status Since 24-Sep-2008

Treated
VF 1
FVT 0
VT 0
AT/AF(Monitor)

Monitored
VT (Off)
VT-NS (>4 beats, >143 bpm) 1
SVT, VT/VF RX Withheld 0
AT/AF 0
Time in AT/AF 0.0 hr/day (0.0%)

Functional Last Week
Patient Activity 3.4 hr/day

Therapy Summary VT/VF AT/AF
Pace-Terminated Episodes 0 0
Shock-Terminated Episodes 1 of 1 0
Total Shocks 1 0
Aborted Charges 0 0

Figure 3.5.3 Example of episode counters.


Only specified episodes shown below.

<table>
<thead>
<tr>
<th>Type</th>
<th>ATP</th>
<th>Shocks</th>
<th>Success</th>
<th>ID#</th>
<th>Date</th>
<th>Time</th>
<th>Duration</th>
<th>Avg</th>
<th>Max</th>
<th>Activity at Onset</th>
</tr>
</thead>
<tbody>
<tr>
<td>VT</td>
<td>1</td>
<td></td>
<td>Yes</td>
<td>14</td>
<td>14-Jan-2009</td>
<td>21:01</td>
<td>:12</td>
<td>81/188</td>
<td>---/188</td>
<td>Rest</td>
</tr>
<tr>
<td>VT</td>
<td>1</td>
<td></td>
<td>Yes</td>
<td>13</td>
<td>14-Jan-2009</td>
<td>20:56</td>
<td>:13</td>
<td>94/188</td>
<td>---/188</td>
<td>Rest</td>
</tr>
<tr>
<td>VT</td>
<td>1</td>
<td></td>
<td>Yes</td>
<td>12</td>
<td>14-Jan-2009</td>
<td>20:50</td>
<td>:12</td>
<td>68/188</td>
<td>---/188</td>
<td>Rest</td>
</tr>
<tr>
<td>VT-NS</td>
<td>11</td>
<td></td>
<td></td>
<td>14-Jan-2009</td>
<td>15:06</td>
<td>:02</td>
<td>47/179</td>
<td>Rest</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Last Programmer Session 14-Jan-2009

| VT   | 3   | 20J    | Yes     | 10    | 13-Jan-2009  | 23:41 | :32      | 87/188 | ---/188| Active          |
| FVT  | 4   | 20J    | Yes     | 9     | 13-Jan-2009  | 23:20 | :38      | 94/188 | ---/188| Rest            |
| VT   | 2   | 20J    | Yes     | 8     | 13-Jan-2009  | 23:15 | :26      | 95/188 | ---/188| Rest            |
| VT   | 3   | 20J    | Yes     | 7     | 13-Jan-2009  | 23:05 | :33      | 94/188 | ---/188| Rest            |
| VT   | 2   | 20J    | Yes     | 6     | 13-Jan-2009  | 21:49 | :27      | 94/188 | ---/188| Rest            |
| VT   | 3   | 20J    | Yes     | 5     | 13-Jan-2009  | 20:35 | :32      | 102/188| ---/188| Rest            |
| VT   | 3   | 20J    | Yes     | 4     | 13-Jan-2009  | 20:16 | :32      | 88/188 | ---/188| Rest            |
| FVT  | 5   | 20J    | Yes     | 3     | 13-Jan-2009  | 19:53 | :45      | 83/188 | ---/194| Rest            |
| VT   | 3   |        | Yes     | 2     | 13-Jan-2009  | 15:50 | :25      | 97/188 | ---/188| Rest            |
| VT   | 3   |        | Yes     | 1     | 13-Jan-2009  | 14:00 | :26      | 94/188 | ---/188| Rest            |

Figure 3.5.4 Example of an event log.
oversensing and not actual arrhythmia. This can often be seen on the stored EGMs and then comparing them to the observed ECG that is obtained with patient in clinic and running a real-time ECG strip.

which is especially helpful when trying to tailor antiarrhythmic or rate control therapy (Fig. 3.5.5). It is important to review these data in the context of each individual patient as some episodes of mode switching can be due to atrial

**Figure 3.5.5** Example of an atrial fibrillation (AF) log with ventricular response during AF. Note the rate excursions in excess of 100 bpm [arrow].
**Percentage of Pacing**

The device will provide a detailed breakdown of the percentages of both atrial and ventricular pacing. These percentages should be reviewed at each visit or remote check to make sure the “goal” of ventricular pacing is being met. Numerous clinical trials have demonstrated the benefit of reducing unnecessary ventricular pacing in patients with sinus node dysfunction (Anderson et al. 1997; Wilkoff et al. 2002; Sweeney et al. 2003; Steinberg et al. 2005; Sweeney and Helcamp 2006; Sweeney et al. 2007) (Fig. 3.5.6). Conversely, alternate large-scale clinical trials have demonstrated the benefit of cardiac resynchronization therapy (Bristow et al. 2004; Cleland et al. 2005), also known as biventricular pacing, where the goal of therapy is to provide 100% biventricular pacing. Understanding the goals of pacing is imperative in assuring quality of care and maximizing positive patient outcomes. Some device vendors provide specialized algorithms that are designed to minimize unnecessary ventricular pacing by extending the AV delay (Medtronic Search AV+® [Medtronic, Inc.], Boston Scientific AV Search Hysteresis® [Boston Scientific, St. Paul, MN]) or provided dedicated AAI pacing with backup DDD pacing in the event of complete heart block (Medtronic MVP® or Biotronik AAIR Safe® [Biotronik, Lake Oswego, OR]). It is important to view these data in the context of each individual patient. For example, a device may record a high percentage of ventricular pacing; however, observation of the real-time EGM reveals pseudofusion (an ineffective pacing stimulus delivered during the absolute refractory period following an intrinsic beat) because the AV delay is programmed close to the patient’s intrinsic PR interval. Although pseudofusion may result in a slight increase in battery drain, it does not alter ongoing ventricular intrinsic activation and may be preferable to a shorter programmed AV delay that causes right ventricular (RV) apical pacing and imposes a left bundle branch block on the patient.

<table>
<thead>
<tr>
<th>Pacing</th>
<th>(% of Time Since 13-Nov-2008)</th>
</tr>
</thead>
<tbody>
<tr>
<td>AS-VS</td>
<td>17.9%</td>
</tr>
<tr>
<td>AS-VP</td>
<td>0.0%</td>
</tr>
<tr>
<td>AP-VS</td>
<td>82.0%</td>
</tr>
<tr>
<td>AP-VP</td>
<td>0.2%</td>
</tr>
<tr>
<td>MVP</td>
<td>On</td>
</tr>
</tbody>
</table>

**Figure 3.5.6** Example of percent pacing. Note that the device is primary providing atrial rate support with minimal ventricular pacing.

**Heart Rate Histograms**

Heart rate histograms provide a visual trending of the percentage of time pacing occurs at a particular heart rate. Many devices provide information on both atrial and ventricular rate binning as well as A–V conduction. The histograms provide useful information with regards to heart rate response to activity, particularly in patients with sinus node dysfunction. The heart rate histogram is also particularly useful in assessing rate control in AF and can be used to tailor rate control therapy (Fig. 3.5.7).

**Lead Trending**

Lead trending data for the atrial, RV, left ventricular (LV), and high-voltage lead systems are particularly useful to monitor lead performance over time. Trending information is derived from lead impedance data and intrinsic value measurements for both the atrial and ventricular leads. An absolute value is often not useful in an individual patient, given the wide range of “normal” values. A thorough review of the trended data allows for abrupt changes in lead data to be easily visualized (Fig. 3.5.8).

**Sensing Integrity Counter (SIC)**

A specific vendor provides information known as the SIC, which is a robust measurement of lead integrity. The device recognizes short non-
Alert Notifications

Most contemporary ICD systems have alert features that monitor device functionality and certain clinical conditions and provide either an audible or vibratory alert that warns the patient of a pending issue. With the advent of the new wireless remote monitoring systems, the alert condition can seamlessly be delivered to the patient’s health care provider via a Web site.
Figure 3.5.9 Example of a sensing integrity counter (SIC). Note the SIC has incremented to 205 short V-V intervals. Coincident with this is an RV lead impedance greater than 2,500 ohms.

fax, e-mail, voicemail, or text message notification. Commonly available device alerts are noted in Table 3.5.4. (Epstein and Shea 2008). If an alert condition is present, it is recommended that the patient be brought in to the office for a full device interrogation and to have the alert condition reset (Fig. 3.5.10).

**ROUTINE DEVICE FOLLOW-UP**

After reviewing the device diagnostics, the next logical progression is testing of lead impedance, intrinsic measurements, and pacing thresholds. Many contemporary devices have an automated follow-up feature or will do the measurements automatically. If the trending data for all the device parameters is stable, this is generally adequate; however, it is typically recommended that semiautomatic and, when indicated, manual measurements be made while the patient is in the office. This gives the practitioner greater assurance that the automated measurements obtained between office visits are accurate.

**Lead Impedance**

The atrial, RV, and LV lead impedances are typically automatically measured by the device. The normal range is 300–800 ohms, although some high-impedance leads can be over 1,000 ohms. High-voltage lead impedances typically range between 30 and 100 ohms, although this can vary depending on the type of lead and its configuration. It is important to evaluate the measured lead impedance as it compares with previous readings to assess for changes over
Table 3.5.4  Device alerts.

- Tachycardia therapies off
- Number of shocks delivered
- All therapies in a zone exhausted
- Low battery voltage
- Excessive charge time
- Device at elective replacement indicator
- Device at end of service
- Atrial fibrillation burden
- Ventricular rate during atrial fibrillation
- Percentage of right ventricular pacing too high
- Percentage of left ventricular pacing too low
- Atrial and ventricular intrinsic amplitude out of range
- Atrial, right ventricular and left ventricular impedances out of range
- Shocking impedance out of range
- Lead integrity alert
- Electrical reset
- Asynchronous pacing mode
- Active Can off without SVC
- Charge circuit time out
- Weight change
- Blood pressure change

Table reprinted from Epstein and Shea (2008).

<table>
<thead>
<tr>
<th>Alert Conditions</th>
<th>Enable-Urgency Tone/Monitor</th>
<th>Threshold</th>
</tr>
</thead>
<tbody>
<tr>
<td>AT/AF Daily Burden Settings</td>
<td>Off/Off</td>
<td>6 hr</td>
</tr>
<tr>
<td>Avg. V. Rate During AT/AF Settings</td>
<td>Off/Off</td>
<td>6 hr at 100 bpm</td>
</tr>
<tr>
<td>Number of Shocks Delivered in an Episode</td>
<td>On-High/Off</td>
<td>1</td>
</tr>
<tr>
<td>All Therapies in a Zone Exhausted</td>
<td>On-High/Off</td>
<td></td>
</tr>
<tr>
<td>A. Pacing Lead Impedance Out of Range</td>
<td>On-High/On</td>
<td>&lt;200 or &gt;1000 ohms</td>
</tr>
<tr>
<td>RV Pacing Lead Impedance Out of Range</td>
<td>On-High/On</td>
<td>&lt;200 or &gt;1000 ohms</td>
</tr>
<tr>
<td>LV Pacing Lead Impedance Out of Range</td>
<td>On-High/On</td>
<td>&lt;200 or &gt;1000 ohms</td>
</tr>
<tr>
<td>RV Defibrillation Lead Impedance Out of Range</td>
<td>On-High/On</td>
<td>&lt;200 or &gt;100 ohms</td>
</tr>
<tr>
<td>SVC Defib Lead Impedance Out of Range</td>
<td>Off/On</td>
<td>&lt;20 or &gt;100 ohms</td>
</tr>
<tr>
<td>Low Battery Voltage RRT</td>
<td>On-High/On</td>
<td>2.62 V(RRT)</td>
</tr>
<tr>
<td>Excessive Charge Time EOS</td>
<td>On-High/On</td>
<td></td>
</tr>
<tr>
<td>VF Detection OFF, 3+ VF or 3+ FVT Rx Off.</td>
<td>On-High/On</td>
<td></td>
</tr>
</tbody>
</table>


<table>
<thead>
<tr>
<th>Date/Time</th>
<th>Event</th>
<th>Threshold</th>
</tr>
</thead>
<tbody>
<tr>
<td>10-Oct-2008 02:15:04</td>
<td>*RV Pacing lead impedance &gt;2500 ohms.</td>
<td>1000 ohms</td>
</tr>
<tr>
<td>10-Oct-2008 02:15:04</td>
<td>*SVC Defib lead impedance Not Taken.</td>
<td></td>
</tr>
</tbody>
</table>

---

Figure 3.5.10  Example of an alert notification.
time, unless the lead impedance has abruptly risen to over 3,000 ohms.

**Intrinsic Measurements**

Intrinsic measurements refer to the device’s ability to recognize a spontaneous atrial (P wave) and ventricular (R wave). In order to obtain a spontaneous measurement, pacing must be temporarily inhibited; therefore, this must be done cautiously in patients whose pacemaker dependence is unknown. For those patients who are known to be dependent, there is no need to attempt a measurement. Many contemporary devices have an automated feature for this, although it is generally recommended to confirm the reliability of the automated measurements by comparing them with manual measurements obtained in the office.

**Pacing Thresholds**

The final component of the routine ICD check is to ascertain the pacing threshold for each chamber. Before checking thresholds, it is important to know if the patient is either functionally dependent in the atrium, ventricle, or both. It is preferable to isolate each chamber being tested, for instance, using the AAI mode for atrial testing and VVI for ventricular testing to avoid confusion around fusing of the intrinsic and paced signal. Some patients dislike the sensation of VVI pacing; therefore, pacing in the DDD mode with a short AV delay is recommended.

**Programming**

The next step of the follow-up process is a systematic review of device programming. Programming is defined as “a non-invasive, stable, reversible change in some of the operating parameters of the device that enable the physician to assess and optimize the device performance and longevity and to tailor these parameters to meet the individual clinical needs of the patient” (Wilkoff et al. 2008a). Typically, the tachycardia parameters established at the time of implant are not changed during a routine clinic visit unless there has been an intervening event and adjustments are warranted. The ventricular sensitivity should not be randomly adjusted unless prior defibrillation threshold (DFT) testing demonstrated that sensing was satisfactory at minimum sensitivity (1.2 mV).

A single VF zone is often reserved for those individuals at risk for polymorphic VT (PMVT) or VF as their presenting arrhythmia (i.e., long QT syndrome [LQTS], Brugada syndrome, or hypertrophic cardiomyopathy [HCM]). Most individuals requiring ICD therapy generally benefit from the addition of a fast VT (FVT) zone where antitachycardia pacing (ATP) is applied as a first-tier therapy. ATP consists of a burst of pacing (generally eight beats) at a rate slightly faster than the clinical VT; the goal being to engage and terminate the tachycardia. The benefit of ATP is that it is typically painless and generally terminates VT, thus sparing the patient a painful shock. Some newer devices allow for ATP during charging in the VF zone.

The PREPARE study (N = 700) examined a specific programming strategy: device detection (FVT and VF NID 30/40), increased tachycardia detection rate (182 bpm), SVT detection discriminators, and ATP delivery (burst, one sequence in the FVT zone) in primary prevention ICD patients. A control group from the MIRACLE ICD and EMPIRIC trials (N = 691) was used for comparison. The study demonstrated that strategic programming of VT/VF
Remote monitoring is rapidly becoming the standard of care for patients with implantable devices and is being fully integrated into most contemporary device clinics. The concept of remote follow-up has evolved into the more sophisticated realm of remote monitoring by providing unprecedented care for patients with implanted cardiac devices. As device technology has continued to evolve with the development of wireless capability and unique patient alert features, remote monitoring is being used to manage not only device functionality but also chronic and often complex clinical conditions as well. Rapid identification of silent or undetected clinical events is essential in preventing untoward events and improved clinical outcomes (Epstein and Shea 2008).

Patients are provided with a home monitoring system (see Fig. 3.5.11) that is connected to an analog phone line. The monitor has a preprogrammed phone number that enables it to transmit the encrypted data automatically to a

**DOCUMENTATION**

Once the device programming is finalized and the device data and diagnostics have been printed, the device counters can be cleared, although this practice will vary from clinic to clinic. A final report is printed for documentation purposes. Many clinics utilize device databases, which serve as a repository for the device data. For some systems, the transfer of data from the programmer to the device database can be done seamlessly via an intranet network connection (Session Sync® [Medtronic]), or by saving the device information to a diskette or USB drive and then importing the data into the database.

The device databases are compatible with many commercial electronic medical record (EMR) systems, thereby creating a flow of data from the device clinic directly into the patient’s EMR. The data are standardized, regardless of device vendor, which makes it more convenient for nondevice practitioners such as anesthesiologists to easily review the data and obtain the information they need in a timely and efficient manner.

**REMOTE FOLLOW-UP**

Remote monitoring is rapidly becoming the standard of care for patients with implantable devices and is being fully integrated into most contemporary device clinics. The concept of remote follow-up has evolved into the more sophisticated realm of remote monitoring by providing unprecedented care for patients with implanted cardiac devices. As device technology has continued to evolve with the development of wireless capability and unique patient alert features, remote monitoring is being used to manage not only device functionality but also chronic and often complex clinical conditions as well. Rapid identification of silent or undetected clinical events is essential in preventing untoward events and improved clinical outcomes (Epstein and Shea 2008).

Patients are provided with a home monitoring system (see Fig. 3.5.11) that is connected to an analog phone line. The monitor has a preprogrammed phone number that enables it to transmit the encrypted data automatically to a
Cardiac Arrhythmia Management

Cardiac Device Monitoring - ICD

Programming per encounter
- Single Lead: $47.25 (-26)
- Dual Lead: $59.51 (-26)
- Multiple Lead: $70.69 (-26)
- Peri-Procedural In Person Only
- Single Lead: $43.64 (-26)
- Dual Lead: $22.72 (-26)
- Any # of leads: $22.00 (-26)

Interrogation
- Single Lead: $32.75 (-26)
- Dual Lead: $14.07 (-26)
- Any # of leads: $36.43

Remote
- Single Lead: $66.36
- Any # of leads: $66.36

Professional Analysis
- Single Lead: $66.36 (G)
- Any # of leads: $36.43

Technical Support
- Single Lead: $66.36 (G)
- Any # of leads: $36.43

Peri-Procedural In Person Only
- Single Lead: $101.71 (G)
- Any # of leads: $101.71 (G)

Figure 3.5.12 2009 Centers for Medicare and Medicaid Services billing codes for cardiac device monitoring: implantable defibrillators. (Reproduced with permission of Medtronic, Inc.)

dedicated server (data repository) for the specific vendor. Once the patient downloads the device data (either wanded or wirelessly depending on the device), the data becomes available to the health care provider on a secure password-enabled Web site, typically within minutes of the transmission. Some vendors are able to transmit data from linked measurement devices such as sphygmomanometers or weight scales, which enable daily measurements of commonly used heart failure indices (Wilkoff et al. 2008a).

Remote monitoring has created a new realm in the area of device follow-up and has changed the paradigm for how patients with implantable devices are currently managed. Most clinics have altered their follow-up schedules so that patients rotate between an office visit and a remote check on a quarterly or biannual basis, according to specific frequency guidelines for billing (Fig. 3.5.12). However, the frequency of visits should be specifically tailored to the patient’s clinical needs and device status.

TROUBLESHOOTING

Device troubleshooting is a critical component of the follow-up process requiring a systematic approach that should include a review of the clinical history, physical evaluation, and comprehensive device assessment.

The clinical history provides many important clues. In situations where the patient has received a shock, particular attention should be paid to any prodromal symptoms such as palpitations, light-headedness, dizziness, chest pain, shortness of breath, presyncope, or syncope. Activity and body position prior to a shock may provide relevant diagnostic clues. The patient’s most recent medication list should be reviewed in detail. Oftentimes, inadvertent therapies may be related to missed doses or discontinuation of certain medications by the patient. In addition, it is important to assess for any related comorbid conditions such as coronary artery disease, heart failure, or atrial fibrillation, which may be contributing factors.

A focused physical examination should be performed to evaluate the device site for any signs of erythema, swelling, drainage, or erosion. The patient should be assessed for any signs of heart failure or any rhythm irregularities that would be suggestive of AF, complete heart block, or nonsustained ventricular arrhythmias.

Lastly, a full device interrogation should be performed. The device and lead(s) model
numbers and serial numbers should be evaluated for any field safety corrective action or safety alert. The initial quick look or summary screen should be scrutinized for any patient alert conditions. A comprehensive review of the device diagnostics should be done to assess for any delivered therapies or nonsustained (NST) events, AF burden and ventricular response, activity level, as well as measures of device functionality such as battery and charge time, lead trending, intrinsic measurements, and threshold values.

Depending on the presenting problem, additional maneuvers may be necessary, such as examining the intracardiac EGM for noise during pocket manipulation, which may be suggestive of a lead fracture or a set screw that was inappropriately or incompletely deployed. Testing at various sensitivity settings can unmask intermittent T wave oversensing. A posterior-anterior (PA) and lateral chest X-ray (CXR) is often recommended if a lead fracture or dislodgement is suspected. It is helpful to compare this with the postimplant CXR, when available, as assessing for lead issues on X-ray can be elusive. Subtle differences may become apparent on comparison. Chest radiographs can also be helpful to identify the brand of ICD by its radiographic signature in situations where the devices’ identity is unknown (Epstein et al. 2007).

OTHER CONSIDERATIONS

Patient education is an integral component of the follow-up process (Epstein et al. 2008; Wilkoff et al. 2008). At the time of implant, the patient, as well as the family or significant other(s), should be educated on the device’s purpose and functionality, incisional care, anticoagulant use, activity restrictions, intimacy, driving recommendations (Epstein et al. 1996, 2007), how to respond to ICD shock (Sears et al. 2005), and routine follow-up care, including the use of remote monitoring. This information should be provided to the patient in a written format prior to discharge (Fig. 3.5.13). Interval office visits provide the opportunity to reinforce this information, provide updates, and respond to specific patient queries at each visit. Including the patient in the process by empowering them to become an active participant in their own care is essential for promoting quality of life and reducing the likelihood for negative or untoward outcomes (Sears et al. 2000; Schroen et al. 2002).

SUMMARY

ICD technology has rapidly evolved since it was first approved by the Food and Drug Administration in 1985. Thirty years of research and development has produced fully programmable, multifunctional devices that provide extensive clinical data and therapy options. As a result, the complexity of patient follow-up has evolved in parallel. Through remote technology, we now have greater access to patient data, earlier identification of silent events, and ultimately, can provide more timely interventions. However, this does not negate the need for periodic in-office visits to assess the human–device interaction.

The paradigm for device follow-up continues to evolve as well. Technological advancements will continue to mold the device clinic of the future. In the not-so-distant future, we will have the ability to reprogram devices remotely. Cellular technology will be integrated into our remote systems, thus providing instant connectivity with patients.

Appropriate follow-up, monitoring, and troubleshooting of ICD devices is critical in promoting patient well-being and mitigating untoward events. A thorough understanding of cardiac electrophysiology, device features, and algorithms, as well as complex patient conditions and comorbidities is paramount. This chapter provided the fundamentals of ICD follow-up as well as a comprehensive review of
Figure 3.5.13 Brigham and Women’s Hospital ICD discharge instructions.
several commonly encountered troubleshooting issues. The allied professional plays a pivotal role in the management of the ICD patient, the importance of which cannot be underestimated, and ultimately, contributes to the success of the therapy.

The following case studies illustrate some common troubleshooting concerns that are often encountered during routine follow-up of ICDs. Each case study will include the diagnostic modalities used as well as the specific management plan.

**Case 3.5.1**

A 47-year-old male with hereditary LQTS status post dual-chamber pacemaker placement in 1993 was admitted in March 2001 after experiencing 4 days of intermittent chest pressure, shortness of breath, and dizziness. He was ruled out for a myocardial infarction. His pacemaker was interrogated and his device had logged two brief episodes of nonsustained torsades de pointes. He subsequently underwent an upgrade of his dual-chamber pacemaker system (Guidant Discovery II 1283) to a dual-chamber ICD (Guidant Ventak Prizm 1861 and Medtronic 6943 dedicated bipolar lead in the right ventricle). This device reached ERI in November 2005 and he underwent an uncomplicated generator change (Guidant Vitality DS T125).

He presented in October of 2008 for a routine in-office device check. Overall, he had been doing well from an arrhythmia perspective and specifically denied any symptoms of light-headedness, dizziness, palpitations, nor syncope. However, upon interrogation of the device, it was noted that he had logged one aborted VF event on September 3, 2008 at 17:21. The stored EGM of the event is shown in Figure 3.5.14. What is the EGM suggestive of? How do you proceed from here?

**Case Discussion**

As mentioned previously, the patient denied any symptoms suggestive of arrhythmia nor had he been ill. He denied any new medication use (either prescription or over-the-counter [OTC]). The event time correlated with the end of his workday, but the patient could not recall anything unusual on that particular date. The stored EGM is suggestive of some type of electromagnetic interference (EMI) on the ventricular channel causing oversensing, as noted by the abnormal signals appearing on all three marker channels simultaneously (atrial, ventricular, and shocking EGMs) (note arrows on Fig. 3.5.14). The patient’s normal rhythm can be seen.
through the artifact. Noise due to a lead fracture would be limited to the affected lead only, as opposed to EMI which appears on both channels, as in this case.

Upon further questioning, the patient recalled carrying a piece of equipment (magnetic ballast; Arri 1,200-W HMI Par [Arri, Munich, Germany]) at work, which likely caused the interference. The patient was provided with education regarding potential interactions between EMI sources and his ICD. No programming changes were made.

Oversensing is defined as “the sensing of inappropriate signals, which can result in pauses, inappropriate mode switching or inappropriate tachycardia detection and/or therapy” (Scher 2003). EMI results from sensing of electrical activity outside the body that is typically nonphysiological (less than 200 ms). Potential sources of EMI are listed in Table 3.5.5.

<table>
<thead>
<tr>
<th>Potential sources of EMI</th>
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<tbody>
<tr>
<td>Electrocautery</td>
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<td>Magnetic resonance imaging</td>
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<tr>
<td>Lithotripsy</td>
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<td>Transcutaneous electrical nerve stimulators (TENS)</td>
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<td>Therapeutic radiation</td>
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<tr>
<td>Electroconvulsive therapy (ECT)</td>
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<tr>
<td>Cellular telephones</td>
</tr>
<tr>
<td>Personal digital assistants (PDA)</td>
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<tr>
<td>Electronic surveillance systems</td>
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<tr>
<td>Battery operated items</td>
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<tr>
<td>Alternator of a running car engine</td>
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<tr>
<td>Slot machines</td>
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<tr>
<td>Arc welding</td>
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<tr>
<td>High-voltage power sources</td>
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<td>Improperly grounded appliances or electrical tools</td>
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Case 3.5.2

A 58-year-old male with coronary artery disease, paroxysmal AF, and an ischemic cardiomyopathy underwent diagnostic electrophysiological testing in October 2001 and was found to have inducible monomorphic VT. He subsequently underwent implantation of a dual-chamber ICD (Medtronic Gem III DR 7275 and 6943 dedicated bipolar lead in the right ventricle). This device reached ERI in June 2005 and was replaced with a Medtronic Maximo DR 7278 device. In June 2008, he underwent routine ICD safety margin testing and an elective cardioversion for AF. His DFT remained stable at 20 J and was tested at minimum sensitivity of 1.2 mV. The shock did not convert the AF; therefore, he did receive a 200 J transthoracic shock that restored sinus rhythm. His original programmed parameters were restored (VF and FVT zones, with a ventricular sensitivity of 0.3 mV). His medical regimen was not changed at the time.

On September 8, 2008, the patient was urgently admitted to an outside hospital after receiving six successive shocks from his device. In the emergency room, he was confirmed to be in sinus tachycardia and his ICD was suspended with a magnet until a Medtronic 2090 programmer was made available. Upon the initial interrogation, he was noted to have logged two FVT events, 16 NST events, and one mode switch event. The FVT events occurred on August 6 and the second event on September 8. The NST events all occurred between August 6 and September 8. The scatter plot and stored EGM of the event on September 8 are shown in Figures 3.5.15 and 3.5.16. What is your interpretation of the data? How would you proceed from here? How can you prevent this from happening again?

Case Discussion

The patient received six sequential shocks for T wave oversensing (TWOS), although it is unclear why this started spontaneously. The scatter plot demonstrates the characteristic “train track” appearance of the V-V signal, which unfortunately fell into the therapy zones. The marker channel on the
stored EGM of the event clearly demonstrates the T waves being inappropriately marked as “tach fast” (TF) events (see Fig. 3.5.16). This inappropriate double counting of the R wave and T wave results in the device meeting the rate cutoff for therapy (the patient’s sinus rate was about 100 bpm) just prior to the event. The device identified the ventricular cycle length as 310 ms.

TWOS is the most common oversensing problem in ICDs due to the oscillating size of the T wave relative to the R wave. Young age, dynamic electrolyte shifts, ischemia, certain metabolic conditions, and exercise can influence the degree of TWOS. TWOS is seen more frequently with integrated bipolar leads as compared with dedicated bipolar leads (Scher 2003; Van Erven and Schalij 2008).

The patient’s ventricular sensitivity was reprogrammed to 0.6 mV at the outside hospital to minimize the likelihood of recurrent TWOS. His medical regimen was not changed and he was discharged home the following day. He was seen in the office 9 days later. Upon interrogation of his device, it was noted that he had not logged any recurrent events. His rate cutoff was increased to 200 bpm

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<th>VT/VF Episode #9 Report Page 1</th>
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**Figure 3.5.15** Scatter plot of T wave oversensing. Note the characteristic “train track” appearance of the ventricular signals (arrows), suggesting oversensing.

**Figure 3.5.16** T wave oversensing resulting in six consecutive shocks. Note the R waves and T waves both being marked as tach fast (TF) events (arrows).
and the ventricular sensitivity was reduced to 0.9 mV (as VF detection was robust at 1.2 mV) to further reduce the likelihood of recurrent TWOS.

Oversensing can be corrected by adjusting the sensitivity or refractory period of the ICD (Epstein et al. 2007, p. 1072). As mentioned previously, the ventricular sensitivity should not be adjusted unless VF detection is robust at the minimum sensitivity of the device (typically around 1.2 mV). Some devices have “autoadjusting” sensitivities, which adjust the sensing between sensed and paced rhythms resulting in oversensing only during ventricular pacing. In patients in whom TWOS is related to ventricular pacing, efforts should be made to minimize ventricular pacing by extending the AV delay or reprogramming to the VVI mode if indicated. Certain devices also offer adjustable thresholds or slope for measuring the T wave, which is particularly useful in those patients prone to TWOS, as may be seen in HCM. Rate control agents are sometimes employed to blunt the heart rate response, thus if double counting occurs, it will not result in the rate reaching a therapy zone. Less frequently, repositioning of the RV lead or even changing the device may be indicated.

Case 3.5.3

An 82-year-old female with a past medical history of hypertension, hyperlipidemia, left bundle branch block, and cardiomyopathy with an ejection fraction of 30% was seen by her primary care physician in March 2007 for complaints of an elevated systolic blood pressure (160–180) and a mild headache. Her pulse was noted to be in the 40s. A 12-lead ECG was obtained, which demonstrated complete heart block with ventricular escape complexes, normal axis, and T wave inversions in aVL, VI, and V3–V5. She was transferred to the hospital for further evaluation. A cardiac catheterization was performed, which revealed a left dominant system and a 50% discrete proximal left anterior descending lesion. The patient subsequently underwent implantation of a Guidant 4473 atrial lead, a Medtronic Fidelis™ 6931 RV lead, a Medtronic 4194 LV lead, and a Medtronic Concerto C154DWK CRTD device. She tolerated the procedure without incident and was discharged from the hospital within a few days.

She was seen in follow-up 2 weeks later and her device was functioning satisfactorily. She was scheduled for a remote device check in 3 months’ time. Due to her age and co-morbidities, she was followed by her local Visiting Nurses Association. During a visit in early April 2007, she was noted to have a pulse in the low 40s, with intermittent light-headed episodes. She subsequently was readmitted and found to have a dislodged RV lead (Fig. 3.5.17). She was taken to the Electrophysiology Lab and had the RV lead repositioned. Due to the appearance of the leads within the pocket, there was some question of twiddler’s syndrome; a condition where the patient, either consciously or unconsciously, spins or flips the pulse generator within the pocket. There is a typical characteristic “twisting” of the leads upon each other (see Fig. 3.5.18).

In October, 2007, Medtronic, Inc. issued an advisory on all its Fidelis leads for premature lead
failure due to fracture (the original event rate was 2.3% at 30 months rising to 6.2% at 40 months). The patient was brought in for the recommended reprogramming and was already set up with wireless remote monitoring at home.

On October 10, 2008, a CareAlert® (Medtronic, Inc.) notification was received on the remote Web site and an alert notification was received via e-mail by the arrhythmia nurse practitioner that morning. Upon viewing the transmission, it was evident that the patient had sustained a fracture of the advisory RV lead and was having periods of intermittent asystole due to inhibition by noise as noted in Figure 3.5.19. She had experienced a spike in her RV lead impedance to over 2,500 ohms and was

Figure 3.5.18 Example of “twiddler’s syndrome.” Note how the leads are twisted upon themselves (arrow). (Photo courtesy of Laurence M. Epstein, MD.)

Figure 3.5.19 Inhibition of ventricular pacing due to noise artifact (arrow).
having multiple nonsustained ventricular tachycardia (NSVT) events due to noise. Her SIC had begun to increment in the past 24 hours (see Fig. 3.5.9). The patient was contacted immediately and advised to call 911. She was taken to a local hospital where she was met by an industry representative who programmed her device to an asynchronous pacing mode, and her tachycardia therapies were disabled. She was transported via ambulance to our facility where she underwent an extraction of the fracture lead, which was twisted upon itself at least eight times. The atrial and LV leads, although twisted, were unharmed and tested out normally. She underwent reimplant of a new Medtronic 6947 Sprint Quattro lead, and considerable efforts were made to secure the lead to prevent further twiddling. It is unclear as to whether or not the RV lead failure was a function of the advisory failure or due to the patient’s twiddling behavior. How would you proceed from here? What would you do to prevent this from happening again?

**Case Discussion**

Lead issues, although uncommon, are generally easily recognizable. A thorough patient history, physical exam, and comprehensive device interrogation provide the necessary data to make the diagnosis. With a lead dislodgment, insulation breech, or fracture, the patient may experience symptoms of shortness of breath, fatigue, light-headedness, dizziness, near syncope, or frank syncope, depending on the amount of pacing support required and in which chamber the affected lead resides. A review of the stored device data may demonstrate several inappropriate tachycardia events due to oversensing or noise, inappropriate therapies, a change in the percentage of pacing, a change in lead trending data (impedances), intrinsic measurements, or pacing thresholds. With either a dislodgment or fracture, the lead impedances will be high (>2,500 ohms) and the capture threshold will be elevated or a lack of capture will be evident. With an insulation breech, the lead impedances will be normal or low (<200 ohms) and the capture threshold will be stable or elevated, depending on the severity of the breech. Sometimes a “make-break” connection will be present, so the breech is not always obvious and may require further investigation.

Manipulation of the device pocket while obtaining a continuous EGM may unmask a lead issue as evidenced by noise on the affected lead. Sometimes, testing the lead at various sensitivities may be required as well as observing the effect of postural changes and deep inspiration and expiration (Van Erven and Schalij 2008). If the lead issue occurs shortly after implant, the provider should be suspect of an inappropriate or incomplete set screw deployment.

A PA and CXR should be obtained, which will clearly demonstrate a lead dislodgment (see Fig. 3.5.17). Occasionally, there can be a “microdislodgment” of the lead; therefore, the CXR should be compared with the postimplant film for any subtle changes in lead position. Lead fractures are often difficult to assess on CXR; however, Figure 3.5.20 demonstrates an obvious fracture and fraying of the lead.

Unlike pacemakers, ICD leads cannot be reprogrammed to the unipolar pacing configuration because of the potential risk for underdetection of VT or VF due to the unipolar pacing signal. If the lead is dislodged, the patient will need to be admitted for lead repositioning. If the lead has an insulation breech or fracture, the patient will need to have the lead extracted and replaced. This procedure is typically performed by an experienced operator in a cardiac operating room due to risk of perforation and tamponade. Alternatively, a separate pace/sense lead can be added to the system. However, if the patient is young, it is generally recommended to remove the damaged lead and replace it; particularly as the patient may require another replacement later on in life.

Device and/or hardware advisories or “safety alerts” present a unique challenge in the management of patients with implantable devices. It is imperative for practitioners involved in device management to be intimately aware of device advisories or “safety alerts” and have a thorough understanding of the mechanism of failure and management recommendations (Epstein et al. 2008).

Each patient’s situation must be considered on an individual basis, with careful attention being given to the patient’s age and clinical status, the type of hardware on advisory, the type of advisory and potential ramifications, and most importantly, the risk/benefit to the patient. Complications of replacing advisory hardware, such as infection, reoperation, and death have been reported (Gould and Krahn 2006). Therefore,
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it is important to consider the estimated risk of the device failure rate combined with the mortality risk from that failure against the risk of proce-
dural morbidity/mortality associated with a device and/or hardware replacement (Epstein et al. 2008).

Case 3.5.4

A 46-year-old woman with a diagnosis of LQTS underwent placement of a permanent pacemaker (St. Jude Medical model 5386 [St. Jude Medical, Sylmar, CA]) in 1995 after suffering a syncopal event while running. In March 2007, she was admitted with complaints of fatigue, palpitations, and dyspnea. Interrogation of her pacemaker revealed asymptomatic AF with rapid ventricular pacing near the upper rate limit; as her AV interval had progressively lengthened, rendering her essentially pacemaker dependent. Her echo revealed normal LV size, with moderate to severe LV dysfunction (ejection fraction of 25) and global hypokinesis. This was felt to be a tachymyopathy. A cardiac catheterization was performed, which showed a right dominant system with clean epicardial coronary arteries and normal filling pressures. Four RV biopsies were taken. Her pacing system was explanted and she underwent implanta-
tion of a dual-chamber ICD (Medtronic 4076 atrial lead; Medtronic 6931 ventricular lead, and a Medtronic Virtuoso D154AWG device).

She did well until November 2008, when she received a shock from her ICD while out jogging and was seen at a local hospital. Her ICD was interrogated and was functioning satisfactorily. Her device diagnostics logged one event that fell into the FVT zone. Examination of the stored EGM (Fig. 3.5.21) revealed a rapidly conducting SVT with a 1:1 relationship and virtually simultaneous AV timing averaging about 200 bpm. This was treated initially with a single burst of ATP, which terminated the tachycardia; however, the tachycardia immediately reinitiated and was terminated with a 35-J shock. Based on evidence of dual AV nodal
Figure 3.5.21  Inappropriate therapy for 1:1 supraventricular tachycardia (SVT) likely AVNRT as evidenced by simultaneous A and V events (arrows).
pathways on her prior 12-lead ECGs over the past few years, this likely represents AV nodal reentry tachycardia (AVNRT). Her beta blocker dose had been recently reduced from 200 mg to 100 mg daily and, combined with heavy exercise and increased adrenergic tone, likely accounted for the onset of the tachycardia.

Her FVT detection zone was increased to 222 bpm and her beta blocker dose was increased by 50 mg. She was referred for catheter ablation of her AVNRT. In December 2008, she underwent a complex SVT ablation. Although she did have dual AV nodal pathways, she had no inducible AVNRT due to lack of retrograde conduction. The slow pathway was modified. An empiric flutter line was placed along the crista terminalis (CTI). She tolerated the procedure well and was discharged home the following day. She has not experienced any recurrent paroxysmal supraventricular tachycardia (PSVT) to date or any recurrent shocks. How would you handle inappropriate shocks for PSVT? What additional steps could you take?

**Case Discussion**

Repeated inappropriate ICD shocks for SVT accounts for approximately 10–24% (Germano et al. 2006) of all ICD therapies. Supraventricular rhythms include sinus tachycardia, AV nodal reentry with 1:1 conduction, accessory pathways with 1:1 conduction, and most commonly, AF or atrial flutter with a rapid ventricular response. Multiple ICD shocks can occur, typically due to repeated detection in the VT, FVT, or VF zones or the inability to terminate the SVT, which can result in significant psychological sequelae such as seen in posttraumatic stress disorder (PSTD) (Sears et al. 2000; Schoen et al. 2002).

Adjustments can be made to the device’s programming to enable the device to discriminate between VT and SVT, thus reducing the likelihood of SVT detection and inappropriate therapy. Several device vendors have developed specialized algorithms to increase the specificity of SVT discrimination without compromising the sensitivity for VT detection (Van Erven and Schalij 2008).

Pharmaceutical agents such as beta blockers, calcium channel blockers, or potent AV nodal blocking agents (adenosine) can be employed for rate control and/or arrhythmia termination. Antiarrhythmic agents can be added to reduce the likelihood of recurrence and for maintenance of sinus rhythm. In some cases, catheter ablation is warranted to prevent further episodes of the SVT.

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**Case 3.5.5**

A 76-year-old male with an ischemic cardiomyopathy was referred for a cardiac resynchronization ICD (CRTD) in August 2002. In May 2007, he underwent a CRTD generator change (Medtronic Concerto C14DWK). In November 2008, he presented with a left infraclavicular pocket infection and erosion to the point where the metal can of the device was visible through the skin (Fig. 3.5.22). The surrounding area was reddened and tender, although he remained afebrile with a normal white blood cell count. Wound and blood cultures were negative. He was admitted for a complete CRTD system extraction and was started on intravenous vancomycin. The patient was prepped for surgery.
in a cardiac operating room/suite. The LV lead was removed with traction alone and the atrial and RV leads were removed with a #16 French laser sheath. The pocket was extensively debrided and he was placed on intravenous antibiotics. Three days later, he underwent implantation of a St. Jude Medical Promote 3207-36 CRTD device on the right. He made an uneventful recovery and was eventually discharged to home on 2 weeks of oral antibiotics. How would you handle device pocket erosion? What additional steps could you take?

Case Discussion

Early complications from ICD implantation can occur during the periprocedure phase, which include pocket hematoma, RV perforation with pericardial effusion and/or tamponade, and pneumothorax. Later complications can include stitch abscess, pocket infection with or without erosion, and thrombophlebitis on the ipsilateral side of the device (Stevenson et al. 2004). Device infections are generally limited to the pocket but can involve the lead(s) as well.

For patients on anticoagulation therapy, extreme care should be used when reinitiating anticoagulation post procedure. The international normalized ratio (INR) or PT/PTT should be monitored frequently to avoid bleeding into the ICD pocket and subsequent hematoma formation. Hematomas can be quite painful, can impede blood flow in the microcirculation of the pocket, and can provide a breeding ground for bacteria, thus increasing the risk of infection. Injectable low-molecular-weight heparins (LMWHs), such as Lovenox® (Sanofi-Aventis, Bridgewater, NJ), should be avoided post implant due to their lack of reversibility.

Infected devices and/or leads typically require a full extraction (removal) of the affected hardware. Antibiotic therapy is typically unsuccessful in salvaging an infected system. The most common organisms responsible for pocket infection include Staphylococcus aureus and Staphylococcus epidermidis and less likely Escherichia coli, Pseudomonas, and Serratia. Blood cultures are typically negative (Wilbur and Marchlinski 1999).

The extraction is usually done in a cardiac operating room/suite by an experienced operator due to the risk of RV perforation/tear, SVC tear, or other vascular complications (estimated at less than 1%). Once the infected system is removed, and antibiotic coverage is deemed appropriate and adequate, a new system can be implanted on the opposite side. The infected pocket may be left open to heal by secondary intention. If indicated, an infectious disease consult should be obtained for recommendations regarding the appropriate antibiotic coverage. Depending on the situation, some patients may need to receive several weeks of antibiotics through a percutaneously inserted central catheter (PICC) before a new ICD system can be implanted. Patients with previously positive blood cultures should have repeat cultures done and reimplanted only when the cultures are negative.

REFERENCES


RESOURCES


INTRODUCTION

Device follow-up today consists of acquiring, interpreting, acting upon, and documenting a large amount of data that is acquired at regular intervals from both the patient and the device. Developing a systematic approach to device follow-up ensures that data will not be missed.

In the past, devices were followed by having patients come to the clinic at regular intervals. In early devices, little diagnostic data was available. Only limited testing could be done, and some devices were nonprogrammable. Battery status on pacemakers was routinely estimated by magnet rate. Today’s devices contain complex technology, which provides a wealth of diagnostic data, and a wide range of programmable parameters. They can be monitored by in-office visits, by telephone, and more recently, by remote monitoring devices that download the device information to a secure Internet site where it is accessed by clinic personnel.

TYPES OF FOLLOW-UP

A long-standing method of follow-up is the telephone pacemaker check. These telephone checks were the earliest form of remote monitoring. They are still in use today for pacemakers that do not have other remote monitoring capability. The checks require the patient to be in telephone contact with a technician. The patient places electrodes on their wrists or legs, then places the telephone in the cradle of a specialized monitoring system. Figure 3.6.1 is an example of the equipment patients may use for these checks. This sends a signal to the technician at the other end of the phone and a rhythm strip is generated. The patient then places a magnet over the pacemaker,
temporarily reprogramming the patient to a nonsensing, nontracking mode. The subsequent rhythm strip will be at the device’s programmed magnet rate. The magnet rate is used to determine battery status.

In addition, many pacemakers perform a threshold margin test when the magnet is first placed on the device. The device delivers three pacing pulses at 100 beats per minute at the programmed voltage. This is followed by one pacing pulse at 75% of the programmed voltage. Failure to capture on the fourth pulse demonstrates that the patient’s device is not programmed to provide a safety margin of two times the capture threshold.

When compared with the amount of data available from a device today, these telephone checks provide limited information. They are still performed today on devices that do not have remote follow-up capabilities.

In recent years, remote monitoring of devices has become an accepted method of device follow-up. There are two types of remote monitoring systems. In the earliest systems, the patient received a remote monitoring system from the manufacturer of the device. Figure 3.6.2 illustrates one type of remote monitoring system. At intervals determined by the clinic, the patient connects the system to a phone line and places an interrogation wand over their device. They press a button, initiating the download, and the system interrogates the device and delivers the information to a secure Internet site. Some devices connect the patient to a technician, who assists the patient to download the data from the device. Clinic personnel access the site using a password and retrieve the information for interpretation.

As devices with the capability to communicate by wireless telemetry came on the market, a second type of remote monitoring system emerged. This system requires the patient to have very little interaction with the system beyond the initial setup. For this system, the patient receives the monitor, places it in their home, and connects it to the phone line. Some manufacturers’ devices are using cell phone technology for this. Once the monitor is turned on and makes initial contact with the patient’s device, it monitors the device on a routine basis without the patient having to initiate an interaction. This allows the monitor to detect problems (increased arrhythmia burden, abnormal heart failure data, or device/lead issues, etc.)
and download the information to a secure Web site, while simultaneously notifying the clinic that a problem exists. These monitors can also be programmed to do routine downloads at set intervals (such as the first Thursday of every third month), allowing the clinic to receive the information without the patient having to schedule a download.

Remote monitoring has changed the face of device follow-up tremendously. Patients no longer need to come into the clinic every month. For patients, this translates into a definite time savings as they do not have to travel to the clinic, find parking, wait for a clinician to download the device, sit through the interrogation, wait at the checkout desk, then return to their parked car. They can have follow-ups done from their own home, at times that are convenient to them. Additionally, for patients whose devices are equipped with wireless technology, there is the added security of knowing the device is being monitored on a daily basis.

The impact of remote device follow-up on a clinic is still evolving. Many clinics find that there is no significant time savings from remote monitoring of devices. Due to the amount of diagnostic data that is downloaded during remote monitoring, there is little time savings involved in interpreting the data. Device testing is not done on remote checks; however, many newer devices perform these tests internally at regular intervals and report the data in trend form. The patient must still be contacted after the information has been analyzed. For a routine download, with no significant problems noted, many clinics opt to do this by mail-out card. If problems are noted, however, a phone call to the patient is in order to assess any problems or symptoms the patient may be experiencing.

Scheduling for wireless devices has also changed. The clinician can program the schedule into the Web site and the device download will automatically take place on the scheduled day, providing the patient is in range of the transmitter. Figure 3.6.3 shows one manufacturer’s scheduling site. Clinic can set up download days in many ways. For a small clinic, it may be more practical to set up all the downloads on one day each week. Larger clinics will likely be receiving downloads every day. In addition, the transmitters can detect device or lead problems on wireless devices and send an alert, complete with a device download to the clinic.

Each clinic will determine what warnings and alerts they will receive from devices. Figure 3.6.4 shows a typical Web site, detailing how the alerts can be configured. In addition, each clinic determines how they receive patient alerts. Some choose to be contacted by fax, others by phone or beeper. Setting up alert conditions and deciding how they will be dealt with when a clinic initiates remote follow-up makes the process easier.

**FREQUENCY OF FOLLOW-UP AND TRACKING PATIENTS**

Device follow-up should occur on a regular schedule. In the Heart Rhythm Society (HRS)/European Heart Rhythm Association (EHRA) Expert Consensus on the Monitoring of Cardiovascular Implantable Electronic Devices (CIEDs): Descriptions, Indications, Personnel, Frequency and Ethical Considerations, the following schedule for follow-up is recommended:

- Within 72 hours of device implantation (in person)
- 2–12 weeks after implantation (in person)
- Every 3–12 for pacemakers and cardiac resynchronization pacemakers (in person or remote)
- Every 3–6 months for implantable cardiac defibrillators (ICDs) and cardiac resynchronization ICDs (in person or remote)
- Annually until battery depletion (in person)
- Every 1–3 months at signs of battery depletion (in person or remote)
As the number of patients with devices increases, tracking patients will become increasingly difficult. Some types of databases are useful to accomplish this task with efficiency. Whether a clinic uses a database they have created to meet the specific needs of their clinic, or one of the commercial databases available for device tracking, is a matter of personal preference. A further consideration for maintaining an efficient tracking system is the ability to quickly locate patients who have particular devices or leads when an alert or recall is announced for that device or lead. In a clinic that follows 50 patients, going through each one by hand to search for a lead or device may not be terribly time consuming. However, in a clinic that

![Figure 3.6.3](image)

**Figure 3.6.3** Example of a scheduling site for remote follow-up. Note that the clinician has several options for scheduling follow-up, as well as for configuring how alerts are handled.
follows several thousand patients, such a process would be overwhelming.

**FOLLOW-UP PROCEDURES**

There are many things that happen during a device follow-up. Device testing is only one of the details to be completed. Others include downloading and reviewing diagnostic data, eliciting information from the patient, identifying problems, troubleshooting, and reprogramming the device. The final step in any patient encounter is always documentation.

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**Gathering and Reviewing the Data**

**Patient History**

When a patient presents for a device follow-up, it can be for a routine, scheduled visit, or because the patient perceives a problem with the device. Remote follow-ups can be regular, scheduled follow-ups for patients who have called with problems or for alerts that have triggered an automatic download from the device.

Do not forget that the information you gather from the patient and family is as important as, if not more important than, the device data. Listen closely to what your patients are telling you and many times you will have a very good idea of what the problem is before you even turn the programmer on. Expert diagnosticians will tell you that most of their time with a patient is spent on taking a good history.

They have narrowed down their differential diagnoses before they ever touch the patient or look at test data. During routine device follow-up, information from the patient may range from patients feeling fine, to multiple complaints, such as chest pain, shortness of breath, fatigue, decreased exercise tolerance, palpitations, twitching, and a whole variety of other problems. This is also the place to ask if there any new medications have been prescribed. Many medications, particularly antiarrhythmics, can affect capture thresholds or defibrillation thresholds.

Good interviewing techniques involve asking open-ended, nonleading questions, and most important, developing good listening habits. If you ask a patient, “Did you feel as if you were going to pass out at 10:00 am last Sunday?” they may answer “Yes” because they think that is the answer you want. A better question is, “Tell me about any worrisome symptoms you have had since you were last here.” It takes a little longer, but it is time well spent.

During remote follow-up, this information will not be immediately available since the
a reference for any programming changes made during the session and serves as a “double check” at the end of the session to make certain that no unintentional changes have been made. Some newer devices print automatically when the device is interrogated. Note, however, that many of these, especially ICDs, do not provide a complete parameter report. Get in the habit of printing the parameters separately at the start of every interrogation.

Device Performance Data

Percent of Pacing

Analysis of this data helps determine if the device is performing the basic tasks it was implanted to perform. Noting the percent of pacing is important for most patients. Recent trials have identified several issues that result from right ventricular (RV) pacing, such as increased atrial fibrillation burden and increased mortality in patients with ICDs. Early identification of increased RV pacing can lead to early intervention and prevention of these problems (Sweeney et al. 2003; Sharma et al. 2005).

A sudden decrease in atrial pacing can be seen in several different situations. It may mean that a patient has stopped or decreased a rate-controlling medication. It may mean a patient has become more active and the atrial rate is now overriding the lower rate limit. Or it can be seen in sudden onset atrial fibrillation. Further investigation of a sudden change in percent pacing is indicated to determine if a true problem exists.

Lead Performance Data

- Impedance trends
- Sensing trends
- Capture trends

Leads are a vital component of the system. They are also vulnerable to injury and failure. Lead technology has evolved rapidly in the last few years, resulting in thinner, more flexible
leads. New materials have increased durability and reliability. The leads, however, remain prone to failure.

Lead dislodgement is a commonly seen lead issue. While more common soon after implant, late lead dislodgement of up to several years is not unheard of. Fractures and insulation failures and damage by external factors, such as a blow to the lead, are other manifestations of lead failure. All of these issues can be evaluated by analyzing the lead performance data.

Review the lead data for significant changes. Minor variations in all of these values are normal. Even one abnormal finding may not be significant, but it should be brought to the attention of the implanting physician and the patient’s chart should be flagged so other clinicians who check the patient will be aware that a problem may exist. A more intense follow-up is often indicated to confirm or disprove a suspected lead problem.

Also note the trends that are available. These indicate changes in lead diagnostics over time and can signal a pending problem. Be sure to print all the lead data for documentation.

**Heart Rate Data**

- Histograms
- Heart rate trending
- Activity trends

Information of the patient’s heart rate is helpful in troubleshooting patients’ complaints. Stacked histograms can be indicative of chronotropic incompetence. Be careful, however, not to assume that the problem is chronotropic. Very inactive patients, such as those who are wheelchair-bound, may not exhibit significant rate changes. The trick is to distinguish whether the patient is inactive because of poor chronotropic response, or whether the histograms are stacked because the patient is inactive. Here again, careful history taking can help the clinician appropriately program the patient’s device.

Heart rate trending data can often help sort out what is happening with a patient’s heart since the trends can be programmed to acquire data during a given time period. They are especially helpful in programming and fine-tuning rate response as they allow the clinician to see the results of programming changes immediately after the changes are made.

Activity trends allow the clinician to look at how active a patient has been over a period of time. These trends vary from device to device, but the data is essentially the same. When put together with the rest of the heart rate data, this can give a more complete picture of the patient’s heart rate response, as well as some indication of the patient’s overall health. Figure 3.6.5 illustrates a typical patient activity trend. The arrow shows where the patient underwent echo optimization of the biventricular pacemaker. Note

![Figure 3.6.5](image-url)  
**Figure 3.6.5** An example of a patient activity graph. Note the sharp upturn in activity hours per day following echo optimization (indicated by the arrow).
the significant increase in the patient’s hours of activity following the optimization. The patient reported feeling much more energetic, and the activity trend provides objective confirmation of what the patient says.

**Heart Failure Data**
- Weight and blood pressure data (in devices where this is available)
- Transthoracic impedance data (in devices where this is available)
- Heart rate variability data

Devices today are capable of collecting data that can assist heart failure specialists in caring for the patient. It is important not only to gather and interpret the information, but, in addition, the information and its interpretation also need to be shared with the appropriate heart failure clinicians. Some of the heart failure data available from today’s devices are blood pressure and weight trending data, transthoracic impedance data that can signal fluid overload, heart rate variability data, and a questionnaire that asks patients to evaluate their heart failure symptoms. Figure 3.6.6 shows one type of heart rate variability data that is based on transthoracic impedance data.

Many of the devices that are equipped for wireless remote follow-up will send automatic alerts to the clinic if heart failure data is out of a programmed range. The intent of these alerts is to signal clinicians before the patient has an acute exacerbation of their heart failure, so intervention can take place in a timely manner and, hopefully, reduce the number of patient admissions for heart failure. Figure 3.6.7 shows an example of alerts received by the clinic for out-of-range weights. Clinic policy should dictate whether these alerts are sent to the heart failure clinic or to the device clinic. Having a written policy saves time getting information to the appropriate provider.

**Arrhythmia Data**
- Number and type of arrhythmias
- Atrial fibrillation burden
- Other supraventricular arrhythmia burden
Interventions for abrupt bradycardic episodes

- Ventricular arrhythmia burden
- For ICDs: number of episodes where therapies were delivered and what therapies were delivered

Devices record all types of arrhythmia data. This information is useful to clinicians in providing timely care. It provides an opportunity to intervene with patients before catastrophic events, such as cardiovascular accidents (CVAs) from silent atrial fibrillation, happen. It is helpful in determining which, if any, of a patient’s symptoms can be attributed to arrhythmic causes. And it can record ventricular arrhythmias that may be harbingers of myocardial ischemia.

Arrhythmia data can be found in several places in the device interrogation. The arrhythmia logs are easily accessible and list all the arrhythmias the device has recorded since the last interrogation. These may be called different things by different manufacturers, but their function is the same. Figure 3.6.8 shows a typical arrhythmia log. Care must be taken in evaluating these episodes. Whenever possible, they should be confirmed by examining the accompanying electrograms (EGMs) to determine whether the device correctly diagnosed the arrhythmia.

Episode length should be noted and can be found in both the episode lists, and in several available graphs, again depending on the manufacturer. Reporting the length of episode is helpful to the attending physician in determining a course of action to be taken for each arrhythmia. For instance, an atrial fibrillation episode of 15 seconds may not warrant consideration of anticoagulation therapy, while an episode of more than 48 hours would certainly warrant that consideration. Note that in some devices, if the mode is DDIR, episode length will not be recorded. Check the manufacturer’s device manual to confirm what, if any, diagnostic information will not be collected in different modes.

Episodes indicating a high atrial rate should be evaluated to determine if they are atrial fibrillation, atrial flutter, or some other supraventricular tachycardia. New onset atrial fibrillation needs to be reported to the patient’s cardiologist and electrophysiologist, and the patient needs to be evaluated for stroke risk and possible anticoagulation therapy. Evaluate the ventricular response to the atrial arrhythmia. Prolonged rapid ventricular response to atrial fibrillation or atrial flutter can predispose a patient to rate-related cardiomyopathies.

For other supraventricular arrhythmias, it is important to note them and what the likely origin of the arrhythmia is. If the patient is not symptomatic, simply noting the arrhythmia is usually enough. If the patient has symptoms that correlate to the arrhythmias, the patient’s cardiologist and electrophysiologist should be notified.

For patients who have therapies programmed to prevent syncope or near syncope for sudden drops in heart rate, note all the episodes...
recorded in the event logs. In many devices, there are no EGMs recorded for these therapies, so it is particularly important to correlate the recorded episodes to patient symptoms in order to document therapy success. If patients are continuing to have syncope or near syncope despite these therapies, device reprogramming may be indicated.

Ventricular tachycardia episodes should be immediately reported to both the patient’s cardiologist and the electrophysiologist. As noted above, whenever possible, these events should be confirmed by EGM. Not being able to view an EGM, however, should not prevent the clinician from notifying the appropriate physician. If possible, patients should be asked if they had symptoms during these episodes.

For patients being monitored remotely, this usually means a phone call to the patient or their family. New ventricular tachycardia in a patient with a pacemaker should prompt evaluation for sudden cardiac death (SCD) risk. Ventricular tachycardia observed in a pacemaker and accompanied by syncope or near syncope can be an indication for immediate hospitalization.

Even in patients who have the protection of an ICD, ventricular tachycardia should not be ignored. Sudden appearance of ventricular tachycardia in a primary prevention patient who has previously had no arrhythmia should be investigated. It may be an indication of ischemia, electrolyte imbalance, or exacerbation of another medical condition.
Chapter 3.6 Pacemaker and ICD Follow-Up

Print and Review Available EGMs

- EGMs recorded automatically from device
- Patient-activated EGM recordings

As noted above, all available EGMs should be reviewed by the clinician. Pay particular attention to whether or not the device has correctly identified the arrhythmia. Some devices record only marker channels. While these are helpful in determining arrhythmia origin, they have some limitations. Figure 3.6.9 shows an episode the device labeled as a high ventricular rate episode. Reviewing the EGM shows that this is likely a supraventricular tachycardia (SVT) incorrectly labeled by the device because the atrial impulses are falling in blanking. If the arrhythmia has been incorrectly identified by the device, make certain that the correct identification is noted in your report.

Review all the episodes recorded from ICDs. Determine if the device appropriately identified the arrhythmia. Note whether therapy was delivered and if it was successful. If it was not successful, note if there was any effect on the arrhythmia from the therapy; that is, did the therapy slow the arrhythmia, did the arrhythmia accelerate as a result of the therapy. Such observations are essential to establishing successful therapy for future episodes.

Note any appearance of noise or “chatter” on stored EGMs as well as on the real-time EGM. Ask questions to determine if the patient has been exposed to any outside electrical sources. This will require a phone call if the interrogation was remote. If the patient is in the office, have him or her perform isometric maneuvers to determine if the noise can be reproduced. Figure 3.6.10 shows the EGM of a 90-year-old patient who downloaded remotely after receiving a shock. Note the noise across both sensing and shocking EGMs, indicating that the shock was likely for electrical magnetic interference (EMI). A phone call to the patient...

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**Figure 3.6.9** Example of incorrectly labeled stored EGM. Note that the episode starts with a premature atrial contraction (PAC) and is likely SVT, rather than VT. The device is “fooled” because the P waves are falling in blanking. While the device cannot “see” these P waves, they are easily discernable to the clinician.
revealed that he had been standing in a pond at his remote gold claim, fixing a pump when he received the shock.

Many pacemakers can be programmed so that the patient can activate an EGM recording by placing a magnet over the device. This can be particularly helpful in correlating patient symptoms to arrhythmias. Note, however, that when this feature is programmed on, the device will not respond to a magnet by reverting to a nonsensing mode. A patient with this feature turned on cannot use the magnet to “blind” the device during procedures such as laser surgery and will need to have the device reprogrammed.

**Device Testing**

During an office visit, the device function and lead integrity must be tested. Lead impedances, sensing, and capture threshold testing are the three tests that should be performed with every
in-clinic visit. These tests are not possible during remote follow-up since testing requires making temporary changes to device programming. Some more modern devices, however, perform some or all of the testing when the device is interrogated, and that data will be available in the remote report. The order of testing is a matter of personal preference. Sensing threshold is often performed first to determine if the patient is pacemaker dependent prior to threshold testing. Although many new devices have algorithms that test impedance, sensing, and capture thresholds on a regular basis, in-clinic testing should still be performed as a routine part of device evaluation whenever the patient is in the clinic.

**Impedance Testing**

Lead impedances are useful in determining the integrity of the leads. Generally, normal lead impedances range from 400 to 1,000 ohms. Some leads on the market are high-impedance leads, and their normal range is over 1,000 ohms. Knowing what lead a patient has helps determine if the lead’s impedance are in range. Lead trends are also helpful in determining lead function. A gradual change in lead impedance may not be noticeable when looking at a single value, particularly if that value is still within the lead’s expected range, whereas the problem becomes glaringly obvious when looking at the trend. Suspected lead problems will require radiological examination to confirm or refute the suspected diagnosis.

Impedance that is climbing raises the concern that a fracture may be developing on the lead. An impedance that is falling may be indicative of insulation failure. Evaluating impedances at every patient encounter allows the clinician to identify problems early.

During lead impedance testing, the device sends a pacing pulse down the lead in order to measure the impedance on that lead. In some devices, this test is performed separately on each lead; in other devices, one button testing is available (pushing the lead impedance test button will send a pacing pulse down all leads in succession). Note any out-of-range numbers.

For many ICDs, the shock impedance is also tested by sending a pacing pulse down the lead, called a painless lead impedance. Some devices, however, deliver a low-voltage shock. These devices should not have shock impedances performed during clinic checks as it is quite painful for the patient. Know how the impedance check is performed on every ICD before attempting to test it.

If a very low impedance is noted, the lead can be changed to unipolar and retested. A lead should never be changed from unipolar to bipolar unless the clinician is absolutely certain that the lead is capable of functioning in a bipolar mode. A unipolar lead that is programmed to bipolar will not be able to complete the electrical circuit, and asystole can occur if the patient is dependent.

**Sensing Thresholds**

On most new devices, sensitivity data is reported as P and R wave measurements. Some devices will test both with the push of one button; other devices require that each lead be tested separately. In each case, the patient’s device is temporarily reprogrammed to allow intrinsic atrial or ventricular activity to be observed. In some older devices, the permanent programming of the device is necessary in order to bring out P or R waves. These are the cases where printouts of the device settings at the beginning are essential. The clinician can compare the final settings with the initial ones to ensure that no unintentional permanent programming has occurred.

Many clinicians test sensitivity before testing capture threshold because it allows them to assess the patient’s underlying rhythm. If no R waves are seen, there is a higher likelihood that
the patient will be symptomatic if ventricular capture is lost for more than a couple of beats. This is a signal to the clinician to be more alert and end capture testing as soon as one beat of lost capture is noticed.

Noting the sensitivity threshold during office testing is only one part of evaluating the lead. The trending data that is available in most new devices should be evaluated just as impedance trends were evaluated. Even if the sensitivity measurement in the office is in range, reviewing trending data can alert the clinician that there is an impending lead problem.

Sensitivity data is vitally important in the ICD. Low-amplitude P waves can impact the ICD’s ability to determine whether a fast heart rate is ventricular or supraventricular in origin and can lead to inappropriate shocks. Low-amplitude R waves can result in delayed identification of ventricular arrhythmias and delay appropriate therapy. In general, R waves lower than 5 mV should be reported to the electrophysiologist. Adjusting sensitivity on an ICD is best done in the electrophysiology (EP) lab where noninvasive programmed stimulation (NIPS) can be performed to ensure that the device can still sense ventricular arrhythmias after the changes are made.

Capture Threshold Testing

Capture threshold testing should be performed at every office visit. Significant changes in capture threshold signal that a lead problem may be on the horizon. A sudden climb in capture threshold can herald a lead dislodgement. Gradual increases in capture threshold may indicate a microdislodgement or exit block. As noted above, monitoring the lead trends allows the clinician to observe the behavior of the lead over time, so that even if the capture threshold is still in a normal range, a gradual increase or decrease stands out in the trend. Unlike impedance or sensitivity testing where some devices allow one-button testing of all leads, during capture threshold testing, each lead must be evaluated separately.

One exception is some older biventricular pacemakers. In these devices, the left ventricular lead is connected by Y connector to the device. To test capture threshold on these devices, amplitude or pulse width are decremented as usual. The rhythm strip is observed for morphology changes as first one of the ventricular leads and then the other loses capture. Usually, it is the left ventricular lead that loses capture first, but the RV lead can be the one that has the higher threshold.

During capture threshold testing, the patient should be placed in a recumbent or semirecumbent position. This will help limit symptoms that can occur during the process of testing the lead. Some patients will be extremely sensitive to loss of capture, especially in the ventricular lead. Others will barely notice. Another group of patients will develop severe pacemaker syndrome during ventricular lead testing, especially if loss of atrioventricular (AV) synchrony occurs. Electrocardiogram (ECG) cables should be used to observe loss of capture. These can be either the cables provided with the device, an ECG machine, or one of the commercially available device programs. Relying on the EGM to determine capture may be quicker since placing the ECG is time consuming, but vital information may be missed. Figure 3.6.11 illustrates this concept. Looking only at the marker and atrial EGM, one might be tempted to conclude that the patient is having bigeminal premature ventricular contractions (PVCs). By combining the information from both the EGM and the ECG, it becomes clear that the ventricular sensing is actually oversensing of the T waves. This is causing the next atrial sensed event, coming in right on time to fall in the device’s refractory period and not be tracked by the device.

It is helpful to note where the last clinician observed capture on each lead. While this should be only considered a guide, it does help point out a starting place. Capture is reported
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as two parameters: voltage and time; that is, 2V at 0.4 ms. Some clinicians prefer to hold voltage stable and decrement milliseconds (pulse width). Others prefer to hold milliseconds stable and decrement voltage. Note how previous clinicians have performed the test and keep it consistent. It is much easier to compare the parameters if the test is performed the same way each time.

The capture threshold of a lead is the lowest voltage at which the impulse from the device produces consistent depolarization. It is not the voltage at which capture is lost. Thus, if capture is lost at 1 V at 0.1 ms, the capture threshold is 1 V at 0.2 ms (assuming pulse width is being decremented). This is an important distinction to make when determining the appropriate setting for permanent programming of the lead outputs. Lead outputs should be programmed to two times the voltage or three times the pulse width to provide an adequate safety margin.

A final observation to make during atrial capture threshold testing is the absence or presence of retrograde atrial conduction. This is a vital piece of information in determining the patient’s risk for pacemaker-mediated tachycardia (PMT). When atrial capture threshold testing is done in DDD mode, loss of atrial capture results in RV capture after the atrial pace. Because there was no atrial depolarization, thus no conduction down the AV node. Because the device delivered an atrial pulse, it will deliver a ventricular pulse.

Ventricular capture is generally very easy to see on the ECG. Atrial capture can be much more problematic, as P waves are relatively small and hard to see. Using a rhythm strip in addition to the EGM greatly enhances the chance of seeing the actual loss of capture. If it is still difficult to determine, a 12-lead ECG can be used to see if P waves are more prominent in one particular lead. If all else fails, echocardiogram can be performed to determine if there is atrial capture.

If the patient has intact AV conduction, test the patient in DDD mode with a long AV delay. This will allow the patient to conduct normally down the AV node following atrial pacing. When atrial capture is lost, there will be no atrial depolarization, thus no conduction down the AV node. Because the device delivered an atrial pulse, it will deliver a ventricular pulse.

Figure 3.6.11 Marker channels on this EGM seem to indicate loss of capture, but correlating this to the surface ECG shows clear T wave oversensing when the patient is paced in the RV.
at the programmed time, resulting in a paced ventricular complex. This paced ventricular complex is where loss of atrial capture occurred. Figure 3.6.12 illustrates this concept. Note that no P waves are visible in the rhythm strip, but a small QRS corresponds with the ventricular sensed (VS) marker. As the voltage is decremented, a ventricular paced (VP) marker is noted, corresponding to a clear ventricular paced complex in the rhythm strip. This is where atrial capture is lost. On this strip, loss of atrial capture can also be confirmed by the appearance of atrial sensed signals on the atrial EGM.

Patients with intact AV conduction can also be tested in AAI mode. The clinician should be absolutely certain that the patient will conduct intrinsically at high rates; otherwise, ventricular asystole could result. When testing in AAI mode, look for loss of AV conduction when atrial capture is lost, resulting in no intrinsic R wave.

It is uncommon to have difficulty determining ventricular capture. However, with many patients now being programmed with long AV delays to promote intrinsic conduction down the AV node, fusion, pseudofusion, and functional noncapture can confuse the issue. Shortening the AV delay to 120ms or less will decrease the possibility of confusing these phenomena for loss of capture. Whenever possible, ventricular threshold testing should be performed in the DDD mode to prevent loss of AV synchrony, which can be very uncomfortable for the patient. In single-chamber devices (or biventricular devices that do not have atrial pacing), be alert for functional noncapture in the nondependent patient.

**Ending the Interrogation**

Before ending the interrogation, *print a final report!* This provides documentation that no parameters have been unintentionally reprogrammed. Compare the final report with the initial report before the patient leaves. If parameters have been unintentionally changed, they can be changed back without having to call the patient back. Some devices provide a print option to print any changes made during the session. This is an easy and quick way to check what has been changed. If that option is not available, note that many device printouts show a hash mark (< or <=) next to a
parameter that has been changed. Review these to be sure that all changes are intentional. One note on final printouts: On some devices, the “final report” does not print all parameters. Always check to be sure that all parameters have been printed, and if necessary, print them separately.

For remote checks, printing the entire check can be optional. The entire device check is on the Web site and available each time the site is accessed. Since it is not possible at the present time to change parameters during a remote encounter, the parameter will not change. Printing diagnostic information and EGMs has more value, as this information will change from check to check. Having a hard copy means it will always be available, even if the Web site or Internet is down.

Some commercially available documentation and tracking systems allow device information to be transferred from the programmer to the documentation system. For some, this is a direct link, and for others, it can be by floppy disk or thumb drive. Some commercial systems also allow remote downloads to be loaded directly in. For in-clinic checks where the data will be saved, be sure it is done prior to clearing the diagnostic data.

The last step before ending the session in a clinic check is to clear the diagnostic data. Some devices will do this automatically within a programmed time frame after ending the session. In other devices, this must be done manually. Clearing the data ensures that the next time the patient is seen, the data will be specific to the time frame in between checks. This is especially important if parameter changes are made as it provides objective data on heart rate, activity level, arrhythmia burden, and so on.

Concluding the Patient Encounter

Once all the data has been gathered and interpreted, the clinician must act on it. In some cases, no specific action other than routine follow-up will be needed. In other cases, drastic action such as immediate hospital admission will be called for. For a majority of follow-ups, minor programming changes will solve any problems identified. These can range from a minor change to the rate response algorithm, to mode and timing changes. Programming changes made to a device should be reviewed by the following physician.

Following remote interrogation, patients will need to be notified of results. Some clinics call every patient; others send out preprinted cards indicating that the download was received, the device is functioning properly, and when the next download will be. When device or lead problems are encountered during a remote follow-up, the patient should be contacted and arrangements made to have the patient come in to the clinic for in-depth follow-up.

In some cases, patients will need referral to an appropriate provider. Who the appropriate provider is will vary from office to office. Having a referral plan in place for such things as new onset atrial fibrillation, ventricular tachycardia on a pacemaker, or abnormal heart failure findings will make the process easier.

Any action taken must be evaluated to determine the outcome of the action. This may mean having the patient follow-up more frequently until the problem is resolved. Sometimes a change can be evaluated immediately in the office, such as when adjustments are made to the rate response algorithm and the patient has obviously improved exercise tolerance on a hall walk. At other times, this will require that the patient have several follow-up visits within a few days or weeks.

Documentation

Finally, each piece of information collected and every action taken must be documented. Personal preference will again dictate whether a clinic uses a commercially available documentation system or one the clinicians developed themselves. Currently, most device printouts
are kept in the patients’ device charts. This may change, as some device manufacturers are exploring ways to have device information from both clinic checks and remote checks transferred directly into patients’ electronic medical records (EMRs). The clinician could then review and interpret the data and write up the patient encounter directly into the EMRs.

The importance of appropriate, timely documentation cannot be overemphasized. Documentation serves several purposes beyond the obvious legal one. Well-documented follow-up visits provide a road map of what works and what does not, helping streamline future visits. Good documentation also records important information about patients (such as a patient who is extremely symptomatic to some portion of the device check). Appropriately documented visits ensure that the next clinician to check the patient’s device will have access to all the information the last clinician had and does not have to discover it all for him or herself.

REFERENCES


RESOURCES


Family Matters: Research and Clinical Management of Psychosocial Issues for ICD Patients and Their Partners

Samuel F. Sears, Kari Kirian, Melissa Matchett, Christie Benton, and Rajasekhar Nekkanti

The success of the implantable cardioverter defibrillator (ICD) in several large-scale randomized trials has established the ICD as the treatment of choice in detecting and treating ventricular arrhythmias (AVID Investigators 1997; Moss et al. 2002; Bardy et al. 2005). Patient-reported outcomes encompassing psychosocial and quality of life (QOL) outcomes have generally been positive and desirable. Nonetheless, living successfully with the risk of spontaneous, potentially life-threatening arrhythmias and the resulting, life-saving high-energy shocks presents challenges to a sizable minority of patients and their families. ICDs save lives in at-risk patients, but the ongoing need for comprehensive, biopsychosocial care plans addressing the range of issues for both patients and families has been a challenge in electrophysiology clinics. Researchers have been stimulated by this modern-day scenario to describe, predict, and treat the potential for general psychological distress (i.e., anxiety and depression) and avoidance behaviors in ICD patients. More recent research has added the perspective of ICD family members, such as partners/spouses/significant others, to better understand the collective adjustment challenges. Preliminary findings indicate that this may be particularly needed as ICD spouses experience significant distress as well. The purpose of this chapter is to review the overall psychosocial and QOL impact of ICDs on patients and their partners. Clinical management strategies and approaches will also be discussed.
PSYCHOSOCIAL IMPACT FOR ICD PATIENTS: COMMON PROBLEMS IN ICD PATIENTS

The success of ICDs has prompted the conversion of a previously acute and terminal condition, such as cardiac arrest, into a chronic and long-term illness managed with technology (Matchett et al. 2009). The realization of the permanence of the cardiac condition and the ICD can be difficult for ICD patients, as they are forced to adjust to a disease state and to the presence of the device. The provision of ICD shocks are the most unique feature that distinguishes ICD patients from other patients coping with heart disease. Patients have described the experience of shock as a swift kick in the chest and have rated it as a “6” on a 0–10 pain scale (Ahmad et al. 2000; Pelletier et al., 2002). Alongside the physical effects of shock, shocks are unexpected and uncontrollable and can occur without perceived symptoms. This may cause a patient to feel vulnerable, frightened, and powerless. Furthermore, as with other cardiac devices, the presence of the ICD may be a constant reminder of one’s own mortality. Finally, the potential to be shocked in a social setting, garnering unwanted attention, may be frightening and embarrassing. While most patients appear to adjust well, patients who experience five or more shocks are more likely to have psychological issues (Namerow et al. 1999; Connolly et al. 2000; Irvine et al. 2002). Below, we review many of the common psychological issues including anxiety, depression, avoidance behaviors, and decrements in QOL.

General Anxiety

Anxiety is the most considerable psychosocial challenge among ICD patients. Anxiety disorders are experienced by roughly 13–38% of ICD patients (Sears et al. 1999b; Sears and Conti 2002). General or ICD-specific anxiety is experienced by approximately 24–87% of ICD patients after implantation (Sears and Conti 2002; Burke et al., 2003; Bilge et al. 2006). Patients with ICDs may be more acutely aware of the beating of their heart and their own mortality, which may contribute to feelings of anxiety. Early research relied heavily on traditional measures of anxiety as used in psychiatric settings. More recently, researchers have progressed to more device-specific adjustment features such as shock anxiety or posttraumatic stress disorder (PTSD) to better explain the device-specific adjustment characteristics.

Shock Anxiety

Separate from general anxiety, some patients further develop anxieties specifically around receiving a shock. Shock anxiety is the fear or anticipation of ICD shock that often results in increased heart-focused anxiety symptoms, as well as the development and maintenance of avoidance behaviors to minimize their perceived risk of shock (Kuhl et al. 2006; Sears et al. 2008b). Never knowing when to expect a shock can result in hyperarousal in ICD patients. More clearly, patients do not know when to expect a shock, so they continuously expect a shock in an attempt to be “prepared.” Their guard is heightened at all times and results in an ongoing, increased level of anxiety regarding the potential specific event of shock. In fact, up to 40–60% of ICD patients may be plagued by shock anxiety (Pauli et al. 1999; Crössman et al. 2007). Further, a significant percentage of ICD patients, 10.3–38.5%, will be confronted with the experience of shock in the first year alone (Connolly et al. 2006).

Psychologists have attempted to provide quantitative tools to assess shock anxiety. The Florida Shock Anxiety Survey (FSAS) has 10 items that can be combined into a two-factor structure, including a Consequence Factor (e.g., fearing creating a scene if the device were to fire) and a Trigger Factor (e.g., fearing sexual activity) (Kuhl et al. 2006). Higher scores on the FSAS reflect unique patient anxieties about the
ability to cope with the impacts of shock, as opposed to device acceptance.

**PTSD**

PTSD is an anxiety disorder that is characterized by avoidance, hyperarousal symptoms, and intrusive thoughts of reexperiencing following a traumatic experience (American Psychiatric Association 2000). It has been studied in other cardiac populations, such as postmyocardial infarction patients, and determined that PTSD symptoms are comorbid in 10–32% of these cardiovascular patients (Bennett et al. 2001; Ginzburg et al. 2002, 2003; Shemesh et al. 2006; Jones et al. 2007; Chung et al. 2008; Wiedemar et al. 2008). Clearly, the ICD patient population could be considered an at-risk population for PTSD related to the perception and possible reality of regular life-threat due to arrhythmias and exposure to high-energy shocks. Both the ICD discharge and the underlying heart disease may meet the Diagnostic and Statistical Manual-IV (DSM-IV) criteria for a life-threatening and traumatic event (American Psychiatric Association 2000). Ladwig et al. (2008) describe the development of PTSD in ICD patients as stemming from the “persistent and enduring” threat to their life and safety (p. 1328). The result of which is a sequela of symptoms including worry and fear regarding treatment and survival. The notable difference with PTSD symptoms in ICD patients, as compared with the more traditional patient with PTSD, is that their fears are rooted in the future, rather than the past (Ladwig et al. 2008).

PTSD has recently acquired some attention in the ICD community. A groundbreaking study conducted by Ladwig et al. (2008) established the potential utility of closely examining PTSD symptoms on prognosis in patients with ICDs. After controlling for depression and anxiety, researchers found that PTSD symptoms significantly affected the mortality risk in ICD patients, such that the relative mortality risk in the PTSD group was 3.45 ($P = 0.002$). This translates to a 77.5% chance that patients with PTSD will die before patients without PTSD. Symptoms of PTSD have a clear negative outcome on survival, and greater research and clinical attention to this phenomenon is indicated.

**Depression**

Depressive symptomatology is reported in 24–33% of ICD patients (Sears et al. 1999b). Bilge et al. (2006) reported an even higher prevalence of depression, with 41% of ICD patients having exhibited depression in a Turkish sample. Multiple theories posit reasons for the development of depression in the ICD patient population. At the forefront of these theories is the behavioral model of learned helplessness. The etiological premise of learned helplessness lies in one’s inability to avoid the aversive stimulus of shock, which may result in feelings of helplessness and the feeling that one cannot control or prevent what happens to them. This is based on landmark studies in animals where animals made to repeatedly endure unpredictable, unpleasant, and unavoidable stimuli (such as shock) eventually display behaviors similar to those evidenced in clinical depression. They have learned that what happens to them is independent of their behavior (Seligman and Maier 1967). This model, applied to humans, results in patients perceiving an absence of control over one’s medical condition and the associated shocks, which leads to a state of learned helplessness that may increase the chances that ICD patients feel vulnerable to depression (Hamner et al. 1999; Sears et al. 1999a; Matchett et al. 2008). Interestingly, results from the Triggers of Ventricular Arrhythmias (TOVA) study indicated the reverse may also be true, whereby risk of shock was associated with depression (Whang et al. 2005).

**Behavioral Avoidance**

In an effort to assert control, some ICD patients may come to avoid certain activities in an
attempt to avoid shock. This marriage of the physiological and psychological experience of shock is clearer when investigated using a theoretical model. Classical conditioning and operant conditioning, among others, have been suggested as explanations of behavior after shock (Sears et al. 1999a). Classical conditioning describes how a previously unconditioned stimulus paired with a neutral stimulus results in a conditioned response to the neutral stimulus. Applied to ICD patients, this may explain why patients come to fear and avoid behaviors they were engaging in prior to shock. The behavior becomes associated with shock, whether it is riding a bicycle or watching television. The result of this conditioned fear is a decrease in the likelihood of engaging in that specific behavior, including activities that patients once enjoyed (Sears et al. 2008a).

Operant conditioning may aid in explaining the maintenance of behavioral avoidance over time by the employment of consequences (or lack thereof) to modify the occurrence of a behavior. Negative reinforcement, or the reward for avoiding an aversive consequence, increases the likelihood that the behavior will be performed again (Godemann et al. 2001). For example, if an ICD patient is not shocked while watching television or engaging in other sedentary activities, he or she may be more likely to engage in those behaviors because he or she believes that these activities are “safe” and the risk of shock is lessened. Approximately 55% of ICD patients regularly avoid people, places, or activities in an attempt to avoid shock (Lemon et al. 2004). Those patients who avoid behaviors that they associate with shock may eventually come to avoid many daily activities to the point in which they are severely limited in their life, and hence, the quality of their life is compromised in an attempt to avoid shock. Whether patients avoid activities that may increase their heart rate or avoid people and places out of fear of embarrassment or helplessness in the event of a shock event, these reasons may inhibit patients from living their life (Matchett et al. 2008). The primary limitation of the major theories to explain distress and shock is that each of the theories is silent on the development of distress in ICD patients who have not been shocked. Clearly, shock is not necessary for distress in ICD patients but this specific scenario points out the limitations of solely implicating shock as the precipitant to distress in all cases.

QOL

QOL has averted a singular definition in the health research literature but most definitions include aspects of a multidimensional health outcome in which biological, psychological, and social functioning are interdependent. QOL indices generally measure the subjective degree of overall well-being that a person experiences. QOL is also influential in quantity of life, as higher QOL in ICD patients has been associated with longer survival (Steinberg et al. 2008).

Health outcome researchers examining ICDs in the context of large clinical trials have generally compared generic QOL measures, such as the Short Form 36 Health Survey (SF-36) subscales, between randomly assigned groups of ICD versus medication patients (Irvine et al. 2002; Schron et al. 2002; Mark et al. 2008). These results have generally indicated that ICD patient-reported QOL is at least equal to, or better than, antiarrhythmic medications on most indicators of QOL (Sears and Conti 2002, Francis et al. 2006). Clearly, a fundamental implication of this conclusion across researchers is that patients opting for an ICD can receive the added protection of the ICD without necessarily compromising QOL. However, the possibility of an ICD shock may change that clinical picture substantially. The occurrence of shock is the clinical “attention stealer” and has generally been implicated in adverse QOL. The actual impact of shock is somewhat more complicated than the simple mantra, “shock is bad.” The evaluation of shock depends in part on the frequency versus the occurrence of
shock. Multiple large-scale trials have recently concluded that ICD patients are able to tolerate one to five shocks with statistically but not meaningful differences in QOL (Passman et al. 2007). However, when shocks amount to five or more, QOL research can show both statistical and clinically significant changes in QOL. In addition, inappropriate shocks (shocks administered for nonlife-threatening arrhythmias) must also be considered in regard to psychological indices of adjustment. The stimulus of shock is identical but the interpretation by the patient is markedly different. Inappropriate shocks can affect how patients perceive the efficacy, value, and safety and warrant clear communication about why they occur and what efforts have been done to prevent them in the future.

SOCIAL SUPPORT AND THE PSYCHOSOCIAL IMPACT OF THE ICD ON PARTNERS: MORE ANXIETY THAN PATIENTS?

Social support plays a key role in the health, QOL, and adjustment of individuals with and without health conditions. Social support is a multidimensional construct that broadly includes the supportive ways that individuals behave in a social network (Helgeson 2003). Quantitatively, social support refers to the mere existence of social relationships, while qualitatively, it refers to emotional, instrumental (tangible assistance), and informational support (Helgeson 2003). Early research demonstrated that it is perceived, rather than actual, social support that yields positive effects on variables of well-being and health. Social support has demonstrated remarkable effects on recovery and QOL in a variety of cardiac patients possibly as a buffer to the effects of stress on cardiovascular functioning (Kamarck et al. 1990; Gerin et al. 1992; Fontana et al. 1999), while low perceived social support is a documented predictor of adverse cardiovascular events, such as heart failure-related readmissions (Welin et al. 2000; Brummett et al. 2005; Tsuchihashi-Makaya et al. 2009). Living alone is related to a twofold risk of recurrent events after myocardial infarction (qualitative study), whereas a plethora of research shows that social support after an infarction is associated with improved QOL (Wingate 1995; Motzer and Stewart 1996; Woloshin et al. 1997; Rankin and Fukuoka 2003). Finally, in a systemic review, Mookadam and Arthur (2004) demonstrated that social isolation is associated with increased mortality and morbidity in cardiac patients, with an odds ratio of 2:3.

Social support research has been extended to patients with ICDs and similar significant trends indicating the importance of spouses. A recent study demonstrated that in the rest period after a mental stress had been endured, ICD patients (82%) who perceived better social support experienced a rapid decrease in cardiac index, heart rate, and arterial blood pressure, as well as a rise in high-frequency heart rate variability (Lache et al. 2007). These results indicate that social support independently predicted better hemodynamic recovery from mental stress in the ICD patient population. By contrast, those patients who lacked poststress hemodynamic recovery reported significantly less perceived social support than patients who reacted normally. The need for social support may prompt ICD patients to engage in support groups. Myers and James (2008) demonstrated that compared with nonattendees of ICD support groups, attendees had significantly more trait anxiety and less satisfaction with their social support, indicating that support groups may serve a vital role for those who lack independent or other resources for recovery.

Research focused on the impact of being an ICD spouse or partner has further highlighted a potentially more difficult adjustment than patient themselves experience. Table 3.7.1 shows the common psychological effects that ICDs have on partners. Across studies,
Table 3.7.1 Common psychological effects of ICD on partners.

<table>
<thead>
<tr>
<th>Authors</th>
<th>N</th>
<th>Psychological end points</th>
<th>Measure</th>
<th>Conclusion</th>
</tr>
</thead>
</table>
| DeGroot et al.   | 284 Patients; n=150 partners; n=134 | Health concern, $P = 0.031$; anxiety, 36%; depression, 24%; nervousness, 66%; need for counseling or support groups, 60% | • Specific ICD-related questionnaires  
• Spielberger State-Trait Anxiety Inventory  
• Rand-38 Mental Health Inventory (MH) | • Partners reported more nervousness and more concern about the health of the ICD patients than the patients themselves |
| Pedersen et al.  | 326 patients; n=182 partners; n=144 | Anxiety, 42%, $P = 0.048$; depression, 29% | • Hospital Anxiety and Depression Scale (HADS)  
• Type D Personality Scale (DS14) | • Partners experienced similar levels of depression (28% vs. 29%) but higher levels of anxiety compared with ICD patients themselves (42% in partners vs. 31% in patients)  
• No differences were evidenced in Type D personality or perceived social support |
| Sowell et al.    | 62 patients; n=40 partners; n=22 | Shock anxiety, $P=0.05$ | • Florida Shock Anxiety Scale (FSAS) | • Partners of ICD patients experience higher levels of shock anxiety than the patients themselves  
• No differences were evidenced in death anxiety, general anxiety, or marital adjustment |
| Jenkins et al.   | 477 patients; n=353 partners; n=124 | Mental summary issues, $r = 0.29-0.36$, $P < 0.03$; quality of life, $r = 0.26$, $P < 0.03$ | • Short-Form 36 (SF-36)  
• Quality of Life Index | • Partners of ICD patients had similar mental summary issues and quality of life as the patients themselves  
• Physical summary scores worsened in partners over time |
researchers have generally demonstrated that spouses have a unique, and possibly more difficult, adjustment burden. Multiple researchers have shown that partners report greater anxiety and health concerns than the ICD patients themselves report (DeGroot et al. 2003; Pedersen et al. 2004). Alternatively, Sowell et al. (2007) found that ICD partners had similar levels of death anxiety, general anxiety, and marital adjustment, but higher levels of shock anxiety. These results suggest that further precision of the measurement of the anxiety might allow for targeted intervention. The “wear and tear” on ICD partners may also be significant as demonstrated by Jenkins et al. (2007), in which physical summary scores of partners of patients with ICDs worsened over time and were similar to the QOL of ICD patients. Taken together, these results indicate that ICD spouses and partners are clearly affected by their loved one’s treatment and may be affected by even higher levels of anxiety and shock anxiety than the patients themselves. Furthermore, psychological distress experienced by spouses could likely interfere with their abilities to lend adequate social support, thereby inhibiting both patient and spousal adjustment (see Fig. 3.7.1). More research is needed in this area to examine the main effects and interactions of ICD patient and partner adjustment and how to efficiently intervene to improve outcomes.

**CLINICAL MANAGEMENT OF ICD PATIENTS AND PARTNERS TO OPTIMIZE OUTCOMES**

Clinicians must primarily direct their clinical attention toward ICD patients to ensure optimal care and outcomes; however, the previously discussed literature also emphasizes the need to care for patients’ support networks, especially spouses. Providing clinical psychosocial attention to spouses of ICD patients may be especially helpful for two major reasons. First, providing care to spouses may equip them to provide improved social support, which may indirectly have positive effects on the recovery and QOL of ICD patients. Second, spouses are the “hidden patients” (Stanley and Frantz 1988) in the care context that are at risk for their own mental and physical health problems if the demands of care are not adequately managed.

Although the need for spousal clinical care is well documented, the typical health care setting provides barriers to addressing their needs due to limitations in awareness of the need, time to address their need, and the financial incentives to complete the tasks. Health care professionals provide well-needed care for ICD patients, but they may not be accustomed to seeing the patient in the context of his or her...
family, albeit research has demonstrated the ill effects (i.e., anxiety) of ICD implantation on spouses. Even if attuned to the needs of partners/spouses of ICD patients, clinicians may not be afforded the time to address them in the hectic pace of the health care system. Further, insurance companies rarely offer coverage for the provisions of clinical care to spouses. This scenario leaves spouses, often critical members of the support network of ICD patients, in the quagmire of minimal or no psychosocial care.

Assessment and Intervention with ICD Patients

The ICD patient population has changed since the Food and Drug Administration (FDA) approval in 1985. ICD patients were initially a secondary prevention population who experienced cardiac arrest and were happy for a second chance at life. Today, ICDs are used in the primary prevention population as well and these patients are attempting to manage their risk of cardiac arrest with the ICD. These different orientations to the ICD may set up sets of psychosocial needs that can be highly variable over time and potentially challenging to busy electrophysiology clinics. This reality should prompt allied providers to perform regular and serial assessments over time of psychosocial functioning. Despite the fact that most ICD patients will adjust well to their device, health professionals need to be prepared to address psychosocial issues. Multiple barriers exist to this ideality of addressing psychosocial risk including time constraints, comfort level, and expertise of providers discussing psychosocial topics. Addressing issues perceived to be beyond one’s realm of expertise can certainly be intimidating. In fact, health care providers generally report feeling most comfortable handling traditional medical issues and least comfortable in handling issues about emotional well-being (Sears et al. 2000; Sears and Conti 2002).

Nonetheless, systematic review of the ICD patient literature highlights the need for both when caring for ICD patients and partners.

Clinical care that integrates both a brief assessment and intervention that is reasonable given the time constraints of an electrophysiology clinic is the goal. Assessment of psychosocial domains need not be exhaustive nor complex. The assessment of areas of functioning spanning psychosocial risk factors, risk behaviors, behavioral observations, and affective assessment can be explored in only a few minutes. It is important to note that if there is reason to believe that a patient’s partner is experiencing psychosocial difficulties, encouragement to schedule an appointment with a mental health professional should be provided.

Assessment Areas

1. **Psychosocial risk factors:** Does the patient have any known risk factors for psychosocial difficulties? Psychosocial risk factors characteristics have been identified and include young patients (less than 50 years of age), women, shock exposure, poor knowledge of ICD and cardiac conditions, multiple comorbidities, and history of psychological difficulties (Sears et al. 1999b).

2. **Psychosocial risk behaviors:** Does the patient or spouse report any explicit or implicit fears and resulting avoidance behaviors from full functioning? Patients who are avoiding certain behaviors (i.e., sex, exercise, leaving the home) or not engaging in activities they previously enjoyed (QOL) are exhibiting risk behaviors that may indicate poor adjustment. Research has indicated that avoidance behaviors are a common response for ICD patients (Lemon et al. 2004).

3. **Psychosocial behavioral observations:** Does the patient exhibit any affective or behavioral clues that could indicate poor psychosocial functioning? What is the patient’s mood and affect? If family members are present for the
visit, what are they saying about the patient’s mood and daily activities? Does this information contradict what the patient is saying? If so, explore this further.

4. **Critical event experience:** Has the patient experienced an adverse event (e.g., recall) or critical clinical event (e.g., multiple shock episode) that may have changed their outlook? Some patient experiences are universally stressful and warrant acknowledgment. Most ICD patients will cope with adverse experiences but some patients will need referral and additional care to synthesize the experience and still feel safe and secure with their ICD.

5. **Brief psychological assessment:** Can I assess the possible signs of distress directly? Depression and anxiety are the most commonly experienced psychological issues for both patients and partners (Sears and Conti 2002). To screen for depression, ask the patient, “During the past month have you felt … (1) down, hopeless, or depressed or (2) less interested or received less enjoyment from doing things you usually take pleasure in? To screen for general anxiety, ask the patient, “Have you been feeling excessively nervous or anxious lately?” Shock anxiety can be assessed by asking, “Do you have continuous fears of being shocked by your device?” Positive answers to any of the depression and/or screening items may indicate a need for a referral to a mental health professional. Finally, suicidal ideation and intent should be assessed with the inquiry, “Have you had thoughts of hurting yourself or ending your life?” Positive answers to this question indicate an immediate need for a formal mental health assessment.

6. **Cultivate and engage a mental health professional collaborator:** Very few mental health practitioners (e.g., psychiatrists, psychologists, or social workers) will have had specific training in electrophysiology or even cardiology. As a result, good working collaborators may need to be developed by your treatment team. Formal training is less important than ongoing development of information and experience. A sizeable literature exists today that would be relevant to the mental health practitioner and integrated into their own existing skill sets of supportive psychotherapy and behavioral change. Furthermore, patients reporting concerns about possible self-injurious or suicidal behavior should be referred for comprehensive assessment of their functioning.

**Brief Interventional Approaches**

Similar to assessment approaches, brief interventions are better than no intervention. Some patients will need much more intervention than simply having a supportive discussion, but many patients will benefit from both the desire to discuss their well-being and reiteration of reassurance of positive expectations. In Table 3.7.2, we highlight some example of common themes of issues for ICD patients. Specifically, ICD patients and partners often seek additional information about the causes and consequences of ICD shocks, managing their concerns about safety and longevity, and issues related to returning to an array of activities. The key message portion of the table displays some of the information that can be reviewed to help patients understand and reassure their return to a desirable QOL. Although this type of intervention will not resolve more serious psychosocial conflicts, these key messages can open discussion and relieve more mild concerns for the typical ICD patient and partner.

**CONCLUSION**

The success of the ICD in preventing mortality in patients at risk for sudden cardiac arrest has produced a need to help patients successfully manage the psychosocial aspects of the cardiac risk and living with an ICD. Research has indicated that QOL outcomes for ICD patients are
### Table 3.7.2 Common themes of issues for ICD patients.

<table>
<thead>
<tr>
<th>Theme of patient question</th>
<th>Frequently asked questions by patient and partners</th>
<th>Key messages</th>
</tr>
</thead>
<tbody>
<tr>
<td>Safety</td>
<td>Am I safe?</td>
<td>Emphasize return to life because of ICD</td>
</tr>
<tr>
<td>Safety</td>
<td>Effectiveness</td>
<td>Highlight scientific efficacy evidence</td>
</tr>
<tr>
<td>Safety</td>
<td>Problem solving</td>
<td>Reiterate multidisciplinary team approach</td>
</tr>
<tr>
<td>Safety</td>
<td>Recall and device malfunction</td>
<td>The chance of recall or device malfunction is rare (see Kirian et al. 2009).</td>
</tr>
<tr>
<td>Shock 101</td>
<td>What does a shock feel like?</td>
<td>Shocks are very quick and have been rated as a “6” on a 0–10 pain scale; only the patient can feel a shock; most people describe a shock as more discomforting than painful</td>
</tr>
<tr>
<td>Shock 101</td>
<td>How likely am I to get a shock?</td>
<td>It is not possible to predict the need for shock. The ICD is implanted to protect you in case you need shock. The most recent research available suggests that between 10–38.5% of ICD patients will receive a shock during the first year (Connolly et al. 2006).</td>
</tr>
<tr>
<td>Shock 101</td>
<td>Is it possible to trigger a shock?</td>
<td>At this time, it is very unlikely that there is any action that you could take that would cause shock. Preventing the need for shock is a key goal for all of us.</td>
</tr>
<tr>
<td>Shock 101</td>
<td>After shock</td>
<td>Follow our shock plan. If you get one shock, please call us to let us know, but it may not be a cause for alarm. If you get more than one shock or do not feel well after a shock, seek emergency care (see, e.g., Sears et al. 2005).</td>
</tr>
<tr>
<td>Return to life</td>
<td>What are my activity restrictions?</td>
<td>Most activities are safe with ICDs. What are you most interested in returning to at this time?</td>
</tr>
<tr>
<td>Return to life</td>
<td>Is sexual activity safe?</td>
<td>Sex is considered a gentle form of exercise.</td>
</tr>
</tbody>
</table>

at least equal to, if not better than, medications alone. However, the experience or exposure of multiple shocks continues to be a source of concern and a potential subtraction from QOL. ICD patients experience the process of living with an ICD together with their partner. Initial research examining partners has indicated that partners may be at even greater risk of anxiety symptoms that potentially impact their own mental and physical health. Collectively, greater
research and clinical attention to the psychosocial adjustment of ICD patients and partners is indicated and may affect health outcomes directly. Brief communications focused on managing shocks, concerns about safety and security, and returning to various activities are usually quite valuable to ICD patients and partners. The prevalence and potential impact of psychosocial adjustment on the QOL in ICD patients justifies the efforts by allied providers.

Case 3.7.1

Case Presentation

Mr. O. is a 61-year-old widowed African American male who presented in an ICD follow-up clinic after receiving a single ICD shock while riding his bicycle. The patient now reports no desire to return to any level of activity. He reports that he believes that increasing his heart rate to any degree will trigger an ICD shock. For this reason, he describes his recent behavior as extremely sedentary and is limited to basically "staying in." He expects to be shocked on a daily basis and expects that he will not live more than 30 days. He believes that his foreshortened sense of future is a clear conclusion because the ICD shock signals "the beginning of the end." At this time, he presents with concerns about whether or not medical treatment is futile.

Case Discussion

Consistent with our clinical assessment approach in the chapter, Mr. O. presented with a positive psychosocial risk factor (shock experience), positive for a psychosocial risk behavior (avoidance of activity), positive behavioral observations in clinic (blunted affect and anhedonia), a critical event (recent shock experience), and possible signs of depression. Each of the suggested assessment areas were highlighted in this case as well as significant misunderstandings about device and heart function and its impact on prognosis or longevity. Clearly, Mr. O. needs additional clinical attention to address these multiple risk factors for poor adjustment and QOL. A reasonable clinical plan in this case could include:

• Emotional impact acknowledgement and empathy: Mr. O. likely wants to be understood about the emotional impact and the feeling of loss of control that shock can produce.

Clinical care should begin with medical stability and then to validation of concerns and emotional impact. The patient is presenting with rather typical post-ICD shock fears such that interpretation about the meaning of shock is occurring. The patient clearly believes that occurrence of an ICD shock is associated with a very poor prognosis and is certainly associated with the need to terminate all physical activities.

• Shock education efforts: Mr. O. would benefit from additional education about why shocks occur and do not occur. His modest daily activity level is not likely to trigger a shock, and education about irregular heart rates versus increased heart rates may be helpful.

• Shock prevention efforts: Mr. O. will likely benefit from reassurance that programming and medication changes made as a result of the shock experience will decrease the chances of a future shock.

• ICD support group: Mr. O. was encouraged to attend a future ICD support group meeting to further normalize his concerns and learn more about the ICD.

• Reengagement in activity: Mr. O. should be encouraged to return to mild physical activity as tolerated. The psychological and physical benefit of activity should be emphasized.

• Considerations for referral:
  a. Cardiac rehabilitation: If Mr. O. demonstrates or reports ongoing activity concerns, consider cardiac rehabilitation referral for the purpose of resuming exercise under supervision to provide a sense of security to Mr. O.
  b. Psychology consultation: If Mr. O. continues to report depressed mood or anhedonia, consideration of a psychology referral for further evaluation and recommendations would be indicated.
Case 3.7.2

Case Presentation

Mr. B. is a 64-year-old male who presented in the clinic with a history of syncope, nonsustained ventricular tachycardia, previous myocardial infarction, and implant of an ICD 6 years prior. The patient was recently hospitalized for repetitive multiple ICD shocks while walking. The multiple shocks were due to suspected supraventricular tachycardia with 1:1 association via interrogation. The patient stated that he wanted the device removed or turned off to not deliver further shock therapies. He reported ambivalence about death but did not want to experience another shock. The electrophysiologist spoke with the patient concerning his fear and performed device parameter changes. The patient was discharged home.

The patient reports to the ICD clinic 4 weeks later for routine ICD interrogation. He stated that as a result of the multiple ICD shocks, his wife has become hypervigilant and would not leave his side. He reported that she does not want him to exercise, drive, or perform activities of daily living. He stated that she has taken temporary leave from her employment to care for him. They are both fearful of potential ICD shocks and that his wife will not "give him a break" from constantly watching him to make sure he is not getting his heart rate up.

Case Discussion

Mr. B. presents with few psychosocial risk factors but positive for a critical event (multiple shock experience due to a nonlife-threatening cause). The patient-and-spouse combination of distress is alarming as a provider charged with their care. It makes routine clinical activities such as device interrogation tedious and cautious. Their attitudes and behavior in clinic increase the self-consciousness of all providers. Although the shock directly affected Mr. B., the increasing hypervigilance and behavioral disengagement/avoidance is easily noted in Mrs. B.

A reasonable clinical plan in this case could include:

- **Acknowledgment of collective emotional impact:** Both Mr. and Mrs. B. need an empathic response to the impact of the cardiac condition and device. The multiple shock experience clearly was distressing and understandably so.
- **Device function and shock prevention efforts:** A major feature of an ICD is its ability to provide highly specific and constant monitoring of Mr. B.’s heart. As much as Mrs. B. cares, she is not as “accurate” as the ICD in reporting his cardiac function! She should be admired for her desire to be “on call” but the ICD does that function well. Instead, her job is to help Mr. B. systematically return to activities in a comfortable and well-tolerated manner versus restraining him.
- **Collective QOL plan:** Mr. and Mrs. B. can also be encouraged to plan a “celebration of life together” plan that affirms their commitment to a desirable QOL together. An example may be a brief overnight stay in a bed and breakfast or a "staycation" at home that involves no housework and carry-out food.
- **Reassessment in short interval:** This brief clinical intervention may only be the start of the needs for this couple to effectively overcome the multiple shock experience. However, the above plan could initiate some degree of hopefulness again that is sufficient. Nonetheless, ongoing reassessment of functioning is needed to determine if a more comprehensive multidisciplinary consultation is required.

REFERENCES


AVID Investigators. 1997. A comparison of antiarrhythmic-drug therapy with implant-


INTRODUCTION

In this chapter, we will examine the role of the associated professional (AP) in preventing or minimizing harm to patients, and limiting legal liability to the organization, when it becomes clear that implantable pacemakers (PM), defibrillators (internal cardioverter defibrillator [ICD]), cardiac resynchronization therapy devices (cardiac resynchronization therapy pacemaker and defibrillator [CRT-P, CRT-D]), or any other implanted components of their systems, such as leads, are or may be faulty. A successful approach to such situations involves not only an organized and careful approach but also attention to detail and thoughtful dealings with patients and families. Before all these things must come planning because recalls are not a possibility or even a probability: With systems as complex as those used in this field, they are a certainty. To a great degree, it is possible to prepare for them and minimize their impact on the rest of the required activities of the practice.

DEFINITION OF RECALL

The Food and Drug Administration (FDA) defines a recall as “a firm’s removal or correction of a marketed product that FDA considers to be in violation of the laws it administers, and against which the Agency would initiate legal action.” FDA characterizes recalls as class I, class II, and class III. Class I includes “a situation in which there is a reasonable probability that the use of, or exposure to, a violative product will cause serious adverse health consequences or death.” Class II means “a situation in which use of, or exposure to, a violative product may
cause temporary or medically reversible adverse health consequences or where the probability of serious adverse health consequences is remote.” A class III recall involves “a situation in which use of, or exposure to, a violative product is not likely to cause adverse health consequences” (FDA 2007).

Needless to say, whether a difficulty with an implanted product ever rises to the level of FDA action, all who care for patients with such products have moral and legal responsibility to see to patient safety in such circumstances. While manufacturers and others have distinct responsibilities of their own, clinicians are very much in the front lines when products fail or are suspect, and the AP role will certainly be an active one.

**READINESS FOR THE RECALL**

An up-to-date database of each and every patient in the practice with implantable rhythm control equipment is vital, and the first step to readiness. Whether this database is one of the available proprietary databases and device management software/hardware packages or a database specifically designed to meet the needs of the organization, it is crucial that the AP take a hand in its design and use.

In relatively small practices, it is sometimes considered safe in times of a recall to rely on identification of involved patients and products by data compiled by manufacturers. At the point of a recall, the manufacturer’s representative would be called or would appear and would search for the desired information, then provide it to the clinician. This method may seem particularly attractive if one manufacturer provides all or most of the implanted products for the organization’s patients. Although it may be better than no method at all, and although it may be a useful adjunct to the organization’s own collected data, to rely on it exclusively runs the risk of missing patients. This can easily happen if, for example, there are a number of potentially affected patients who are now followed in the practice but whose devices were implanted elsewhere and are unknown to the manufacturer’s representative.

Instead, it is preferable for the AP to assure that these data are compiled and maintained in the practice. Quite elaborate ways of configuring database software are possible so that detailed reports of many kinds can be made. However, a simple database program is all that is required, allowing rapid search of relevant fields.

The fields for the primary database should include at least the patient’s last name, first name and middle initial, gender, date of birth, organization ID number, type of device (PM, ICD, CRT-P/CRT-D), manufacturer, model name, model number, serial number, device implant date; atrial lead manufacturer, model number, serial number, lead implant date; right ventricular lead manufacturer, model number, serial number, right ventricle (RV) lead implant date; left ventricular lead (if any) manufacturer, model number, serial number, and left ventricle (LV) lead implant date. Also included should be all demographic data, each within a separately named field to allow independent search. In addition, there should be fields to indicate the implanting, following, and referring physician, whether the patient is PM dependent, the principal reason for implant, whether as of the most recent visit the patient is living or deceased, and the date of that most recent visit.

If the database is intended to provide for generation of visit reports, then it should also include fields for findings in threshold testing, battery voltage, charge time (if applicable), various lead impedances, and free text fields to allow descriptive documentation.

Once this primary database exists and its fields have been populated, a second “recall-ready” database can more easily be created, to be filled in from the fields of the primary database upon notification that a recall has occurred. This simpler collection and display instrument would have fields only for the
patient identification data and demographics, implanted products, implanting/referring/following physician information, visit recency, PM dependency, and status as living or deceased. There should be free text fields as well, which can include “action taken/date” and “actions pending” to provide a way to track what has been done in individual cases.

**STEPS TO TAKE UPON NOTIFICATION OF RECALL**

Commonly, first word of the difficulty will be delivered by a representative of the manufacturer. The representative will be prepared to discuss the problem, products involved, potential consequences, and the likely percentage of products affected. Usually, the representative will have a copy of the company’s official “Dear Doctor” letter by which the physicians in the practice will be formally notified of the action. This action may not be termed “recall” if it is done entirely at the manufacturer’s initiative; it may instead be termed “safety alert,” “product advisory,” or called by some other name. Frequently, a representative who is familiar with the practice will deliver this copy of the notification letter directly to the AP, knowing that he or she is the person most likely to begin work on the matter.

It is important for the AP never to assume, however, that the practice’s physicians are already aware of the problem, and to bring copies of the letter to each of them personally without delay as a first step. Each may be conscious of particular issues the problem might present for individual patients and may be able to direct the AP in prioritizing visits for these patients.

The second step should be a discussion between all involved physicians and APs in the practice about the recall and priorities for the response. Difficulties in assembling a group meeting being what they are, it may be necessary to have this discussion by conference call or e-mail. Topics on the agenda might include the nature of the problem and the suspect products, the number of patients potentially affected, and the general likelihood that action beyond heightened observation and increased visit frequency might be needed, as well as what that action might include. Because it may not be practical for all to participate directly in the meeting, to save time one physician should assume leadership in the matter, with the help of one AP, either being able to delegate some responsibilities to his or her associates.

Another of the things to be decided in the meeting should be the way in which patients will be notified. You may be sure that enterprising plaintiff attorneys in the community and beyond will very shortly be doing their part to make the recall known to the public. In days, patients will be calling about newspaper articles titled “Attorney Brown Decries Irresponsible Manufacturer of XYZ Defibrillator” and the like. It will be far less disturbing to them if first word of the recall comes from a trustworthy source: the office of their own physician. Planning that first contact in detail will take some discussion, but opening the topic should be a high priority.

It goes without saying that the last order of business should be setting the next meeting for a discussion of progress. This next discussion should take place within a day or two.

The third step after notification should be creation of the database of potentially involved patients and a review of their records. This will possibly take more than 1–2 days or even more, but it must begin as soon as possible to allow for prioritizing patient contacts and visits. Since it is crucial to success, completion of this step may even require shifting of other duties, even patient appointments, to other times. If more than one AP can be made available to speed this third step’s completion, although it may be at the expense of other matters, it will be time well spent.

The fourth step is delivery of the database to all physicians and APs in the practice. This step
should very quickly be followed by the fifth step: another meeting, live or virtual, to review which patients must be seen first and how to proceed according to the visits’ findings.

Next comes decision making about the way patients will first be informed of the recall. While this first notification may certainly take the form of a telephone call to the patient from the AP, the content should be carefully designed and even scripted at a meeting between physician leadership and APs. Even when the product action concerns a problem unlikely to cause harm or risk to patients, such a call may be quite disturbing to receive. With a list of prepared “talking points” in hand, the AP may be more easily able to communicate the message of calm concern, acceptance of the patient’s worries, and ability and intent to investigate and deal with the problem as it applies to this patient in particular. The reader will find a sample script for this initial patient contact as below.

Hello, Mr. Smith. This is John Doe from the ABC Device Clinic, where you have your defibrillator checked. Dr. Jones asked me to call you today to give you some information. Is this a good time to talk, or should I call back at a different time?

We’ve just received word from the XYZ Corporation, the company which makes your defibrillator, that they’ve noticed that a few of the defibrillators they make haven’t all been working perfectly. One of the models they make happens to be the same as yours, and Dr. Jones has decided she’d feel better checking on yours by asking you to come in to the office this week, if possible. That way, we’ll know for sure right away that yours is working well now. Do you think we could make an appointment for you this week?

Dr. Jones is going to write a letter to you to tell you everything about what the company said. When something like this happens, it’s often called a "recall" and that can make some people think it means the doctor will have to replace your defibrillator. We want to be sure to tell you that replacing it will almost certainly not be needed. The problem the company has seen is that some of the batteries in the same model defibrillator as yours lose their power sooner than they were expecting. All over the country, more than 200,000 of this same model have been given to patients, but the company has seen this problem with only seven of them. So far, it seems as if only a very few individual defibrillators will have the problem. The ones with the problem are so far devices that were made between October and December of last year.

They spotted the difficulty because doctors called them to report it in those seven cases when the patients had their office checks. The doctors and the staff in the clinics noticed that the batteries were wearing down more rapidly than usual. No one has been hurt, and only two of the defibrillators needed to be replaced.

Dr. Jones feels that it’s best to check your defibrillator, even though yours was made quite a while before the ones with the problem, and that’s why we’re asking if you can come in this week to see us. As I said, she’s writing a letter to you with more information and if she’s in the clinic when you come, she’ll want to speak with you personally. If she isn’t here, she’s going to call you within a day after you come in.

We also want to tell you that we’re going to arrange home monitoring of your defibrillator so we can see how everything is with it even in between visits. The company makes special monitor machines for this, and we’ll ask them to send one to your house. There’s no charge to you for the service. The monitor connects to your telephone and once it’s plugged in, it will automatically collect information from your defibrillator and send it to the Internet, so we can see all the information in the clinic. Of course, we’re still going to see you in the office for regular visits, too. We’ll tell you more about home monitoring when we see you here.

I want to make an appointment for an office visit, but right now, do you have some questions for me?
The AP who makes this call must remember that a recall notification, no matter how well phrased, can be a very disturbing event to patient and family. Expect that in many cases you will need to repeat portions of the message several times, even points which may seem quite simple. Expect an emotional, even frightened or angry, response, and prepare yourself to be at your most compassionate and patient. When patients you have called come in for clinic visits and speak with the physician, they are likely to have forgotten much or all of what you have told them by phone and may report you have given them no information at all. This is an entirely normal reaction to what may be an alarming situation for them.

As always, take care to be absolutely truthful in this first contact, but at the same time limit the details if possible so as to not overload the patient with information. Be cautious not to give medical advice or to speak for your physician colleagues; do not make promises that no action such as product explantation will be needed. Remember that patients will take their firmest reassurance from speaking with the physician since he or she will be able to speak more definitively about further follow-up and other actions. Be sure to give them ways to contact the practice and pass on their questions to the physician staff, and make every effort to arrange time in clinic for patients and families to speak with them directly.

A letter directly from the physician who sees each individual patient should be sent out in very short order, best within 3 days of notification of the recall. Not only will it provide fast reassurance that the problem is being addressed, but it will also help in limiting the practice’s legal liability. Some organizations send these letters with return receipt requested or even signature on delivery to be sure they have arrived safely; others regard this step as more likely to alarm patients and families, and simply phone afterward to see whether the letter has arrived. A sample letter is provided below.

ABC Device Clinic
123 Main Street
Anytown, USA 00000
(003) 090-2000
(Date)

Dear (Patient):
I am writing to you to discuss the announcement by the XYZ Corporation, which made your defibrillator, that it has noticed in a very few cases that some of its models lose their battery power more rapidly than expected.

Our records show that your defibrillator is the same model as one of those with this problem, and we ask that you visit our clinic so we may check its performance. At your appointment, I'll try to meet with you personally to discuss the meaning of this problem in your particular case. If I can’t see you myself on that day, within a day afterwards I'll phone you to talk about these issues.

I want to stress that from information we have now, only seven cases of this problem have been seen out of about 200,000 models like yours implanted in the United States. Still, I ask that you allow us to examine your defibrillator so that I can know more. In addition, as we’ll explain in detail in the office, I want to arrange for the company to enroll you at no charge in its home monitoring program. A monitor you'll have at home will automatically collect information about the performance of your defibrillator and the program will make that information available to us here in the clinic. In this way, we’ll be able to get information between visits.

Again, let me reassure you, the chances are very good that no further action, other than the office and home monitoring checks, will be needed. With the
data they provide I’ll have much more information to help me know everything I need to know about your defibrillator’s performance.

Perhaps our staff has already contacted you to request an appointment. If not, would you please call our office at your first convenience to make the arrangement? If you have questions, please don’t hesitate to leave a message for me. I’m looking forward to seeing you in clinic.

Sincerely,

Jane Jones, MD
Director, ABC Device Clinic

If circumstances permit, a review of this written communication by an attorney with risk management experience is wise before mailing it. While it may seem cynical to think defensively, it is foolish to miss an opportunity to meet every responsibility to patients, and lawyers focus on this very issue most effectively.

**AFTER THE INITIAL POSTNOTIFICATION VISIT**

The AP or delegated staff must keep careful records on which patients have and have not been contacted and come to clinic, and the AP must pay particular attention to the quick scheduling of any procedures, such as product explantation or replacement, found necessary. If an actual or potential product defect is judged to require such action for individual patients, they will be most comforted if it is plain to them that their needs are being seen to without delay. More routine procedures might even need to be postponed to allow completion of recall-related matters.

APs should encourage the practice to strongly consider remote monitoring (RM) of involved devices if possible. Although RM is a less-than-ideal method of follow-up for some patients, it may provide an additional source of data for clinicians to confirm the continued good function of devices between visits. In addition, patients and families may find it reassuring to know their device function is being monitored at a distance.

Referring physicians should not be forgotten in the recall process. In many instances, they will not receive manufacturer notifications and indeed in the worst cases may learn of the recall along with the general public, or even be informed by their own patients. Reasonably, they expect to be kept informed about the care of their patients in other practices. The most courteous approach is for physician leadership in the group to telephone them personally, with the charts at hand of those patients they have referred for rhythm management. The AP can greatly facilitate this process by gathering this information. This simple and respectful gesture, together with occasional concise updates, can go a long way toward preserving and even improving good will among referring physicians.

After the initial flurry of recall-related activity, interest within the practice may begin to fade once patients are being seen and the potential defect gradually ruled out in the patient population. It is at just this point that the AP must encourage the continued communication that ensures every patient is seen, and every patient in need of action receives it promptly. In a busy practice, e-mail may be the best-tolerated form of such contact and also provides a paper trail to prevent misunderstandings.

A final review of the recall should take place between the AP or APs involved and physician leadership once the last patient has been seen and the last procedure scheduled. This debriefing will serve as a launching platform for ideas for management of the inevitable next recall.
CONCLUSION

The AP has a vital role in management of recall activities, including planning, database creation and management, meeting and communication organization, drafting of notifications to patients, arrangement for appointments, procedures, and remote follow-up, continuing communication within the practice, and debriefing to improve performance in recalls to come.

REFERENCE


RESOURCES

Device Evaluation in Special Circumstances

Lynne D. Foreman and Deepak Bhakta

Given the broader indications for device therapy, implanted cardiac devices are being applied in greater numbers and in a wider of spectrum of patients. This has led to a remarkably heterogeneous patient population who receive such devices, sometimes resulting in unique presentations when device-related problems are encountered.

Regardless of the situation in which a device patient presents, a thorough, organized, and stepwise evaluation must be used. This assures that all aspects of care are addressed, both of the patient and their device. All too common, caregivers center their awareness on an obvious device-related problem or finding, and divert attention away from the consequences exerted on the patient at hand. The goal of evaluation is to determine whether an individual’s symptoms are due to a spontaneously occurring rhythm disorder, and the integrity and appropriateness of device behavior given the clinical circumstances. Many of the steps of evaluation may be performed simultaneously to allow for expeditious diagnosis and treatment of a patient with device malfunction or an ongoing rhythm disturbance.

Any device evaluation should begin with a thorough history. Historical details to be obtained include the nature of symptoms, many of which may be nonspecific. While palpitations may be a vague symptom in an individual without an implanted device, they may indicate nonphysiological pacing resulting in tachycardia in a device patient. Similarly, fatigue and effort intolerance may represent loss of atrioventricular (AV) synchrony due to asynchronous ventricular pacing, absence of rate-adaptive pacing in a chronotropically incompetent patient, or loss of left ventricular capture in a biventricular system. Neck or head pulsations may suggest asynchronous ventricular pacing due to loss of atrial sensing or
capture in a dual-chamber system, or loss of ventricular capture and resulting junctional or ventricular escape rhythm in a patient with complete heart block. Ischemic and heart failure symptoms may also yield important clues. Perhaps most ominous is the presence of pre-syncope or overt syncope, hallmarks of a hemodynamically-significant arrhythmia that may accompany implantable cardioverter defibrillator (ICD) shock or catastrophic loss of capture without underlying escape rhythm. Other details that may be helpful include activity performed at the time of symptom onset and timing and duration of symptoms. Physical examination should be directed toward determining the potential etiology of symptoms and uncovering consequences of a rhythm disturbance or device malfunction. Hemodynamic status can be readily assessed measuring blood pressure and heart rate. The presence of pallor or cool extremities may also indicate reduced cardiac output, as can be seen with ongoing tachyarrhythmias or bradyarrhythmias, the latter suggesting pacing failure. Signs of heart failure such as neck vein distension, inspiratory rales, and edema may indicate underlying acute cardiac failure, possibly acting as a trigger for ventricular arrhythmias resulting in ICD discharge, or reduced cardiac output due to pacing failure or inappropriate pacemaker programming. Cannon A waves in the jugular venous pulse may be observed with continuing ventricular arrhythmias or with asynchronous ventricular pacing (as with loss of atrial capture or sensing). Finally, examination of the implant site including device manipulation within the pocket, particularly with recent implantation, may also explain abnormal findings, or induce them at the time of evaluation. Ongoing rhythm disturbances and hemodynamic derangements should be addressed during assessment.

Device interrogation should follow the history and physical examination. Initial programmed parameters should be reviewed, followed by obtaining characteristics of battery performance (battery voltage and, where applicable, magnet rate or charge time), measurement of pacing threshold and sensing performance in all channels, pacing impedance, and if relevant, shock lead impedance. A review of stored arrhythmia logs and determination of underlying rhythm complete the analysis. Appropriate corrective steps and reprogramming can then be performed based on observed findings.

Upon conclusion of the evaluation and along with beginning any treatment, the patient should be reassured regarding the status of their device and rhythm disorder, and have their questions answered.

The special situations in which a device patient can present have been categorized into various acute and periprocedural presentations. Further classification within these categories is discussed.

ACUTE PRESENTATIONS

Office Presentation

In light of the complex cardiovascular and arrhythmic history seen in device patients, office presentation is generally encouraged, given access to personnel most experienced at diagnosing and treating cardiac rhythm and device-related problems. Findings observed in this setting may vary considerably, ranging from appropriate ICD therapy corresponding to a patient’s perceived shock, to incidentally discovered, asymptomatic lead abnormalities. While a general history and physical examination are still advocated, their yield may be attenuated, particularly in asymptomatic individuals. Examples of abnormalities observed in the office are shown in Figures 3.9.1 and 3.9.2.

Emergency Department (ED)

While office presentation is often the most suitable for device patients, the unpredictable nature of arrhythmic problems may preclude
Figure 3.9.1  Arrhythmia episode recorded during routine in-office ICD interrogation in a 33-year-old man with prophylactic biventricular ICD implantation for severe nonischemic cardiomyopathy and congestive heart failure. Tracings from top to bottom represent atrium, ventricle, and shock lead recordings. Panel (A) demonstrates triggering premature ventricular complex followed by ventricular fibrillation (VF) (B). Defibrillatory shock is seen at the left of panel (C), with ultimate restoration of sinus rhythm. This event occurred during sleep; hence, the patient was completely asymptomatic.
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Figure 3.9.2 Mode switch episode obtained during in-office dual-chamber pacemaker interrogation in an asymptomatic woman. Tracings from top to bottom are atrium, ventricle, and marker channel and intervals. Noise oversensing is observed solely in the atrial channel, triggering inappropriate mode switch. This patient had a unipolar atrial lead, and skeletal muscle potentials were invoked as the cause of interference.

Remote Monitoring

Considerable advances have been made with regard to remote device monitoring. Transtelephonic pacemaker evaluation provides the clinician with information regarding pacemaker magnet rate (and therefore, a limited evaluation of battery performance) and adequacy of pacing and capture. Newer, manufacturer-specific remote monitoring systems are available. Initially developed for ICD monitor-
Chapter 3.9 Device Evaluation in Special Circumstances

Treated VT/VF Episode #43

<table>
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<tr>
<th>Type</th>
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<th>Shocks</th>
<th>Success ID#</th>
<th>Date</th>
<th>Time hh:mm:ss</th>
<th>Duration hh:mm:ss</th>
<th>Avg bpm</th>
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<td>VF</td>
<td>0</td>
<td>6</td>
<td>No</td>
<td>43</td>
<td>27-Jan-2009</td>
<td>02:04</td>
<td>-03:06</td>
<td>80/300</td>
</tr>
</tbody>
</table>

Detection

- V-V
- A-A

**VF = 320 ms**

Figure 3.9.3  Emergency room interrogation from a 78-year-old man with prophylactic biventricular ICD implantation for ischemic cardiomyopathy and congestive heart failure who presented with 52 ICD discharges without associated symptoms. Panel (A) shows an interval plot from one episode, demonstrating sinus rhythm (marked by open squares) and erratic ventricular rhythm (black dots). A total of six maximum energy shocks were delivered for this particular episode. Panel (B) shows corresponding electrogram (from top to bottom, atrium, ventricle, and marker channel with intervals). Sinus rhythm is seen with apparently irregular ventricular rhythm. The latter is due to electrical noise from “make-break” contact from suspected lead conductor fracture. Asterisks and arrows denote actual conducted and paced QRS complexes, respectively.

...ing, at least one manufacturer has support available for pacemakers. While portions of the evaluation described above, such as physical examination, are obviously omitted with remote monitoring, these systems offer considerable advantages and have become a valuable tool in the care of device patients. These include patient convenience, comprehensive interrogation of device performance, programmed parameters, and stored arrhythmia episodes.
Figure 3.9.3 (Continued) Panel [C] shows a lead impedance trend; a sharp increase to 1,600 Ω is observed on the day of presentation, confirming abrupt lead conductor fracture.

These systems also afford more intensive device monitoring, a particular benefit to allow for earlier detection of potentially serious device and lead malfunction. Figures 3.9.5–3.9.7 demonstrate findings recorded with the help of remote monitoring systems.

PERIPROCEDURAL EVALUATION

A variety of situations may arise where an implanted cardiac rhythm device’s function can be affected by iatrogenic sources. The classical
permanent corruption of device circuitry and programming, with self-programming to rudimentary functioning. Several situations deserve special consideration, however, due to unique device interactions in addition to those already mentioned, and are discussed below.

**Surgical Electrocautery**

The archetype of iatrogenic device interaction can be seen with surgical electrocautery. The nature and frequency of signals generated by electrocautery strongly resemble that seen with EMI, with the typical observed responses as outlined above. Inhibition of pacing, device reset, and false atrial and ventricular arrhythmia detection may occur, the latter resulting in inappropriate ICD therapy delivery (Lamas 1986; Cassavant 1998). Although uncommonly

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**Figure 3.9.5** Transtelephonic pacemaker monitoring report in a patient with a dual-chamber epicardial pacing system. The top tracing shows the presenting cardiogram, indicating normal sensing. The bottom tracing demonstrates normal magnet response with loss of capture on a single paced beat (arrow). In this particular device, the third paced beat following magnet application is delivered at 80% programmed output, indicating ventricular pacing threshold in this patient.
Figure 3.9.6  Routine remote ICD transmission from a patient with biventricular ICD implantation for nonischemic cardiomyopathy, congestive heart failure, complete heart block, and complaints of dizziness. Orientation is the same as in Figure 3.9.3. (A) Low-amplitude electrical activity is seen in the ventricular channel, resulting in oversensing and inhibition of ventricular pacing. (B) Corresponding interval plot. In both panels, note transient reduction in sinus cycle length following oversensing of short ventricular intervals causing inhibition of ventricular pacing/asystole.
Figure 3.9.7 Home ICD transmission in a patient with dual-chamber ICD for ischemic cardiomyopathy and sinus node dysfunction who was awakened by ICD shock. Orientation is the same as in Figure 3.9.3. (A) The onset of irregular ventricular tachycardia is seen (asterisk) followed by attempted termination with antitachycardia pacing (arrow). This results in acceleration to ventricular fibrillation (VF) (dagger) and successful defibrillatory shock, seen in panel (B).
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In pacemaker-dependent individuals, an asynchronous pacing mode (DOO, VOO, or AOO in single-chamber atrial pacing systems) can be used to prevent electrocautery-induced inhibition of pacing resulting in asystole. In ICD patients, inactivation of arrhythmia detection and therapies can be programmed to prevent spurious therapy delivery (see Figs. 3.9.9 and 3.9.10). Regardless of the approach used, continuous electrocardiogram (ECG) telemetry and prompt access to defibrillatory and resuscitation equipment is mandatory, and device-trained personnel must be readily available. Postprocedural evaluation is recommended in all cases to assure restoration of original programmed parameters, proper device and lead function, and although rare, to exclude direct electrocautery- or surgery-related device damage.

Radiation Therapy

While a surprisingly rare phenomenon, device evaluation in radiation therapy warrants mention in light of potentially serious device interaction and malfunction (Kapa 2008). Device failure can occur via two primary mechanisms: through EMI (generally a transient phenomenon observed acutely during therapy delivery)
Figure 3.9.9  Device-determined atrial arrhythmia episode retrieved from a dual-chamber pacemaker implanted in a patient with sinus node dysfunction who underwent shoulder surgery. Atrial and ventricular electrogram channels show evidence of intraoperative electrocautery usage.

Figure 3.9.10  Arrhythmia episode recorded in a patient with cardiomyopathy who had prior single-chamber ICD implantation. This stored arrhythmia episode is representative of numerous episodes from the day of coronary artery bypass surgery. (A) Right ventricular (top) and shock lead electrogram (bottom) show bursts of high-frequency electrical activity corresponding with electrocautery usage. (B) Enough short intervals are present to result in false arrhythmia detection and eventual delivery of antitachycardia pacing.
and more serious permanent damage to device circuitry resulting in complete device failure (Calfee 1982; Rodriguez 1991; Marbach 1994; Last 1998; Hurkmans 2005). There are clear adverse effects of ionizing radiation on pacemakers, including EMI during radiation therapy. More serious malfunction and failure are also evident, including telemetry failure, variations in pulse width, markedly rapid pacing (i.e., “runaway pacemaker”), and complete (occasionally abrupt) loss of function (Adamec 1982; Rodriguez 1991; Souliman 1994; Last 1998). These effects appear to be more likely with direct exposure, although the effects of scatter radiation should not be overlooked (Hurkmans 2005; Kapa 2008). Less data are available regarding ICDs. In vitro studies evaluating direct radiation exposure on ICD function have demonstrated battery depletion, prolongation of capacitor charge time, abnormal delivery of lower-than-programmed shock energy, aberrations in pacing and sensing, and overt failure (Rodriguez 1991; Hurkmans 2005). EMI during acute irradiation has also been reported (Hurkmans 2005). A larger series did not show any evidence of in vitro ICD failure in response to scatter radiation (absence of reset, change in programmed parameters, or loss of telemetry). In this series, a retrospective analysis of a small number of implanted ICDs exposed to scatter radiation also did not show any abnormalities or device failure (Kapa 2008).

These seemingly disparate results lead to equally divergent management recommendations. Some manufacturers recommend lead shielding of the device, while others advise against this to prevent backscatter. Additionally, some manufacturers have recommended upper limit of radiation doses, whereas others do not (Solan 2004). Finally, recommendations as to the need and frequency of device interrogation vary by manufacturer, and no evidence exists to support this practice (Solan 2004; Kapa 2008).

Management of the device patient surrounding the time of radiation therapy should be individualized, again based on the features mentioned above. All individuals should have telemetry monitoring during treatment. Temporary reprogramming to an asynchronous pacing mode should be considered, particularly in those who are pacemaker dependent. Magnet application should suffice in most that are not pacemaker dependent. ICD patients should have arrhythmia detection and/or therapy temporarily disabled either via magnet application or temporary reprogramming. Access to defibrillation and device-trained personnel is mandatory, and communication between the radiation oncologist and device caregiver may help prevent adverse consequences.

It is clear that direct radiation exposure may result in catastrophic device failure; in this situation, moving the device or reimplantation to a site outside the radiation field is recommended (Adamec 1982; Marbach 1994; Solan 2004; Kapa 2008). The decision to shield the device should be manufacturer-specific (Solan 2004). Interrogation following the first several treatment sessions seems appropriate, while further interrogation should be governed by individual patient and device characteristics. This may be simplified by using remote monitoring systems. While the benefit of serial interrogation is uncertain, the effects of ionizing radiation appear to be cumulative (i.e., greater probability of device failure with progressive exposure), and abrupt failure has been reported at relatively low radiation doses (Calfee 1982; Rodriguez 1991; Solan 2004; Hurkmans 2005; Kapa 2008). Consequently, serial follow-up of some form is indicated. In all cases, the device manufacturer should be consulted for specific recommendations (Solan 2004).

**Transcutaneous Electrical Nerve Stimulation (TENS)**

TENS is used to treat a variety of chronic pain, neurological, and cardiovascular conditions. By applying low-level, cyclical electrical current, the potential for adverse device effects is appar-
ent through pacing inhibition and false arrhythmia detection (Eriksson 1978). In one report, no adverse interaction was seen with initial telemetry during TENS use, with evidence for interference with chronic application. Pacemaker reprogramming to a less sensitive setting eliminated interaction (Chen 1990). A larger series of pacemaker patients showed no ill effects (Rasmussen 1988). The effects in ICDs have been less well studied. Two prospective, observational series studied the potential effects of TENS specifically in ICD patients utilizing a range of delivered pulse widths, current strengths, and frequencies. Both showed clear evidence of interaction in a significant number of individuals (37–53%), with falsely detected ventricular arrhythmias due to TENS application in several subjects that would have resulted in the inappropriate delivery of ICD therapy. Significant interaction appeared to be more likely when TENS application occurred in closer proximity to the ICD system (neck and chest), although interference was also seen when applied to the lower extremities. Interference was also more common in those with integrated (as opposed to dedicated) bipolar leads (Crevenna 2003, 2004; Holmgren 2008).

While the potential for device interaction due to TENS is clear, the clinical significance of such interaction should be weighed against the potential benefit of its use. The decision to recommend TENS use in a device patient should be individualized, and the factors to be taken into account are the same as those when dealing with surgical electrocautery. TENS use may be considered in carefully selected pacemaker patients with bipolar systems who are not pacemaker dependent, where no evidence of interaction is seen when device interrogation is performed during application. If observed, programmed sensitivity adjustments may eliminate such interaction in pacemaker patients, but should probably be avoided in ICD patients given the risk of underdetection of ventricular arrhythmias (Chen 1990). In those who are pacemaker dependent or with unreliable underlying escape rhythm, TENS may be considered with simultaneous magnet application, or if asynchronous pacing mode can be programmed during use. ICD patients should probably avoid TENS application altogether, unless arrhythmia detection can be inactivated (either via reprogramming or magnet application) and ECG telemetry can be maintained during use (rendering home use nearly impossible). In all cases, periodic device evaluation should be performed to assess for adverse interaction and to assure system integrity.

**Extracorporeal Shock Wave Lithotripsy (ESWL)**

By utilizing a hydraulic shock wave generated from vaporized fluid, ESWL is most commonly used as an effective treatment for upper urinary tract calculi. The possibility of adverse device interaction through EMI and mechanically induced structural damage has led to its limited use in device patients. Over the years, several series have studied the effects and safety of ESWL on cardiac devices. *In vitro* testing of pacemakers subjected to ESWL in one such study demonstrated inhibition of pacing in 50% of devices (regardless of unipolar or bipolar pacing configuration), without evidence of structural pacemaker damage (Langberg 1987). A similar study used single- and dual-chamber pacemaker pulse generators strapped to the abdominal wall of patients undergoing clinically indicated ESWL, and a separate analysis using pulse generators suspended in a water bath. Lithotripter output was delivered in both a synchronized (i.e., delivered synchronously to the timing of atrial pacing stimulus) and unsynchronized manner, with interaction observed in all systems. Oversensing of lithotripter output in transient inhibition of pacing was seen when oversensing occurred in the ventricular channel, and inappropriate delivery of ventricular stimuli was recorded when output was detected in the atrial channel.
Cardiac Arrhythmia Management

While it appears that many device patients may safely have ESWL, precautions must be taken to limit the risk of adverse device interaction, although clear recommendations have not been published. All device patients undergoing ESWL should have preprocedural and postprocedural device evaluation. Continuous ECG monitoring should be used in all patients. If possible, the ESWL focal point should be placed a minimum of 15–18 cm away from the implanted device, with lithotripter output synchronized to the QRS complex. In pacemaker-dependent patients, the device should be reprogrammed to an asynchronous pacing mode (VOO, or AOO in single-chamber atrial pacing systems); for those with a stable underlying rhythm, the VVI mode (or AAI mode in single-chamber atrial pacing systems) should be selected. Rate-adaptive pacing should also be temporarily inactivated. For those with ICDs, arrhythmia detection and/or therapies should be inhibited, with access to defibrillatory and resuscitation equipment and device-trained personnel. A full interrogation should be performed following ESWL in all cases, with restoration of previous programmed parameters. Careful attention should be given to evidence of noise reversion and arrhythmia logs/counters as these may be clues to device interaction and may assist future management if ESWL is required. Radiography should be used if mechanical damage is suspected. Alternate management should be considered for those with abdominal pacemaker or ICD systems. Finally, the device’s manufacturer should be consulted for device-specific recommendations (Platonov 2007).

Ventricular Assist Devices (VADs)

The advent of prophylactic ICD implantation in those with systolic left ventricular dysfunction has resulted in an expanding population of ICD recipients with heart failure (Moss 2002; Bardy 2005). In turn, the progressive nature of heart failure and subsequent development of advanced heart failure has led to the growing
use of mechanical circulatory support devices (Miller 2007). With these technological advancements, the unprecedented application of combined device therapy in this complex patient population has raised the issue of potential device interaction. Two reports have documented significant interaction between one particular ICD model (St. Jude Atlas V-193, St. Jude Medical, Sunnyvale, CA) and VAD model (Thoratec HeartMate II, Thoratec Corporation, Pleasanton, CA); in both cases, manifesting as failure to communicate with the ICD. EMI emitted by the VAD at a similar frequency (7.2 kHz) as that used by the ICD to perform interrogation and reprogramming (8 kHz) was invoked in both situations. In both cases, pulse generator replacement to a newer generation device with different telemetry frequency (64 kHz) was undertaken to alleviate this interaction (Matthews 2007; Mehta 2007). Temporary reduction in VAD voltage input has also reportedly been effective to circumvent this problem (Mehta 2008). The authors have observed this same interaction between these devices in two patients, and successfully thwarted this problem by applying ferromagnetic material between the two devices, effectively shielding one from the other (unpublished data). While such interactions may seem rare, the growth and overlap of severe heart failure and ICD patient groups along with further developments in device therapy may yield similar, more complicated interactions in the future.

POSTMORTEM EVALUATION

While often overlooked, the postmortem evaluation can offer valuable information regarding details surrounding a device recipient’s death. Admittedly, the information to be gained may be limited, although careful review of arrhythmia logs with approximate correlation to the time of death can confirm or exclude the presence of arrhythmic death, evaluate for possible device or lead failure leading to death, and assist in medical-legal proceedings. Interrogation must be carried out soon after death, as explantation and handling of the device and lead(s) as part of funeral preparations often result in artifact; such artifact can obscure findings or be misinterpreted as arrhythmia. Data on the frequency and utility of postmortem device interrogation is unfortunately lacking, although it may provide valuable insights into the cause of death in device recipients (see Figs. 3.9.11 and 3.9.12).

Figure 3.9.11  Postmortem interrogation from a patient found by his family. This is the same patient featured in Figure 3.9.1, approximately 1 year later. Orientation is the same as in Figure 3.9.1. (A) Sinus tachycardia with frequent premature ventricular complexes is seen followed by initiation of ventricular tachycardia.
(B) Relatively organized ventricular flutter results followed by degeneration to ventricular fibrillation (VF) (C). (D) All therapies delivered thus far failed to terminate VF. The eighth and final shock (arrow) also fails, with cessation of further therapy attempts and VF prevailing as the terminal rhythm.

Figure 3.9.11 (Continued)
**Figure 3.9.12**  Postmortem interrogation obtained soon after death from a patient with ischemic cardiomyopathy with prophylactic single-chamber ICD. ICD lead failure was suspected as possibly contributing to death. (A) No sustained arrhythmia episodes were detected surrounding the time of death. (B) Normal premortem lead measurements were recorded by the device. These findings excluded arrhythmia and device/lead failure as potential causes of death.

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**Counters Report**

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**Status Report**

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**Device Status**

Charge Circuit is OK
CONCLUSION

Evaluation of the device patient should be done in a thoughtful, routine manner and should remain consistent despite differing circumstances. Observations can be tailored to specific situations, but care must be taken to avoid narrowing focus only to the device or one particular feature or finding. Communication between the device expert, patient, and other involved caregivers is essential to assure that all device-related issues and patient needs are addressed.

Case 3.9.1

Case Presentation

A 14-year-old girl with complex congenital heart disease presents to the office with complaints of severe presyncope, exertional dyspnea, and effort intolerance for 2 weeks. She underwent implantation of a dual-chamber epicardial pacemaker system at age 8 for heart block following reparative cardiac surgery. Physical examination is significant for blood pressure 75/40 mmHg, pulse 55/minute, cannon A waves in the jugular venous pulse, harsh grade 3/6 holosystolic murmur at the left sternal border with parasternal heave, and cool, pale extremities. An abdominal pacemaker incision site is well healed. ECG is shown in Figure 3.9.13.

Case Discussion

The in-office ECG shows sinus tachycardia (100/minute) with complete heart block and junctional escape rhythm (55/minute). Sinus P waves are followed by ventricular pacing stimuli best seen in lead V1 rhythm strip (arrows) without resulting capture. Pacemaker interrogation confirmed abnormal ventricular lead function, and urgent ventricular lead reimplantation was performed, once again using an epicardial route.

Figure 3.9.13 Electrocardiogram shows sinus tachycardia with complete heart block and junctional escape rhythm.
Case 3.9.2

Case Presentation

A 28-year-old man had dual-chamber pacemaker implantation for congenital complete heart block accompanied by progressive fatigue. Symptomatic improvement was noted on a follow-up visit 1 week after implantation, with normal interrogation and device function. One week later, he returned to the ED with palpitations and fatigue, similar to his pre-implant status. Physical examination showed cannon A waves in the jugular venous pulse and variable S1 intensity. The pacemaker incision site in the left infraclavicular fossa was well healing. A recording taken from the pacemaker interrogation is shown in Figure 3.9.14.

Case Discussion

Figure 3.9.14 shows the absence of both atrial and ventricular capture, with normal atrial sensing, suggesting lead dislodgement, which was confirmed with chest radiograph. On closer questioning, the patient admitted lifting and moving a heavy church podium several days prior to emergency room presentation. As satisfactory capture could not be achieved with programming adjustments, lead revision was performed. Postoperative instructions included a strong reminder to restrict ipsilateral arm movement. Follow-up visit 2 weeks later showed normal pacemaker function with resolution of fatigue.

Figure 3.9.14 Absence of both atrial and ventricular capture, with normal atrial sensing.
Case 3.9.3

Case Report

A 59-year-old man was brought to the ED following a minor motor vehicle accident. He reported syncope while driving into an intersection and struck the car in front of him. He denied ICD discharge. No injuries were sustained in the crash. The patient had prophylactic biventricular ICD implantation 4 weeks prior to the accident for nonischemic cardiomyopathy, left bundle branch block, and advanced congestive heart failure. Physical examination showed mild chest wall tenderness, clear lungs, grade 1/6 apical holosystolic murmur, S4 gallop, with well-healed left shoulder incision. ICD interrogation was performed and showed normal device and lead function. One arrhythmia episode had been recorded and corresponded to the time of the accident (Fig. 3.9.15).

Case Discussion

The figure demonstrates ventricular fibrillation successfully treated with a single 31-J shock. The patient’s syncopal episode was attributed to this rhythm disturbance and explains why ICD shock was not perceived. Management included increasing beta blocker dose and shortening arrhythmia detection criteria to possibly prevent syncope with arrhythmia recurrence. The patient was counseled to avoid driving for a minimum of 6 months.

REFERENCES


RESOURCES
Section 4
Syncope and Sudden Cardiac Death
INTRODUCTION

Although a common medical complaint, syncope or the “common faint” is not commonly understood. Syncope, defined as the transient loss of consciousness and postural tone with spontaneous recovery, is associated with a wide variety of medical conditions that manage to perplex and frustrate even the most astute clinician (Grubb and Olshansky 2005). The earliest description of syncope dates back to the beginning of time, when Hippocrates coined the term “syncopen”—meaning to “cut short” (Grubb and Kosinski 2002). Even Florence Nightingale, the founder of modern nursing, is reported to have suffered from episodes of syncope associated with chronic fatigue and debilitation as a result of a persistent brucellosis infection contracted during the Crimean War (Young 1995).

The depiction of syncope or “fainting” in art and film conjure up images of beautiful young women who conveniently “swoon” into the eager arms of bystanders. This characterization has perhaps contributed many of the current myths about syncope, namely, that it is insignificant, and usually found in emotional, weak, grieving, or “hysterical” young females (Dixon 1995). On the contrary, syncope occurs in males and females, the young and old, and in all racial groups. Syncope is anything but glamorous; in fact, the outcome can be dangerous, even lethal. Regarded as both a sign and a symptom, the wide spectrum of disorders that contribute to syncope range from the more serious, lethal, and often cardiac causes, to the less worrisome—but nonetheless troublesome—chronic, intermittent, and benign or self-limiting conditions.

Considering the misconceptions about syncope, the lack of clear understanding of the multiple etiologies associated with syncopal episodes, and the labor-intensive investigation into the potential causes of syncope, it is not surprising that many practitioners find the
topic truly overwhelming. Thus, this chapter is intended to provide the reader with a concise overview of the epidemiology and significance of syncope, the classification of syncope and the most common causes of syncope, the patient evaluation including the history, physical, and diagnostic testing, and guidelines for management of this complex medical problem.

**EPIDEMIOLOGY AND SIGNIFICANCE**

The frequency of syncope and its associated mortality varies with age, gender, and etiology (Olshansky 2005). Estimates of syncope in the general population suggest that anywhere from 20 to 40% of adults will experience a syncopal episode some time in their life (Manolis et al. 1990; Kenny and Kapoor 2003; Chen et al. 2006). In the United States each year, over 1 million individuals are evaluated and treated for recurrent syncope (Grubb and Kosinski 2002; Olshansky 2005). As well, experts explain that the numbers are a conservative calculation, likely secondary to underreporting by patients. Usually, these patients are believed to suffer from isolated, self-limiting episodes of neurocardiogenic syncope (NCS), or an autonomic nervous system (ANS) cause (Olshansky 2005). Data from the Framingham Study demonstrate that approximately 3% of adult men and 3.5% of women experience syncope, with over twice the frequency (5.6%) in individuals older than 65 years of age (Savage et al. 1985). In the same study, the incidence of syncope was greater in younger females, but somewhat higher in elderly males (Soteriades et al. 2002).

Approximately 3–5% of all emergency room visits are due to syncope, and of these patients, 35% are admitted to the hospital (Day et al. 1982). In addition, 1–6% of all hospital admissions are for syncope as a primary diagnosis (Kapoor 1990). The costs associated with recurrent syncope are staggering; in the United States alone, it is estimated that evaluations for syncope and treatment are over $750 million dollars a year (Olshansky 2005). The data on an average cost per diagnosis of an undirected syncope evaluation were reported by one study to be $16,000 a year (Calkins et al. 1993), yet in another study, depending on the diagnostic studies ordered, the cost per syncope diagnosis was estimated to be as high as $78,000 (Olshansky 2005). Estimated US hospital costs associated with syncope exceed $10 billion annually (Kenny and Kapoor 2003).

Without doubt, the consequences from syncope can be devastating. This is especially true in the elderly, who have the highest incidence of syncope. Falls secondary to syncope result in a substantial burden to the individual, families, health care, and society. In adults, 10% of falls are due to syncope, and 50% of falls in the elderly are attributed to syncope (Cambell et al. 1981). Even so, this estimate is apt to be much higher due to the changing demographics of an aging population. In the United States, an older adult is treated in the emergency room every 18 seconds due to a fall (Centers for Disease Control and Prevention, National Center for Injury Prevention and Control 2008). Over one-third of adults age 65 and older fall each year, and among these adults, falls are the leading cause of nonfatal and fatal injuries (Centers for Disease Control and Prevention, National Center for Injury Prevention and Control 2006; Stevens 2006). In a study of 52 community living adults aged 70 years and older, 40% of the subjects reported falling in a 12-month period (Hausdorff et al. 2001). Falls are the leading cause of traumatic brain injury in adults age 65 and older, and the occurrence of fall-related deaths has significantly increased in the last decade (Stevens 2006).

While controversial, researchers believe there is a considerable “amount of overlap” between falls and syncope in the elderly (Shaw and Kenny 1997; Parry and Kenny 2005). It is hard to determine the cause of falls in the elderly due to problems with recall of the event and lack of an eyewitness account of the event.
who suffer from epilepsy (Santhouse et al. 2007), lung disease, rheumatoid arthritis, chronic back pain, and heart failure (Barón-Esquivias et al. 2003).

Syncope commonly occurs in children and adolescents, and this group may be especially susceptible to altered family, school, and peer relationships (Grubb and Friedman 2005; Rollinson 2005). No studies have specifically addressed quality of life in children and adolescents with syncope, but it is likely that this group is not immune from its detrimental effects. The syndromes associated with recurrent syncope and near syncope in pediatric patients may be comparable to other life-altering, chronic medical disorders, such as pediatric cancer. It is known that pediatric cancer patient survivors suffer significant impairment in quality of life (Eiser 2007; Maurice-Stam et al. 2007) and have been found to experience posttraumatic stress syndrome (PTSS) in young adulthood (Lee et al. 2007; Rourke et al. 2007). Postural tachycardia syndrome (POTS), a condition that results in recurrent syncope, is a syndrome frequently encountered in adolescents (Grubb 2008). In one study of adults with POTS (mean age 34.2 years), the participants demonstrated significant impairment in physical functioning, role functioning, bodily pain, general health, vitality, and social functioning when compared with healthy controls and similar to quality of life reported in patients with chronic obstructive pulmonary disease and congestive heart failure (Benrud-Larson et al. 2002).

CLASSIFICATION OF SYNCOPE

Syncope may result from a multitude of conditions that require a steadfast approach and diagnostic acumen. The quest to delineate between benign and lethal causes of syncope often instills a considerable amount of anxiety in even the most experienced clinician, the patient, and the family. When no clear etiology
is found, most clinicians order a battery of unnecessary diagnostic tests that are expensive, pose a risk for the patient, are usually unrewarding, and generally do not increase diagnostic yield (Calkins et al. 1993; Olshansky 2005). Given this, the clinician must be prepared to accept a certain amount of ambiguity in dealing with these patients and must adopt an open mind to other possible diagnoses should the original diagnosis not fit. Often it is necessary to revisit the patient history and family history for clues, reconsider secondary contributing causes, and perhaps treat based on clinical judgment and a “leap of faith” (Grubb and Kosinski 2002). As one can imagine, patients without a clearly identified cause for syncope experience great psychological distress, and during this time patients and families need a considerable amount of support and guidance.

Any phenomena that disrupt cerebral blood flow for 8–10 seconds or cause a significant drop in mean cerebral arterial blood pressure below 40mm Hg can result in syncope. The reticular activating system in the brain stem is responsible for alertness, body posture, and reflex vasomotor and cardiovagal mechanisms; accordingly, the reduction of cerebral blood flow to the reticular activating system in the brain stem results in the loss of consciousness, which in most cases is usually transient (Kaufmann 2004).

To make things easier, researchers have separated the daunting list of differential diagnoses for syncope into three primary groups, including cardiovascular, noncardiovascular, and syncope of unknown origin. This grouping provides direction for the diagnostic workup, management, and prognostic significance (Table 4.1.1).

Other researchers have further separated the three major groups into common and uncommon causes of syncope, and cause of syncope per age group (Grubb and Kosinski 2002; Olshansky 2005) (Tables 4.1.2–4.1.4).

Table 4.1.1 Classification of syncope.

### Various causes and types of syncope

<table>
<thead>
<tr>
<th>Cardiovascular</th>
<th>Noncardiovascular</th>
</tr>
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<tbody>
<tr>
<td>Arrhythmias</td>
<td>Reflex mechanisms</td>
</tr>
<tr>
<td>AV block with bradycardia</td>
<td>Neurocardiogenic</td>
</tr>
<tr>
<td>Sinus bradycardia</td>
<td>Micturition</td>
</tr>
<tr>
<td>VT</td>
<td>Defecation</td>
</tr>
<tr>
<td>Nonarrhythmic</td>
<td>Cough</td>
</tr>
<tr>
<td>HCM</td>
<td>Deglutiition</td>
</tr>
<tr>
<td>AS</td>
<td>Postprandial</td>
</tr>
<tr>
<td></td>
<td>OH</td>
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<tr>
<td></td>
<td>Dysautonomia</td>
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<tr>
<td></td>
<td>Fluid depletion</td>
</tr>
<tr>
<td></td>
<td>Bed rest, debilitation</td>
</tr>
<tr>
<td></td>
<td>Drugs</td>
</tr>
<tr>
<td></td>
<td>Psychogenic</td>
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<tr>
<td></td>
<td>Conversion reactions</td>
</tr>
<tr>
<td></td>
<td>Hysteria</td>
</tr>
<tr>
<td></td>
<td>Panic/anxiety disorder</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Conditions that can be mistaken for syncope</th>
</tr>
</thead>
<tbody>
<tr>
<td>Epileptic seizures</td>
</tr>
<tr>
<td>Hypoglycemia</td>
</tr>
<tr>
<td>Stroke</td>
</tr>
<tr>
<td>Drug-induced (alcohol, narcotics)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Syncope of unknown origin</th>
</tr>
</thead>
</table>
| There is a considerable amount of discrepancy in the number of patients with syncope attributed to an unknown cause. Initial studies suggest that syncope related to an unknown cause was as high as 40–50%, but a greater understanding of autonomic disorders and new diagnostic modalities such as the head-upright tilt-table test (HUTT) and implantable loop recorders (ILRs) have reduced this esti-
### Table 4.1.2 Common causes of syncope.

<table>
<thead>
<tr>
<th>Cardiovascular disease</th>
<th>Noncardiovascular disease</th>
<th>Other</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arrhythmic</td>
<td>Reflex mechanisms</td>
<td>Syncope of unknown origin</td>
</tr>
<tr>
<td>AV block with bradycardia</td>
<td>Vasodepressor “neurocardiogenic”</td>
<td>About 50% of all syncope patients</td>
</tr>
<tr>
<td>Sinus pauses/bradycardia</td>
<td>Micturition</td>
<td>Undiagnosed seizures</td>
</tr>
<tr>
<td>VT and structural heart disease</td>
<td>Deglutition</td>
<td>Improperly diagnosed syncope</td>
</tr>
<tr>
<td>Nonarrhythmic—hemodynamic</td>
<td>OH</td>
<td>Confusional states due to hypoglycemia, stroke, etc.</td>
</tr>
<tr>
<td>HCM</td>
<td>Dysautonomia</td>
<td>Drug induced</td>
</tr>
<tr>
<td>AS</td>
<td>Fluid depletion</td>
<td>Alcohol</td>
</tr>
<tr>
<td></td>
<td>Illness, bed rest</td>
<td>Illicit drugs</td>
</tr>
<tr>
<td></td>
<td>Drugs</td>
<td>Prescribed drugs (especially the elderly)</td>
</tr>
<tr>
<td></td>
<td>Psychogenic</td>
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<tr>
<td></td>
<td>Hysterical</td>
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<td></td>
<td>Panic disorder</td>
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<td></td>
<td>Anxiety disorder</td>
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</tbody>
</table>

### Table 4.1.3 Uncommon causes for syncope.

<table>
<thead>
<tr>
<th>Cardiovascular disease</th>
<th>Noncardiovascular disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arrhythmic etiology</td>
<td>Reflexes</td>
</tr>
<tr>
<td>SVT</td>
<td>Posttussive</td>
</tr>
<tr>
<td>Long QT interval syndrome</td>
<td>Defecation</td>
</tr>
<tr>
<td>Brugada syndrome</td>
<td>Glossopharyngeal</td>
</tr>
<tr>
<td>Arrhythmogenic right ventricular cardiomyopathy</td>
<td>Postprandial</td>
</tr>
<tr>
<td>Idiopathic VT(s)</td>
<td>CSH</td>
</tr>
<tr>
<td>MI (and bradycardia/tachycardia)</td>
<td>Hyperventilation</td>
</tr>
<tr>
<td>Nonarrhythmic etiology</td>
<td>Migraine</td>
</tr>
<tr>
<td>Pulmonary embolus</td>
<td>Carcinoid syndrome</td>
</tr>
<tr>
<td>Pulmonary hypertension</td>
<td>Systemic mastocytosis</td>
</tr>
<tr>
<td>Dissecting aortic aneurysm</td>
<td>Metabolic</td>
</tr>
<tr>
<td>Subclavian steal</td>
<td>Hypoglycemia</td>
</tr>
<tr>
<td>Atrial myxoma</td>
<td>Hypoxia</td>
</tr>
<tr>
<td>Cardiac tamponade</td>
<td>Multivessel</td>
</tr>
</tbody>
</table>

### Table 4.1.4 Causes of syncope by age.

- **Young patients (<35 years)**
  - Neurocardiogenic
  - Psychiatric
  - Situational
  - Epileptic seizures*
  - Long QT syndrome*
  - HCM*
  - SVT*
- **Midlife (35–65 years)**
  - Neurocardiogenic
  - Cardiac arrhythmias
- **Older patient**
  - Cardiac
  - Hemodynamic
  - Arrhythmic
  - Dysautonomic (OH)
  - Drug-induced
  - Multifactorial
  - Reflex syncope
  - CSH
  - Micturition/defecation
  - Neurocardiogenic

*Important but less common causes.
Cardiac conditions that may present with syncope include ventricular tachycardia (VT) and ventricular fibrillation (VF) in the setting of postmyocardial infarction (MI), heart block, HCM, dilated cardiomyopathy, long QT syndrome, arrhythmogenic right ventricular dysplasia (ARVD), and anomalous coronary arteries.

Understandably, one of the most vital tasks of the clinician when assessing syncope is to determine if the heart is structurally normal. Indeed, the significance of the patient history and physical (H & P) cannot be understated, because syncope in the setting of heart disease is associated with higher mortality. Not unexpectedly, a cardiovascular cause for syncope is more common in the elderly, due to the higher incidence of cardiovascular disease. In patients presenting with syncope, if heart disease is suspected or diagnosed, the syncope is more likely to be related to cardiac causes. One study of patients with syncope (n = 341) found that in patients with suspected or confirmed cardiac disease, 34% of the cases were found to have syncope related to cardiac causes, compared with 3% without suspected or diagnosed cardiac disease. Thus, after the initial evaluation of syncope, confirmed of suspected heart disease is a significant predictor of a cardiac cause for syncope (Alboni et al. 2001).

Cardiovascular Causes of Syncope

It is noted that in patients with syncope, 10–30% will have a cardiac cause (Day et al. 1982; Soteriades et al. 2002; Olshansky 2005). This group has the highest rate of death, reported as high as 20–30% (Grubb and Kosinski 2002). Syncope associated with sudden death was first recognized as far back as Hippocrates when he observed, “those who suffer from frequent and severe fainting often die suddenly” (Hippocrates Aphorisms 2.41 as reported in Grubb and Olshansky 2005). In the United States alone, there are 400,000 sudden deaths annually, and in 45% of the cases, the only warning was syncope. Sudden cardiac death (SCD) in athletes occurs every 3 days, or a total of 100 deaths per year in the United States (Maron et al. 2006).

Although sudden death in athletes is not common, many of the tragic, highly publicized deaths in athletes symbolize the important need to differentiate among cardiac and non-cardiac causes of syncope. Several retrospective studies of sudden death in pediatric patients and young athletes have found a significant number who experienced syncope prior to their death (Maron 1998, 2003; Hedrich et al. 2005). Hypertrophic cardiomyopathy (HCM) is the most common cardiovascular cause of sudden death in young athletes, and in one study, more common in African Americans (52%) and uncommon in females (3%) (Maron et al. 2006). In view of this, syncope during exercise or exertion and associated with comorbid underlying heart disease is a poor prognostic indicator in children and adults, and may foreshadow SCD (Hedrich et al. 2005). The most frequent lethal cardiac conditions that may present with syncope include ventricular tachycardia (VT) and ventricular fibrillation (VF) in the setting of postmyocardial infarction (MI), heart block, HCM, dilated cardiomyopathy, long QT syndrome, arrhythmogenic right ventricular dysplasia (ARVD), and anomalous coronary arteries.

Understandably, one of the most vital tasks of the clinician when assessing syncope is to determine if the heart is structurally normal. Indeed, the significance of the patient history and physical (H & P) cannot be understated, because syncope in the setting of heart disease is associated with higher mortality. Not unexpectedly, a cardiovascular cause for syncope is more common in the elderly, due to the higher incidence of cardiovascular disease. In patients presenting with syncope, if heart disease is suspected or diagnosed, the syncope is more likely to be related to cardiac causes. One study of patients with syncope (n = 341) found that in patients with suspected or confirmed cardiac disease, 34% of the cases were found to have syncope related to cardiac causes, compared with 3% without suspected or diagnosed cardiac disease. Thus, after the initial evaluation of syncope, confirmed of suspected heart disease is a significant predictor of a cardiac cause for syncope (Alboni et al. 2001). Cardiovascular causes of syncope can be divided into arrhythmic and nonarrhythmic etiology. Although a brief description follows, a more detailed discussion of the cardiovascular disorders can be found elsewhere in this book.

Dysrhythmias and Syncope

Arrhythmogenic causes of syncope such as tachyarrhythmias and bradyarrhythmias are commonly known to result in syncope and may be benign or lethal. In all patients with syncope, tachyarrhythmias account for 25% of the cases, and VT is the primary underlying rhythm 85% of the time (Pelosi and Morady 2005). The
significantly clinical characteristics that predicted the rhythm of severe bradycardia or asystole included syncope associated with convulsions, loss of consciousness ≥10 minutes, and no prodrome or aura preceding the event; palpitations, however, did not predict bradycardia and/or asystole (Kanjwal et al. 2009). Palpitations are a nonspecific sign of a cardiac rhythm disturbance as a cause for syncope. However, if the palpitations precede a syncope episode, they are more likely to represent a rhythm disturbance (Olshansky 2005). In adults and children, SVT, especially AV nodal reentry tachycardia (ANRT), rarely leads to syncope. On the other hand, if syncope occurs during peak exercise, this pattern is commonly seen in VT or SVT (Grubb and Friedman 2005).

### Table 4.1.5 Conditions that warrant more detailed evaluation of syncope.

| 1. | Syncope that occurs during peak exertion |
| 2. | Seizure/convulsive activity |
| 3. | Recurrent syncope (more than two to three times) |
| 4. | Chest pain that precedes syncope |
| 5. | Syncope resulting in significant injury |
| 6. | Abnormal cardiac examination |
| 7. | Repaired or palliated structural disease |
| 8. | Abnormal cardiovascular examination |
| 9. | Family history of sudden death |

Although the middle-age patients and the elderly have a greater incidence of rhythm disturbances as a cause for syncope and subsequent increase in risk of mortality, there are several arrhythmias associated with syncope in the pediatric patient that are nonetheless significant. In children and adolescents, congenital heart disease is the primary cause of rhythm disturbances (Grubb and Friedman 2005), and syncope in this setting should be explored further. Less common, but potentially life-threatening rhythm disturbances that may cause syncope in the pediatric patient include long QT syndrome, Brugada syndrome, Wolf–Parkinson–White (WPW) syndrome and other supraventricular tachycardias (SVTs) (Olshansky 2005). In pediatric patients with syncope, conditions that warrant a more detailed exam are found in Table 4.1.5.

In patients with recurrent syncope of unknown origin, prolonged monitoring of heart rate with ILRs has demonstrated that bradycardia is more common than tachycardia (Inamdar et al. 2006). In a small study (n = 19), researchers looked at predictors of asystole and bradycardia responses as a cause of syncope in patients using ILR monitoring. Major findings based on this preliminary study note that the

### Nonarrhythmic Cardiac Causes of Syncope

Of the nonarrhythmic cardiac causes of syncope, two of the more common and important conditions are aortic stenosis (AS) and HCM.

AS is an obstructive cause of syncope in both children (congenital) and adults (rheumatic and senile or degenerative). The disorder may progress to aortic valve malformation and dysfunction, and subsequently a reduction in cardiac output. In the majority of children with congenital AS, the disorder goes undetected throughout childhood without evidence of cardiac compromise or developmental delay.
(Grubb and Friedman 2005). Over time, the diseased valve thickens and may cause an obstruction to cardiac outflow. These children may experience exertional syncope, which signals advanced disease and a high risk for sudden death (Braunwald et al. 1963). In adults, particularly the elderly, the aortic valve may undergo degenerative changes that result in sclerosis, calcification, and fibrosis. Senile or degenerative AS is the most frequent nonarrhythmic cardiac cause of syncope in the elderly and the frequency increases with advancing age (Grubb and Kanjwal 2005). Like children with AS, many adults may be asymptomatic; however, 25% may experience syncope with exertion, exposure to heat (vasodilation), or rapid position changes (Grubb and Kanjwal 2005).

HCM is another nonarrhythmic cardiac condition that may result in syncope; in fact, 15–20% of adult patients with HCM experience syncope. HCM is a heterogeneous, inherited group of cardiac disorders that result in gene mutation and misregulation of the proteins that code for the cardiac sarcomere, the muscle unit responsible for myocardial contractility (Nishimura and Holmes 2004). Myocyte derangement and fibrosis cause massive left ventricular hypertrophy (LVH) that may lead to diastolic dysfunction, myocardial ischemia, arrhythmias (primarily VT and atrial fibrillation), left ventricular tract outflow obstruction, and decreased cardiac output (Nishimura and Holmes 2004). Patients with HCM may live long asymptomatic lives; experience dyspnea, chest pain, syncope, and chronic heart failure symptoms; or suffer SCD (Maron et al. 1995; Nishimura and Holmes 2004). As stated, HCM is the primary cause of sudden death in children, young adults, and competitive athletes (Maron 2002; Maron et al. 2006).

Although pediatric patients with HCM experience syncope less often than adults, when present, it is an ominous sign (Grubb and Friedman 2005). Major and minor risk factors for SCD from HCM include previous cardiac arrest with VF, sustained VT, family medical history of sudden death, unexplained syncope, left ventricular wall thickness >30 mm, and hypotensive response to exercise (please refer to Nishimura and Holmes 2004 for a more detailed discussion). Syncope, in patients with HCM, can occur with or without exertion and requires a further diagnostic workup (Grubb and Kanjwal 2005).

**Noncardiac Causes of Syncope**

Unlike cardiac causes for syncope, in the patient who has a noncardiac-related cause for syncope, the death rate ranges from 1 to 6% over a 1- to 2-year period (Grubb and Kosinski 2002). Fortunately, noncardiac causes of syncope contribute to the bulk of the diagnoses, and although benign, patients with recurrent syncope may experience a number of physical, social, interpersonal, and emotional difficulties (Grubb 2007). Several of the noncardiac causes of syncope (e.g., seizure, hypoglycemia, drug reactions, stroke) are actually other conditions that can be mistaken for syncope and will not be addressed here. However, the majority of noncardiac causes of syncope appear to stem from difficulty with regulation of the ANS. One of the primary functions of the ANS is to control heart rate and blood pressure, especially in response to upright posture or gravitational stress and postural changes (Hamill and Shapiro 2004). Patients with disorders of the ANS experience transient disturbances in autonomic function that may result in hypotension, diminished cerebral perfusion, and ultimately, loss of consciousness or syncope (Fouad-Tarazi 2004; Grubb 2005).

During the last few decades or so, our knowledge of disorders associated with the ANS has greatly expanded (Grubb 2005). This exciting new era of research has led to a greater appreciation of symptom complexes that were previously not well understood, and to
MECHANISMS OF BLOOD PRESSURE CONTROL

The human nervous system is divided into two intricately connected systems, namely the central nervous system (includes the brain and spinal cord) and the peripheral nervous system. The peripheral nervous system can be further subdivided into the ANS and the somatic nervous system. The ANS is the conduit between the internal and external environment, and “has the daunting task of ensuring the survival and procreation of the species” (Hamill and Shapiro 2004, p. 20). Indeed, the ANS is responsible for bodily functions essential for life itself including heart rate, respiratory and blood pressure regulation, thermoregulatory control, gastrointestinal function and motility, genitourinary function, sexual function, metabolic...
and endocrine function, and adaptation to stress (Hamill and Shapiro 2004; Grubb 2005).

The ANS includes the sympathetic nervous system (SNS), parasympathetic nervous system (PNS), and enteric divisions. The organization of the SNS is such that it allows for rapid integration and processing of stimuli to produce a physiological response, hence, the “fight or flight.” The SNS is responsible for involuntary activities that permit energy expenditure, whereas the PNS, another system not under conscious or voluntary control, is responsible for the conservation of energy, or the “rest and digest” system. The enteric system is concerned with gastrointestinal tract functioning (Camilleri 2004). The primary target organs of the SNS, PNS, and the enteric system include the visceral smooth muscles of the heart, lungs, gastrointestinal and genitourinary organs, endocrine glands, arterioles, and skeletal muscle afferents involved in autonomic reflexes. The amalgamation and synchronization of the vast neuronal groups of the sympathetic and parasympathetic systems serve to regulate the majority of body functions in such a way that a constant internal balance or “homeostasis” is achieved.

One of the critical, primal functions of the ANS is to counteract the gravitational effects of the change in blood volume that occurs when an individual assumes an upright posture. The coordinated short-term and intermediate response of the ANS to a shift in blood volume is essential for adequate cerebral perfusion, and any alteration in the responses may result in orthostasis, near syncope, or syncope (Grubb et al. 2008). In the supine position, almost 30% of the blood volume lies in the thorax. When upright, gravity displaces approximately 500–800 cc of blood downward to the abdomen and lower extremities (especially the splanchnic and cutaneous venous beds). The decrease in central blood volume leads to a decrease in venous return to the heart and an ensuing reduction in stroke volume, perhaps as much as 40% (Grubb et al. 2008). The lower stroke volume and preload activates the high-pressure receptor cells (carotid and aortic/sinus arch) and low-pressure mechanoreceptors in the heart and lungs, which relay impulses via afferents to the central autonomic control center. This central awareness creates an increase in sympathetic efferent outflow and decrease in vagal tone that allow for an instantaneous increase in peripheral vasoconstriction (mainly venoconstriction), heart rate, and contractility (Grubb et al. 2008). While an individual continues to remain upright, the long-term hydrostatic effects on the vasculature result in an increase in extracellular fluid volume and an ensuing activation of the neurohumoral mechanisms of the renin-angiotension system (Fouad-Tarazi 2004). The following discussion will focus on the distinct syndromes of OI under autonomic control, but researchers now know there is considerable overlap among NCS, POTS, and pure autonomic failure (PAF) as shown in Figure 4.1.2.

**ANS DISORDERS OF OI**

**Reflex Syncopes**

The reflex syncopes include NCS, carotid sinus hypersensitivity (CSH), and the situational syncope. Germane to these disorders is the
interprets the massive influx of impulses as hypotension; subsequently, the neuronal reflex efferent response of vasodilation and bradycardia follows, resulting in syncope. Other not well-understood triggers include pain, fear, and strong emotions, and in some, the source of provocation is undetermined (Grubb 2005).

NCS is a common cause of syncope in younger individuals and is usually characterized by a distinctive prodrome of lightheadedness, pallor, dizziness, nausea, diaphoresis, and visual disturbances (Kaufmann 2004; Grubb 2005). The loss of consciousness is usually brief, and recovery is short without postictal confusion. A supine position results in restored cerebral perfusion and return to consciousness. Most patients with NCS experience infrequent or intermittent episodes but unfortunately some experience recurrent syncope (Raj and Robertson 2004). Although NCS is considered a benign cause of syncope, in patients with little prodrome or warning, a lone syncopal episode can have disastrous consequences (fractures, head trauma, auto accidents, work-related accidents). Patients with NCS are usually asymptomatic between episodes and rarely exhibit other signs of autonomic compromise (temperature intolerance, gastrointestinal difficulties, urinary difficulties, and altered sweat patterns) (Kaufmann 2004). Thus, unlike patients with other autonomic disorders, the ANS appears to be spared despite the “peculiar,” transient disruption of normal autonomic baroreflex responses (Grubb 2005).

CSH

As noted, CSH frequently results in syncope, and like NCS, the exact primary abnormality has yet to be elucidated (Parry and Kenny 2005). The prevalence of CSH increases with age, and is more common in males with comorbid coronary artery disease and hypertension (Parry and Kenny 2005). Conditions that precipitate syncope include compression on the carotid sinus, tight collars, sudden head turning, and shaving. Other sources of provocation
include prolonged standing, meals, and certain vagal maneuvers. The prodome consists of dizziness and lightheadedness, and is usually much more sudden than NCS. As a consequence, the lack of warning may result in significant trauma from falls (Parry and Kenny 2005).

**Autonomic Failure Syndromes**

Autonomic failure syndromes encompass a group of disorders that range from the mild (yet significant) POTS, to the chronic, degenerative autonomic failure syndromes including PAF and the more serious multiple system atrophy (MSA). These disorders of OI share similar symptoms, but are nonetheless distinctive syndromes of dysautonomia (defined as ANS dysfunction that results in impairment). One of the primary pathophysiological mechanisms that relate to syncope common to all of the disorders is the impairment of autonomic peripheral nerve functioning that leads to excessive venous pooling. In addition, these syndromes also share global disturbances in autonomic nerve functioning and, in most people, lead to daily symptoms of autonomic compromise. Like other chronic multisystem disorders such as diabetes and heart failure, patients with dysautonomia present with a constellation of subjective (but nonetheless debilitating) symptoms and daily functional impairment that can be extremely taxing not only for the patient, but also for families and caregivers. Thus, these patients require a comprehensive, holistic approach to management that addresses physical, emotional, social, and spiritual needs.

**POTS**

Over a decade ago, autonomic researchers began to describe a syndrome of OI that did not behave like classic OH or NCS (Hoeldtke and David 1991; Schondorf and Low 1993; Khurana 1995; Low et al. 1995; Grubb et al. 1997; Karas et al. 2000). Indeed, these patients appeared to have global autonomic difficulties like other autonomic disorders, but most striking was the marked upright postural tachycardia and only modest decrease in blood pressure. As well, these patients exhibited severe OI, defined as the provocation of symptoms upright, primarily relieved by recumbence (Low et al. 1995; Grubb et al. 1997). Symptoms of OI include debilitating exercise tolerance, extreme fatigue, lightheadedness, cognitive impairment, anxiety, tremulousness, near syncope, and syncope (Grubb 2008).

This syndrome was later named POTS and is believed to be a heterogeneous group of disorders with similar characteristics (Grubb 2008). POTS is currently defined as the presence of symptoms of OI associated with a heart rate increase of 30 bpm (or a rate that exceeds 120 bpm) that occurs within the first 10 minutes of standing or upright tilt, not associated with other chronic debilitating conditions known to reduce vascular tone (Grubb 2008). Aside from the symptoms of OI, many POTS patients experience profound disruption in daily functioning and are unable to attend to even basic daily tasks. As a result, these patients are often labeled as having conversion disorders, psychogenic illness, anxiety, and depression (Grubb 2008).

Although there are a variety of primary and secondary forms of POTS, in the majority of the syndromes the patients appear to suffer from a failure of the peripheral vasculature to maintain adequate vascular resistance in the face of orthostatic stress (Grubb 2008). Secondary POTS describes conditions and disease states that alter peripheral autonomic nerve function including chronic diabetes mellitus, amyloidosis, sarcoidosis, and autoimmune diseases, to name a few. The majority of patients with primary POTS are women (5:1), and the onset frequently occurs after viral illness (Low 2004), infections, pregnancy, surgeries, trauma, and sepsis; furthermore, many researchers believe POTS may be autoimmune in nature (Low 2004; Grubb 2008).
In POTS, when an individual stands, this results in a greater than normal degree of blood pooling in the dependant areas of the body, that is, the legs, lower arms, and mesenteric vasculature. The decrease in peripheral vascular resistance (PVR) results in a compensatory upright tachycardia. Over time, the venous pooling exceeds the compensatory heart rate increase, and this leads to a decline in venous return. A domino effect then ensues, and subsequently the heart can only pump what it receives; the low cardiac output results in low cerebral perfusion and ultimately near syncope and syncope. POTS physiology, then, can be comparable to heart failure. As a basic recall, poor heart function decreases contractility, which results in a decrease in cardiac output. In heart failure, the increase in PVR is one of the hallmark compensatory mechanisms that ensure adequate cerebral and vital organ perfusion (Ledoux et al. 2003). In POTS, PVR is decreased, with venous pooling; hence, the compensation mechanism is the hallmark persistent sinus tachycardia, with heart rates while standing that may approach 160 bpm. Often these patients are confused with inappropriate sinus tachycardia and undergo radiofrequency ablation; thus, this demonstrates how important it is to recognize the syndrome (Grubb and Kosinski 2002). One of the simplest diagnostic tests used to discern POTS is to obtain the blood pressure and heart rate lying, sitting, immediately standing, and then 2, 5, and 10 minutes upright (Grubb and Karabin 2008). Any heart rate increase 30 bpm above baseline, as described previously, may be diagnostic (the patient should be free of medications that would blunt the heart rate response such as beta blockers, antidepressants, hypertensive medications). A more extensive review of POTS can be found elsewhere (Grubb 2008).

**PAF and MSA**

Patients with PAF and MSA exhibit a progressive, degenerative form of ANS dysfunction (dysautonomia) that manifests as chronic OH, which can be dramatic and severe enough to result in syncope. As well, these patients have a generalized state of autonomic failure resulting in disturbed bowel, bladder, sudomotor, and sexual functioning (Grubb et al. 2008). Current researches have postulated that these syndromes may result from degeneration of the peripheral autonomic nerves (Grubb and Kosinski 2002). A significant distinction between PAF and MSA is the apparent lack of somatic nerve involvement in PAF. Although the two are relatively difficult to distinguish initially due to the overlap of symptoms, MSA is much more progressive and severe, and as the disorder progresses, somatic nervous system involvement increases, as do defects in the central nervous system (Grubb 2008). They both share common clinical characteristics of vague orthostatic weakness, lightheadedness, fatigue, dry mouth, early satiety and bloating, constipation, inability to sweat, temperature intolerance, erectile dysfunction, diminished libido, urinary incontinence, and urinary retention (symptoms usually passed off as insignificant by clinicians) (Kaufmann and Schatz 2004). In MSA, unlike PAF, patients may have sensory, cerebellar, extrapyramidal deficits with gait disturbance, motor disturbances, tremor, rigidity, and rapid eye movement-associated behavior disorder (RBD) (Quinn 2004). Both PAF and MSA tend to occur after the fifth decade of life, and occur in males twice as frequently as females (Grubb 2008).

In patients with PAF and MSA, one of the most debilitating symptoms is OH and is often the reason patients seek medical attention. OH is the result of autonomic neurocirculatory dysfunction in the absence of any underlying cause (decreased blood volume, bed rest, medications) (Grubb et al. 2008). Although OH is classically defined as the drop in systolic blood pressure > 20 mm Hg or drop in diastolic blood pressure > 10 mm Hg while upright 2–3 minutes, in many patients the fall in blood pressure is much more gradual, may be less dramatic, yet
Without question, a detailed H&P examination in a patient with syncope is essential to help elucidate the cause. No specific battery of tests is ever indicated, and the history should guide the diagnostic workup. A variety of clinical algorithms have been proposed, which may be confusing (Olshansky 2005). The most critical place to start, as mentioned, is to determine if heart disease exists, and a brief algorithm can be found in Figure 4.1.3.

The history must address patient age, previous cardiovascular conditions, any previous episodes of syncope and medical records if available, comorbid conditions, medications, supplements, and illicit or recreational drug

### Diagnostic Evaluation of Syncope

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Outcome</th>
<th>Next Step</th>
</tr>
</thead>
<tbody>
<tr>
<td>History</td>
<td>Abnormal</td>
<td>Further testing of treatment dependent upon data obtained</td>
</tr>
<tr>
<td>Physical exam</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Electrocardiogram</td>
<td>Normal</td>
<td>Further testing or treatment dependent on data obtained</td>
</tr>
<tr>
<td>Electrocardiography</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Stress testing</td>
<td></td>
<td></td>
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<tr>
<td>Tilt-table testing</td>
<td></td>
<td></td>
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<tr>
<td>Event recorder</td>
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</table>

In "select" patients proceed with EPS, coronary arteriography, or prolonged ambulatory monitoring

**Figure 4.1.3** Diagnostic evaluation of syncope.
use. Questions should be directed to information about recent infections, febrile illness, surgeries, trauma, menarche, childhood growth and development, adolescent growth spurt, pregnancy and lactation, menopause, change in cognitive functioning, and recent travel. Family history should include information on history of syncope, seizures, autoimmune disease, congenital heart disease, arrhythmias, cardiovascular disease, unexplained sudden death, and sudden infant death syndrome (SIDS). Also vital to the examination is information related to the review of systems, especially related to autonomic function (neurological, cardiovascular, gastrointestinal, temperature regulation, sweat patterns, urinary function, sexual function). Important pieces of history are summarized in Table 4.1.6.

Characteristics of the syncopal episode include number and frequency of the episodes (new onset, isolated, intermittent, recurrent, changing pattern in established syncope, and last episode); associated triggers (prolonged standing and sitting, heat, shower/bathing, migraine or other headache, menstrual cycle, emotions, immobility, change in exercise pattern, cough, swallowing, micturition, defecation, blood draw, fright, and during sleep, sexual intercourse, or swimming); relationship to body position, exercise, posture, and exertion (standing, sitting, driving, work-related, rapid head movement, shaving, peak exercise, postexercise, and other exertion); relationship to meals, alcohol, medications (postprandial, early morning, new medication, dose increase, medication withdrawal, e.g., selective serotonin reuptake inhibitor [SSRI], selective norepinephrine reuptake inhibitor [SNRI], central stimulants, and hypertensive medications); prodromal symptoms (length of prodrome, visual disturbances, dizziness, lightheadedness, anxiety, confusion, tremulousness, cognitive impairment, headache, “coat hanger” neck pain, muffled hearing, palpitations, exercise intolerance, weakness, fatigue, pallor, clamminess, flushing, sweating, paresthesia, nausea, diarrhea, cough, shortness of breath, and chest pain); presence or absence of convulsive movements (consider convulsive syncope in light of negative neurological workup); length of syncopal episodes (brief < few seconds; short 1–3 minutes; prolonged > 10 minutes); and postsyncopal symptoms (transient confusion, rapid recovery, full return to previous activity, prolonged confusion, fatigue, amnesia, headache, diarrhea, vomiting, and any associated trauma).

It is important to remember that many autonomic conditions other than NCS cause daily symptoms of OI, and not frank syncope. Indeed, in POTS, only 50% of the patients experience

<table>
<thead>
<tr>
<th>Table 4.1.6 History: important data to obtain.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Witnesses</td>
</tr>
<tr>
<td>Situation</td>
</tr>
<tr>
<td>Age elderly (&gt;65 years)</td>
</tr>
<tr>
<td>Age young (&lt;40 years)</td>
</tr>
<tr>
<td>Heart disease</td>
</tr>
<tr>
<td>Family history of sudden death</td>
</tr>
<tr>
<td>Number of episodes</td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td>Previous evaluation</td>
</tr>
<tr>
<td>Medications</td>
</tr>
</tbody>
</table>
Cardiac Arrhythmia Management

Sympotm management should be considered first, because although autonomic failure syndromes occur primarily in the elderly, this cohort is also at higher risk for cardiovascular disease. Symptoms related to syncopal episodes are summarized in Table 4.1.7.

The physical examination may guide diagnosis, and should highlight the vital signs, cardiovascular exam, and the neurological exam. Vital signs including supine and upright heart rate and blood pressure (as detailed earlier) are important. The cardiovascular exam, including the presence of neck vein distention, pulses, murmurs, gallops, bruits,

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Probable cause</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nausea, diaphoresis, fear</td>
<td>Neurocardiogenic</td>
</tr>
<tr>
<td>Aura</td>
<td>Seizure</td>
</tr>
<tr>
<td>Palpitations</td>
<td>Tachycardia (nonspecific finding)</td>
</tr>
<tr>
<td>Exercise-related</td>
<td>VT or SVT, hypotension/bradycardia</td>
</tr>
<tr>
<td>Posture-related</td>
<td>OH, volume depletion, dysautonomia</td>
</tr>
<tr>
<td>Urination, defecation, eating, coughing</td>
<td>Vagal-induced hypotension, bradycardia</td>
</tr>
<tr>
<td>Diarrhea, vomiting</td>
<td>Hypovolemia, hypokalemic-induced arrhythmia, vagal-induced hypotension, bradycardia</td>
</tr>
<tr>
<td>Melena</td>
<td>Gastrointestinal bleed</td>
</tr>
<tr>
<td>Visual change, neurological abnormality</td>
<td>Stroke (unlikely presentation), seizure, migraine</td>
</tr>
<tr>
<td>Headaches</td>
<td>Migraine, intracerebral bleed</td>
</tr>
<tr>
<td>Chest pain</td>
<td>Ischemia-induced arrhythmia</td>
</tr>
<tr>
<td>Dyspnea</td>
<td>Pulmonary embolus, pneumothorax, hyperventilation (hysteria)</td>
</tr>
<tr>
<td>Abdominal pain</td>
<td>Aortic aneurysm, gastrointestinal bleed, peritonitis acute abdomen, trauma</td>
</tr>
<tr>
<td>Back pain</td>
<td>Dissecting aneurysm, trauma</td>
</tr>
<tr>
<td>Flushing</td>
<td>Carcinoid syndrome</td>
</tr>
<tr>
<td>Prolonged syncope</td>
<td>AS, seizure, neurological or metabolic cause</td>
</tr>
<tr>
<td>Slow recovery</td>
<td>Seizure, drug, ethanol intoxication, hypoglycemia, sepsis</td>
</tr>
<tr>
<td>Injury</td>
<td>Arrhythmia, cardiac cause, neurocardiogenic</td>
</tr>
<tr>
<td>Confusion</td>
<td>Stroke, transient ischemic attack, intoxication, hypoglycemia</td>
</tr>
<tr>
<td>Prolonged weakness</td>
<td>NCS</td>
</tr>
<tr>
<td>Skin color</td>
<td>Pallor—neurocardiogenic; blue—cardiac; red—carbon monoxide</td>
</tr>
</tbody>
</table>
and peripheral edema, is important. The presence of tremor; altered reflexes; rigidity; gait and balance disturbance; and sensory, temperature, and pain deficits should be noted. Skin color changes, pallor, acral cyanosis (mottled bluish extremities in dependent position due to peripheral venous pooling), skin hyperextensibility (suggestive of joint hypermobility syndrome), and rashes may aid in diagnosis. Other physical findings are found in Table 4.1.8.

**Diagnostic Testing**

**Electrocardiogram**

Most clinicians agree that the electrocardiogram (EKG) should be a routine test ordered when a patient presents with syncope (Grubb and Kosinski 2002; Olshansky 2005). The diagnostic yield is not completely known, but some reports suggest anywhere from 5 to 10% (Olshansky 2005). In the pediatric patient, look for evidence of ventricular pre-excitation, delta waves (WPW), long QT interval, accelerated AV conduction or block, epsilon waves (ARVD), or HCM. In the older patient, evaluate for evidence of coronary artery disease (old Q waves, and new ST-T changes), conduction disturbances, atrial fibrillation, and hypertensive heart disease (LVH).

**Echocardiogram**

Based on the H&P and EKG findings, the next test that may provide diagnostic clues is the echocardiogram. In children and adults, the ability to assess right and left ventricular size and function (HCM, right ventricular dysplasia), cardiac valves (AS), and other structural abnormalities may be diagnostic. Although the diagnostic yield is reported to be low, consensus agrees that it is reasonable for clinicians to obtain nonurgent echocardiograms based on clinical judgment derived from the H&P (Grubb and Kosinski 2002).

**Table 4.1.8** Physical findings: key points.

<table>
<thead>
<tr>
<th>Finding</th>
<th>Implication</th>
</tr>
</thead>
<tbody>
<tr>
<td>Heart rate—slow, fast</td>
<td>Arrhythmic cause for syncope, acute illness, gastrointestinal bleed</td>
</tr>
<tr>
<td>Respiration rate—slow, fast</td>
<td>Hyper/hypoventilation, pneumothorax, heart failure</td>
</tr>
<tr>
<td>Carotid massage</td>
<td>Carotid hypersensitivity</td>
</tr>
<tr>
<td>Blood pressure</td>
<td>OH, drug-induced hypotension, volume depletion</td>
</tr>
<tr>
<td>Neck vein distension</td>
<td>Pulmonary embolus, congestive failure, cardiac causes</td>
</tr>
<tr>
<td>Skin pallor</td>
<td>Blood loss, neurocardiogenic cause</td>
</tr>
<tr>
<td>Carotid bruits</td>
<td>Concomitant heart disease. Unlikely, primary cause for syncope</td>
</tr>
<tr>
<td>Heart murmur</td>
<td>Obstructive or other cardiac syncope</td>
</tr>
<tr>
<td>Left ventricular lift</td>
<td>Heart failure with cardiac syncope</td>
</tr>
<tr>
<td>S3 gallop</td>
<td>Heart failure with cardiac syncope</td>
</tr>
<tr>
<td>Rash</td>
<td>Anaphylaxis causing syncope</td>
</tr>
<tr>
<td>Abdominal tenderness</td>
<td>Blood loss, or hypotensive cause for syncope</td>
</tr>
<tr>
<td>Absent or variable pulses</td>
<td>Dissecting aneurysm, subclavian steal</td>
</tr>
<tr>
<td>Neurological findings</td>
<td>Seizure, stroke, transient ischemic attack</td>
</tr>
<tr>
<td>Stool guaiac</td>
<td>Blood loss</td>
</tr>
</tbody>
</table>
Exercise Stress Testing

In some, ischemia may be suspected as the cause of syncope, and thus stress testing may be appropriate. Other patients that should be considered for exercise stress testing include syncope occurrence during exercise or exertion. Exercise-induced NCS is reported by several researchers to be the leading cause of syncope associated with exercise (Sakaguchi et al. 1995; Grubb and Kosinski 1996; Kosinski et al. 1996, 2000). A number of worrisome cardiac conditions that may be discovered on exercise stress testing are postexercise asystole, long QT syndrome, postexercise hypotension, ischemia, and nonischemic exercise-induced arrhythmias, and anomalous coronary artery distribution (Kosinski et al. 2000; Grubb and Kosinski 2002).

Holter Monitors/Event Recorders/ILRs

Holter monitoring is generally not useful in syncope unless the patient has daily recurrent episodes. It is highly unlikely that the symptom–rhythm correlation would be captured, and several large studies have documented the correlation of rhythm disturbance to syncope using Holter monitoring was less than 5% (Olshansky 2008). Interestingly, DiMarco and Philbrick (1990) reviewed a large number of studies of Holter monitors in patients with syncope and found that 4–30% of the significant arrhythmias found on monitoring did not correlate with symptoms, and symptom rhythm correlation occurred in only 22% of the patients. Event recorders (external loop recorders) provide extended monitoring (30 days), and in studies using both Holter and event monitoring, the diagnostic yield is less than 40% (Gibson and Heitzman 1984; Zimetbaum and Josephson 1999).

In contrast to external monitoring devices, ILRs have emerged as a very useful diagnostic tool used to assess for arrhythmias associated with syncope. These devices have a higher diagnostic yield than external monitors, and allow for long-term monitoring (up to 3-year battery life) (Krahm et al. 2005). ILRs have made possible the detection of infrequent, elusive arrhythmias associated with syncope that most likely would have previously been undetected. As noted (see arrhythmia and syncope) thus far, only one study has looked at clinical characteristics that may be indicated for implantation, which include prolonged loss of consciousness (>10 minutes), no prodrome (a condition that is usually associated with trauma), and convulsive movements (Kanjwal et al. 2009). Most units now have the capacity to allow for home monitoring; thus, patients may download episodes without the need for an office visit. The devices are small (about the size of a flash drive stick) and are implanted in the subcutaneous tissue in the left precordium under local anesthesia (Krahm et al. 2005). The procedure time and recovery time are negligible, and the complication rate (infection) is low. We experienced one patient who developed an allergic skin reaction to the metal employed; thus, patients should be screened for nickel or metal allergy.

Laboratory Tests

In some instances, laboratory tests can reveal the cause of syncope, although routine blood tests are not always necessary or fruitful (Olshansky 2008). As with other diagnostic tests, the blood work should be guided by the H&P. The complete blood count (CBC) may detect anemia or acute blood loss, electrolyte panel may reveal hydration, and a chemistry panel, thyroid stimulating hormone (TSH), serum drug levels, and pregnancy test may also be useful. Other more specific blood work and diagnostic testing for autonomic function are beyond the scope of this chapter, and may be found elsewhere.

HUTT

HUTT is a well-established diagnostic test used by clinicians to assess patients who present
with syncope (Benditt et al. 1996). One of the early beginnings of HUTT use was by the National Aeronautics and Space Administration (NASA) over 50 years ago to assess the effects of prolonged weightlessness on orthostatic tolerance during upright posture of astronauts in space flights, and it emerged in cardiology in the late 1980s to investigate syncope (NASA 1999; Barón-Esquivias et al. 2003). When an individual stands and gravity displaces blood downward, the venous system expands to hold the excess blood volume. Reflex activation of the sympathetic system results in an increase in PVR and a concurrent increase in skeletal muscle tone. The skeletal muscle pump is an essential component in the maintenance of upright posture, as it aids venous return to the heart and ultimately the cerebral circulation (Brignole 2005). HUTT was designed to simulate the effects of upright posture and orthostatic stress on an individual by inhibition of the skeletal muscle pump in the dependent extremities. When an individual is tilted upright and venous pooling begins, the normal compensatory increase in skeletal muscle contraction does not occur. Individuals with excessive venous pooling (e.g., POTS) or who have difficulty with PVR (e.g., peripheral autonomic neuropathy), or are sensitive to diminished venous return (e.g., NCS), may have difficulty maintaining upright posture on HUTT because of the heavy reliance on the skeletal muscle pump. Thus, the tilt may reproduce patient symptoms and may demonstrate changes in heart rate and blood pressure response, depending on the syndrome.

There are a number of HUTT protocols proposed, but the primary outcome is to induce venous pooling using passive upright position, or provocation with vasodilators such as isoproterenol, nitroglycerin, or adenosine, and to inhibit the innate skeletal muscle pump. The reader is referred to a more extensive discussion elsewhere (see Benditt et al. 1996; Brignole 2005). Other commonly employed tests during HUTT assess for CSH with carotid massage and to differentiate between seizure disorder and convulsive syncope using tilt-induced electroencephalograph (EEG). HUTT results are highly reproducible, and the diagnostic yield of HUTT ranges from 60 to 70%, with a specificity greater than 85–90% (Benditt et al. 1996; Barón-Esquivias et al. 2003). There are very few side effects and the complication rate is low (Barón-Esquivias et al. 2003; Brignole 2005).

Indications for HUTT have been well detailed by consensus guidelines lead by Benditt et al. (1996) from the American College of Cardiology, and may be referred to for an in-depth discussion. Briefly, from the guidelines, conditions that warrant HUTT are recurrent syncope or a single episode in a high-risk patient; no evidence of structural heart disease; structural heart disease only if other causes have been excluded by diagnostic testing; part of the evaluation for exercise-induced syncope; and further evaluation in patients for which a syncopal cause has been delineated (e.g., asystole), but where the susceptibility to NCS would guide treatment. Conversely, conditions in which HUTT are not warranted are single syncopal episode without injury and not in a high-risk setting; and syncope with identified cause and identification of NCS would not alter treatment. Finally, conditions with relative contraindications to HUTT are syncope with severe ventricular outflow obstruction; critical mitral stenosis; critical known coronary artery stenosis; and known critical cerebral vascular stenosis.

Responses to HUTT are beyond the scope of this chapter; however, interested readers can refer to Grubb and Olshansky (2005) for further details.

**EPS**

Depending on the underlying cardiac condition, the EPS may be employed in select patients with syncope. Guidelines from the American College of Cardiology report that EPS is indicated (class I) for patients with syncope and underlying cardiac disease in which the cause
of syncope remains unexplained despite an appropriate initial evaluation (Zipes et al. 1995). In unexplained syncope without structural heart disease, the guidelines are less defined, and class II guidelines provide EPS for patients with recurrent syncope without structural heart disease with negative HUTT (Zipes et al. 1995).

**TREATMENT**

**Conservative Treatment**

As with many conditions, treatment will depend on the subtype or cause of syncope; accordingly, any reversible conditions that contribute should be addressed (anemia, volume loss, medications).

If a secondary cause for syncope is discovered (sarcoidosis, neoplasm), the appropriate referrals should be made. Aggravating factors or triggers should be identified and avoided if possible (e.g., dehydration, alcohol, heat, hot baths/showers, prolonged standing/sitting). Fluid intake should approach 2 L daily, and if no underlying hypertension, salt intake may be increased to 3–5 g daily. All patients should slowly start a reconditioning program, and should work up to at least 20–30 minutes, 3–4 days weekly. Resistance training to help build the skeletal muscle pump is important. Patients normally do well with guided aquatic therapy, physical therapy, or cardiac rehabilitation, especially if other medical conditions, safety, or motivation is an issue. Bilateral waist-high compression stockings at least 30–40 mmHg counterpressure is recommended, but may be cumbersome for the elderly and heat-intolerant individuals. Sleeping with the head upright 6–12 inches may help, as does biofeedback. Patients should all be cautioned on driving, and in most states, any loss of consciousness will restrict driving privileges for a period of time, and the reader is directed to the Web site www.aamva.org.

One of the greatest challenges for clinicians who work with patients who have recurrent syncope and chronic autonomic disorders is dealing with family, education, and employment concerns. The clinician will devote a tremendous amount of time and energy to help patients with these issues. By far, these issues can be much more troubling for the patient than the symptoms alone. Often the syndromes are acquired, or not present from birth and can profoundly disrupt or “shatter” the world of an individual. As well, most patients do not look ill; in fact, many of the syndromes can be considered “hidden” disabilities. Thus, because they are hidden, these patients often face discrimination and skepticism by some educators and other health care providers that ironically, should help them the most. Diminished cognitive function, memory impairment, headaches, and nausea—the daily symptoms of OI—limit participation considerably. Altered employment can be devastating for families, and at least 25% of patients may be disabled. Children and adolescents with these syndromes may not be able to participate fully in the educational system, and reduced school schedules are common. It goes without saying that these syndromes dramatically change family, peer, and social relationships. As a consequence, the patient may be at a high risk for depression and maladaptive coping skills. We recommend all of our patients and families to see a mental health counselor, preferably one with experience in chronic illness. One organization that is particularly helpful for children and adolescents with dysautonomia can be found online at www.dynakids.org.

**Pharmacotherapy**

Often conservative measures may be all that is necessary to return an individual to functioning; however, pharmacotherapy may be needed and is tailored to the individual patient based on cause of syncope and underlying comorbid conditions. In general, patients with NCS and autonomic causes for syncope respond well to fludrocortisone, a mineralcorticoid that
causes sodium and fluid retention, which results in expanded blood volume (Scott et al. 1995), and increases vasoconstriction by sensitizing the blood vessels to norepinephrine. It may cause potassium and magnesium depletion; thus, we monitor these levels periodically. In some patients, hair loss, peripheral edema, acne, and depression have been noted (Calkins 1999). Beta blockers have been reported to be effective in several studies, although randomized studies did not show a benefit in NCS. In our practice, we mainly employ beta blockers in the hyperadrenergic form of POTS (Grubb 2005).

In many of the disorders, individuals have difficulty with peripheral vasoconstriction; thus, the agent midodrine has been successfully employed (Perez-Lugones et al. 2001). Midodrine is a peripherally acting alpha agonist that increases vasoconstriction. It has virtually no cardiac effects, and is relatively safe, but extreme caution must be employed when using in patients with underlying hypertension. It has a short half-life and must be given every 4–6 hours. Side effects include scalp paresthesia (not harmful) and supine hypertension; thus, the last dose should not be given 4 hours before bedtime.

Animal and human studies have demonstrated that serotonin (5-hydroxytryptamine) plays an essential role in the central autonomic regulation of heart rate and blood pressure, and some autonomic researchers postulate that patients with dysautonomia have altered central serotonin levels. Selective serotonin reuptake inhibitors are effective in NCS and OH (Grubb et al. 1997; Girolamo et al. 1999).

A more extensive review of medications for postural tachycardia can be found in Grubb (2008); NCS in Grubb (2005); and OH/dysautonomia in Grubb et al. (2008).

**CONCLUSION**

Unraveling the mystery of syncope and its potential causes requires meticulous attention to the H&P. The yield of diagnostic studies increases with a well thought, individualized patient approach, and thus tests should be used sparingly. Cardiac causes of syncope, although less frequent, are associated with significant mortality. Even syncope due to a benign cause may be debilitating, cause trauma, and negatively impact an individual’s quality of life. Understandably, patients with recurrent syncope and their families require our patience, understanding, and advocacy. As our knowledge of autonomic disorders increases, patients who previously may have had their syncope attributed to “anxiety,” “psychogenic,” or “unknown” cause may be given new hope.

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**Case 4.1.1**

**Presentation of Case**

Mr. T is a 65-year-old male with a history of diabetes mellitus, tobacco abuse, and family history of premature coronary artery disease who presented with 3 days of intermittent chest pain. Initial ECG demonstrated sinus rhythm with ST elevations in V1 through V4 and Q waves inferiorly. He was given aspirin and metoprolol, and the cardiac catheterization laboratory was called in. He subsequently developed sustained polymorphic VT and lost consciousness. He was promptly resuscitated with an external 200J biphasic shock.

Cardiac catheterization demonstrated a 100% proximal left anterior descending artery (LAD), 50% middle part of the left circumflex artery (mLCx), and 100% midright coronary artery (RCA) disease with a left ventricular ejection fraction of 20%. He underwent angioplasty and stenting of the proximal LAD. He was admitted to the coronary care unit (CCU) and then had an uneventful hospital course. He was started on an aspirin, plavix, beta blocker, angiotension-converting enzyme (ACE) inhibitor, and statin. He was also referred for
cardiac rehabilitation and smoking cessation counseling. Is Mr. T a candidate for implantable cardioverter defibrillator (ICD) implantation?

**Discussion**

On presentation, Mr. T has many of the risk factors for ventricular tachyarrhythmias and SCD. He has coronary artery disease and has just suffered an MI with evidence of a possible previous MI with a chronically occluded RCA. His ejection fraction is severely reduced, and he suffered a cardiac arrest with documented polymorphic VT. However, during the acute phase of an MI and ischemia, there are substantial, reversible electrophysiological changes that can lead to ventricular arrhythmias. After the infarction or ischemia is addressed and/or healed, and his heart disease is treated with appropriate medications, the risks of subsequent ventricular arrhythmias can be substantially reduced. This is due to remodeling or healing of the ventricular myocardium with reperfusion of the heart with intervention and aggressive medical therapy. The DINAMIT study looked at the utility of ICD implantation early after MI when severe left ventricular dysfunction was documented. All cause mortality was not reduced with ICD implantation.

Current guidelines recommend reassessing his ejection fraction in 40 days after an MI. If his ejection fraction remains severely reduced, the scars related to his infarctions and coronary artery disease place him at risk for SCD as per the MADIT II criteria. In the intervening period between the infarction and 40 days, he may be at risk for ventricular arrhythmias, and he should be counseled to be aware of symptoms of dangerous arrhythmias such as palpitations, dizziness, and syncope. If he develops these symptoms and/or has documented repetitive nonsustained VT, an electrophysiologist should evaluate him as soon as possible.

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**Case 4.1.2**

**Presentation of Case**

Mr. X is a 27-year-old male without a significant past medical history who presented after an episode of syncope. His wife heard a thump after he walked up a flight of stairs with a laundry basket. She found him unresponsive and emergency medical services (EMS) was called. EMS found him in sinus rhythm with a blood pressure of 100/70. He spontaneously regained consciousness without complaint. Retrospectively, he has had intermittent palpitations, sometimes associated with dizziness. Evaluation included a normal EKG (Fig. 4.1.4), normal echocardiogram, and exercise stress test. Telemetry documented one burst of polymorphic VT in the middle of the night. Electrophysiological testing was performed. Normal baseline conduction was documented and testing did not demonstrate sustained VT. Procainamide infusion was then performed. EKGs before and after procainamide infusion are shown in Figure 4.1.5. Downsloping ST elevations in V1 and V2 were demonstrated. Mr. X was given the diagnosis of Brugada syndrome and an ICD was implanted.

**Discussion**

Brugada syndrome is a genetic defect of the sodium channel that can lead to electrical instability, polymorphic VT, and SCD. The patient has a structurally normal heart, and life-threatening arrhythmias are often the first sign of the disease. Diagnosis is made purely by an EKG with either spontaneous changes in V1 and V2 or changes unmasked by sodium channel blocking drugs, such as procainamide, flecainide, or amjaline. Though electrophysiology testing may provide some additional information, symptoms of arrhythmia or family history in patients with documented Brugada EKG provide the most prognostic value. Mr. X’s symptoms of syncope and symptomatic palpitations indicate that he is at high risk for SCD and ICD implantation is warranted.

Genetic electrophysiological defects often occur in structurally normal hearts and are diagnosed by EKG changes or arrhythmias documented on rhythm monitors. It is crucial that changes such as a long QT interval or ST changes consistent with Brugada are recognized by all health care providers so that these patients are identified early and appropriately treated.
REFERENCES


associated with exercise, a manifestation of neurally mediated syncope. Am J Cardiol 75: 476–481.

RESOURCES
Sudden Cardiac Death

Steven C. Hao and Katie Morganti

INTRODUCTION

Sudden cardiac death (SCD), defined by the World Health Organization as an unexpected death due to cardiac causes occurring within 1 hour of symptom onset, is a major medical and public health issue. “Sudden cardiac death (SCD) causes approximately 450,000 deaths annually in the United States and nearly 50% of all cardiovascular mortality worldwide” (Goldberger and Lampert 2006). More people die from SCD than AIDS, breast cancer, and lung cancer combined (Fig. 4.2.1).

Advances in cardiopulmonary resuscitation (CPR), external defibrillation, and emergency medical services have provided lifesaving techniques to provide a bridge to hospitalization and ultimate survival. However, despite these advances, the chance for survival to hospital discharge has been shown to be as low as 4% (Dorian et al. 2002). In addition, SCD is often the first sign or symptom of cardiac disease. Thus, identification of patients at risk for life-threatening arrhythmias, before their first event, is crucial to impact the incidence and mortality associated with SCD. The development of the implantable cardioverter defibrillator (ICD) and appropriate implementation in patients at risk has been a critical piece in saving lives.

The goals of this chapter are to summarize the information regarding the etiology and risk factors for SCD, the landmark trials in the treatment of SCD, as well as the current guidelines regarding ICD implantation.

ETIOLOGIES OF SCD

SCD most often occurs in the setting of underlying structural heart disease, namely coronary artery disease (CAD) or heart failure (HF) (Fig. 4.2.2). It is important to note, however, that in addition to the underlying etiologies discussed below, there can be an acute inciting event, either ischemia, electrolyte imbalance,
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CAD and scarring of the myocardium following a myocardial infarction (MI) may predispose the heart to electrical conduction abnormalities. Electrophysiological changes due to ischemia are attributable to alteration of both conduction and repolarization patterns that provide the environment for reentrant arrhythmias, specifically ventricular tachycardia (VT).

Studies reviewing autopsy results of victims of SCD report that a recent occlusive coronary thrombus is present in up to 95% of patients (Reddy and Ruskin 2005). Thus, appropriate, aggressive management of risk factors for CAD, including diabetes mellitus, hypertension, hyperlipidemia, and smoking, has a crucial role in impacting the incidence of SCD.

### Cardiomyopathy

Cardiomyopathy, a weakening of the heart muscle or disorder of cardiac muscle structure, is an important risk factor for SCD. Most cardiomyopathies are due to a MI, but cardiomyopathies due to a nonischemic, dilated cardiomyopathy (DCM), or hypertrophic cardiomyopathy (HCM) account for approximately 15% of all SCD events (Myerburg et al. 2004). Fibrosis or myofibrillar disarray related to these disorders provides the substrate for ventricular arrhythmias. HCM is an inherited disease that leads to idiopathic left ventricular (LV) hypertrophy, which can be an independent risk factor for SCD. DCM is often idiopathic but can be related to myocarditis, alcohol, toxins, or rarely a genetic cause. Diffuse fibrosis associated with DCM predisposes patients to both arrhythmias and pump failure (Newton and Newton-Church 2008).

The greater extent of myocardial dysfunction, likely related to greater fibrosis, is related to a higher risk of SCD. Moderate to severe reduction in ejection fraction (EF) holds the higher risk. In addition to LV dysfunction, functional capacity, as assessed by the New York Heart Association (NYHA) scale, has also shown to have prognostic value in identifying potentially proarrhythmic medications, or autonomic nervous system dysfunction that precipitates SCD.

### CAD

An overwhelming majority of SCD events, approximately 75–80%, are attributable to underlying CAD. Ischemia from multivessel CAD and scarring of the myocardium following a myocardial infarction (MI) may predispose the heart to electrical conduction abnormalities. Electrophysiological changes due to ischemia are attributable to alteration of both conduction and repolarization patterns that provide the environment for reentrant arrhythmias, specifically ventricular tachycardia (VT).

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those at highest risk for SCD. Functional class III and IV patients have been identified as having a higher mortality risk.

**Primary Electrophysiological Disorders**

The remaining 5% of SCD events are due to a primary electrophysiological defect, most commonly the channelopathies, which include long QT syndrome (LQTS), short QT syndrome, and Brugada syndrome. These genetic disorders are associated with a high incidence of malignant ventricular arrhythmias and SCD in patients with otherwise normal hearts. Although many genotypic and phenotypic differences exist between LQTS, short QT syndrome, and Brugada syndrome, these genetic disorders result in mutations of the ion channel(s), which can contribute to life-threatening arrhythmias. LQTS causes early afterdepolarizations (EADs) that initiate torsades de pointes (TdP), whereas Brugada syndrome causes a dispersion of repolarization that leads to phase 2 reentrant arrhythmias, such as VT (Brugada et al. 2004).

In addition to the channelopathies, other genetic defects such as catecholaminergic polymorphic ventricular tachycardia (CPVT), arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C), and Wolf–Parkinson–White (WPW) syndrome may lead to SCD. Patients with CPVT have a defect in calcium handling within the cell and ARVD/C patients have issues with cell-to-cell adhesion. Both defects can lead to ventricular arrhythmias and SCD. Rapid anterograde conduction down an accessory pathway in WPW with atrial fibrillation can result in very fast ventricular rates and degenerate into ventricular fibrillation.

**MECHANISMS OF SCD**

Irrespective of the underlying etiology, the primary mechanism of SCD is a ventricular tachyarrhythmia—either VT or VF—that quickly leads to hemodynamic collapse (Fig. 4.2.3). Other mechanisms include TdP, bradycardia, or asystole. Data that have been collected from Holter monitors reveal that VT is the most common inceptive arrhythmia (Reddy and Ruskin 2005). The relatively high probability that the initial rhythm is VT or VF ultimately impacts the success of lifesaving antitachycardia strategies, including ICDs and automatic external defibrillators (AEDs).

**ICD**

The invention and early development of the ICD by Michael Mirowski were crucial, paradigm-shifting milestones in affecting the impact of SCD. Initial human implants were rudimentary, large devices that need to be implanted surgically in the abdomen with direct placement of patches on the epicardial surface of the heart. Implantation was associated with a high mortality and the devices had limited utility, only shock boxes with short life spans. Subsequent development led to defibrillating leads that could be placed endovascularly and smaller devices that could be placed pectorally (Fig. 4.2.4). These advances led to safer implants, and further evolution added additional utility such as the ability to also act as a pacemaker, painless antitachycardia pacing for termination of VT, and resynchronization therapy.
REVIEW OF THE LITERATURE

Multiple trials have definitively demonstrated the efficacy of the ICD decreasing mortality associated with SCD and defined the patients who benefit from ICDs. The results from trials such as the Antiarrhythmics versus Implantable Defibrillators (AVID) Trial, the Multicenter Automatic Defibrillator Implantation Trial (MADIT), the Multicenter Automatic Defibrillator Implantation Trial II (MADIT II), and the Sudden Cardiac Death in Heart Failure Trial (SCD-HeFT) addressed patients who were at the highest risk for SCD (Table 4.2.1). These patients were deemed to be at the highest risk for SCD, having survived an SCD event or severe cardiomyopathy associated with CAD and/or HF. The current guideline recommendations for selecting patients eligible for ICD implant are based largely on the entry criteria of these landmark trials.

Initially, ICDs were exclusively used for the secondary prevention of SCD, that is, patients who had been successfully resuscitated from a previous SCD event. The AVID Trial was one of the earliest studies to evaluate the efficacy of ICD therapy in the prevention of SCD. The study compared the impact of ICD therapy with antiarrhythmic drug therapy on overall

Table 4.2.1 Randomized controlled trials of ICDs in SCD.

<table>
<thead>
<tr>
<th>Study</th>
<th>Patient population</th>
<th>Randomization</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>AVID</td>
<td>SCD survivors or sustained VT; LVEF ≤40%</td>
<td>ICD versus amiodarone or sotalol</td>
<td>33% RRR; P &lt; 0.02</td>
</tr>
<tr>
<td>MADIT</td>
<td>MI; LVEF ≤35%; NSVT; inducible MMVT</td>
<td>ICD versus amiodarone or sotalol</td>
<td>54% RRR; P &lt; 0.009</td>
</tr>
<tr>
<td>MADIT II</td>
<td>MI; LVEF ≤30%</td>
<td>ICD versus optimal medical Rx</td>
<td>31% RRR; P = 0.016</td>
</tr>
<tr>
<td>SCD-HeFT</td>
<td>NYHA II–III; LVEF ≤35%</td>
<td>Placebo versus amiodarone versus ICD</td>
<td>23% RRR (ICD vs. placebo); P = 0.007</td>
</tr>
</tbody>
</table>

AVID, Antiarrhythmics versus Implantable Defibrillators; MADIT, Multicenter Automatic Defibrillator Implantation Trial; MADIT II, Multicenter Automatic Defibrillator Implantation Trial II; SCD-HeFT, Sudden Cardiac Death in Heart Failure Trial; VT, ventricular tachycardia; NSVT, nonsustained ventricular tachycardia; LVEF, left ventricular EF; MI, myocardial infarction; NYHA, New York Heart Association congestive heart failure class; Rx, therapy; RRR, relative risk reduction.
The study findings highlight the superiority of ICD therapy over conventional therapy in reducing overall mortality in patients with a history of MI and cardiomyopathy.

The SCD-HeFT trial conducted by Bardy et al. (2005) was designed to evaluate the effectiveness of amiodarone therapy and/or ICD therapy, as compared with placebo, on decreasing overall mortality in a population of patients with moderate systolic congestive HF. Inclusion criteria consisted of NYHA class II or III congestive HF and an LVEF of 35% or less. All three groups were equally required to receive optimal medical therapy for HF consisting of an ACE inhibitor, beta blocker, aldosterone antagonist, statin, and aspirin when appropriate. Results exhibited a 23% decreased risk of overall mortality with ICD therapy ($P = 0.007$). Amiodarone, when used for primary prevention purposes, did not show any survival benefit relative to placebo.

**RISK STRATIFICATION**

The studies reviewed and other data support the use of ICDs to prevent SCD in patients deemed to be at highest risk. However, the vast majority of SCD occurs in patients who do not have diagnosed HF, CAD, or any symptoms of arrhythmia. It has been reported that 30% of all SCD events occur as the first clinical event of CAD and that furthermore, 33% of all SCD events occur in those patients considered to be at low risk (Myerburg et al. 2004).

The AVID trial was stopped early when analysis revealed that the difference in the primary end point of overall mortality in the two treatment groups had crossed the statistical boundary for early termination.

The MADIT was a primary prevention trial that evaluated the efficacy of ICDs versus standard medical therapy in patients with previous MI, LV dysfunction (LVEF ≤35%), spontaneous nonsustained VT (NSVT), and inducible of sustained VT during electrophysiological testing. The sample consisted of 196 patients throughout the United States and Europe who were followed for an average of 27 months. The study was terminated early when interim analysis demonstrated a substantial improvement in mortality of ICDs versus medical therapy primarily with beta blockers and amiodarone (54% reduction in all cause mortality, $P < 0.009$) (Moss et al. 1996).

The MADIT II conducted by Moss and colleagues (2002) sought to evaluate the potential survival benefit of prophylactic ICD implantation in patients with a prior MI and LV dysfunction, defined as an EF of 30% or less without the necessity of documenting NSVT or inducible sustained VT during electrophysiological testing. The sample consisted of 1,232 patients from 76 hospitals who met the inclusion criteria of age greater than 21 years, a history of MI 1 month or more prior to enrollment, and an EF of 30% or less within 3 months prior to entry. The primary end point was all cause mortality. Results revealed an overall mortality reduction of 31% in those patients who were treated with ICD therapy versus standard medical therapy.
autonomic balance. Hopefully, future studies will identify more patients at risk and provide more impact in the numbers of patients who die every year from SCD.

**CONCLUSION**

SCD is a major public health issue. The identification and thorough evaluation of those patients who may be at risk for SCD, and the appropriate implementation of the guidelines in order that all patients at risk receive optimal therapy, is critical to reduce the rate of SCD. The evidence presented highlights the paramount importance of rapid resuscitation with defibrillation in preventing SCD, and provides compelling evidence that ICDs improve survival in this patient population. This evidence has led to guidelines that clearly identify appropriate risk stratification and implantation of ICDs. Future research continues to search for better methods of risk stratification to provide appropriate therapy for a larger population at risk.

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RESOURCES

Section 5
Care of the Pediatric Arrhythmia Patient
Children are different. The pediatric patient with a device presents many unique challenges related to age, size, and growth. The heart diseases and indications for device implantation differ from those of adult patients as well. Children enjoy playing and tend to be much more physically active than their adult counterparts with devices. For those of us who care for children, balancing their need to play with our goal to keep them safe can be difficult at times. Their level of cognitive development and psychosocial functioning must also be assessed and dealt with appropriately. Ideally, infants and young children with a device should be managed by health care professionals with training and experience in the specialty area of pediatrics. This chapter will address the specific challenges pertaining to the care of the pediatric patient with a pacemaker or an implantable cardioverter defibrillator (ICD). Indications for device therapy, implantation techniques, device selection, programming considerations, and specific issues regarding the care and follow-up of infants, children, and adolescents will be discussed.

**PACEMAKERS**

**Indications**

The indications for pacemaker implantation in children are similar to those for adults, but the underlying etiologies differ. The American College of Cardiology/American Heart Association Task Force on Practice Guidelines recently published updated indications for permanent pacing in children, adolescents, and patients with congenital heart disease (see Table 5.1.1) (Epstein et al. 2008). Basically, pacemakers are recommended for conduction disorders involving the atrioventricular (AV) node or sinus node. AV block may take the form of
Cardiac Arrhythmia Management

Table 5.1.1: Indications for permanent pacing in children and adolescents.

Class I—Permanent pacemaker implantation is indicated for:
1. Advanced second- or third-degree AV block associated with symptomatic bradycardia, ventricular dysfunction, or low cardiac output
2. Sinus node dysfunction with correlation of symptoms during age-inappropriate bradycardia
3. Postoperative advanced second- or third-degree AV block that is not expected to resolve or that persists at least 7 days after cardiac surgery
4. Congenital third-degree AV block with a wide QRS escape rhythm, complex ventricular ectopy, or ventricular dysfunction
5. Congenital third-degree AV block in the infant with a ventricular rate less than 55 bpm or with congenital heart disease and a ventricular rate less than 70 bpm

Class IIa—Permanent pacemaker implantation is reasonable for:
1. Congenital heart disease and sinus bradycardia (intrinsic or secondary to antiarrhythmic treatment) for the prevention of recurrent episodes of intra-atrial reentrant tachycardia. Congenital third-degree AV block beyond the first year of life with an average heart rate less than 50 bpm, abrupt pauses in ventricular rate that are 2–3 times the basic cycle length, or associated with symptoms due to chronotropic incompetence
2. Sinus bradycardia with complex congenital heart disease with a resting heart rate less than 40 bpm or pauses in ventricular rate longer than 3 seconds. Congenital heart disease and impaired hemodynamics due to sinus bradycardia or loss of AV synchrony. Unexplained syncope in the patient with prior congenital heart surgery complicated by transient complete heart block with residual fascicular block after a careful evaluation to exclude other causes of syncope

Class IIb—Permanent pacemaker implantation may be considered for:
1. Transient postoperative third-degree AV block that reverts to sinus rhythm with residual bifascicular block. Congenital third-degree AV block in asymptomatic children or adolescents with an acceptable rate, a narrow QRS complex, and normal ventricular function. Asymptomatic sinus bradycardia after biventricular repair of congenital heart disease with a resting heart rate less than 40 bpm or pauses in ventricular rate longer than 3 seconds.

Class III—Permanent pacemaker implantation is not indicated for:
1. Transient postoperative AV block with return of normal AV conduction in the otherwise asymptomatic patient. Asymptomatic bifascicular block with or without first-degree AV block after surgery for congenital heart disease in the absence of prior transient complete AV block
2. Asymptomatic type I second-degree AV block. Asymptomatic sinus bradycardia with the longest relative risk interval less than 3 seconds and a minimum heart rate more than 40 bpm.

Source: Epstein et al. (2008).

Fontan procedure for single ventricles, such as hypoplastic right heart or hypoplastic left heart syndromes. The atrial scarring associated with these procedures often creates a substrate for intra-atrial reentry tachycardia. This potential sequela needs to be considered when selecting and programming pacemakers for these patients.

Congenital complete AV block (CCAVB), an AV block that presents in the infant, or even in
most pediatric patients will require an implanted pacemaker indefinitely, many prefer to preserve venous access for later in life.

Some patients with congenital heart disease require an epicardial system because of their cardiovascular anatomy. Those with single ventricle physiology are, by convention, limited to an epicardial system. These patients undergo staged repairs including a Glenn shunt, in which the superior vena cava (SVC) is transected and anastomosed to the right pulmonary artery, followed by a Fontan procedure, tunneling the inferior vena cava up to the underside of the pulmonary artery. Thus, the vena cavae are connected directly to the pulmonary arteries, precluding venous access to the heart. Epicardial implantation is also recommended for those with residual intracardiac shunts or prosthetic tricuspid valves.

The surgical approach for epicardial systems may be via a sternotomy, thoracotomy, or subxiphoid incision. Epicardial leads are often implanted during concomitant cardiac surgery. Suture-on steroid-eluting leads, unipolar or bipolar, are employed most of the time. Typically, lead lengths of 15, 25, and 35 cm are used. The shorter leads are ideal for pediatric use, but even if still functional, may need to be replaced down the road as the child grows. When unipolar leads are employed, a parylene-coated generator is recommended to reduce the risk of muscle stimulation. Whether epicardial or transvenous leads are used, bipolar leads offer the advantage of minimizing the risk for phrenic or diaphragmatic stimulation or muscle twitching around the pacemaker pocket that may occur with unipolar lead systems.

**Implantation Techniques**

A major difference between pediatric and adult patients is the route of pacemaker implantation. In adults, the majority of pacing leads are implanted transvenously, whereas in the pediatric population, about half of the patients will have an epicardial system (Berul and Barrett 2001). Factors such as size, growth, and cardiovascular anatomy are considered when choosing the route of implantation.

An epicardial approach is commonly used in children weighing less than 10–20 kg (corresponding to an age of 2 years). With an epicardial system, the redundant loops of lead allow for somatic growth, barring any lead trapping by adhesions. In addition, considering that

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>N = 338</th>
</tr>
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<tbody>
<tr>
<td>Surgical or acquired AV block</td>
<td>125 (37%)</td>
</tr>
<tr>
<td>Sinus node dysfunction</td>
<td>100 (30%)</td>
</tr>
<tr>
<td>Congenital AV block</td>
<td>75 (22%)</td>
</tr>
<tr>
<td>2° Mobitz II AV block</td>
<td>29 (8%)</td>
</tr>
<tr>
<td>Long QT syndrome</td>
<td>4 (1%)</td>
</tr>
<tr>
<td>Conduction system disorder</td>
<td>3 (1%)</td>
</tr>
<tr>
<td>Syncope</td>
<td>3 (1%)</td>
</tr>
</tbody>
</table>

*Table 5.1.2 Distribution of diagnoses among pediatric pacemaker recipients at Lucile Packard Children’s Hospital at Stanford.*
Cardiac Arrhythmia Management

Although some innovative strategies to implant a transvenous system in infants have been reported (Gillette et al. 1992; Molina et al. 1995), concern exists over long-term consequences, such as venous thrombosis or fibrosis, resulting in vessel occlusion (Bar-Cohen et al. 2006). Newer, thinner transvenous leads have reduced these complications. Lead stretch, fracture, or dislodgement due to somatic growth may occur as well when leads are implanted prior to puberty (Fortescue et al. 2004). To avoid such complications, leads are usually implanted with a generous amount of slack. Some pediatric electrophysiologists advocate leaving a redundant loop of lead in the right atrium, calculating that roughly 10 mm of lead length are needed for each year of growth (Gheissare et al. 1991; Antretter et al. 2003). However, care must be taken to avoid ectopy or tricuspid regurgitation caused by the excess lead moving within the atrium or prolapsing across the AV valve. Lead fibrosis with adherence to the endocardium is a concern as well (Serwer and Law 2006).

Device Selection

There are no devices made specifically for children, presumably because pediatric implants represent only about 1% of all device implants (Zhan et al. 2008). Features that are desirable for pediatric implantation include small size and programmable high pacing rates. Unfortunately, some of the smallest pacemakers currently available are not capable of pacing at the high rates needed to provide physiological pacing. Also, smaller devices and faster rates come at the expense of battery longevity. While 7–10 years is often the predicted battery life for pacemaker generators, in young children 3–5 years is more realistic. Dual-chamber pacing is preferred over single-chamber ventricular pacing in children of all ages with AV block. However, because of size constraints, a single-chamber device may be employed initially with plans to upgrade later to a dual-chamber device after the child has grown further.
Programming Considerations

The mode of pacing is determined by the child's underlying rhythm defect and whether a single- or dual-chamber device is implanted. The base pacing rate needs to be appropriate for the child's age. In very young children with little stroke volume reserve, cardiac output is dependent on heart rate. Although a rate of 100–130 bpm is commonly used for babies, some neonates or critically ill postoperative infants may require a much higher rate. An example of a neonate with long QT syndrome who required atrial pacing at a high rate to overcome a functional 2:1 AV block is illustrated in Figure 5.1.1A,B. As the child ages, the base pacing rate is gradually decreased. In dual-chamber devices, the upper tracking rate should be high enough to avoid a Wenckebach or 2:1 block response during sinus tachycardia. Normally, an individual should be able to achieve a heart rate during exercise of 220 bpm minus their age in years. Therefore, in the setting of DDD pacing for AV block, a child under the age of 10 years ideally should be able to attain a heart rate of at least 210 bpm with 1:1 atrial tracking and ventricular pacing. Currently, only a few pacemaker models have upper rate limits exceeding 200 ppm. For children with single-chamber devices or chronotropic incompetence, rate responsive sensors may be used. However, these sensors were basically designed for adults. Because children differ not only in body mass, but also in the location of the pulse generator (abdomen vs. pectoral area) and the types of activities they perform, these sensors operate suboptimally, but can generate adequate heart rates for moderate exercise. A child's functioning sinus node is the best physiological sensor and should be used whenever possible.

In addition to using rate limits appropriate for pediatric patients, it is imperative that the individual programming the pacemaker be aware of the need to shorten the postventricular atrial refractory period (PVARP) to allow for a reasonable Wenckebach window between the upper rate limit (or maximum tracking rate) and the 2:1 block point. The 2:1 block point is calculated by dividing the sum of the AV delay plus the PVARP (total atrial refractory period [TARP]) into 60,000. Unfortunately, because of limited programmability, some children do reach their 2:1 block point during exercise, at which time they may become symptomatic and need to stop their activity. Along with the PVARP, the AV delay should be programmed shorter to accommodate faster heart rates. Activating a rate-adaptive AV delay is helpful in mimicking the physiological shortening of the PR interval that is normally seen with increasing heart rates and will allow a higher 2:1 block point to be attained.

Some older children and young adults with congenital heart disease, particularly those with transposition of the great arteries who have had a Mustard or Senning procedure, or those with a single ventricle following a Fontan procedure, are prone to develop atrial tachyarrhythmias. Mode switching may be a beneficial feature to turn on in these patients, but care must be taken to program the mode switch rate well above their sinus tachycardia rate to avoid inappropriate mode switching during exercise. Not all pacemakers offer the desired range of programmability.

As with adults, children with pacemakers should have serial pacing and sensing threshold measurements that direct how the outputs and sensitivities are programmed. Battery conservation is important, but programming to ensure an adequate safety margin takes precedence.

ICDS

Indications

With devices becoming smaller and easier to implant, the indications for ICD therapy in young patients are increasingly more liberal. Device size is rarely a contraindication any
Figure 5.1.1 A and B. ECG of a newborn with long QT syndrome. Notice every other P wave falling between the QRS and T wave resulting in a functional 2:1 block due to ventricular refractoriness. The infant was treated with an implanted pacemaker programmed to DDD mode at a base pacing rate of 150 ppm. The fast atrial pacing rate helped to shorten the QT interval enough to permit 1:1 AV conduction, as shown here in this pacemaker rhythm strip.
more. As with pacemakers, the indications for pediatric ICD implantation are similar to those for adults but are often related to different underlying pathologies. Aborted sudden cardiac death or symptomatic ventricular tachycardia (VT) clearly warrants ICD therapy. ICD implantation for primary prevention is increasingly more acceptable in vulnerable children. In 2008, the American College of Cardiology/American Heart Association Task Force on Practice Guidelines published updated indications for ICDs in pediatric patients and patients with congenital heart disease (see Table 5.1.3).

Many pediatric patients have ICDs for primary or secondary prevention following surgery for congenital heart disease, particularly tetralogy of Fallot and aortic stenosis. In these patients, prior VT or cardiac arrest merits ICD placement if there are no coexisting hemodynamic abnormalities amenable to further surgery. Because QRS duration greater than 180 ms has been shown to be a predictor of sudden cardiac death in patients following tetralogy of Fallot repair (Gatzoulis et al. 1995), ICD implantation might be considered for primary prevention in such individuals. Additionally, cardiac resynchronization with RV pacing may be beneficial in tetralogy of Fallot patients with right bundle branch block (Dubin et al. 2008).

Another large group of ICD candidates includes those with cardiomyopathy, mostly dilated or hypertrophic cardiomyopathy. The ICD is used as a bridge to transplant for many of these young patients.

Children with inherited arrhythmic syndromes, such as long QT syndrome, Brugada

<table>
<thead>
<tr>
<th>Table 5.1.3</th>
<th>Indications for ICDs in children and adolescents.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Class I—ICD implantation is indicated for:</td>
<td></td>
</tr>
<tr>
<td>1. Survivor of cardiac arrest after evaluation to define the cause of the event and to exclude any reversible causes</td>
<td></td>
</tr>
<tr>
<td>2. Patients with symptomatic sustained VT in association with congenital heart disease who have undergone hemodynamic and electrophysiological evaluation</td>
<td></td>
</tr>
<tr>
<td>Class IIa—ICD implantation is reasonable for:</td>
<td></td>
</tr>
<tr>
<td>1. Congenital heart disease with recurrent syncope of undetermined origin in the presence of either ventricular dysfunction or inducible ventricular arrhythmias at electrophysiological study</td>
<td></td>
</tr>
<tr>
<td>Class IIb—ICD implantation may be considered for:</td>
<td></td>
</tr>
<tr>
<td>1. Recurrent syncope associated with complex congenital heart disease and advanced systemic ventricular dysfunction when thorough invasive and noninvasive investigations have failed to define a cause</td>
<td></td>
</tr>
<tr>
<td>Class III—ICD implantation is not indicated for:</td>
<td></td>
</tr>
<tr>
<td>1. Patients who do not have a reasonable expectation of survival with an acceptable functional status for at least 1 year</td>
<td></td>
</tr>
<tr>
<td>2. Inncessant VT or VF</td>
<td></td>
</tr>
<tr>
<td>3. Patients with significant psychiatric illnesses that may be aggravated by device implantation</td>
<td></td>
</tr>
<tr>
<td>4. NYHA Class IV patients with drug-refractory congestive heart failure who are not candidates for cardiac transplantation or CRT-D</td>
<td></td>
</tr>
<tr>
<td>5. Syncope of undetermined cause in a patient without inducible ventricular tachyarrhythmias and without structural heart disease</td>
<td></td>
</tr>
<tr>
<td>6. VF or VT that is amenable to surgical or catheter ablation</td>
<td></td>
</tr>
<tr>
<td>7. Ventricular tachyarrhythmias due to a completely reversible disorder in the absence of structural heart disease</td>
<td></td>
</tr>
</tbody>
</table>

Source: Epstein et al. (2008).
syndrome, arrhythmogenic right ventricular dysplasia (ARVD), or catecholaminergic polymorphic ventricular tachycardia (CPVT), may receive ICDs for either primary or secondary prevention. ICD implantation is clearly indicated following cardiac arrest or documented VT. It is an increasingly more accepted practice to implant ICDs in first-degree relatives of these victims for primary prevention when genetic testing confirms the diagnosis (Table 5.1.4).

**Implantation Techniques**

As with pacemakers, there are no ICDs designed specifically for pediatric use. The relatively larger size of ICDs compared with pacemakers and the need for a defibrillation coil or subcutaneous array add to the technical challenges of ICD implantation in small children. Several clinical reports have highlighted the skill and innovation of pediatric cardiac surgeons who have successfully implanted ICDs in young infants using novel approaches (Gasparini et al. 2005; Kriebel et al. 2006; Kugler and Erickson 2006; Stephenson et al. 2006; Agarwal et al. 2007; Berul 2008; Blom 2008). However, long-term complications related to growth and fibrosis have been reported, resulting in some recent modifications (Berul et al. 2008; Tomaske et al. 2008).

Defibrillator patches are rarely used in the current era due to fibrosis and the potential for a restrictive cardiomyopathy to develop as the child grows. A subcutaneous coil or array has been shown to work well in young children. In the very small child, a short SVC coil or array has been shown to work well in young children. In the very small child, a short SVC coil may be implanted pericardially and tunneled, along with the epicardial pace/sense lead(s), down to the abdominally implanted ICD. Suturing the distal end of the coil to the pericardium helps prevent slippage as the child grows (see Fig. 5.1.2). The heart should lie between the coil or array and the ICD can to provide a good shocking vector. Experiments with computer modeling may prove useful in identifying optimal configurations for electrode placement in patients requiring unconventional defibrillator implantation (Jolley et al. 2008).

**Table 5.1.4** Distribution of diagnoses among pediatric ICD recipients at Lucile Packard Children’s Hospital at Stanford.

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>N = 172</th>
</tr>
</thead>
<tbody>
<tr>
<td>VT*</td>
<td>48 (28%)</td>
</tr>
<tr>
<td>Long QT syndrome</td>
<td>36 (21%)</td>
</tr>
<tr>
<td>Dilated cardiomyopathy</td>
<td>32 (19%)</td>
</tr>
<tr>
<td>Hypertrophic cardiomyopathy</td>
<td>21 (12%)</td>
</tr>
<tr>
<td>Secondary cardiomyopathy</td>
<td>15 (9%)</td>
</tr>
<tr>
<td>VF</td>
<td>11 (6%)</td>
</tr>
<tr>
<td>Cardiac arrest</td>
<td>7 (4%)</td>
</tr>
<tr>
<td>Brugada syndrome</td>
<td>1 (0.5%)</td>
</tr>
<tr>
<td>CPVT</td>
<td>1 (0.5%)</td>
</tr>
</tbody>
</table>

* Two patients were also diagnosed with a cardiac tumor.
** Cardiomyopathy developed following surgery for congenital heart disease or secondary to chronic pacing for congenital AV block.

**Figure 5.1.2** Chest X-ray demonstrating an epicardial dual-chamber ICD in a 2-week-old with Timothy syndrome. Note the pericardial placement of an "SVC" coil.
A transvenous ICD system is preferred, when possible. This approach offers the advantage of avoiding a thoracotomy. Also, transvenous leads are associated with lower pacing thresholds and a lower risk of lead fracture. With transvenous implantation, the decision to use a single-coil versus dual-coil lead is often based on the intercoil distance, which may be too long for some children. Also, adhesions may develop around the SVC coil, making future extractions difficult. For these reasons, single-coil defibrillation leads are usually preferred unless high defibrillation thresholds (DFTs) are anticipated because of the child’s body mass or diagnosis.

**Device Selection**

With small children, size does matter. Obviously, smaller, thinner ICDs with rounded edges are desirable. Perhaps more importantly, though, when choosing a device, careful consideration must be given to the programmable features. When bradycardia support is not necessary and atrial tachyarrhythmias are not an issue, a simple single-chamber ICD can be implanted. However, dual-chamber ICDs are preferred when pacing support is needed or when rhythm discrimination may be of value. For infants requiring pacing, such as a long QT syndrome patient with functional 2:1 AV block, a high base pacing rate may be important. Many ICDs have a maximum tracking rate of only 150 bpm, which is appropriate for older adults, but may be inadequate for young patients. A few of the newer ICDs can be programmed to track up to 180 bpm. Additionally, the shocking energy must be considered. For example, a 5-J shock may be more than sufficient in an infant. An ICD with the option of programming low-energy therapy for the initial and subsequent shocks should be used in smaller children. This capability, however, may not necessarily be found in the smallest ICDs. One must look thoughtfully at these features to select an appropriate defibrillator model.

**Programming Considerations**

In addition to the previously mentioned recommendations for pacemaker programming, when programming an ICD, utmost care must be given to minimize the potential for inappropriate shocks. Unlike the older adult, a child or teen may be quite active, and with exercise, may easily achieve normal sinus tachycardia rates up to 180–200 bpm or so. VT and ventricular fibrillation (VF) detection rates should be programmed high enough to avoid any overlap with normal sinus tachycardia. The use of VT discrimination features, such as PR Logic (Medtronic, Inc., Minneapolis, MN), is helpful as well, especially in patients with a history of intra-atrial reentrant tachycardia.

A VT monitoring zone may be a useful surveillance tool to assess the occurrence of ventricular ectopy that might indicate a need to adjust antiarrhythmic medications as the child grows. A monitoring zone may also help identify noncompliance with drug therapy, such as prescribed beta blockers.

The programming of ICD therapies should be based on the child’s diagnosis and likely response to overdrive pacing. If potentially effective, pace-termination should be included as an early therapy for VT to help avoid the trauma of receiving a shock.

**CARDIAC RESYNCHRONIZATION THERAPY**

Recently, cardiac resynchronization therapy (CRT) has been adapted for children with mechanical dyssynchrony and poor ventricular function. In a recent multicenter study, CRT was shown to improve ejection fractions in a cohort of 103 pediatric patients. Diagnoses included CAVB (13%), cardiomyopathy (16%), and complex congenital heart disease (71%), including single-ventricle physiology (7%) (Dubin et al. 2005). More than half of these
patients required an epicardial or hybrid system due to size or anatomical constraints. For many young patients, CRT serves as a bridge to transplantation, while a small number of responders improve sufficiently to be removed from the transplant waiting list.

**CARE AND FOLLOW-UP**

The child’s initial device implantation and subsequent revisions represent major events for the family. It is important to assess both the parents’ and the child’s level of cognitive function and their attitudes toward device implantation. Preparation for the parents should begin as early as possible. The child’s preparation should be individually tailored and age appropriate (LeRoy et al. 2003). For preschoolers, a short teaching session the day before surgery is generally best, using simple pictures and medical play that focus on what the child will experience. School-age children tend to cope better when given about a week’s notice of pending surgery with information given at their level of understanding. Offering concrete explanations along with seeing an actual model of a pacemaker or ICD can be helpful in allaying their anxiety. Teenagers vary in their level of maturity and their ability to cope with stressful situations, but usually prefer to have several weeks, when possible, to prepare for procedures. The circumstances surrounding the device implant may factor into their acceptance. Choices should be given whenever possible to allow the pediatric patient to feel some sense of control. Pairing a new device patient (and family) with an experienced one can help them see they are not alone, and provides a unique opportunity for them to ask questions about life with a device that health care providers may not be able to answer. Also, if time allows, the child and his or her family may benefit by meeting with a mental health specialist. This counseling may be especially important for older children and teens prior to ICD implantation. Parents may benefit by counseling as well.

Along with describing the surgical procedure and expectations, the preoperative discussions with the family should address activity restrictions. Activity recommendations are based on the child’s diagnosis, risk factors, and the individual preferences of the child’s health care provider. Though practice patterns vary (Lampert et al. 2006), most pediatric electrophysiologists recommend avoiding all contact or collision-type sports that might result in a lead fracture. Imposing such restrictions on older children and teens may lead to feelings of anxiety, anger, and depression (Eicken et al. 2006). It may be helpful to identify activities that the patient is allowed to perform rather than dwell on those that will no longer be permitted. In some cases, a protector can be devised to wear over the implanted device to allow them to participate in certain activities. Cardiopulmonary resuscitation classes are highly recommended for the family members of pacemaker-dependent children and ICD recipients. Enrollment in MedicAlert™ (MedicAlert Foundation, Turlock, CA) is strongly encouraged as well.

As school is a significant part of the child’s world, school issues should be addressed with not only the patient and family, but also the school nurse, teachers, and relevant coaches. Restricted or modified physical education will likely be prescribed. Backpacks should not be carried over the affected shoulder. To minimize the weight to be carried, duplicate sets of books, one for home and one for school, may be requested. The clinician may be asked about the safety of industrial arts classes, vocational education programs, certain science experiments, field trips to science or exploratory museums, amusement parks, or electronic games. Schools often request that the health care provider draft care plans, especially for children who are pacemaker dependent or have ICDs.

The psychosocial adjustment of children and adolescents to device implantation has been explored, but much more needs to be learned. Common themes reported by young patients include anxiety, depression, anger, parental
Chapter 5.1 Care of the Pediatric Patient with a Device

overprotectiveness, concerns related to body image and peer acceptance, and issues related to the future, including employment and parenthood (Zeigler and Corbett 1995; Sears et al. 2001; DeMaso et al. 2004; Eicken et al. 2006). Local support groups or Internet-based groups, like the Sudden Arrhythmic Death Syndrome Foundation, may offer education or reassurance to some teens and their families. Also, cardiac camps can provide helpful opportunities for children and teens to interact with other device recipients. Occasionally, a referral to counseling services may be warranted.

Frequent follow-up in the first 2–3 months is helpful in not only assessing the implanted device and wound healing, but also in gauging how the child and family are dealing with the newly implanted pacemaker or ICD. These visits provide opportunities for family members to ask questions and for the clinician to reinforce teaching points.

Infants, toddlers, and preschoolers often present challenges to the device specialist attempting to perform a follow-up evaluation. Keeping a parent nearby, perhaps even having the parent hold their child on their lap, is essential. For infants, dimming the lights and having them feed during the evaluation may result in a more cooperative patient. The use of distraction, such as a DVD player or cable TV with cartoon favorites, may be helpful with young children. Some of the device companies have created special covers for their programming wands to make them more “kid friendly” and much less threatening. Wireless technology, though currently available for ICDs only, solves the problem of trying to maintain a good signal with the programming head over a device in a fidgety youngster.

The basic process for device follow-up is the same with children as adults but, obviously, much more patience is required when dealing with the little ones. The evaluation should include a baseline electrocardiogram (ECG) and, if possible, a recording of the underlying ECG. It may be reassuring for the parents to see the underlying rhythm, if present. Rate histograms and event counters will reveal the appropriateness of pacing and uncover episodes of ectopy or arrhythmias. Sensing and pacing thresholds should be measured. If a child is not used to being paced, the sensation during threshold testing may be uncomfortable, so an advanced warning may be appreciated. With unipolar leads, the potential for muscle twitching or diaphragmatic pacing should be assessed, especially in the child who may not be able to verbalize symptoms. A review of lead impedance trends can identify early signs of impending lead problems. As mentioned previously, the upper rate limit needs to be programmed to a reasonably high tracking rate, while the AV delay and PVARP should be shortened to provide an adequate Wenckebach window between the upper tracking rate and the 2:1 pacemaker block point. Unless the tracking of atrial arrhythmias is a concern, the upper tracking rate should be programmed as close as possible to 220 bpm minus the patient’s age. For young children programmed to rate-responsive modes, adjustments can be based on heart rates achieved with walking and running in the clinic hallway until they reach an age when more formal exercise testing is possible, usually around age 6 or 7. When children are pacemaker dependent, such as those with surgical AV block, care must be taken to identify impending battery depletion and to arrange replacement before the device trips the elective replacement indicator (ERI). If normally paced in a dual-chamber mode, VVI pacing at a slower ERI rate may lead to symptoms in a young child, especially if ventricular function is poor.

A major difference between device follow-up for children versus adults is the need to assess the pacing leads serially for tension due to somatic growth. Every 1–2 years until the end of puberty, a chest radiograph should be obtained. If a pacing lead appears taut, the child will need to undergo surgery to advance the lead or replace it. Sometimes adhesions will trap sections of the lead, so the entire length of the lead
should be examined for any signs of stretch. Twiddler’s syndrome has been reported in children and should be ruled out by X-ray as well (Abrams and Peart 1995; Berul et al. 1997). ICD coils should be evaluated radiographically to determine whether the shocking vector has changed with growth. Repeat DFT testing and lead revision may be necessary. Significant changes in body mass may warrant follow-up DFT testing as well.

The overall goal of device therapy in the pediatric patient is to provide the care and support needed to facilitate a positive outcome, both medically and psychosocially, for the child and family. Safety of the child must be a priority. Ideally, infants and children with a device should be managed by health care professionals with training and expertise in the specialty area of pediatric cardiology. Additionally, knowledge of the unique aspects of pediatric device implantation, programming, and follow-up is essential.

REFERENCES


INTRODUCTION

Supraventricular tachycardia (SVT) is the most common significant arrhythmia in the pediatric population. While it accounts for 95% of arrhythmias diagnosed in children with structurally normal hearts, SVT may also affect children with congenital and acquired heart disease (Campbell et al. 1984) and is a source of troublesome morbidity following complex congenital heart defect (CHD) repair. Although SVT in children with normal cardiac anatomy is rarely life threatening, this rhythm disturbance can cause great concern for children and their families. Nurses and associated professionals encounter pediatric SVT across all health care settings, including the primary care office, the emergency department, and the intensive care unit.

Definition

SVT is a general term for any abnormal rhythm that originates in the bundle of His or above, or incorporates atrial tissue. A tachycardia arising from the ventricular muscle or Purkinje fibers is never correctly classified as SVT, but the term does apply to some arrhythmias that pass through the ventricle in addition to atrial tissues. It is common in clinical practice to use the abbreviation “SVT” to mean regular, rapid, narrow complex tachycardia, but some SVT types actually have wide complexes, and others are irregular.
Incidence/Prevalence

The exact incidence of SVT in the pediatric population is estimated to be between 1 in 25,000 and 1 in 250 (Chun and Van Hare 2004). Complex atrial arrhythmias remain a common complication after surgical repair for CHDs, a population that is growing rapidly due to significant advances in cardiovascular surgery. Arrhythmias in children occur with equal frequency for males and females.

Etiology

No structural heart disease is present in most SVT patients, particularly young infants. The incidence of Wolff–Parkinson–White (WPW) syndrome is increased in patients with Ebstein’s anomaly, corrected transposition of the great arteries, and hypertrophic cardiomyopathy. The small scars and suture lines from cardiac surgery create an environment susceptible to SVT, which may occur shortly or many years following operation.

CLINICAL MANIFESTATIONS

History

Neonates with arrhythmias of various kinds, including SVT, may be described as listless or fussy, but the specific onset of the symptoms is often vague. In children that are school age and older, the history should be directed at distinguishing palpitations due to SVT from those due to autonomic changes, anxiety, and normal physiology. Some historical data include total number of episodes, frequency, duration, what initiates the event, how the child feels during the event, the child’s appearance to a parent during event, any associated symptoms, and what terminates the episode. It is important to note that dizziness, headaches, fatigue with exercise, chest pain, and fainting are complaints unlikely to be related to SVT.

Age-Specific Signs and Symptoms

SVT of the most common forms (accessory pathway [AP] and AV node reentry, or AVNRT) is characterized by an abrupt onset and cessation of an abnormally and fairly constant fast rate. Atrial ectopic tachycardia (AET) may have a more gradual onset and offset. Typical SVT symptoms and complaints by age are as follows:

- **Neonates**: irritability, poor feeding, tachypnea, diaphoresis and poor color; parent report of heart rate too fast to count; may mimic sepsis, congenital heart disease, or respiratory conditions
- **Infants/toddlers**: irritability, poor feeding, tachypnea, diaphoresis and poor color; observed palpitations such as fluttering of shirt or neck vessels; parent report of heart rate too fast to count
- **School-age child**: “My heart hurts/feels funny/is jumping/is beeping/is racing,” vague palpitations, general malaise, chest pressure; parent may report, “that the child’s heart rate was too fast to count”
- **Adolescents**: generally can provide more detailed history; able to describe setting which symptoms began (with activity or at rest); gradual versus sudden onset; rapid, regular palpitations; heart rate rapid (often too fast to count); dizziness (syncope rare); chest pain/pressure; fatigue; shortness of breath; occasionally no symptoms are reported: SVT may rarely be an incidental finding.

Differential Diagnosis of Palpitations and Elevated Heart Rate

SVT must be clinically distinguished from:
**Sinus Tachycardia**
- A normal P wave precedes each QRS with normal PR interval. Associated with: congestive heart failure, fever/sepsis, hypovolemia, anemia, hyperthyroidism, anxiety, pain, exertion, and certain medications (stimulants, bronchodilators)

**Ventricular Tachycardia**
- SVT usually has narrow QRS complexes
- Many SVT types terminate with intravenous (IV) adenosine

**DIAGNOSTIC TOOLS**

**Twelve-Lead Electrocardiogram**
The 12-lead electrocardiogram (ECG) remains a useful test, even when the arrhythmia is not present at the time it is performed. It may reveal:

- Evidence of AP conduction, such as WPW syndrome, where “delta” waves are seen (see Fig. 5.2.3)
- Evidence of congenital or acquired cardiac disease
- The arrhythmia of concern, when occurring at the time of ECG
- The response of a heart rhythm to medication
- The child’s age should always be indicated on the ECG as it is essential in accurate diagnosis

Various monitors can be used to record the heart rhythm, as summarized in Table 5.2.1.

**Exercise Testing**
Exercise testing may be useful in evaluating rhythm disturbances that are triggered by activity, though its diagnostic yield is lower than that of event monitors, and it is impractical in children under age 5. If a teen says she experiences palpitations with soccer practice, she is more likely to reproduce her symptoms wearing a monitor to practice than running on a treadmill.

**CLINICAL AND ECG PATTERNS OF SVT (SEE TABLE 5.2.2)**

**AP-Mediated Tachycardias**
AP-mediated tachycardia is the most prevalent SVT type in infants and smaller children. It is often known as atrioventricular reentry tachycardia (AVRT). Heart rates are typically between 200 and 300 bpm, with infants having higher rates and young adults demonstrating lower rates within that range (see Figs. 5.2.1 and 5.2.2). The QRS is narrow during this tachycardia.

Most AP-mediated SVT propagates through the heart as follows:

- Beat goes through AV node and normal His-Purkinje fibers
- Activation continues through ventricle normally
- Activation crosses back up to the atrium via the AP
- The atria are activated starting where the AP inserts (not from the sinus node), and then excite the AV node
- This circular activation sequence continues

**WPW and Its Risks**
WPW syndrome is the combination of episodes of SVT and the finding of pre-excitation on the ECG, or “delta waves,” when the heart rate is normal (see Fig. 5.2.3). Patients with this ECG pattern, however, are at risk not only for SVT, but also for degeneration, first to atrial fibrillation, and then to unstable ventricular
<table>
<thead>
<tr>
<th>Monitoring devices used in pediatric SVT diagnosis.</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Usual duration</strong></td>
</tr>
<tr>
<td>Holter monitor</td>
</tr>
<tr>
<td>Looping event monitor</td>
</tr>
<tr>
<td>Postsymptom event monitor</td>
</tr>
<tr>
<td>Ambulatory telemetry</td>
</tr>
<tr>
<td>Ambulatory monitoring devices</td>
</tr>
</tbody>
</table>

**Benefits**
- Continuous recording of rhythm (48 hours also available)
- Usually multiple (2-3) leads
- Document frequent arrhythmia, tabulate ectopy totals, evaluate heart rate range, nighttime pauses, etc.

**Disadvantages**
- Patients may not comply with daily wearing of monitor; skin irritation due to adhesive stickers
- Patients forget to keep monitor with them; short events not recorded if device not handy

**Diagnostic application**
- Any arrhythmia investigation
- Document frequent arrhythmia
**Table 5.2.2** Summary of SVT types and their characteristics.

<table>
<thead>
<tr>
<th>Type</th>
<th>Typical rate</th>
<th>Rhythm/pulse</th>
<th>P wave</th>
<th>PR interval</th>
<th>QRS complex</th>
<th>Response to adenosine</th>
<th>Medical treatment</th>
<th>Interventional treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sinus tachycardia</td>
<td>Up to 225 in infants</td>
<td>Regular</td>
<td>Normal</td>
<td>Normal</td>
<td>Narrow*</td>
<td>Transient slowing</td>
<td>Manage cause</td>
<td>None</td>
</tr>
<tr>
<td>AVNRT</td>
<td>150–280</td>
<td>Regular</td>
<td>Not seen or very shortly after QRS; P wave within QRS; very short RP interval (&lt;70ms)</td>
<td>Narrow*</td>
<td>Terminates</td>
<td></td>
<td>See Table 5.2.3</td>
<td>Ablation</td>
</tr>
<tr>
<td>AP (AVRT)</td>
<td>180–300</td>
<td>Regular</td>
<td>Often negative and shortly following QRS</td>
<td>N/A</td>
<td>Narrow*</td>
<td>Terminates</td>
<td>See Table 5.2.3</td>
<td>Ablation</td>
</tr>
<tr>
<td>PJRT</td>
<td>110–250</td>
<td>Regular</td>
<td>negative P waves in leads II, III, aVF, and the left lateral leads</td>
<td>Normal PR (long RP); always 1:1 A:V relationship</td>
<td>Narrow*</td>
<td>Terminates and re-initiates</td>
<td>Second-line agents in Table 5.2.3</td>
<td>Ablation</td>
</tr>
<tr>
<td>IART/Atrial Flutter</td>
<td>100–300</td>
<td>usually regular</td>
<td>Sawtooth pattern; postoperative less distinct</td>
<td>Variable</td>
<td>Narrow*</td>
<td>AV node conduction blocked, flutter continues</td>
<td>Ablation, maze surgery, anti-tachycardia pacing</td>
<td>Ablation</td>
</tr>
<tr>
<td>Ectopic atrial tachycardia</td>
<td>150–300</td>
<td>Variable; warm-up, warm-down</td>
<td>May be notched, inverted, or more subtly different from sinus P-wave</td>
<td>Variable</td>
<td>Narrow*</td>
<td>Causes brief AV block allowing P-waves to be seen; may terminate</td>
<td>See Table 5.2.3</td>
<td>Ablation</td>
</tr>
<tr>
<td>Junctional tachycardia</td>
<td>150–225</td>
<td>Regular</td>
<td>large amplitude distinct p waves; very slow atrial rate</td>
<td>not applicable due to AV dissociation</td>
<td>Narrow*</td>
<td>Some slowing; VA block</td>
<td>Amiodarone</td>
<td></td>
</tr>
<tr>
<td>AVNRT, atypical</td>
<td>150–220</td>
<td>Regular</td>
<td>negative P waves in leads II, III, aVF</td>
<td>normal (or slightly long) and with a long RP interval</td>
<td>Narrow*</td>
<td>Terminates</td>
<td>See Table 5.2.3</td>
<td>Ablation</td>
</tr>
<tr>
<td>MAT</td>
<td>100–300</td>
<td>Irregular</td>
<td>Discrete with three or more p wave morphologies</td>
<td>Variable</td>
<td>Narrow*</td>
<td>Transient slowing</td>
<td>Flecaainde, amiodarone</td>
<td>None</td>
</tr>
<tr>
<td>Antidromic AVRT (AP)</td>
<td>180–300</td>
<td>Regular</td>
<td>Not seen</td>
<td>N/A</td>
<td>Wide</td>
<td>Terminates</td>
<td>See Table 5.2.3</td>
<td>Ablation</td>
</tr>
<tr>
<td>Atrial fibrillation</td>
<td>80–200</td>
<td>Irregular</td>
<td>usually low-voltage, irreg. difficult to discern discrete p waves</td>
<td>N/A</td>
<td>Narrow*</td>
<td>Some slowing</td>
<td>Various</td>
<td>Ablation, maze surgery</td>
</tr>
</tbody>
</table>

* Narrow QRS (<120ms in adolescents, or 100ms in younger children), except with rate-related aberrancy or pre-existing bundle branch block.
tachycardia (VT) due to rapid conduction to the ventricle of atrial beats via the AP. This life-threatening phenomenon is uncommon and is considered very rare in infants and small children. It is usual practice to perform ablation when feasible for the patient with WPW who is approaching adolescence.

Of note, most patients with AVRT episodes do not have pre-excitation, so they can have SVT episodes without being at risk for more malignant arrhythmias.

**Clinical Context and Symptoms**

Children will most typically describe episodes beginning unexpectedly and not associated with their level of activity; some may report onset of tachycardia with bending over or other positional changes, with activity, while others experience events while sedentary.

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**Figure 5.2.1** ECGs of AP-mediated SVT (also known as AVRT). Panel A reveals an event monitor tracing of SVT in an adolescent who had complained in clinic of episodic palpitations. Panel B demonstrates rapid SVT in a stable neonate, at a rate of approximately 300 bpm, due to an AP. Red circles identify the inverted P waves, which follow the QRS complexes.

**Figure 5.2.2** Course of activation of cardiac chambers during AP-mediated tachycardia. The pictured AP crosses the mitral annulus in this patient, allowing the atrium to be activated after the ventricles. For this reason, the tachycardia is often referred to as AVRT. The reverse circuit is also sometimes seen.

**Figure 5.2.3** WPW during normal sinus rhythm. Pre-excitation or delta waves can be identified in lead II as marked by arrows.
AP-mediated tachycardia is by far the most common form of infant SVT, but approximately half of infants who have it will have complete resolution of the SVT episodes and the pathway conduction, as AP presence in infants is often transient.

**Antidromic Reciprocating Tachycardia**

Antidromic reciprocating tachycardia (ART) is a type of SVT in which conduction occurs from atrium to ventricle via the AP, and retrograde conduction travels to the atria via the AV node. ART accounts for less than 10% of SVT associated with accessory connections (always in patients with WPW syndrome). The QRS morphology is wide and resembles pre-excited QRS morphology during normal sinus rhythm (see Table 5.2.2).

**Persistent Junctional Reciprocating Tachycardia**

A special type of AP allows for the clinical entity of “persistent junctional reciprocating tachycardia” (PJRT), a still-used misnomer for an AP-mediated tachycardia that is typically slower but more persistent (see Fig. 5.2.4). PJRT may start without any particular trigger, from sinus rhythm. It may terminate and reinitiate, such that throughout the day the predominant rhythm is PJRT, with brief, interspersed intervals of sinus beats (see Table 5.2.2). Some rate variability can occur with exertion or stress due to catecholamine sensitivity. Because it may not produce symptoms, PJRT may be an insidious cause of ventricular dysfunction.

**AVNRT**

AVNRT is a form of SVT whose circuit revolves around the AV nodal tissues. Its clinical presentation is similar to AP tachycardias; however, there are key distinctions. Particularly in adolescents, the onset of AVNRT episodes is most commonly with exertion, such as sports activities or dance.

AVNRT is the second most common reentrant SVT in children; it is rarely seen in infants, but by age 6–10 years, accounts for approximately 30% of SVT (Deal et al. 2004). In teenagers, AVNRT is the most likely mechanism of episodic palpitations due to arrhythmia. The atypical forms of AVNRT are rare in children.

**Intra-Atrial Reentrant Tachycardia and Atrial Flutter**

Atrial flutter is the common term for a tachycardia within the atrium, and results from a circular (reentrant) activation pattern. The classic “flutter waves” (see Fig. 5.2.5), having a sawtooth appearance, are usually only seen in patients who have normal hearts, and only rarely in young patients. The neonate with elevated heart rate may have atrial flutter and usually does not recur after conversion to sinus rhythm.

Atypical atrial flutter, also known as intra-atrial reentry tachycardia, is a troublesome
MAT often occurs in healthy infants. It is irregular, with pauses and varying P wave morphology. It does not respond to adenosine or cardioversion.

Multifocal Atrial Tachycardia

Multifocal atrial tachycardia (MAT), also known as chaotic atrial rhythm, is rare in children (healthy children as well as those with heart disease). As noted in Table 5.2.2, MAT is defined by P waves of three or more morphologies and irregular P-P intervals at heart rates >100bpm. It is likely that MAT results from premature beats arising from multiple separate sites in the atrium, resulting in an irregular rhythm. While often benign, serious cardiac compromise may occur when the rate is rapid and the rhythm is present for days or longer (Bradley 2006, p. 135) (Fig. 5.2.7).
Chapter 5.2 Care of the Pediatric Patient with SVT

**Junctional Ectopic Tachycardia**

Junctional ectopic tachycardia (JET) occurs most frequently in the first days following cardiac surgery in infants (ventricular septal defect, tetralogy of Fallot). The rapid beats originate in the His bundle and may be dissociated from atrial contractions. JET can last 48–72 hours and is usually limited, but its effects on cardiac output, particularly in the recovering heart, contribute to morbidity and mortality in children if not quickly identified and managed (Zeigler et al. 2001). Amiodarone is the most common medication administered for JET. A congenital form of JET rarely occurs, but when treated with amiodarone or ablation, it responds with variable results.

**Atrial Fibrillation**

Atrial fibrillation in children is also rare and suggests an underlying problem such as:

- WPW syndrome
- Hypertrophic cardiomyopathy
- A genetic abnormality of atrial cells

The most critical of these is WPW syndrome, as discussed above.

**PREVENTIVE MANAGEMENT AND TREATMENT**

**Wait and Watch**

For children who experience infrequent episodes and have no evidence of cardiomyopathy or syncope, it may be acceptable to provide only education and reassurance. This “no therapy” or “watch and wait” approach may also be a good option if self-conversion of tachycardia episodes is effective using vagal maneuvers. For some SVT types (PJRT, atrial flutter), it is not appropriate.

**Vagal Maneuvers**

“Vagal maneuvers,” so called because they are believed to produce inhibition via the vagal nerve, are an excellent first-line treatment for episodes of SVT dependent on the AV node (AVRT, AVNRT). They are relatively noninvasive and work by inducing transient slowing or block of conduction through the AV node. Vagal maneuvers are ineffective in AET, MAT, and JET.

**Vagal Maneuvers**

- Ice bag or cold cloth to the face for 5–10 seconds, repeat as needed (for infants)
- Hold child upside down for up to 1 minute, repeat as needed (for infants)
- Performing a headstand (by patient report, the most effective)
- Holding breath and coughing
- Bearing down as if to have a bowel movement
- Stimulation of gag reflex
- Blowing against a thumb placed over closed lips

**Synchronized Cardioversion**

For young children in SVT and with signs of compromise, direct current (DC) synchronized cardioversion should be performed. The energy dose, 0.5–2 J/kg, should be delivered with continuous cardiac monitoring and sedation if possible. A rational assessment of a child whose heart rate is 200–300 bpm is essential; even at these high rates, critical hemodynamic compromise is uncommon in otherwise healthy children.

Synchronized DC cardioversion is often the best way to convert a patient from atrial flutter or fibrillation to sinus rhythm. DC cardioversion is ineffective for the conversion of ectopic or automatic tachycardias, such as MAT and AET.
Cardiac Arrhythmia Management

Adenosine

Adenosine can be used both therapeutically and diagnostically. It has an extremely short half-life, and requires expert IV administration:

- Obtain IV access
- Obtain pertinent history (such as asthma)
- Place on cardiac monitor and place ECG leads (if possible)
- Through the same IV port, rapidly flush adenosine (0.1–0.3 mg/kg) followed immediately by the IV flush
- If ineffective, follow with an increased dose

Even when unsuccessful at normalizing the rhythm, adenosine may have diagnostic value. While no sedation is required, reassure older children regarding the brief but unpleasant side effects of the medication. Adenosine side effects include:

- Transient AV block and pronounced sinus slowing (pauses in heart rate of one to several seconds may occur)
- Facial flushing
- Presyncope
- Chest pain
- Hypotension (transient)
- Wheezing

Other Acute Medical Therapies

Children, particularly infants, may present with AP tachycardia (AVRT), which is difficult to terminate with adenosine. Procainamide can be administered IV in this context. Amiodarone may also be used.

Oral agents used to control SVT in the outpatient setting are summarized in Table 5.2.3. The role of medication treatment has been much reduced in the era of radiofrequency and cryoablation techniques. Side effects are common and incomplete arrhythmia control is often associated with medical therapy.

Electrophysiology Study and Ablation

Pediatric heart centers have active programs offering ablation, often as the first specific treatment for children with recurrent episodes of SVT of all types. Specialized catheters exist for the application of these techniques to even small toddlers and, when necessary, infants.

Cryoablation techniques, while taking somewhat longer than radiofrequency ablation, appear to have comparable success rates and allow for very safe ablation near the AV node.

Pediatric-Specific Considerations

Knowledge of pediatric behavior and cognitive developmental stages is essential to guiding the age-appropriate care of a pediatric SVT patient. A useful guideline statement was recently published and can serve as a valuable reference for nurses and associated professionals (Leroy et al. 2003).

Nursing and Associated Professional Implications

A coordinated approach to caring for the pediatric patient with SVT is essential. The pediatric cardiology attending (electrophysiologist), pediatric cardiology fellow, advanced practice nurses (APN), EP registered nurses, EP lab technicians, ECG/Holter/telemetry technicians, and administrative support staff contribute to the safety and effectiveness of diagnosis and treatment. Clarity of team member roles instills confidence in the patient’s family and sets the stage for a successful procedure and positive experience.

Role of APN

APNs are suited to provide optimal care of the pediatric SVT patient. Their knowledge combined with practical bedside nursing experience allows for shrewd assessment and selective
• Patient appropriateness for sedation or anesthesia
• Confirmation of any history and physical data previously collected
• Identify key diagnostic recordings pertinent to the study

The APN may visit the EP lab and provide regular updates on the procedure, to break up the sometimes lengthy waiting period for the child’s family.

Postprocedure

Patient care after the procedure should include:
• Assessment of potential complications
• Education and follow-up plan for what may include same-day discharge
• Phone follow-up within 24–48 hours

Table 5.2.3 Medications used in pediatric SVT.

<table>
<thead>
<tr>
<th>Class</th>
<th>Dose (mg/kg/day)</th>
<th>Dose interval (hour)</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Digoxin Glycoside</td>
<td>8–10 mcg</td>
<td>12</td>
<td>Decreasing in use, minimal antiarrhythmic effect*</td>
</tr>
<tr>
<td>Propranolol Beta blocker</td>
<td>1–4</td>
<td>8</td>
<td>Depression, fatigue may occur</td>
</tr>
<tr>
<td>Atenolol Beta blocker</td>
<td>0.5–1.5</td>
<td>12–24</td>
<td>Fewer central nervous system side effects than propranolol</td>
</tr>
<tr>
<td>Nadolol Beta blocker</td>
<td>1–2.5</td>
<td>12–24</td>
<td>Can be reconstituted as oral suspension</td>
</tr>
<tr>
<td>Diltiazem Calcium channel blocker</td>
<td>1.5–3.5</td>
<td>6–8</td>
<td>Sustained-release form available*</td>
</tr>
<tr>
<td>Flecainide Sodium channel blocker</td>
<td>1–5</td>
<td>8–12</td>
<td>Structural heart disease a contraindication</td>
</tr>
<tr>
<td>Sotalol Beta blocker and Class III antiarrhythmic</td>
<td>2</td>
<td>8–12</td>
<td>Contraindicated if QTc prolongation</td>
</tr>
<tr>
<td>Amiodarone Blocks potassium and other channels</td>
<td>5</td>
<td>24</td>
<td>Potential end organ toxicity long term</td>
</tr>
<tr>
<td>Dofetilide Class III antiarrhythmic</td>
<td>Not defined</td>
<td>12</td>
<td>Admission and cardiac monitoring prior to initiating mandatory</td>
</tr>
</tbody>
</table>

*Considered unsafe for use in WPW syndrome.

communication of a very complex field and its treatment to patients with varying levels of understanding.

Preprocedure

Figure 5.2.8 illustrates the nursing process that an APN can use to guide the care of the pediatric patient with SVT in preparation for EP study and ablation. Time taking the patient’s history, teaching, listening, and answering questions with the patient and family can greatly enhance the procedure day experience.

Day of Procedure

On the day of a pediatric EP study and ablation, the APN assessment should be focused on:
Communication of Holter result (if relevant)

Common postprocedure concerns are summarized in Table 5.2.4.

**PROGNOSIS**

Prognosis is dependent on the patient’s SVT type and treatment. An infant with SVT may have troublesome tachycardia requiring many medication trials and a challenging ablation, or may have spontaneous resolution without treatment. Most patients who have undergone SVT ablations have no recurrence and require few follow-up visits. The patient with congenital heart disease and complex flutter, or intra-atrial reentrant tachycardia (IART), may have an incomplete response to both medications and ablation, and years of troublesome symptoms.

**COMPLICATIONS**

If left undiagnosed and untreated, prolonged SVT, even if not very rapid, can result in diminished heart function with ventricular dilation. This *tachycardia-induced cardiomyopathy* appears to be a result of prolonged elevated heart rates. The child may have rela-
Table 5.2.4  Complications of pediatric SVT ablation and their management.

<table>
<thead>
<tr>
<th>Complaint</th>
<th>Timing</th>
<th>Differential diagnosis</th>
<th>Evaluation</th>
<th>Management</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hematoma</td>
<td>Groin pain, swelling</td>
<td>1–3 days</td>
<td>Ecchymosis; normal cath site healing</td>
<td>Phone triage or clinic assessment</td>
</tr>
<tr>
<td>Arrhythmia recurrence</td>
<td>Racing heartbeats</td>
<td>24–48 hours</td>
<td>Premature atrial beats; anxiety</td>
<td>Arrhythmia monitor</td>
</tr>
<tr>
<td>Heart block</td>
<td>Slow beats, fatigue, or asymptomatic</td>
<td>&lt;24 hours</td>
<td>Transient AV nodal injury, sinus bradycardia</td>
<td>12-lead ECG; Holter monitor</td>
</tr>
<tr>
<td>Premature atrial beats</td>
<td>Skipped beats or extra, hard beats</td>
<td>1–2 weeks postop</td>
<td>Pathway recurrence; anxiety</td>
<td>Phone triage and Holter</td>
</tr>
<tr>
<td>Bleeding from catheter site</td>
<td>Blood, warmth from groin sites</td>
<td>&lt;24 hours</td>
<td>Skin oozing; Betadine stain</td>
<td>During recovery or phone triage</td>
</tr>
<tr>
<td>Pericardial effusion</td>
<td>Dyspnea, fatigue</td>
<td>&lt;24 hours</td>
<td>Intercurrent illness</td>
<td>Echocardiogram</td>
</tr>
</tbody>
</table>
tively vague symptoms of fatigue despite severely diminished left ventricular ejection fraction. Improvement after arrhythmia treatment, usually with ablation, is excellent. PJRT and AET are the types of SVT most commonly implicated in this condition. Medication may be used to control the rhythm temporarily if immediate ablation cannot be performed.

Complications following ablation treatment of SVT are described in Table 5.2.4.

**CONCLUSION**

Pediatric SVT is by far the most common arrhythmia that children experience. Thorough knowledge of age-specific considerations, types of SVT, and management strategies is essential. As part of a multidisciplinary team, nurses and associated professionals are fundamental to the delivery of expert care and can enhance positive outcomes.

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**Case 5.2.1**

**Case Presentation**

At 32 weeks gestation, baby girl Jones was diagnosed with fetal SVT at a routine prenatal visit. No treatment was needed and she was born without complications at 38 weeks gestation. Her postnatal course was unremarkable and she was discharged home at day of life 2. She presented at age 2 weeks to the Pediatric Cardiology Clinic for a follow-up visit. She appeared well in her mother’s arms in no apparent distress. Her 12-lead ECG reveals a narrow complex rhythm with a rate of 268 bpm (see Fig. 5.2.9). What course of action should be taken for this patient?

**Case Discussion**

This little girl had been feeding well without respiratory distress and is gaining weight appropriately. Her mother reported that on the previous evening she did note that her chest wall seemed to be moving more than usual. On examination, she was alert, well appearing, and in no apparent distress. Her radial pulse remained fixed at 268 bpm. In the clinic, vagal maneuvers—knee-chest position, positioning upside down, bag of ice to the face three times—failed to convert her to normal sinus rhythm. She was transported to the Electrophysiology Laboratory and a peripheral IV was placed. IV conscious sedation was administered. Transesophageal pacing was successful in converting her SVT to normal sinus rhythm. She was started on nadolol (1 mg/kg/day) therapy. At 6 weeks follow-up, she was asymptomatic, and at 9 months of age, medications were stopped without subsequent recurrence.

![Figure 5.2.9](image-url) ECG of an asymptomatic infant with AVRT. This AP-mediated tachycardia is the most common form of SVT experienced during infancy.
Case 5.2.2
Case Presentation
Patient L.F. is a 5-year-old boy with a history of regular SVT beginning on day of life 1. His ECG is normal (no pre-excitation) and his episodes were originally treated with digoxin. Over the past year, he has had multiple breakthrough SVT episodes. His parents elected to pursue electrophysiology (EP) study and ablation in hopes of eliminating further SVT events and the need for daily medication. What type of SVT is typical for this age patient? What treatment was deemed appropriate? What follow-up care is necessary?

Case Discussion
Under general anesthesia, EP study was performed using four intracardiac catheters. Testing revealed a concealed left lateral AP. His AVRT had a cycle length of 280 ms (rate of 214 bpm). Transseptal puncture was performed, and radiofrequency ablation of the AP successfully eliminated the tachycardia. Postprocedure echocardiogram was normal: no impairment of the mitral valve or pericardial effusion. His ECG was also within normal limits. He was discharged home after 4 hours in the recovery room. Due to the theoretical risk of a stroke from the healing transseptal puncture, he was prescribed aspirin 81 mg daily for 6 weeks.

Case 5.2.3
Case Presentation
A 15-year-old patient had previously undergone EP study and cryoablation of AET from the right atrium. He was asymptomatic without recurrence of arrhythmia at his 6-week follow-up evaluation. At his 1-year return visit, his ECG revealed a heart rate of 160 bpm while supine (see Fig. 5.2.6). He denied any sensation of palpitations, but noted ongoing exertional fatigue for the past few months. Echocardiogram revealed severely depressed left ventricular systolic function without intracardiac thrombus or effusions. What is this rare complication of untreated SVT? What is the best approach to treatment of this patient?

Case Discussion
Tachycardia-induced cardiomyopathy is unusual, but does occur in the setting of sustained, untreated SVT, even when the rate is not severely elevated. This patient underwent repeat EP study and radiofrequency ablation 2 days later. His procedure was successful in eliminating his AET, and follow-up echocardiogram demonstrated complete recovery of cardiac function.

Case 5.2.4
Case Presentation
A 19-month-old boy presented with l-transposition of the great arteries status post combined atrial and arterial switch operation. He underwent dual-chamber permanent pacemaker placement due to the complication of surgical complete heart block. He developed recurrent atrial flutter and has required multiple electrical cardioversions. Originally, rhythm control was achieved with amiodarone; however, despite escalating amiodarone dosing, he now experiences frequent atrial flutter.
episodes. Home monitoring transmissions from his pacemaker reveal a rapid atrial rate and a pacing rate half as fast as the atrial beats. What other medications could be considered for this patient? Would antitachycardia pacing be potentially beneficial for this patient? Is anticoagulation therapy appropriate?

Case Discussion
This case demonstrates some of the complexities of infant cardiac care and surgery for congenital defects. Amiodarone has dose-related adverse effects on the thyroid gland, liver, skin, and lung tissues; it interacts with many other medications, requiring alterations in usual doses. But it remains effective for a variety of arrhythmias and can be tolerated without negative effects in infants for short- and medium-length courses. Alternatives for this patient include other medications (flecainide, sotalol, and propafenone), antitachycardia pacemakers, and ablation. This patient achieved better control on a higher dose of amiodarone, but his long-term course remains to be seen. Note that anticoagulation was not initiated for this patient.

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RESOURCES
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Section 6
Additional Topics in Cardiac Arrhythmia Management
INTRODUCTION

The past 20 years have seen the focus of cardiac electrophysiology (EP) laboratories evolve from providing diagnostic information to being primarily involved in interventional EP procedures. The ablation of arrhythmias began in the late 1980s with direct current ablation of the atrioventricular (AV) node or His bundle to achieve complete heart block. The ability to ablate arrhythmicogenic tissue became easier and safer with the advent of radiofrequency (RF) energy as a source for cardiac ablation. Initially, RF was used for the ablation of such arrhythmias as Wolff–Parkinson–White (WPW) syndrome and AV nodal reentrant tachycardia (AVNRT). These early ablations were performed using standard EP catheters and physiological recording systems that displayed intracardiac electrograms. The catheter ablation of typical atrial flutter became routine during the mid-1990s (Cosio et al. 1993). EP physicians began to attempt to mimic the surgical Maze procedure for atrial fibrillation using catheters and RF energy in the mid-1990s. This investigation led to an appreciation of the importance of the left atrium and pulmonary veins in the initiation and perpetuation of atrial fibrillation (Haissaguerre et al. 1998). The importance of cardiac anatomy as it relates to arrhythmogenesis became increasingly apparent during this time.

Standard EP catheters using fluoroscopic imaging along with the digital display of intracardiac electrograms are sufficient tools for routine ablation procedures; however, they have limitations for complex interventional EP procedures. The mid to late 1990s saw the introduction of nonfluoroscopic electroanatomical mapping systems that could assist with complex anatomically based mapping and ablation procedures. Current mapping systems allow the
operator to accurately correlate an intracardiac electrogram with its exact anatomical location, either endocardial or epicardial. In addition, mapping systems are able to create three-dimensional graphic representations of cardiac chambers. Mapping systems have rapidly become critical equipment in EP labs for routine and complex interventional EP procedures. The goal of the present review is to discuss the most commonly used mapping systems and their clinical utility. In addition, the energy sources used for catheter ablation and the biophysics of ablation will be reviewed.

**CARTO MAPPING SYSTEM**

The initial published description of a clinical use of electroanatomical mapping technology to assist with catheter ablation was in 1996 (Ben-Haim et al. 1996). This technology evolved to become the CARTO (Biosense Webster, Diamond Bar, CA) electroanatomical mapping system. This technology has since been used extensively worldwide for mapping of atrial and ventricular cardiac arrhythmias. The CARTO system uses electromagnetic fields to allow for visualization and cataloging of catheter locations, along with the corresponding intracardiac electrograms. The system involves a locator pad placed underneath the operating table, as well as a magnetic sensor embedded in the tip of the mapping catheter. The locator pad includes three coils, which each generate an ultralow magnetic field that decays in strength as a function of distance from the coil. The tip sensor in the mapping catheter measures the strength of each magnetic field, thus enabling determination of the catheter distance from each coil. The intersection of the distance from the three coils determines the catheter tip location in space. The catheter tip location is determined relative to the location of a reference patch on the patient’s back, thereby compensating for minor movement of the patient.

Intracardiac electrograms, as well as the corresponding catheter location in the cardiac chamber, are displayed and recorded on the mapping system. As the catheter is sequentially moved throughout the chamber, a three-dimensional map is created. The three-dimensional map graphically shows the chamber geometry, as well as the electroanatomical information. The electroanatomical data is color-coded as activation timing of the intracardiac electrogram on the mapping catheter relative to a timing reference, which can be either an electrocardiographic (ECG) channel or an intracardiac electrogram. For each map, an interval of interest for each cardiac cycle is determined relative to the timing reference. This interval typically approximates the tachycardia cycle length. In addition to activation timing, the CARTO map allows the display of propagation movies of sequential activation, as well as voltage maps that show the maximum voltage for each catheter location. Voltage maps can be useful in the ablation of substrate-based tachyarrhythmias, such as ischemic ventricular tachycardia or atypical atrial flutter in the setting of congenital heart disease.

**Clinical Utility of CARTO**

All cardiac arrhythmias were initially studied using conventional EP catheters with the intracardiac electrograms displayed on physiological recording systems. The CARTO electroanatomical mapping system has subsequently been used and validated for most atrial and ventricular tachyarrhythmias.

The mapping and ablation of focal or ectopic atrial tachycardia has been performed with the CARTO system since the introduction of the mapping technology (Natale 1998). Highly detailed maps demonstrating earliest activation during tachycardia can be acquired. The maps typically show an early location, with propagation conducting centrifugally from the earliest site. The CARTO mapping system has been shown to be highly effective in guiding
the ablation of focal atrial tachycardia, with a relatively small number of ablation lesions. Typical isthmus-dependent atrial flutter has been understood and successfully ablated with conventional EP catheters since the mid-1990s. The circuit is a stable reentrant circuit in the right atrium that preferentially uses the isthmus between the tricuspid valve annulus and the orifice of the inferior vena cava as a critical protected zone of conduction. The ablation strategy involves creating a complete line of conduction block in the cavo-tricuspid isthmus. The CARTO electroanatomical mapping system has been used to graphically demonstrate the anatomical circuit using activation mapping, as well as mark ablation lesions on the three-dimensional map. Importantly, the CARTO system can be used to assess for bidirectional isthmus block and to guide in identifying gaps in the linear lesion set (Nakagawa and Jackman 1998).

The catheter-based ablation of atrial fibrillation has led to a significant increase in the number of interventional EP procedures being performed worldwide. The CARTO system has been shown to be an adjunctive tool for electrophysiologists performing atrial fibrillation ablations (Pappone et al. 1999). The current strategy for the curative ablation of atrial fibrillation in most laboratories involves circumferential ablation of the pulmonary vein antra. The creation of pulmonary vein electrical isolation is considered one of the critical end points for the procedure. The CARTO mapping system allows a graphic three-dimensional representation of the left atrium and pulmonary venous anatomy, as well as the corresponding activation and voltage for each anatomical catheter position. Additionally, the specific anatomical location of ablation sites can be shown on the left atrial three-dimensional geometry. Damage to adjacent structures, such as the esophagus and phrenic nerves, has become a well-recognized potential complication of atrial fibrillation ablation. The CARTO system allows for the representation of these adjacent structures on the three-dimensional geometry, thus allowing the operator to avoid ablation in those regions, thereby reducing the possibility of collateral damage. Currently, previously acquired computed tomography (CT) or magnetic resonance imaging (MRI) images of the left atrial and pulmonary venous anatomy can be integrated into the CARTO system using the CARTOMERGE software. Once the images are registered to the CARTO map, the CT or MRI geometry becomes the three-dimensional map used for catheter navigation and ablation lesion representation.

Electrophysiologists have attempted to use catheter-based ablation technology to replicate the success that cardiac surgeons had with the ablation of ventricular tachycardia in the setting of a prior myocardial infarction. Mapping of ischemic ventricular tachycardia using entrainment with concealed fusion has proven to be successful in patients with hemodynamically stable ventricular tachycardia. The CARTO mapping system has been utilized to acquire sequential point-by-point maps during ventricular tachycardia. The system has been used to map and ablate idiopathic ventricular tachycardia from the right ventricular outflow tract (Nadamanee and Kosar 1998). CARTO can be used in the ablation of ischemic ventricular tachycardia to mark ablation lesions and points of interest. The maps can correlate anatomy with local electrogram amplitude in voltage or “scar” maps, thus identifying the potential substrate for ventricular tachycardia in patients with a prior myocardial infarction. Potential channels within or near the scar can be identified by manipulating the voltage calipers of the CARTO system (Fig. 6.1.1). Targeting these
channels with ablation lesions has been shown to be effective in decreasing ventricular tachycardia episodes (Hsia et al. 2006).

There are several limitations to the CARTO electroanatomical mapping system. Because it requires sequential point-by-point map acquisition, mapping can be time consuming. Infrequent or rapidly changing tachycardias can be difficult to map using this technology. Due to the location reference patch being located on the patient’s back, significant patient movement can negatively affect the accuracy of the catheter’s anatomical location and navigation on the map.

**ENSITE MAPPING SYSTEM**

The EnSite System (St. Jude Medical, St. Paul, MN) was introduced in the United States in the late 1990s (Gornick et al. 1999). The system has two technologies that facilitate electrophysiological mapping and ablation procedures. The EnSite Array catheter and computer-based system allows for noncontact mapping of electrical depolarization throughout a cardiac chamber in a single cycle of the tachycardia. The EnSite NavX system allows for three-dimensional graphic representation of cardiac chambers and catheter navigation using a combination of cutaneous patches and EP catheters. The NavX system can perform isochronal activation mapping and display electrogram voltage for the identification of arrhythmogenic substrates, such as scar in patients with prior myocardial infarction or congenital heart defects. Both the Array and NavX systems can assist in mapping of complex tachyarrhythmias and allow catheter navigation with limited fluoroscopic exposure for patient or physician.
EnSite Array Catheter and System

The EnSite Array mapping system is a computerized system that creates three-dimensional electroanatomical maps without the need for point-by-point contact electrograms. A noncontact balloon catheter creates virtual unipolar electrograms using a mathematical inverse solution to estimate the electrical potentials.

The EnSite system consists of a 9-French multielectrode array catheter mounted on a 7.5-mL balloon, amplifiers, and a computer workstation. There are 64 insulated 0.003-in.-diameter wires around the circumference of the balloon. Each wire has a 0.025-in. break in the insulation that serves as the unipolar electrode. The array size, when inflated, is 1.8 × 4.6 cm. The system has the capability of localizing conventional electrode catheters by emitting a low-current locator signal from the catheter tip electrode and then sensed by two-ring electrodes located on the array. The locator signal is processed to create a three-dimensional model of the cardiac chamber, while the mapping catheter is moved throughout the chamber. With the multielectrode array in position in the chamber, far-field potentials are recorded, amplified, digitized, sampled at 1.2 kHz, and then filtered at 0.1–300 Hz. The resulting signals are used to create 3,360 virtual unipolar electrograms. The computed isopotential or voltage data from the virtual electrograms is displayed on a three-dimensional endocardial map. Isochronal maps can also be created, which represent the progression of activation throughout the chamber relative to a user-defined reference timing point.

Several investigators have demonstrated and validated the EnSite Array system for mapping of atrial and ventricular tachycardias of focal origin (Friedman et al. 2002; Higa et al. 2004). The advent of three-dimensional systems has added greatly to the ability of electrophysiologists to successfully map and ablate complex reentrant tachycardias. Typical and atypical atrial flutter, as well as ventricular tachycardia in patients with a prior myocardial infarction, can be difficult reentrant arrhythmias to map using conventional techniques. Entrainment techniques require a sustained arrhythmia,
which can be difficult in the case of hemodynamically unstable ventricular tachycardia. Noncontact mapping allows for mapping of a single cycle of the tachycardia without the requirement of a sustained arrhythmia. Noncontact mapping has been used to confirm or demonstrate the anatomical location of a reentrant circuit. Because of global mapping capability, entire reentrant circuits can be demonstrated, including isthmus and exit locations (Della Bella et al. 2002; Klemm et al. 2007). Due to its ability to record from multiple sites simultaneously, noncontact mapping is able to identify gaps in previously placed linear lesion sets, such as in the cavo-tricuspid isthmus for typical atrial flutter.

The EnSite Array noncontact system allows for two ways of determining the location of the critical isthmus of slow or protected conduction in reentrant arrhythmias, such as macroreentrant atrial flutter or ventricular tachycardia in patients with a prior myocardial infarction. The standard use of virtual electrogram data displayed on a three-dimensional map may be used for reentrant tachycardias, as previously described for focal tachycardias. The display can demonstrate the propagation of wavefronts, and locate areas with unipolar QS morphology. The unipolar QS electrogram that precedes the onset of the surface P wave or QRS for atrial or ventricular rhythms, respectively, is considered the source of activation from which the depolarizing wavefront spreads to the rest of the chamber. This location is considered to be the exit site from the critical isthmus of slow conduction. Following the identification of the exit site, isthmus conduction can be traced back in time through the diastolic segment of the cardiac cycle. The earliest site of endocardial activation prior to the exit can be identified and marked on the anatomical geometry. Several investigators have published their experience using the EnSite Array for mapping and ablation of reentrant tachycardias. The second component of the EnSite Array system that can be applied to the identification of critical substrates in reentrant tachycardias is an automated voltage program called “Dynamic Substrate Mapping” (DSM). The DSM software program is used in conjunction with the noncontact array and the unipolar virtual electrogram information. DSM is a fully automated program that can be used to characterize signals during electrical diastole. The anatomical area with the greatest voltage during this interval should represent conduction through the critical isthmus or diastolic pathway. Investigators have demonstrated the utility of this tool for the mapping of substrate-mediated tachycardias (Kaltman et al. 2006).

The EnSite Array has several limitations in clinical practice. The balloon catheter can be difficult to deploy in some chambers due to its size or elongated shape. In the right ventricular outflow tract, the balloon can be unstable and cause ventricular ectopy. This can make mapping of clinical ectopy challenging. The accuracy of the virtual electrogram information can be limited in large cardiac chambers. Investigators have validated the accuracy of noncontact mapping if the distance from the center of the balloon array is <4 cm. At distances >4 cm, low-amplitude signals may not be detected. Due to the morphology of the balloon catheter, electrogram information from the polar ends of the array can be less reliable than data from the circumferential or transverse direction. A well-known limitation of unipolar mapping is the recording of far-field signals, which may make the identification of local activation difficult. EnSite’s ability to allow adjustment of the filtering of the signals can alleviate much of this problem.

**EnSite NavX Mapping and Navigation**

The NavX mapping and navigation system received Food and Drug Administration (FDA) approval in the United States in 2003. NavX uses conventional catheters and cutaneous
patches to provide three-dimensional catheter tracking and mapping (Krum et al. 2005). Three pairs of patches are placed along three orthogonal axes, comprising a three-dimensional coordinate system. A low-amplitude 5.7-kHz signal is emitted from the patches and received by catheters within the heart. Catheter location is determined by measuring the resulting electrical potential or field strength received by the catheters.

The system has several features that are useful in mapping and ablation procedures. The system has capabilities similar to conventional EP recording systems, including the display and recording of surface ECG and intracardiac electrograms. The system has the ability to display up to 12 catheters and a total of 64 electrodes on the three-dimensional map, thus allowing catheter navigation with reduced fluoroscopic imaging. The graphic display of multiple catheters can be especially useful in atrial fibrillation ablations, as catheters in the heart and esophagus can be visualized. The NavX system can track the location of conventional EP catheters as they are maneuvered within a cardiac chamber, and a geometric model of the chamber can be created. Activation and voltage data can be acquired and displayed on the three-dimensional geometric model. Ablation lesions can be marked on the surface of the geometric model or as three-dimensional lesions if they are not adjacent to a created geometry. Three-dimensional CT or MRI images can be imported and fused with the created anatomical geometry, which can facilitate anatomically based ablation procedures. Several investigators have published their experience with the NavX, showing a reduction in fluoroscopy times when compared with conventional mapping (Earley et al. 2006; Papagiannis et al. 2006). Recently, studies have been published showing the ability to perform atrial fibrillation mapping and ablation procedures with the NavX using very limited or no fluoroscopy (Takahashi et al. 2005; Estner et al. 2006; Ferguson et al. 2009). The current approach for the mapping and ablation of some arrhythmias, such as atrial fibrillation and scar-mediated ventricular tachycardia, is anatomically based. The NavX system allows for the creation and display of multiple three-dimensional geometric representations of cardiac chambers. The creation of anatomical geometries is achieved by collecting three-dimensional locations as a catheter is maneuvered within a cardiac chamber.

Prior to geometry creation, a positional reference must be determined. The displayed position of all catheters is relative to the location of the positional reference. The preferred positional reference is an intracardiac catheter that will remain in a stable location throughout the procedure. The coronary sinus catheter is often used because of its stability, especially when placed from the superior approach. The effects of respiration on navigation can also be minimized by using the Respiration Compensation tool.

An advantage of the NavX system is that any catheter can be utilized as the Active EnGuide and used for creating geometry. Geometry formation using multiple electrodes from a catheter can decrease the amount of time necessary to create an accurate geometry. NavX allows for the creation of up to 16 different chamber geometries. Multiple geometry formation is especially useful in atrial fibrillation procedures, when the creation of pulmonary vein and atrial anatomies is necessary. The “Reassign” feature allows for points collected as part of one geometry to be added to a different chamber. This can be useful when creating left atrial and pulmonary vein geometries to accurately display the location of the pulmonary vein ostia.

The Diagnostic Landmarking tool allows for the display of electrophysiological data on a three-dimensional map (Mangrum et al. 2005). As the mapping catheter is maneuvered throughout the cardiac chamber, the three-dimensional location, as well as voltage and timing data of each position, is saved. The
corresponding voltage or activation timing information can be displayed as a color map. A single set of collected points can be used to display the different map types.

The Local Activation Time (LAT) feature allows for a color-coded isochronal map of activation times for each collected catheter location. A surface ECG or intracardiac electrogram is determined by the user to be reference waveform for activation time mapping. The local activation time is determined by the relative timing of the local electrogram on the mapping catheter, as compared with the electrogram timing on the reference electrogram. Activation times are displayed on the map as colors from white to purple, corresponding to earliest to latest activation. Similar to activation maps obtained with the CARTO system, NavX maps of focal tachycardias show centrifugal conduction away from an early point. The NavX and CARTO systems similarly show activation of reentrant rhythms utilizing the entire tachycardia cycle length in the chamber and the “early-meets-late” phenomenon (Fig. 6.1.2).

The Diagnostic Landmarking tool also has the capability of displaying the voltage data for each collected catheter location. This feature can be helpful when mapping substrate-mediated tachycardias, such as ventricular tachycardia in patients with a prior myocardial infarction.

**Figure 6.1.2** EnSite NavX Diagnostic Landmarking tool showing an activation map of left atrial flutter. The patient had undergone a previous ablation for atrial fibrillation. The left panel shows the “early-meets-late” phenomenon on the septal mitral valve annulus. With entrainment, conduction around the annulus of the mitral valve appeared to be within the reentrant circuit. The right panel shows a linear lesion set performed between the mitral valve and the left inferior pulmonary vein ostium. This line resulted in mitral isthmus block.
A technique currently being investigated for the ablation of atrial fibrillation involves targeting areas of complex fractionated atrial electrograms. The NavX system includes an automated program to determine and display electrogram fractionation characteristics. The complex fractionated electrogram (CFE) maps can display a fractionation index based on the intervals between multiple, discrete, and local potentials. The NavX system allows for three-dimensional models created from CT or MRI to be incorporated into the system for display with a feature called Verismo. Three-dimensional images that have been previously created on CT or MRI systems can be loaded or the Verismo software can be used to segment or create three-dimensional images from digital “raw” data from CT or MRI. The currently available system allows registration of CT or MRI images with the created geometry and catheter navigation to be displayed on the anatomically accurate three-dimensional map (Fig. 6.1.3).

There are limitations to the EnSite NavX system. Due to the sequential point-by-point mapping, it requires a stable arrhythmia to map. NavX has the capability of simultaneously acquiring electroanatomical data from multiple catheters, which may reduce the requisite duration of the tachycardia. Because it utilizes an internal positional reference, such as the coronary sinus catheter, patient movement can negatively affect the anatomical accuracy of the three-dimensional map. Respiratory motion...
can reduce the positional accuracy of the catheter location and navigation; however, the Respiratory Compensation feature can largely overcome this limitation.

**ABLATION TECHNOLOGY**

The success of ablation procedures is dependent on the production of lesions that block the initiation of focal tachycardias or the propagation of reentrant rhythms. RF energy has been used for catheter ablation of arrhythmias since 1990. It has been shown to be safe and effective for the ablation of cardiac arrhythmias. Alternative energy sources have also been investigated. This section will review the physics of RF lesion formation, as well as briefly describe alternatives to standard RF.

RF energy is a form of alternating current; therefore, the ablation system consists of the ablation catheter as well as a return or dispersive electrode placed on the patient’s skin. The effect of the RF energy occurs at the site of the smaller electrode and highest resistance, that is, the area of contact between the catheter tip and the tissue. RF energy delivered through the tip of an ablation catheter causes thermal injury to the tissue due to resistive heating. This resistive heating occurs in a very superficial region adjacent to the catheter tip, usually 1–2 mm in depth. Further heating is caused by conduction of heat to the surrounding tissue. In addition, there is convective cooling of the catheter tip and the endocardial surface due to blood flow. Permanent tissue injury occurs when tissue temperatures reach 50°C or greater. Tissue heating is indicated by an increase in the catheter tip temperature and a decrease in measured impedance of 5–15 ohms. Standard RF lesions are typically 4–5 mm in width and depth. Lesion size is determined by a number of variables, such as electrode size, amount of energy, and electrode–tissue contact. Larger ablation electrodes (8 or 10 mm) allow greater energy delivery as compared with standard electrodes (4 or 5 mm) due to the larger surface area and increased convective cooling, thereby creating larger volume lesions.

The lesion size with standard RF ablation is limited by the temperature at the interface between the electrode tip and the tissue. At tip temperatures above 70°C, heating is limited by coagulation of blood proteins on the catheter tip. Therefore, current RF systems allow the operator to set a target temperature, with the amount of power being titrated to achieve the desired temperature. If the measured temperature exceeds the target, power delivery will be limited. Irrigated-tip RF ablation catheters allow more energy to be delivered without a significant increase in the electrode–tissue interface temperature. Due to cooling at the tissue–electrode interface, energy is delivered farther into the tissue, thus creating a deeper lesion. Irrigated-tip RF may be advantageous in areas of thick muscle, such as the left ventricle, or in areas with limited blood flow, such as the epicardial space. There are two types of irrigated-tip electrodes currently available: internal irrigation that circulates fluid in a closed system through the catheter tip, and open irrigation that infuses saline through pores in the catheter tip electrode.

Several alternative energy sources have been evaluated for the ablation of cardiac arrhythmias. These include cryoablation, ultrasound, microwave, and laser ablation. Cryoablation involves cooling of the catheter and, subsequently, the tissue to achieve permanent loss of conduction in that region. Cryoablation has been extensively utilized in the ablation of supraventricular tachycardias, especially those using septal accessory pathways. Cryoablation energy has potential advantages over RF energy for certain arrhythmias. It allows for brief, reversible lesions to be delivered, thus reducing the likelihood of permanent heart block when ablating septal accessory pathways. Cryoablation may be less likely to cause collateral damage to adjacent structures when performing left atrial ablation for atrial fibrillation.
High-intensity focused ultrasound and laser energy using balloon systems are currently undergoing investigation for achieving pulmonary vein isolation (Reddy et al. 2008).

**CONCLUSION**

Interventional EP has become a rapidly-growing field of cardiology. Advanced mapping systems and ablation technologies have facilitated the ability to successfully treat routine and complex cardiac arrhythmias.

**REFERENCES**


INTRODUCTION

Procedural sedation, also referred to as moderate sedation, is increasingly becoming a nursing task after being exclusively performed by anesthesiologists.

The Joint Commission on Accreditation of Healthcare Organizations (JCAHO) (2005) defines procedural sedation (moderate sedation/analgesia) as “a drug-induced depression of consciousness during which individuals served respond purposefully to verbal commands either alone or accompanied by light tactile stimulation. No interventions are required to maintain a patent airway, and spontaneous ventilation is adequate. Cardiovascular function is usually maintained.”

It is important to recognize that sedation is part of a continuum that progresses from a minimal level of sedation to a deeper one. Thus, close and diligent care is important to prevent the patient from progressing into a deep sedation state. Meanwhile, the nurse administering sedation and analgesia should be prepared to intervene appropriately if such a situation arises (American Society of Anesthesiologists (ASA) 2004).

Procedural sedation allows the patient to tolerate an unpleasant procedure while maintaining his or her own reflexes and breathing. During invasive electrophysiology (EP) procedures, moderate sedation is used to prevent patients from excessive movements, thus allowing the procedure to be performed with a high degree of safety.

Procedural sedation could also be effectively utilized combined with local anesthesia in a good number of procedures that are uncomfortable or painful, such as pacemaker and defibrillator implants.
Medications utilized for moderate procedural sedation are selected to achieve the following objectives:

- To provide analgesia by elevating the patient’s pain threshold
- To reduce the patient’s level of anxiety and induce sleep
- To allow safe titration to desired level of consciousness

It is very important to distinguish between sedation and analgesia. Medications that provide sedation may decrease the patient’s anxiety without any effects on pain tolerance. Agents used during procedural sedation should be combined to provide the desired effects of reducing anxiety as well as eliminating or reducing painful experiences.

NURSING CARE OF PATIENTS UNDERGOING PROCEDURAL SEDATION

Administration of procedural sedation requires vigilant assessment prior, during, and after administration of sedatives and narcotics to insure safe drug administration and good procedural outcomes.

Patient assessment before administration of procedural sedation is critical to ensure patient’s safety during and after sedation. The goals of preprocedure assessment are as follows:

1. To identify any risk factors that may place the patient at increased risk of complications. These factors are, for example, severe sleep apnea or cardiovascular disease such as severe cardiomyopathy or coronary artery disease. These conditions may complicate achieving successful sedation and analgesia.
2. To identify patients with a high level of anxiety or patients who suffer from addiction to narcotics. These patients may be difficult to manage during the procedure and may be managed better by an anesthesiologist (ASA 2009).

The Joint Commission requires certain standards for presedation assessment that should be performed on each patient prior to beginning procedural sedation. Thorough assessment of the cardiac, respiratory, and neurological systems is a must. All medications administered during procedural sedation will significantly affect these systems. Failure to assess the patient prior to administering these medications may put the patient at a significant risk of serious complications (JCAHO 2005).

The patient’s age, height, weight, body surface area (BSA), and preexisting medical diagnosis should be noted. A physical examination should be performed by a qualified health practitioner immediately prior to the procedure.

Airway Assessment

Procedures for Sedation and Analgesia (ASA 2002)

Positive pressure ventilation, with or without tracheal intubation, may be necessary if respiratory compromise develops during sedation–analgesia. This may be more difficult in patients with atypical airway anatomy. In addition, some airway abnormalities may increase the likelihood of airway obstruction during spontaneous ventilation. Some factors that may be associated with difficulty in airway management are the following:

- **History:** previous problems with anesthesia or sedation; stridor, snoring, or sleep apnea; advanced rheumatoid arthritis; chromosomal abnormality (e.g., trisomy 21).
- **Physical examination:**
  - **Habitus:** significant obesity (especially involving the neck and facial structures)
  - **Head and neck:** short neck; limited neck extension; decreased hyoid–mental dis-
tance (3 cm in an adult); neck mass; cervical spine disease or trauma; tracheal deviation; dysmorphic facial features (e.g., Pierre Robin syndrome)

- **Mouth:** small opening (<3 cm in an adult); edentulous; protruding incisors; loose or capped teeth; dental appliances; high, arched palate; macroglossia; tonsillar hypertrophy; nonvisible uvula
- **Jaw:** micrognathia; retrognathia; trismus; significant malocclusion

### Airway Assessment Utilizing the Mallampati Scale

The Mallampati Scale identifies any structural abnormalities in the airway that may represent difficulties in maintaining an open airway while the patient is sedated. The scale also allows the assessment of the degree of difficulty if intubation is needed.

- The examination is performed while the patient is in the sitting position
- The patient’s head is maintained in a neutral position, and the mouth is opened 5–6 cm and the tongue is protruded.
- Classification of the patient’s airway is based on a description of the anatomical area visualized (see Fig. 6.2.1)

### Preprocedure Instructions to Patients and Their Families

- Nothing per mouth (NPO) status
- Arrival time
- The estimated time and duration for the procedure
- The estimated time for discharge from the hospital, if applicable
- If the patient will be discharged, an adult should be present to accompany the patient
- Patient should know that he or she will not drive for 24 hours after discharge
- Instruct the patient in writing if certain medications should be discontinued prior to the procedure

The patient **must** sign an informed consent prior to administration of any sedative or narcotic agents. Consents signed after administration of sedatives or narcotics are not valid.

Baseline information to be obtained prior to administration of sedation:

1. Vital signs (HR, respiration, and blood pressure)
2. Neurological status
3. Pulse oximetry and end tidal CO₂ (ETCO₂)

This information should be documented before the first dose of sedation is given.

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**Figure 6.2.1** The Mallampati classifications range between 1 and 4.

![Class 1: The entire uvula, soft palate, fauces, and anterior and posterior pillars are visible. This is a sign of an easy-to-manage airway.](image1)

![Class 2: All or most of the uvula is visible, but other structures are less visible. These are basically easy airways to manage.](image2)

![Class 3: Only the base of the uvula can be seen.](image3)

![Class 4: The soft palate and uvula are not visible at all.](image4)
PATIENT MONITORING DURING PROCEDURAL SEDATION

During the entire duration of the procedure, the most important responsibilities of the nurse administering sedation and analgesia are the following:

- Insure that the patient is comfortable and that the level of pain is appropriate and the procedure is well tolerated.
- Insure the patient’s safety by monitoring vital signs, PO\textsubscript{2}, and ETCO\textsubscript{2}.
- Keep an accurate record of the procedure, medications given, and patient’s response to medications.
- Remain with patient during the entire time.
- The nurse administering sedation and analgesia should not participate in any other aspects of the procedure and should not allow any distraction to interfere with monitoring the patient during procedural sedation.

Further Precedation Assessment of Patient

The ASA (2009) has developed a Risk Classification Scale (see Table 6.2.1) for patients undergoing anesthesia.

- Patients in Class 1 and 2 are considered good candidates for sedation procedures.
- Patients in Class 3 and 4 carry higher risks.

Emergency Equipment That Must be Available Prior to Initiation of Procedural Sedation

1. Well-stocked crash cart should be within easy reach.
2. Established protocol on how to summon additional resources in case of emergency. All personnel involved in the procedure should be able to follow that protocol. Who to summon? What method will be used such as pagers or phone numbers, which should be posted in the procedure room?
3. All equipment should be age- and size-appropriate for the patient receiving procedural sedation.
4. All equipment should be checked daily for proper functioning, and all staff members in the room should be well trained on how to use these equipment in case of emergency (Table 6.2.2).

Monitoring of the Cardiac Function

Patients undergoing procedural sedation should be monitored for heart rhythm and rate. The Joint Commission requires this as standard of care (JCAHO 2005). In EP labs, although heart rhythm and rate are continuously monitored through other recording and mapping systems, it is the responsibility of the nurse administering sedation to monitor the rate and rhythm independently and alert the physician and the other EP staff to any changes. It would also be the responsibility of the nurse administering sedation to cardiovert/defibrillate if needed. It is a standard practice in most EP labs to apply remote defibrillation patches to all patients undergoing diagnostic and interventional procedures in the EP lab.

Table 6.2.1  ASA classification of physical status.

<table>
<thead>
<tr>
<th>Class</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Normal healthy patient</td>
</tr>
<tr>
<td>2</td>
<td>Mild systemic disease</td>
</tr>
<tr>
<td>3</td>
<td>Severe systemic disease that limits activity, but is not incapacitating</td>
</tr>
<tr>
<td>4</td>
<td>Incapacitating systemic disease that is a constant threat to life</td>
</tr>
<tr>
<td>5</td>
<td>Moribund patient not expected to survive 24 hours with or without operation</td>
</tr>
</tbody>
</table>
cardiomyopathy or in patients with known low pressure.

With automatic noninvasive blood pressure monitoring, the frequency of the measuring blood pressure can be set to obtain a reading as needed. It is advisable though that blood pressure readings should not be monitored more than every 15 minutes and should be recorded accordingly. Appropriate cuff size should be utilized according to the patient’s age and arm size.

Any blood pressure reading that is widely different from baseline should immediately be reported to the physician. It is expected that with sedation and analgesia, blood pressure may drop; however, a mean blood pressure should always be maintained at 60 or higher to insure proper organ perfusion (ASA 2009).

### Table 6.2.2  Emergency equipment list.

<table>
<thead>
<tr>
<th>Standard equipment</th>
<th>Noninvasive and Invasive Blood Pressure Monitoring</th>
</tr>
</thead>
<tbody>
<tr>
<td>• IV access equipment</td>
<td>• Method to summon help</td>
</tr>
<tr>
<td>• Oxygen delivery system</td>
<td>• Policy and procedure manual</td>
</tr>
<tr>
<td>• Cardiac monitoring system</td>
<td>• Documentation forms</td>
</tr>
<tr>
<td>• Pulse oximetry</td>
<td>• Emergency supplies (see below)</td>
</tr>
<tr>
<td>• Blood pressure monitoring device</td>
<td>• Gloves</td>
</tr>
<tr>
<td>• IV access equipment</td>
<td>• Medications (see list below)</td>
</tr>
</tbody>
</table>

**Emergency airway supplies**

- Nasopharyngeal airways (appropriate sizes)
- Oropharyngeal airways (appropriate sizes)
- Tongue blades
- Laryngoscope handle
- Macintosh blades (appropriate sizes)
- Miller blades (appropriate sizes)
- ET tubes (appropriate sizes)
- Bag valve mask device (appropriate sizes)
- Face masks (appropriate sizes)

**Emergency medications**

- Oxygen source with gauge
- Glucose
- Atropine
- Lidocaine
- Epinephrine (1:1,000 and/or 1:10,000)
- Magill forceps of appropriate size
- Sterile lubricant
- Suction equipment
- Oxygen supply
- Spare lightbulbs
- Yankauer suction catheters
- Suction catheters
- Stylets
- Dopamine
- Dobutamine
- Sodium bicarbonate
- Naloxone
- Methylprednisolone

Note: These medications and equipment may be part of the emergency cart. IV, intravenous; ET, endotracheal.
Pulse Oximetry

Pulse oximetry monitoring has been an essential tool to assess the respiratory function of patients undergoing procedural sedation, and it is required by the Joint Commission as standard of care. It allows the monitoring of hemoglobin saturation as a method of detection of impending hypoxia, thus allowing rapid intervention to prevent further deterioration of the patient’s respiratory status.

Pulse oximetry probes are relatively easy to use and can be placed either on the fingers, toes, or even earlobes. However, it has several disadvantages when used in patients with poor peripheral circulation, severe hypotension, hypothermia, or severe anemia.

Pulse oximetry also may not be accurate when PO\(_2\) drops to levels below 60%, and it is not a good tool to indicate the patient’s ventilatory status. The ventilatory status is the ability of the lungs to take in oxygen and expel CO\(_2\). Rising level of CO\(_2\) in the blood cannot be detected by pulse oximetry (ASA 2009).

ETCO\(_2\) Monitoring

ETCO\(_2\) monitoring measures expired carbon dioxide and provides information on the patient’s ventilation. This type of monitoring is most commonly utilized with deep sedation and general anesthesia; however, it is now widely used during procedural sedation. Rising levels of CO\(_2\) in the blood is detected much earlier than the fall of oxygen saturation detected by PO\(_2\), allowing for early intervention before hypoxemia becomes severe (ASA 2009).

Administration of Supplemental Oxygen during Procedural Sedation

Most EP lab policies indicate that all patients receiving procedural sedation will receive supplemental oxygen. Oxygen therapy devices are advantageous for a number of reasons.

1. Use of sedation medications interfere with the patient’s ability to breathe adequately in response to falling level of oxygenation
2. The patient may not be able to institute compensatory measures during this period.
3. Oxygenation level may fall to a dangerously low level in patients with comorbidities such as lung or heart disease.

Oxygen therapy devices utilized in procedural sedation range from simple (nasal cannula) to the more complex (nonrebreather masks and Venturi masks). Each device must be selected according to the patient’s preexisting conditions, age, and level of sedation achieved.

Table 6.2.3 summarizes few of the oxygen delivery devices that may be used during procedural sedation. Each device is suitable for a certain situation. Selection is based on the patient’s condition, length of procedure, and level of sedation achieved.

Restlessness and agitation during procedural sedation should always be considered as signs of hypoxemia rather than pain or discomfort. Careful assessment should be performed to rule out hypoxemia since deterioration can occur very rapidly. The patient should be stimulated so level of consciousness can be determined. Deeper level of consciousness should be corrected immediately with reversal agents. However, if the cause of agitation is pain or discomfort, the patient should be able to indicate so verbally.

Assessment of vital signs may be done as frequently as every 5 minutes, but should not be longer than every 15 minutes. Assessment should also be performed 5 minutes after administration of any sedation dose to determine the patient’s response to that dose (American Nurses Association (ANA) 1991; JCAHO 2005).

Frequent assessment of the airway should also be performed. During procedural sedation,
breath sounds should be clear to auscultation. Two of the main risks during sedation are airway obstruction and, in some patients, accumulation of fluids in the lungs leading to pulmonary edema. During long EP procedures, considerable volume of fluids may be infused. Patients with low ejection fraction may not be able to tolerate these levels of fluids. Administration of diuretics intravenously may be helpful to prevent deterioration of the respiratory status of the patient.

**Assessment of the Level of Consciousness during Procedural Sedation**

The Ramsay Sedation Score is the most used scale to assess the patient’s level of consciousness. The Modified Ramsay Sedation Score (Table 6.2.4) is used in the intensive care unit (ICU) (American Nurses Association (ANA) 1991), and may be used in the EP lab for the same purpose.

**Care of Patients after Receiving Procedural Sedation**

At the end of the EP procedure and after the last dose of sedation has been administered, the patient should continue to be monitored until all vital signs and neurological status have returned to baseline.

- The patient will be transferred to a holding or recovery area where monitoring should be continued for a few hours.
- The nurse who has administered procedural sedation should accompany the patient and give a full detailed report to the receiving nurse in the recovery area. This report should include all preprocedure baseline information as well as any event that may have happened during the procedure.

### Table 6.2.3  Suggested oxygen therapy devices.

<table>
<thead>
<tr>
<th>Type</th>
<th>FiO&lt;sub&gt;2&lt;/sub&gt;</th>
<th>Flow (L/min)</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nasal cannula</td>
<td>24–40%</td>
<td>3–6</td>
<td>Comfortable, provides low flow; may not be suitable for long procedures</td>
</tr>
<tr>
<td>Simple face mask</td>
<td>25–55%</td>
<td>5–8</td>
<td>Allows for moderate flow; must be used with humidification</td>
</tr>
<tr>
<td>Nonrebreather mask</td>
<td>40–100%</td>
<td>6–15</td>
<td>Mask with a reservoir bag and one-way valve, achieves high concentration of FiO&lt;sub&gt;2&lt;/sub&gt;; most suitable for EP procedures</td>
</tr>
<tr>
<td>Venturi mask</td>
<td>24–55%</td>
<td>10–15</td>
<td>Adjustable flow; suitable for shorter procedures not needing high flow of O&lt;sub&gt;2&lt;/sub&gt;</td>
</tr>
</tbody>
</table>

Note: FiO<sub>2</sub>, fractionated inhaled oxygen.

### Table 6.2.4  Modified Ramsay Scale.

<table>
<thead>
<tr>
<th>Awake states</th>
<th>Sleep states</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Patient anxious, agitated, or restless</td>
<td>1. Patient asleep, sluggish response to loud auditory stimulus</td>
</tr>
<tr>
<td>2. Patient cooperative, oriented, tranquil</td>
<td>2. Patient has no response to loud auditory stimulus but does respond to painful stimulus</td>
</tr>
<tr>
<td>3. Patient asleep, brisk response to loud auditory stimulus</td>
<td>3. Patient does not respond to painful stimulus</td>
</tr>
</tbody>
</table>

The nurse who has administered procedural sedation should accompany the patient and give a full detailed report to the receiving nurse in the recovery area. This report should include all preprocedure baseline information as well as any event that may have happened during the procedure.
The receiving nurse should be aware of any potential complications that may take place during the recovery period.

Finally, the postprocedure orders should be explained thoroughly to the receiving nurse, including when to allow the patient eat or drink.

The patient may need reorientation to time, date, and place. Some of the medications used for sedation induce a degree of amnesia, such as Midazolam. It is not uncommon in elderly patients that amnesia or confusion could occur due to sedation. This group of patients may need longer recovery time.

**DOCUMENTATION**

Documentation of the events of the procedure is critical from the clinical and legal points of view. Good and accurate documentation allows continuity of care after leaving the procedure room and serves as future reference to the patient’s response to the medications and the procedure.

Documentation should include the following:

- Preprocedure assessment and baseline information, including premedications, preprocedure vital signs, and neurological examination
- Intraprocedural log of vital signs, sedatives and narcotics administered, and the patient response to these medications; any adverse events or reversal agents given during the procedure
- Postprocedure status, vital signs, and neurological status
- Patient status at time of discharge from recovery area and destination
- Discharge instructions that were given to the patient, family, or accompanying adults; it is recommended that these instructions to be given in the presence of another accompanying adult since the patient may not retain all the information given due to the effect of the sedatives and narcotics given

**POTENTIAL COMPLICATIONS DURING PROCEDURAL SEDATION**

Although adverse events and complications are rare, procedural sedation is not without potential risks (ASA 2004). The most common complications are related to preexisting conditions such as sleep apnea, cardiomyopathy, and severe pulmonary problems. Thorough preprocedure assessment and diligent monitoring can reduce or eliminate these complications.

Complications of procedural sedation include:

- Over- or undersedation: These perhaps are the most common
- Respiratory insufficiency or arrest: Since in elderly patients some sedatives and/or narcotics, especially opiates and barbiturates, tend to suppress their respiratory drive, care should be taken to slowly titrate these drug’s dosages to avoid such complication
- Cardiac arrest
- Airway obstruction due to increased secretion or due to the tongue falling back
- Nausea and vomiting
- Hemodynamic instability due to hypotension or hypoxia
- Malignant hyperthermia
- Aspiration of oral secretions or vomitus
- Dysrhythmia, usually bradycardia
- Urinary retention, especially in the elderly

Certain groups of patients are at increased risk of the above mentioned complications:

1. Patients who are either very young or very old
2. Patients with reduced cardiac function as in severe cardiomyopathy or congestive heart failure
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3. Patients who are obese who usually suffer from sleep apnea
4. Patients with liver and/or renal disease
5. Patients with severe emphysema or other underlying pulmonary disease

SUMMARY

Procedural sedation, or also known as moderate sedation, is generally safe when administered by nonanesthesiologists during EP procedures. The Joint Commission and the ASA have provided standards and guidelines for registered nurses to administer moderate procedural sedation with patient safety in mind. It is important to realize that “sedation and analgesia” comprise a continuum of states ranging from minimal sedation through deep state or general anesthesia (ASA 2002).

The ability of the registered nurse administering sedation to recognize the patient’s transition from one state to the other is of paramount importance. This is accomplished by thorough and vigilant assessment and by being prepared to intervene should any adverse events arise. Thorough knowledge of the pharmacological agents used and adhering to the established protocols will insure that the patient experiences good care and procedure outcomes.
INTRODUCTION

The implantable electronic cardiac pacemaker was first used in a human about 50 years ago, and the implantable cardioverter defibrillator (ICD) was first used in a human about 30 years ago (Kusumoto and Goldschlager 2002). Since then, the indications for and use of pacemakers and ICDs have increased greatly (Birnie et al. 2006; Goldberger and Lampert 2006; Mond et al. 2008). For example, during 2001 alone, about 220,000 pacemakers and 120,000 ICDs were implanted into patients in the United States (Mond et al. 2008). Because of these trends, millions of Americans now have implantable cardiac devices (Maisel et al. 2006; Mond et al. 2008). Therefore, it is likely that clinicians (i.e., physicians, physician assistants, nurse practitioners, nurses, and others) will care for increasing numbers of patients with implantable cardiac devices and be involved in device-related clinical ethical dilemmas. For example, many patients with devices are elders (Mond et al. 2008), and since most deaths occur among elders (US Bureau of the Census 2000), clinicians may encounter dying patients who request device deactivation (i.e., reprogramming the device so that it is nonfunctional) in order to avoid uncomfortable ICD shocks and other reasons. The clinician, however, may view carrying out such a request as unethical. Hence, clinicians should be familiar with the ethical dilemmas commonly encountered when caring for patients who have indications for device therapies or who have implantable cardiac devices. This chapter reviews these dilemmas and approaches to resolving them using illustrative cases. Established guidelines (Epstein et al. 2008) were used to categorize device therapy indication classes.
Table 6.3.1  Principles of ethics.

<table>
<thead>
<tr>
<th>Principle</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Beneficence</td>
<td>Duty to promote patient interests</td>
</tr>
<tr>
<td>Nonmaleficence</td>
<td>Duty to prevent or do no harm</td>
</tr>
<tr>
<td>Respect for patient autonomy</td>
<td>Duty to respect patients and their health care-related values, preferences, and goals</td>
</tr>
<tr>
<td>Justice</td>
<td>Duty to treat patients fairly (i.e., based on medical need, not on patient characteristics such as gender, age, or race)</td>
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**PRINCIPLES OF ETHICS**

Clinical ethics is a “discipline that provides a structural approach for identifying, analyzing, and resolving” moral problems that arise in the care of patients (Jonsen et al. 2002, p. 1). Four prima facie ethics principles encompass most ethical concerns in clinical practice: beneficence, nonmaleficence, respect for patient autonomy, and justice (Table 6.3.1) (Beauchamp and Childress 2009). In general, no principle has priority over another; priority depends on the circumstances of a given clinical scenario. In addition, these principles may be at odds with each other. For example, a clinician’s desire to help a patient (e.g., implantation of an ICD to prevent sudden death) may be at odds with the patient’s health care-related values and preferences (e.g., refusal of device implantation).

**ETHICAL ISSUES IN IMPLANTABLE CARDIAC DEVICE-RELATED CLINICAL PRACTICE**

**Fostering Beneficence**

The principle of beneficence refers to the clinician’s duty to promote the best interests of the patient (Beauchamp and Childress 2009). This duty takes precedence over the clinician’s self-interests. Beneficent clinicians maintain competence and strive for excellence. Beneficence requires that clinicians share their assessments and recommendations with patients completely and clearly. Beneficent clinicians present options for treatment not as a menu of choices, but as a hierarchy based on safety, efficacy, and patients’ health care-related values and goals. Finally, beneficent clinicians ensure that patients understand their assessments and recommendations.

**Case 6.3.1**

A 77-year-old woman with metastatic breast cancer who just completed a course of chemotherapy is admitted to the hospital after an episode of syncope. Evaluation reveals third-degree (complete) heart block. Just prior to admission, she was contemplating hospice care. Many patients benefit from device therapies, especially those with class I indications. The patient in the case has a class I indication for a permanent pacemaker. However, she is also suffering from cancer and contemplating hospice care. Nevertheless, she still may benefit from pacemaker implantation and therapy. In situations such as these, beneficent clinicians avoid making unilateral and paternalistic treatment decisions. Instead, these decisions should be patient-centered. The clinician caring for the patient in the case should describe the benefits and risks of pacemaker implantation and therapy. If pacemaker implantation and therapy are consistent with the patient’s health care-related values and goals, then the clinician should proceed with device implantation. If pacemaker therapy is not expected to benefit the patient (e.g., longevity), or is inconsistent with the patient’s values and preferences (e.g., avoidance of invasive procedures), then device implantation should be foregone and the patient should be offered care that minimizes
symptoms related to her rhythm abnormality and, given her cancer, consultation with a palliative medicine specialist.

**Promoting Nonmaleficence**

The principle of nonmaleficence refers to the clinician’s duty to not harm patients (Beauchamp and Childress 2009). This principle is closely allied with the principle of beneficence. Indeed, weighing the potential benefits and harms of a treatment is common in clinical practice. Regarding implantable cardiac devices and therapies, clinicians should inform patients not only of the benefits of such treatments but also of the harms (e.g., ICD shocks, device replacements, burdens of monitoring). If the treatment is used, then clinicians should attempt to minimize harms associated with it. Conflicts of interest should not compromise the clinician’s nonmaleficence duties (ABIM Foundation et al. 2002). For example, a clinician’s relationships with industry should not interfere with decisions that are in patients’ best interests.

**Case 6.3.2**

A 28-year-old woman with long QT syndrome who has an ICD because of a history of ventricular fibrillation (VF) arrest (class I indication) is brought by ambulance to the hospital after experiencing an ICD shock. She is nonadherent with her medications and device follow-up. In fact, she is seen only after being admitted to the hospital following appropriate ICD shocks. The patient is verbally abusive to the care team and the team wonders if she should be dismissed from the practice.

Nonmaleficence also encompasses the clinician’s duty not to abandon patients (Quill and Cassel 1995). In the case, the care team understandably is frustrated with the patient’s behavior. Nevertheless, despite the patient’s behavior, the team is obligated to care for her until an alternative team is identified and the patient and the alternative team agree to the transfer of care. In the meantime, the care team should attempt to discern and address the reasons for the patient’s behavior. Social workers, chaplains, and psychologists can be very helpful in these situations.

**Obtaining Adequate Informed Consent**

Informed consent (and refusal) derives from the principle of respect for patient autonomy. Patient autonomy is maximized when patients understand their diagnoses and treatment options and participate fully in clinical decision making. Clinicians should ensure that patients are informed about their diagnoses and treatment options; codes of ethics endorse this obligation (American Medical Association Council on Ethical and Judicial Affairs 2002; Snyder and Leffler 2005).

Clinicians also have legal obligations related to informed consent. It was the 1957 court case, *Salgo v. Stanford University*, in which the term “informed consent” was first used. The patient, who developed paralysis following an invasive procedure, claimed he was not informed of the procedure’s risks. The court agreed and declared that clinicians violate their duties to patients if they withhold from their patients facts required to make informed decisions (*Salgo v. Leland Stanford Jr. Univ. Bd. of Trustees* 1957). Later cases defined the amount of information patients should receive in order for consent to be “informed.” *Canterbury v. Spence* established the “reasonable patient” standard; that is, what a “reasonable patient” would need to know in order to make an informed decision about a proposed intervention (*Canterbury v. Spence* 1972; Meisel and Kuczewski 1996). Today, the “reasonable patient” standard is widely used.
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regard device therapies as life-sustaining (Mueller et al. 2008). Clinicians, however, should recognize that patients have the right to refuse or request the withdrawal of unwanted treatments. Patients also have the right to refuse or request the withdrawal of treatments to which they previously consented (Quill et al. 1994). These rights derive from the principle of respect for patient autonomy. Regardless of the clinician’s intent, commencing or continuing a treatment that a patient has refused is unethical and, from a legal standpoint, may be viewed as battery (Snyder and Leffler 2005).

Nevertheless, some clinicians may be reluctant to honor such requests because of perceived nonmaleficence duties (i.e., not to harm) and legal concerns (e.g., prosecution for unlawful death). A number of prominent court decisions, however, have clarified patients’ rights to refuse or request the withdrawal of life-sustaining treatments. For example, the New Jersey Supreme Court, in the Quinlan case, ruled that the right to privacy includes the right to refuse unwanted treatments including life-sustaining treatments (In re Quinlan 1976). The US Supreme Court, in the Cruzan case, affirmed that competent patients have the right to refuse unwanted treatments and that incompetent patients have the same right (as exercised through previously expressed wishes and surrogate decision makers) (Cruzan v. Director, Missouri Dept. of Health 1990). Notably, there are no ethical or legal differences between refusing and withdrawing treatments (Fairman 1992; Gostin 1997; Snyder and Leffler 2005). Finally, honoring a patient’s refusal of or carrying out a patient’s request to withdraw a life-sustaining treatment is not so much respecting a “right to die,” but rather respecting a right to be left alone (Gostin 1997).

In addition, honoring refusals of and carrying out requests to withdraw life-sustaining treatments are not forms of assisted suicide or euthanasia. In assisted suicide, the patient ter-

Case 6.3.3
Following a myocardial infarction, a 73-year-old man develops persistent second-degree heart block at the atrioventricular (AV) node level. He has no symptoms related to this rhythm abnormality. A cardiology trainee sees the patient, tells him he needs a pacemaker (class IIb indication), hands the patient a consent form, asks him to read and sign it, and leaves the patient’s room. The trainee returns 1 hour later to retrieve the signed form.

The three essential elements of informed consent are information (e.g., diagnosis, treatment options, risks and benefits of these options), patient decision-making capacity, and patient voluntariness (Marsh 1986). A signed consent form is not the same as informed consent. While a signed form may be needed for compliance purposes, it is not a substitute for a detailed and documented discussion with the patient. The patient in the case may read and sign the consent form, but may not understand it, much less the indications for, risks and benefits of, and alternatives to pacemaker placement.

Clinicians should obtain informed consent for most interventions. Under certain circumstances, however, clinicians cannot obtain informed consent (e.g., emergencies) or must obtain it from surrogates (e.g., for patients who lack decision-making capacity).

Managing Refusals and Requests for Withdrawal of Device Therapies
Patients commonly refuse treatments and many patients—especially those who are dying—request the withdrawal life-sustaining treatments (e.g., mechanical ventilation, hemodialysis, tube feeding). Most clinicians who care for patients with implantable cardiac devices regard device therapies as life-sustaining (Mueller et al. 2008). Clinicians, however, should recognize that patients have the right to refuse or request the withdrawal of unwanted treatments. Patients also have the right to refuse or request the withdrawal of treatments to which they previously consented (Quill et al. 1994). These rights derive from the principle of respect for patient autonomy. Regardless of the clinician’s intent, commencing or continuing a treatment that a patient has refused is unethical and, from a legal standpoint, may be viewed as battery (Snyder and Leffler 2005).

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In addition, honoring refusals of and carrying out requests to withdraw life-sustaining treatments are not forms of assisted suicide or euthanasia. In assisted suicide, the patient ter-
minates his or her life using a method prescribed by a clinician (e.g., lethal prescription). In euthanasia, the clinician terminates the patient’s life (e.g., lethal injection). In assisted suicide and euthanasia, the cause of death is the means prescribed or administered by the clinician; the intent is patient death. When a patient dies after refusal or withdrawal of a life-sustaining treatment, the cause of death is the underlying disease; the intent is freedom from a treatment the patient perceives as burdensome (Meisel et al. 2000; Rhymes et al. 2000; Snyder and Leffler 2005). If, however, the clinician’s intent is to hasten or cause a patient’s death by withdrawal of the life-sustaining treatment (e.g., device deactivation), then the act is akin to euthanasia.

Case 6.3.4
An 81-year-old man with a history of myocardial infarction and a left ventricular ejection fraction (LVEF) of 30% underwent ICD implantation for primary prevention of sudden cardiac death (SCD) (class I indication) 5 years ago. He has been appropriately shocked once. He undergoes routine device interrogation, which indicates that the device’s battery has reached depletion indicators. Pulse generator replacement is recommended. Citing worry over being shocked, the bother associated with monitoring, and a desire to be left alone, he declines.

Not surprisingly, some patients who have indications for device therapies refuse device implantation. As with other treatments, forcing a patient to undergo device implantation is unethical and illegal. Instead, clinicians should ensure that such refusals are informed and, if so, respect them. Likewise, patients may refuse treatments to which they previously consented. The patient in the case, who has an ICD, refuses device replacement. The clinician caring for the patient should ensure that the patient’s decision is informed and, if so, respect it.

Case 6.3.5
A 72-year-old woman with metastatic breast cancer is hospitalized for pain control. Three years ago, she underwent ICD implantation for primary prevention of SCD in the setting of ischemic cardiomyopathy and an LVEF of 30% (class I indication). She lapses into a coma and within the last hour has received two appropriate ICD shocks for ventricular arrhythmias. Her family requests device deactivation.

In this case, the clinician should determine if carrying out the family’s request is consistent with the patient’s previously expressed wishes and, if so, carry out the request. ICD deactivation will prevent uncomfortable shocks during the last hours of the patient’s life. In this setting, ICD deactivation prevents harm. When death occurs, the cause of death is the patient’s underlying disease, not ICD deactivation. Clinicians should ensure that terminally ill patients who undergo device deactivations have adequate palliative care. Notably, a majority of clinicians who care for patients with implantable cardiac devices have deactivated pacemakers and ICDs in terminally ill patients (Mueller et al. 2008).

In 2010, the Heart Rhythm Society (HRS) released an expert consensus statement on the management of implantable cardiac devices in patients requesting withdrawal of therapy or nearing the end of life (Lampert et al. 2010). This statement outlines the ethical, legal, and religious principles that underlie the permissibility of withdrawing life-sustaining treatments from patients who no longer want the treatments including device therapies. The statement also provides guidance on the man-
agement of patients who have devices and are nearing the end of life (e.g., logistics of device deactivation, the role of palliative medicine specialists). Clinicians who care for patients with implantable cardiac devices should be familiar with this helpful document.

At times, a clinician may conscientiously object to a patient’s request to withdrawal life-sustaining treatments including deactivating implanted cardiac devices, especially pacemakers in pacemaker-dependent patients (Mueller et al. 2008). Clinicians should not be compelled to participate in practices they find morally unacceptable. In these situations, the clinician should transfer care of the patient to another clinician (Quill et al. 1994; Snyder and Leffler 2005; Lampert et al. 2010).

**Engaging Patients with Implantable Cardiac Devices in Advance Care Planning**

Advance care planning is a process in which a patient identifies his or her values and preferences regarding future health care and a surrogate decision maker in the event the patient loses capacity to make decisions (Snyder and Leffler 2005). This process includes discussing the patient’s values and preferences with his or her loved ones, potential surrogates, and clinicians, documenting the discussions in the patient’s medical record, and completing an advance directive (AD).

In general, there are two types of ADs: the living will and the durable power of attorney for health care. The living will allows a patient to list his or her health care-related values and treatment preferences. The durable power of attorney for health care allows a patient to specify a surrogate in the event the he or she loses the capacity to make decisions. Some ADs have features of both a living will and a durable power of attorney for health care. Clinicians should view the AD as an extension of the autonomous patient.

Few patients discuss end-of-life issues with their clinicians (Layson et al. 1994). In addition, few patients have completed their own ADs. In one study (Hanson and Rodgman 1996), only 10% of decedents in the United States had ADs. Likewise, advance care planning is uncommon among patients with implantable cardiac devices. For example, few patients with ICDs discuss ICD deactivation with their clinicians or know that the device can be reprogrammed in order to avoid shocks (Goldstein et al. 2007). Studies have shown that 30–60% of patients with pacemakers and ICDs have ADs; very few of their ADs, however, specifically mention the device (Berger et al. 2006; Tajouri et al. 2010a,b). Unfortunately, some dying patients with ICDs—including patients in hospices—experience shocks during the last hours of life (Goldstein et al. 2004, 2010; Lewis et al. 2006). Patients with ICDs who have participated in advance care planning, however, are less likely to experience shocks during the dying process (Lewis et al. 2006).

### Case 6.3.6

A 68-year-old widow presents with involuntary weight loss; evaluation reveals metastatic renal cell carcinoma. Three years ago, she underwent implantation of a permanent pacemaker for second-degree AV block with associated symptomatic bradycardia (class I indication). The patient has decision-making capacity. She previously completed an AD, in which she named a sister as her surrogate decision maker (if necessary). She also included a statement in her directive that she did “not wish to be kept alive artificially if there is no hope for survival.”

Most patients welcome advance care planning but prefer that their clinicians initiate the process (Layson et al. 1994). Patients who are undergoing device implantation or have devices should be encouraged to engage in
advance care planning and execute their own ADs (Lampert et al. 2010). Patients should specify their preferences regarding device management at the end of life. The patient in the case has engaged in advance care planning and executed an AD. At this point in time, while she has decision-making capacity, she should be asked to clarify (and document) what she means by the statement in her AD, “not wish to be kept alive artificially if there is no hope for survival,” particularly as it pertains to the pacemaker. If she loses decision-making capacity, her surrogate would be forced to interpret the statement.

**Assuring Proper Surrogate Decision Making**

Clinicians commonly care for patients who lack decision-making capacity. Under these circumstances, clinicians must rely on surrogates to make decisions. If the patient has an AD and identifies a surrogate in the AD, that choice should be honored (Snyder and Leffler 2005). If the patient does not have an AD, clinicians must identify the appropriate surrogate. While the ideal surrogate is one who best understands the patient’s health care-related values and preferences (Hayley et al. 1996), many states have laws that specify a hierarchy of surrogates (e.g., spouse followed by adult child). Clinicians should adhere with these laws.

**Case 6.3.7**

An 80-year-old man with respiratory failure due to trilobar pneumonia is admitted to the intensive care unit. He has an ICD because of history of sustained ventricular tachycardia (VT) in the setting of structural heart disease (class I indication). He rapidly develops respiratory failure and is intubated and mechanically ventilated; he also loses decision-making capacity. He does not have an AD. After several weeks of intensive care, the patient’s wife, citing her husband’s previously expressed wishes, requests withdrawal of ventilator therapy and ICD deactivation to avoid shocks. The patient’s daughter—an attorney—arrives and disagrees with her mother.

In this case, the most appropriate surrogate is the wife since most states—in the absence of an AD—legally specify her as such. In the absence of such laws, she likely remains the most appropriate surrogate given her relationship with the patient and her knowledge of his health care-related values and preferences.

When making decisions, a surrogate should adhere with the instructions in the patient’s AD (if one exists) and base decisions on the patient’s—not the surrogate’s—values and preferences if known (i.e., “substituted judgment”) (Hayley et al. 1996). If necessary, clinicians can remind surrogates of this obligation by stating, “If the patient could wake up for 15 minutes and understand his or her condition fully, and then had to return to it, what would he or she tell you to do?” (Quill 2005, p. 1,633). If unknown, surrogates should base their decisions on clinical, quality of life, and other factors (i.e., “best interest”) (Hayley et al. 1996).

**Dealing with Device Advisories**

Device advisories (also known as recalls and safety alerts) are not uncommon; between 1990 and 2000, the Food and Drug Administration (FDA) issued 52 advisories involving more than 500,000 patients (Maisel et al. 2001; Maisel 2004). Manufacturers usually notify clinicians about advisories by letter; these letters describe the problems, devices affected, risks of malfunction, and management recommendations. Management decisions, however, rest with clinicians and their affected patients (Maisel...
Most patients learn about advisories from the media, but prefer to learn about them from their clinicians (Stutts et al. 2007). Patients with devices under advisory may experience several types of harm: harm due to device malfunction (i.e., failure to deliver therapy and inappropriate delivery of therapy), harm associated with device replacement after an advisory, and psychological harm due to the advisory itself. Of the 2.25 million pacemakers and more than 415,000 ICDs implanted in the United States during the years 1990–2002, more than 17,000 devices (8,834 pacemakers and 8,489 ICDs) were explanted due to confirmed malfunctions and 61 deaths were attributed to malfunctions (Maisel et al. 2006). A majority of physician respondents to one survey reported that they had replaced at least one device because of an advisory or malfunction within the three previous years (Maisel 2004).

Recent device advisories cited failure risks between 0.009 and 2.6% (Kapa et al. 2007). However, the risk of complications associated with device replacement is not negligible: 1.2–8.1% (Gould et al. 2006; Kapa et al. 2007). These complications include pocket infection, hematoma requiring reoperation, and death. Thus, while clinicians may desire to prevent advisory-related harms to their patients who have devices, replacing a device is not without risk.

Clinicians should explain the reasons for the device advisory using language that is understandable and discern patients’ concerns. Throughout the discussion, clinicians should check for patient comprehension. Clinicians should base their recommendations on available evidence, weighing the risks of replacing versus the risks of retaining the device (i.e., clinicians should not categorically recommend device replacement after all advisories since such an approach would cause excessive harm). Clinicians should also base their recommendations on patient factors (e.g., comorbid illness), the indications for device therapy, the nature of the advisory, and published guidelines (Carlson et al. 2006). Notably, mathematical models may facilitate patient–clinician discussions and decision making in response to device advisories (Amin et al. 2006; Gula et al. 2007).

### Case 6.3.8

Two years ago, a 54-year-old man underwent implantation of a permanent pacemaker for second-degree heart block at the AV node level (class IIb indication). He is not pacemaker dependent. He now presents to your office demanding device replacement following the release of an FDA advisory regarding his device.

In this case, if the risks of continuing with the device on advisory outweigh the risks of device replacement, then the clinician should recommend device replacement. If the risks of device replacement outweigh the risks of continuing with the current device, then the clinician should recommend device retention and explain the reasons for the recommendation and how the patient and device will be monitored. Patients, typically, will follow their clinician’s recommendation. If, despite the aforementioned discussion, the patient remains steadfast in demanding device replacement, then the clinician has several options including proceeding with device replacement and seeking a second opinion from a colleague.

### Handling Requests for Interventions

Patients or their surrogates often request treatments and other interventions. Clinicians should grant such requests if they are reasonable and within standards of care. However, clinicians are not obliged to carry out requests
for treatments that are contraindicated, non-beneficial, or violate his or her conscience (Weijer et al. 1998). Nevertheless, the clinician should attempt to understand the reasons for such requests.

**Case 6.3.9**

A 78-year-old woman with metastatic breast cancer is admitted to the hospital after falling at home; she spends most of the day in bed because of bone pain. She is admitted to a cardiac telemetry unit. Her heart rate is consistently less than 40 beats per minute. On several occasions, she had pauses lasting 3–3.4 seconds. She comments that she is often light-headed; however, she has never experienced syncope. The telemetry findings prompt a cardiology consultation. Before the cardiologist has a chance to interview and examine the patient, members of the patient’s family—which includes health care providers—demand that she undergo pacemaker implantation (class IIb indication).

Patients or their surrogates may also request treatments of questionable efficacy that support an uncontroversial end (Weijer et al. 1998). In the case, pacemaker therapy would be questionably effective for the observed cardiac rhythm abnormality. The most likely motive for the family’s demand, however, is prolongation of the patient’s life—an uncontroversial end. In situations such as this, the clinician should assess the facts of the case and—despite the family’s demands—discern the patient’s health care-related values and goals. If the patient’s goal is to prolong life, then the clinician should inform the patient that pacemaker therapy is questionably effective for the cardiac rhythm abnormality and, therefore, will not prolong life, especially given the patient’s underlying malignancy. It is likely, however, the patient is aware of her prognosis and that maximizing comfort is an important goal for her. The only way to discern this goal is to ask the patient.

Patients or their surrogates may also make requests for treatments that are effective, yet support a controversial end (Weijer et al. 1998). If the patient in the case had symptomatic complete heart block, then pacemaker therapy would be effective in keeping her alive and she and her family might desire pacemaker therapy for that reason alone. However, the clinician may view pacemaker therapy as futile because it will not reverse the patient’s underlying malignancy. In other words, what clinicians regard as futile may not be regarded as such by patients or surrogates. Indeed, medical futility is hard to define (American Medical Association Council on Ethical and Judicial Affairs 2002). Rather than declaring device therapy futile, the clinician should determine the patient’s health care-related values and preferences and explain how device therapy is in alignment with them. This process usually keeps patients’, surrogates’, and clinicians’ perspectives in alignment and may prevent ethical dilemmas. However, should a patient or surrogate remain steadfast in their request and carrying out the request violates the clinician’s conscience, then the clinician should arrange for a second opinion and, if necessary, ethics consultation (Snyder and Leffler 2005).

**Addressing Confidentiality Issues**

For thousands of years, oaths and codes of ethics have declared the clinician’s duty to maintain patient confidentiality (American Medical Association Council on Ethical and Judicial Affairs 2002; Snyder and Leffler 2005). Respect for patient autonomy is the ethics principle that underlies the clinician’s duty to maintain patient confidentiality. Furthermore, maintaining confidentiality is necessary for the
proper evaluation and treatment of patients (American Medical Association Council on Ethical and Judicial Affairs 2002). Clinicians need to be free to ask questions about sensitive matters (e.g., sexual history) and conduct thorough physical examinations in order to assess and treat patients properly. Patients must trust that clinicians will not divulge patients’ personal and medical information. Indeed, clinicians should not divulge patients’ information without patients’ permission. Notably, clinicians are also legally obliged to maintain patient confidentiality.

Under certain circumstances, clinicians are obligated ethically and legally to breach patient confidentiality. For example, statutory and case law may require that clinicians breach confidentiality in order to protect the best interests of others (e.g., mandatory reporting of infectious diseases). Here, the clinician’s duty to protect the public’s health overrides the duty to maintain the individual patient’s confidentiality. Clinicians are also obligated to breach confidentiality when the patient poses a risk of harm to himself, herself, or others. The patient in the case has the right to refuse unwanted treatments. However, she poses a risk to herself and others if she drives. Hence, the patient’s clinicians should advise that she not drive. If she refuses, or if there is evidence that she is continuing to drive, then the clinician should breach confidentiality in order to protect others by reporting the patient to the appropriate authorities. Understandably, clinicians may feel uncomfortable in these situations; colleagues such as social workers and health care institution-based attorneys can provide valuable assistance.

Notably, guidelines exist for advising patients who undergo cardiac device implantation and therapies about driving (Epstein et al. 1996, 2007). In addition, each state in the United States has its own regulations restricting driving in persons with cardiac rhythm disorders (Finch et al. 1997). Clinicians should be aware of these regulations for their jurisdictions.

Case 6.3.10

A 34-year-old woman presents with recurrent spells of sudden light-headedness, the last episode resulting in a car accident. Evaluation reveals recurrent polymorphic VT in the setting of long QT syndrome. Medications and implantation of an ICD are recommended (class I indication). Preferring “natural” therapies to allopathic treatments, she declines. The care team wonders if the patient should be allowed to drive.

Avoiding Inappropriate Bedside Allocation of Health Care Resources

Justice is the ethics principle that underlies the clinician’s duty to treat patients fairly (Beauchamp and Childress 2009). Clinicians should avoid making clinical decisions based on patients’ characteristics such as race, gender, affluence, ethnicity, religion, or other social category; injustice occurs when clinicians base their decisions on these factors rather than on medical need (American Medical Association Council on Ethical and Judicial Affairs 2002).

Case 6.3.11

A 52-year-old man is brought to the emergency room after collapsing at a homeless shelter. He is resuscitated and brought to the hospital. He is found to have ischemic cardiomyopathy and recurrent polymorphic VT. Implantation of an ICD is recommended (class I indication). He was laid off from his job more than a year ago and has no insurance or assets.
In this case example, the clinician’s duty is to act on behalf of the patient. In situations such as this one, the clinician may be the patient’s only advocate. In addition, the clinician should not abandon the patient. Rather than unilaterally withholding the preferred treatment from the patient, the clinician should discuss the treatment options with the patient and engage allied health colleagues (e.g., social workers, hospital administrators) to assist with resolution of the dilemma. Indeed, this patient may qualify for government-based assistance. In addition, many health care institutions and nonprofit organizations have benevolence programs to assist uninsured patients; pharmaceutical companies and device manufactures may have similar programs.

**Case 6.3.12**

A 76-year-old woman returns for routine follow-up of ischemic cardiomyopathy. Her LVEF is 25% and she is in New York Heart Association functional class II. She is adherent with her medications, diet, and exercise program. Her cardiologist contemplates recommending ICD implantation for primary prophylaxis (class I indication). The cardiologist, however, does not offer the patient this therapy, believing such therapy to be a wasteful use of scarce health care resources.

This case is an example of possible bedside rationing based on age, gender, or both. Indeed, ICD implantation and therapy is less likely to be provided to women and racial/ethnic minorities than Caucasian men in the United States (Redberg 2007). The cardiologist’s reason for withholding device therapy from the patient is wrong for several reasons. First, it is not in accordance with guidelines (Epstein et al. 2008). Second, the patient may derive benefits from device therapy beyond prolongation of life (e.g., psychological benefits). Third, the patient may actually decline device therapy; not offering device therapy deprives the patient the opportunity to decline it. Fourth, bedside rationing— withholding device therapy from the patient— falsely presumes the cost savings will be applied to health care needs elsewhere. If consistent with evidence-based guidelines and barring relevant contraindications, the cardiologist should offer the patient ICD implantation and therapy.

**Maintaining Proper Relationships with Industry-Employed Allied Professionals (IEAPs)**

IEAPs provide valuable technical assistance to clinicians who care for patients with implanted cardiac devices. This assistance occurs during device implantation, programming, and follow-up. In a policy statement, HRS recognizes that IEAPs often provide valuable support to clinicians, particularly since clinicians cannot be familiar with every aspect of each manufacturer’s product. Furthermore, the HRS policy statement articulates appropriate roles for IEAPs in the clinical setting (Lindsay et al. 2008).

**Case 6.3.13**

A 77-year-old male farmer, terminally ill due to severe chronic obstructive pulmonary disease, is under home hospice care. He has a history of VF arrest and has an ICD (class I indication). He has received appropriate shocks twice. He does not want shocks during the dying process. His primary physician recommends deactivation of the ICD. The patient agrees. However, the primary physician has neither the knowledge nor the equipment to carry out a deactivation. The patient requests that the local device manufacturer field representative, with whom he has developed a cordial relationship, come to his home to deactivate the ICD. He lives 75 miles from the nearest hospital.
This case illustrates a relatively common scenario: IEAP involvement in device deactivations. Indeed, in a recent survey, the “industry representative” was cited as the individual “who deactivates devices most of the time” (Mueller et al. 2008). The involvement of IEAPs in device deactivations, however, raises a number of ethical concerns. First, IEAPs represent their companies and at the same time they represent the ordering physician when carrying out a device deactivation; that is, they have dual agency (Mueller et al. 2008). Second, reprogramming a device is a medical procedure and most IEAPs are not licensed clinicians (Lindsay et al. 2008). Third, although the IEAP may have a cordial relationship with the patient, the responsibility of caring for the patient belongs to the clinician (Snyder and Leffler 2005).

The HRS policy statement states that device reprogramming should be “carried out only at the request and under the direction of a qualified physician” with the physician nearby and “IEAPs should not provide technical assistance in a patient’s home in the absence of a responsible physician” except in “rare and emergent circumstances” (Lindsay et al. 2008, p. e8). Device deactivation in terminally ill patients has been described as an emergency procedure (Pinski 2000). The HRS policy statement addresses this issue: “The patient’s comfort is a legitimate consideration when ICDs are reprogrammed not to shock a terminally ill patient who is at home or in a hospice” (Lindsay et al. 2008, e9).

The patient in the case is under home hospice care. Requiring him to travel to the nearest hospital for ICD deactivation would be inconvenient and inconsistent with goals of hospice care. Therefore, device deactivation by the IEAP at the patient’s home may be reasonable. Consistent with HRS policy, the IEAP should “only provide service remotely under written and direct order by the physician.” The physician should generate a written order and document the events in the patient’s medical record (Lindsay et al. 2008). Notably, many IEAPs are uncomfortable with deactivating devices (Mueller et al. 2010). If the IEAP objects to deactivating the patient’s device, the primary physician should arrange for another means of carrying out the deactivation.

**AVOIDING ETHICAL DILEMMAS IN CARDIAC ELECTROPHYSIOLOGY PRACTICE**

Effective patient–clinician communication is associated with improved patient satisfaction, adherence with treatment plans, and health outcomes (Stewart 1995; Barrier et al. 2003). Effective patient–clinician communication may also prevent ethical dilemmas. Fortunately, clinicians can improve their communication skills (Barrier et al. 2003). First, when seeing a patient for the first time or follow-up, the clinician should attempt to learn about the patient (e.g., their health care-related values and preferences) (Platt et al. 2001). When gathering information, the clinician should allow patients to list all of their concerns; this process typically takes only 1 minute (Beckman and Frankel 1984). Clinicians should elicit all of the patient’s concerns by asking questions such as “Do you have any other questions?” Notably, it is often useful to gauge patients’ understanding of their issue or condition using a statement such as “What is your understanding of your heart condition?” Such statements allow for correction of misinformation. After the patient articulates all of his or her concerns, the patient and clinician should jointly prioritize the concerns. Clinicians can build relationships with patients by using statements such as “We will solve this problem together,” “It sounds like you have a lot of concerns about your pacemaker,” “Many people with defibrillators have similar concerns,” “I will be with you throughout the procedure,” and “Here is how you can reach my team if you
have any questions.” When conveying results of tests and other information, clinicians should use jargon-free language and frequently assess patient comprehension by asking questions such as “Am I making sense?” Finally, clinicians should summarize the visit and establish a treatment and follow-up plan (Barrier et al. 2003).

ETHICS CONSULTATION

Even under ideal circumstances, clinicians who care for patients with implantable cardiac devices will encounter challenging ethical dilemmas. Most health care institutions have forms of ethics consultation to help resolve these dilemmas (Swetz et al. 2007). Indeed, the Joint Commission requires health care institutions to have a process for addressing ethical concerns that arise while caring for patients (Joint Commission 2010). Clinicians should engage this process when appropriate.

CONCLUSIONS

The indications for implantable cardiac devices are growing. Likewise, the number of patients who have implantable cardiac devices is growing. Thus, it is reasonable to expect that clinicians will encounter an increasing number of device-related ethical challenges. Clinicians who care for patients with implantable cardiac devices should be familiar with commonly encountered ethical dilemmas in cardiac electrophysiology practice and approaches to resolving these dilemmas. Nevertheless, even under ideal circumstances, daunting ethical dilemmas occur. In these situations, ethics consultation can help resolve these dilemmas.

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