Cardiac Pacing and Defibrillation in Pediatric and Congenital Heart Disease

Cardiac Pacing and Defibrillation in Paediatric and Congenital Heart Disease

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This book is dedicated to my family, especially my parents who taught me that nothing is impossible, my husband Shaun for making everything possible, and my son Neil who reminds me every day of the endless possibilities that lie ahead.

I am deeply grateful to Dr. Larry Rhodes for his mentorship, Dr. K.M Cherian for believing in me when I emerged fresh out of fellowship, Dr. Frank Cecchin for imparting his exemplary technical skills, Dr. Paul M. Weinberg for his rigorous teachings of cardiac anatomy, and Dr. Victoria Vetter for her support in creating a Device Implantation & Lead Extraction program at The Children's Hospital of Philadelphia. I am also very grateful to my Division of Cardiology Chief, Dr. Robert Shaddy and Department of Pediatrics Chair, Dr. Joseph St. Geme for their support of a sabbatical which allowed me to complete this project. Last but not the least, this book is dedicated to the Cardiology and Electrophysiology fellows who motivate me every day to learn more and become a better teacher.

–Maully Shah

Thanks to Dr. Ed Walsh for teaching me Electrophysiology, Dr. Vickie Vetter for letting me practice Electrophysiology, my fellows for making Electrophysiology fun, and my family for putting up with me.

–Larry Rhodes

I would like to thank my mentors, Larry Rhodes and Gail Pearson, my EP colleagues and friends and most importantly, Becca, Maddie and Jacob.

–Jonathan Kaltman

Contents

Foreword

Children represent only about 5% of the market for implantable cardiac rhythm management devices and adults with congenital heart diease certainly form an even smaller segment. It is easy to forget, therefore, that much of the impetus for the development of pacemakers in the early days was to answer a major problem: namely the occurrence of a complete atrioventricular block as a complication of open heart surgery to close septal defects. Unique clinical needs often drive innovation in medicine, and those developmemnts are then noticed and put to use by the larger community of practitioners. The endless variation in heart size and anatomy seen in children with congenital heart disease, along with the rapid development of new approaches to surgical repair, mean that we will continue to have a need for innovation in this field.

The field of pediatric electrophysiology has matured from a small group of practitioners trying to fit a square peg into a round hole into a true international professional body called PACES (Pediatric and Congenital Electrophysiology Society) that is innovating and advocating for pediatric and congenital heart disease specific device therapy. The editors have put together a group of authors that are leaders in this field, providing in this book their knowledge and experience, which is the round peg.

Doctors Shah, Rhodes, and Kaltman have edited and provided us with a textbook that fills a major gap and a pressing need focused on device management in children given the absence of specific leads or device features that meet the challenges of implantation and extraction of devices in children as a result of their smaller size, anticipation of growth, unique activity spectrum, and the relative overrepresentation of anatomic variants and congenital heart disease in the pediatric group. These challenges have been addressed by experienced implanters with idiosyncratic technical modifications at implant or device programming, and until now, there has been no targeted work that summarizes the approaches best suited for pediatric device practice.

Due to the comprehensive nature of this textbook it will find its home in a broad range of readers. For the trainee it will be a syllabus, for the the general practitioner it will provide the background and detail needed to comanage these patients and for the practicing electrophysiologist it will be a daily reader.

This textbook importantly explains the technical approaches in the normal pediatric heart as well as devoted sections for device management in congenital heart disease. Innovative and useful features that enhance the value of this textbook include a pacemaker and ICD glossary and a dedicated companion website that helps test the readers' knowledge and understanding as well as videos that help implanters in picturing the special needs in children. I have no doubt that *Cardiac Pacing and Defibrillation in Pediatric and Congenital Heart Disease* will be a must-have on the shelves of all of us who help manage rhythm disturbances in children and the adult congenital heart disease population.

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Preface

This book is written to address the unique issues of pacemaker, resynchronization and defibrillation therapy in children and young adults with special emphasis on patient size, growth, development, lifestyle, and co-existent congenital heart disease (CHD). The first functional external battery operated pacemaker was implanted in a child with post-operative heart block following repair of a ventricular septal defect in 1957. During the ensuing six decades, the field of cardiac pacing has seen ground-breaking innovations that have served as a foundation for other advanced life saving device therapies such as implantable cardioverter defibrillator (ICD) and cardiac resynchronization therapy (CRT). Although current devices and leads are designed and marketed predominantly for an adult patient population, the myriad applications in pediatric patients are well accepted. As pediatric cardiologists, we recognize that a child is not a miniature adult and we must therefore continue to pioneer, modify, and adapt techniques in accordance with the specific characteristics of our patients. Clearly, there is still significant opportunity for the scientific, engineering and regulatory organizations to manufacture cardiac rhythm devices and leads that are more suitable and efficacious for the pediatric patient.

This book addresses the need for articulation of current concepts, principles and clinical practices that underlie device management in children and patients with CHD. It is our hope that this book will serve as a comprehensive and informative resource to trainees as wells as practicing cardiologists and electrophysiologists, especially those involved in the care of CHD patients with rhythm disorders. We also hope that this book will serve as a guide to physicians who are faced with the challenges of pediatric device implantation and management in parts of the world where pediatric electrophysiologists are scarce.

The content of the book follows a logical progression starting with a brief history describing the brilliant innovations of several inventors to create the first implantable pacemaker. From there we proceed to the fundamental principles of pacing and defibrillation, a description of clinical concepts and indications, device implantation techniques, and subsequent management with detailed sections on troubleshooting, complications and follow-up. We have briefly included new technologies such as the totally sub-cutaneous ICD and the leadless pacemakers. Instructive device electrograms recordings and x-ray images are presented throughout the book. Finally, the website version has select videos and chapters 2–18 have interactive multiple choice questions.

This book could not have been completed without the encouragement and enthusiastic support of its contributors and several others. Contributors have been eager and motivated from the start and we thank them for their time, patience, and expertise. The staff at Wiley-Blackwell Publishing Company, especially Thomas V. Hartman who initiated the project and Claire Bonnett who facilitated its completion, deserve our thanks for the efficiency and meticulous care they have brought to the book's preparation. Carrie Stackhouse has been an intellectual and technical resource whom we cannot thank enough for her constant readiness to tackle difficult device programming and

troubleshooting questions. Our trainees continue to inspire us with their thirst for knowledge. In many ways this book is a testament to our passion for teaching and learning with them. Most important, we thank the countless patients and families who have entrusted us with their care. It has been our privilege to learn something from each and every one of them.

Maully Shah
Larry Rhodes
Jonathan Kaltman

About the Companion Website

This book is accompanied by a companion website:

www.wiley.com/go/shah/cardiac_pacing

The website includes:

- Interactive multiple choice questions (MCQs)
- Videos

PART 1
Introduction

1

CHAPTER 1

History of cardiac pacing and defibrillation in the young

Larry Rhodes[1] *and Robert Campbell*[2]

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The earliest years of cardiac pacing predate the birth of many current pediatric cardiac electrophysiologists. An old saying states that "failure to understand history dooms one to repeat it." In contrast, understanding this history of successful collaboration between pioneering physicians and engineering partners allows us to marvel at the developments that were to follow rapidly over the next 50 years, and potentially repeat this formula in years to come.

Benjamin Franklin harnessed electricity from lightning using a kite in 1752. An early "medical" use of electricity was not to augment life but to document the end of it with patients receiving an electrical shock to prove they were dead. In 1774, electrical energy was applied to resuscitate a child using a transthoracic approach.[1] As early as 1899, the *British Medical Journal* published a report of experiments demonstrating that application of electrical impulses to the human heart would lead to ventricular contractions.[2] In 1926, Dr. Mark C. Lidwell and physicist Edgar H. Booth of Sydney developed a device with pacing rates of 80–120 bpm and outputs varying from 1.5 to 120 V.[3] This "pacer" was described as being a portable device "plugged into a lighting point." One pole was connected to a pad soaked in strong salt solution and applied to the skin and the other, "a needle insulated except at its point, was plunged into the appropriate cardiac chamber." In 1928, this apparatus was used to revive a stillborn infant whose heart continued to beat after 10 minutes of stimulation.[4]

During the 1930s, Dr. Albert Hyman noted that the success of intracardiac delivery of medications for cardiac arrest was likely independent of the medication but was instead related to the needle stick leading to alteration in electrical potentials and myocardial contraction. Knowing that multiple needle sticks would be impractical and dangerous, he developed a generator to deliver electrical impulses via needle electrodes.[5]

Following World War II there was a significant interest in pacemakers generated by investigations in the use of general hypothermia for cardiac surgery. Cardiac arrest was noted during hypothermia and adequate heart rate was required to maintain adequate hemodynamics during rewarming. John A. Hopps, an engineer at the National Research Council of Canada developed a pacemaker that produced impulses at a desired rate

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through an electrode placed in the area of the sinus node.[6]

In 1952 Dr. Paul M. Zoll used an external pacemaker coupled with transcutaneous needle electrodes to rescue a patient suffering from Stokes-Adams attacks following a myocardial infarction.[7,8] The patient continued to experience ventricular asystole despite being administered 34 intracardiac injections of adrenaline over a 4-hour period. Dr. Zoll applied "external electrical stimulation" and successfully paced this patient's heart over the next 25 minutes.[8] The patient developed cardiac tamponade secondary to perforation of a cardiac vein during the intracardiac injections. Dr. Zoll then successfully paced a 65-year-old man with episodes of ventricular standstill for 5 days by external electrical stimulation at which time he developed an idioventricular rhythm at 44 bpm and was discharged.[9]

In the mid-1950s, open heart surgery was becoming a reality. Although for the first time in history, intracardiac palliation of structural heart disease was possible, the complication of surgical heart block was a significant morbidity. Dr. W. Lillehei, Dr. W. Weirich, and others at the University of Minnesota demonstrated that pacing could be performed by connecting a pulse generator to a wire electrode attached directly to the heart of a dog.[10,11] In January 1957, Lillehei used this pacing system in the first human patient, a child with post-operative heart block following repair of a ventricular septal defect. The pacer was programmed to a pulse width of 2 ms and a voltage ranging from 1.5 to 4.5 V (Figure 1.1).[12]

The generators used by both Zoll and Lillehei were devices which transformed alternating current into direct current to pace the heart. In 1957, following a power failure in Minneapolis in which patients could not be paced, Dr. Lillehei enlisted the help of Earl Bakken and Medtronic for battery backup for AC pacemakers. Silicon transistors had become commercially available in 1956 leading to the potential for development of smaller and more practical pacemakers. The original transistorized, zinc oxide battery-powered external pacemaker was developed by Mr. Bakken in 1957; the device was smaller and thus applicable for pediatric patients.[13,14] This, the first wearable external pacemaker, was housed in a small plastic box, with controls to allow adjustment of pacing rate and voltage (Figure 1.2).

Figure 1.1 *Patient pushing pacemaker cart (1958). (Source: Reproduced with permission of Medtronic, Inc.)*

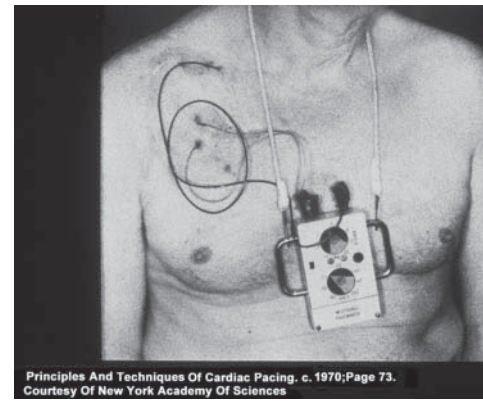

Figure 1.2 *Wearable pulse generator (1958). (Source: Reproduced with permission of Medtronic, Inc.)*

Although novel and potentially lifesaving, the advances described here were not a long term solution in that there was a significant risk of infection and external pacing was uncomfortable and impractical. There was a definite need for implantable pacing systems. Ake Senning, a Swedish surgeon, in collaboration with engineer Rune Elmqvist, developed a permanent implantable pulse generator with the first clinical implantation in 1958.[15] This device failed after three hours. A second device was implanted and lasted 2 days. The patient, Arne Larsson, went on to receive 26 different pacemakers until his death in 2001 at the age of 86 (Figure 1.3).[16]

Figure 1.3 *History – First "permanent" implantable pacemaker and bipolar Hunter–Roth lead (1958). (Source: Reproduced with permission of Medtronic, Inc.)*

During that same year, Seymour Furman introduced temporary transvenous pacing using the recently described Seldinger technique.[17] In 1962 Ekestrom, Johannson, and Lagergren reported the first non-thoracotomy pacemaker implantation by introducing the electrode transvenously into the right ventricle.[18]

By the end of 1960 virtually all pacemakers used mercury-zinc cells as the power supply, but battery life expectancy was generally less than 2 years on average. A greater problem was that because the batteries emitted hydrogen gas, the pulse generator could not be sealed to protect from contamination with body fluids.[19] Dissatisfaction with this power source generated interest in alternatives that included, but were not limited to, bioenergy sources (using piezoelectric transducers that generated electricity based on the expansion and contraction of the abdominal aorta, or motion of the diaphragm), nuclear generators, and, by the mid-1970s, lithium batteries.[20] There was significant interest in the use of nuclear powered pacers because they offered a remarkable lifespan (10–20 years) and reliability. A number of drawbacks related to radiation exposure in case of a capsule leak and disposal hindered their acceptance.

Lithium-iodide power sources persist as the battery of choice today. Voltage output of the lithium-iodine cell showed gradual decline rather than the abrupt drop associated with the mercury zinc during battery depletion. This new battery generated no gas byproduct allowing the entire pulse generator to be hermetically sealed in a titanium case, which was initially accomplished in 1969 by Telectronics and then by Cardiac Pacemakers, Inc., (Minneapolis, MN), in 1972. Battery life was significantly increased to greater than 5 years on average.

Leads

In the early 1960s it became routine practice to manage patients with temporary transvenous leads and an external pulse generator to relieve congestive heart failure. These served as a bridge to a thoracotomy for placement of a permanent pacemaker and lead system. Permanent transvenous pacing, which first appeared in the early 1960s, gained widespread acceptance by the end of the decade.[21,22] Initial leads were unipolar in design, but gradually gave way to a bipolar preference. Coaxial leads allowed for smaller lead diameter and greater durability. Smaller surface electrodes were designed to reduce energy consumption. Greater surface areas were achieved allowing improved lead function. Steroid-eluting leads were designed as a mechanism to reduce fibrosis at the epicardium-electrode interface, thus avoiding chronic rise in stimulation thresholds. Lead fixation, using passive or active mechanisms, were designed to prevent the previously high incidence of lead dislodgement. Silicone insulation gradually gave way to a preference for polyurethane. These newer leads had a generally smaller diameter than previous silicone leads, which facilitated the introduction of the implantation of two leads through a single vein, associated with the implementation of dual chamber pacing.

Pacing modes

The first implanted pacemakers were fixed rate ventricular systems, which competed with intrinsic ventricular activation. Unfortunately, the theoretical risk of inadvertent induction of ventricular fibrillation was in fact documented electrocardiographically.[23] Additionally, studies determined that fixed rate asynchronous pacing at times had an adverse hemodynamic impact on patients with myocardial dysfunction. Thus, the

impetus for development of a demand pacemaker which could sense intrinsic ventricular activity was heralded. Virtually simultaneously, two companies debuted demand ventricular pacemakers. In 1966 the Medtronic system functioned in a true demand mode, with inhibition of ventricular pacing during sensed intrinsic ventricular activity.[24] The Cordis "standby" pacemaker functioned in the ventricular triggered mode, such that a sensed R-wave triggered the pacer stimulus with no AV delay so that it fell within the refractory period of the intrinsic QRS complex.[25] These modes were, respectively, termed VVI and VVT, both non-competitive modes.

The first pacemakers to permit atrial synchronization with the ventricle depended upon new sensing technology to detect intrinsic atrial activity,[26] and the triggering of a paced ventricular response after a programmed AV interval. These devices were bulky because of the complexity of the circuitry, and also demonstrated a significant reduction in battery life. Problems with erratic sensing of the intrinsic atrial activity and abrupt drops in pacing rates that occurred when upper rate limits were reached also limited the acceptance of these early dual chamber systems.

In the early 1980s a third generation of dual chamber pacemakers was introduced. These generators had long-lived lithium batteries and generally incorporated new dual endocardial leads. Pacemaker systems were able to both sense and pace in both the atrium and ventricle allowing physiologic rates and AV synchrony. The development of leads which could be used for atrial stimulation, as well as atrial sensing, enhanced the functionality of these early dual chamber systems.

Rate adaptive pacing, for patients with chronotropic incompetence, permitted rate responsive pacing that augmented heart rate response when intrinsic sinus node function was inadequate.[27] A more recent breakthrough mode was antitachycardia pacing, applicable especially for postoperative congenital heart disease patients with recurrent medically-refractory intraatrial reentry tachycardias.[28]

Non-invasive programmability

Seymour Furman and associates reported in 1969 the first techniques for routine transtelephonic monitoring of pacemaker function.[29,30] Subsequent advances included the ability of the system to estimate battery longevity. Continuous advancement in these non-invasive technologies has finally led to the ability to provide non-invasive electrogram analysis for tachycardia detection, tachycardia termination, and antitachycardia defibrillation systems.

Multiprogrammability

By the mid-1960s, the early non-invasively programmable pacemakers had advanced to multiprogrammable units dependent upon bidirectional telemetry. In 1978, Intermedics introduced a pacemaker for whom pacing rates, pulse width, and sensitivity could be programmed; this system was a result of collaboration between engineer Robert Brownlee and physician G. Frank Tyers.[31] Dual chamber pacemakers also permitted programmability of pacing mode, in the event of recovery of intrinsic AV nodal function (allowing atrial pacing alone) or ventricular pacing only in the event of failure of the atrial lead (pacing and/or sensing capabilities).

Miniaturization

Initial external pacemaker systems required portable carts (Figure 1.1). By the early 1960s when permanent implantable systems were in place, the pulse generators were still bulky. Advanced pacemaker and software technologies allowed further miniaturization, but often at the expense of battery life. Smaller generators had a unique implant role for the smallest of neonates and pediatric patients, but required frequent generator changes due to battery depletion. Further decrease in lead size allowed implantation of multiple leads within a single vein, even in the smallest patients, but electrodes were still relatively large. Even these small lead systems were associated with a high incidence of venous obstruction/occlusion.

Pacemaker codes

The Inter-Society Commission for Heart Disease Resources (ICHD)[32] proposed a three-position

"conversational" pacemaker code in 1974 to distinguish pacemakers according to three fundamental attributes:

Position 1. Chamber or chambers paced:
- V – ventricle paced
- A – atrium paced
- D (dual) – both atrium and ventricle paced
- O – neither atrium or ventricle paced

Position 2. Chamber or chambers in which native cardiac events were sensed:
- V – ventricle sensed
- A – atrium sensed
- D (dual) – both atrium and ventricle sensed
- O – neither atrium or ventricle sensed

Position 3. Pacemaker response to sensing a spontaneous chamber depolarization:
- T – triggered
- I – inhibited
- D (dual) – both triggered and inhibited
- O – none

Subsequent revisions paralleled development of pacemaker capabilities. The most recent revision of this original three-position code was published in 2000 incorporating a five-position code.[33] Position 4 is used only to indicate the presence (R) or absence (O) of a rate adaptive mechanism, used to compensate for patients with chronotropic incompetence. Position 5 indicates whether multi-site pacing is present in none of the cardiac chambers (O); in one or both of the atria (A) with stimulation sites in each atrium or more than one stimulation site in either atrium; in one or both of the ventricles (V), the stimulation sites in both ventricles or more than one stimulation in either ventricle; or in dual chambers (D), in one or both of the atria and in one or both of the ventricles. This most recent coding was endorsed by both the North American Society for Pacing and Electrophysiology (NASPE), (now known as the Heart Rhythm Society: HRS), and the British Pacing and Electrophysiology Group (BPEG).

Guidelines for implantation of cardiac pacemakers and antiarrhythmia devices

The first guidelines were introduced in 1984 through a joint subcommittee of the American College of Cardiology and the American Heart Association.[34] Pediatric cardiac pacing was represented by Dr. Paul Gillette. Guidelines were grouped according to the following classifications – class 1: conditions for which there is general agreement that permanent pacemakers and antitachycardia devices should be implanted; class 2: conditions for which permanent pacemakers and antitachycardia devices are frequently used but there is a divergence of opinion with respect to the necessity of their insertion; class 3: conditions for which there is general agreement that permanent pacemakers and antitachycardia devices are unnecessary. Multiple revisions have occurred, coincident with advances in technologies of these devices. The most recent guidelines were issued in 2008 through the American College of Cardiology, American Heart Association, and Heart Rhythm Society, and were developed in collaboration with the American Association for Thoracic Surgery and the Society of Thoracic Surgeons.[35] Guidelines for pediatric pacing were generated with the input of Dr. Mike Silka. In addition to the class 1, 2, and 3 indications for pacemaker implantation, guidelines were also rated according to evidence to support the guidelines. Levels of evidence A: data derived from multiple randomized clinical trials or meta-analysis; B: data derived from a single randomized trial or nonrandomized studies; and C: only consensus opinion of experts, case studies, or standard of care. These guidelines continue to dynamically evolve, but are widely regarded by consensus as detailing the appropriate use of devices in both adult and pediatric patients.

North American Society of Pacing and Electropysiology (NASPE)

Senior pacing physicians during the 1970s founded the *Journal of Pacing and Clinical Electrophysiology* (*PACE*) and organized a supporting professional society, NASPE.[36,37] NASPE arose out of a concern for the growing complexity of pacemaker systems and implantation techniques, the maintenance of quality control and good manufacturing practices by companies, and the proper post-implantation care of an ever expanding patient population. As lead technology advanced, the non-invasive transmission of intracardiac electrograms allowed increased patient diagnostic surveillance and

treatment, and paralleled the explosive development of intracardiac electrophysiologic testing in advance of cardiac ablative therapies. NASPE (HRS since 2004) currently has over 5400 cardiac pacing and electrophysiology professionals worldwide and is the international leader in science, education and advocacy for cardiac arrhythmia professionals and patients, and the primary information resource on heart rhythm disorders. Its mission is to improve the care of patients by promoting research, education, and optimal health care policies and standards.

The implantable cardioverter defibrillator (ICD)

Prior to discussing the history of implantable cardioverter defibrillators, a brief review of defibrillation is in order. At the turn of the twentieth century, Prevost and Batelli researched ventricular fibrillation in dogs describing methods to fibrillate the heart using alternating (AC) and direct (DC) electrical currents. They noted it took stronger currents to defibrillate than to fibrillate the heart.[38] In 1947, Dr. Claude Beck performed the first successful human defibrillation using internal cardiac paddles on a 14-year-old boy who developed VF during elective chest surgery.[39] The device used on this patient, made by James Rand, had silver paddles the size of large tablespoons that could be directly applied to the heart. In 1956, Paul Zoll used a more powerful unit to perform the first closed-chest defibrillation of a human.[40]

The remarkable technical advances that occurred in clinical electrophysiology and pacemaker technologies through the 1960s and 1970s established the groundwork for the development of the implantable cardioverter defibrillator (ICD). External cardiac defibrillation was proven to be an effective method for terminating potentially life-threatening cardiac rhythm disturbances, including unstable ventricular tachycardia and ventricular fibrillation. In contrast to the pioneering collaborative efforts of multiple teams of physicians and engineers responsible for pacemaker development through the 1960s and 1970s, the development of the ICD is attributed almost single-handedly to the unwavering determination of Dr. Michael Mirowski in Baltimore, and his engineering collaborator, Dr. Morton Mower. In a 1970 publication, Mirowski and Morton described the elements of an early ICD device, which would be required to quickly diagnose and treat ventricular fibrillation using a unit small enough for subcutaneous implantation.[41] Extended battery life would be a key component given the high output demands anticipated. Ventricular fibrillation detection techniques were initially dependent upon right ventricular pressure transducers, with a drop in blood pressure in post myocardial infarction patients triggering the device.[42] This unreliable sensing method was upgraded to the use of an intracardiac electrogram feature and a complex probability density algorithm distinguishing ventricular fibrillation from sinus rhythm. Initial device design used a hybrid endocardial and epicardial lead system with a single right ventricular transvenous lead and a subcutaneous defibrillation patch in the anterior chest wall. Subsequent iterations included a shock vector from a superior vena cava coil to apical patch. A completely transvenous system ultimately consisted of a right ventricular apical coil electrode with a second electrode in the superior vena cava or right atrium.

Initial animal studies demonstrated the efficacy of the device to terminate electrophysiologic induced ventricular fibrillation. Despite initial encouraging published results, there were vigorous dissenters who disqualified the device and the concept of the approach. The first human implantation occurred in 1980 at Johns Hopkins Hospital.[43] The device was non-programmable, committed, and had no telemetry capabilities. There was also no antitachycardia pacing option for patients with unstable ventricular tachycardia. Second generation defibrillators incorporated an epicardial right ventricular electrode for ventricular tachycardia detection.

Generator device and battery advancements have continuously developed. A significant design modification resulted in a new lead design in 1988, allowing for the first complete transvenous implantation[44] consisting of proximal and distal shocking coils.

The concept of tiered therapy was introduced in the early 1990s. A progressive therapy for ventricular tachycardia allowed for initial programmed bursts of antitachycardia pacing, followed by a low

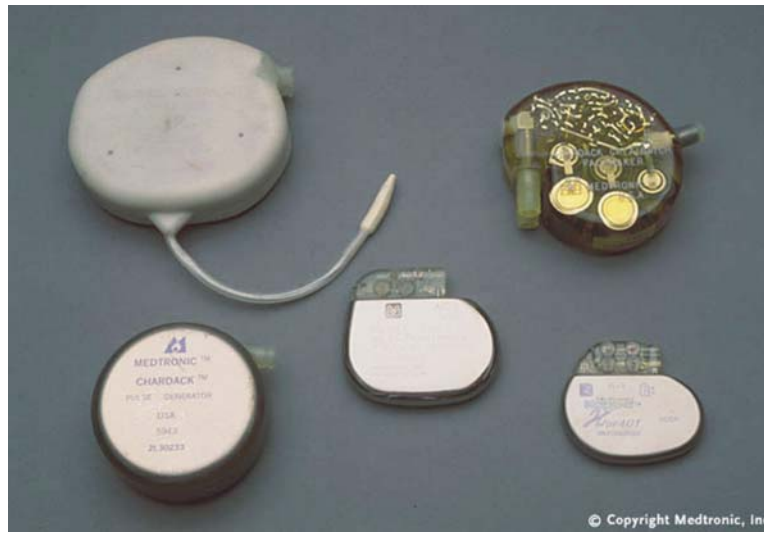

Figure 1.4 *Early implantable single chamber device to current dual chamber Kappa. (Source: Reproduced with permission of Medtronic, Inc.)*

energy shock for unstable VT, culminating in a high energy shock for unstable VT not terminated using step 2 or for tachycardia that had degenerated to ventricular fibrillation.

Advancement in devices and patches has allowed the successful implantation of ICD therapy in even young patients, and those with complex congenital heart disease anatomy limiting ICD lead placement (endocardial and/or epicardial) and generator positioning (thoracic or abdominal). Current guidelines for ICD implantation are likewise detailed in the 2008 ACC/AHA/HRS Guidelines for Device Based Therapy of Cardiac Rhythm Abnormalities.[28]

Summary

It is difficult to find a better example in medicine where the development of technology was driven in large part by the needs of children than that seen in pacer therapy. This occurred secondary to the fact that symptomatic bradycardia frequently presents in childhood as congenital heart block or a consequence of congenital heart disease. The primary motivation for successful cardiac pacing paralleled the development of open heart procedures for patients with congenital heart disease. This new era of palliation of children previously doomed to a life of disability could not to be derailed by a heart rate that did not maintain an adequate cardiac output. The commitment of Dr. C. Walter Lillehei and other pioneers at the University of Minnesota in the 1950s to continue with their heroic efforts to offer these children the potential for a normal life led to Earl Balken developing what is now Medtronics in a small garage in Minnesota. Throughout the last 60 years, the needs of children relative to size and anatomy have led to the development of smaller pacers and leads that have, in turn, continued to advance the field for patients of all sizes and ages (see Figure 1.4).

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2

CHAPTER 2

Clinically relevant basics of pacing and defibrillation

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Basic concepts in cardiac pacing

The fundamental principle of artificial cardiac pacing involves delivery of an electrical impulse of sufficient strength from an electrode to cause excitation of a critical mass of cells. Since the heart is a syncytium, once the critical volume of cells is excited, the conduction propagates to the rest of the myocardium.[1, 2] Clinically relevant pacemaker features and terminology are described next.

Stimulation threshold

The minimal energy required to produce myocardial depolarization is called *stimulation threshold*. There are two components of stimulation: *pulse amplitude* (measured in volts, V) and *pulse duration* (measured in milliseconds, ms). Current pacemaker systems are constant voltage systems and the resultant strength-duration curve is hyperbolic in shape suggesting an exponential relationship between stimulus amplitude and duration (Figure 2.1). At short pulse durations, a small change in pulse duration is associated with a significant change in pulse amplitude required to produce myocardial depolarization. At long pulse durations, a small change in the pulse duration has little effect on threshold amplitude. There are two important points on the strength-duration curve: *rheobase*, which is the smallest amplitude that stimulates the myocardium at infinitely long pulse duration and *chronaxie*, which is the threshold pulse duration at twice the stimulation amplitude. The latter approximates the point of

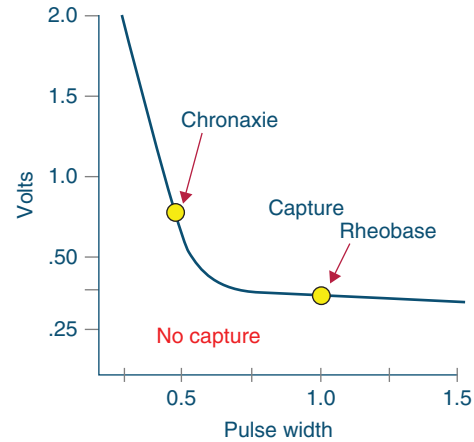

Figure 2.1 *Representation of chronic ventricular strength-duration relationships. Rheobase is the threshold at infinitely long pulse duration. Chronaxie is pulse duration at twice rheobase.*

Cardiac Pacing and Defibrillation in Pediatric and Congenital Heart Disease, First Edition.
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minimum threshold energy (microjoules) required for myocardial depolarization.[1, 2]

Stimulation thresholds typically oscillate in the ensuing weeks after implantation and are highly dependent on lead design, electrode-myocardial interface and patient factors, but chronic thresholds are typically reached by 3 months. With steroid eluting pacing leads, stimulation thresholds do not rise rapidly after implantation as with earlier generation non-steroid leads, but, tend to decrease to acute threshold values following a slight initial increase.[3]

Transvenous pacing leads with acute fixation mechanisms often have relatively high immediate pacing thresholds at implantation secondary to hyperacute injury due to advancement of the screw into the myocardium and frequently decline within the first 5–30 minutes.[4] The implanter should keep this in mind and wait a few minutes and re-check thresholds before repositioning the pacing lead.

Implantation of a pacing lead results in acute injury to the myocardial cellular membrane resulting in an acute inflammatory response, and this tissue becomes fibrotic over time. As a consequence, the distance between the electrode and excitable myocardial tissue is increased and may result in increased stimulation thresholds (typically 4–8 weeks after implantation) and a decrease in the sensed endocardial signal. This phenomenon is known as *lead maturation*. With increasing time, the size of the edematous capsule shrinks and stimulation thresholds decrease and stabilize chronically (typically by 12 weeks after implantation). Steroid eluting leads improve lead maturation by minimizing fibrous capsule formation and reducing energy consumption along with maintenance of stimulation and sensing thresholds as well as lead impedance values (Figures 2.2 and 2.3).[3, 5–9] *Exit block* is manifested by progressive rise in threshold over time due to fibrous tissue at the lead myocardial interface resulting in capture threshold that exceeds the programmed output of the pacemaker.[4]

Post implantation, stimulation thresholds may be altered by various factors. An increase in thresholds is encountered during sleep, hyperglycemia, hypoxemia, acidosis, acute illnesses, electrolyte disturbances, and certain cardiac drugs (Table 2.1).[10–17]

Pacemaker sensing

Intrinsic cardiac electrical signals are produced by electric activation in the myocardium. As the

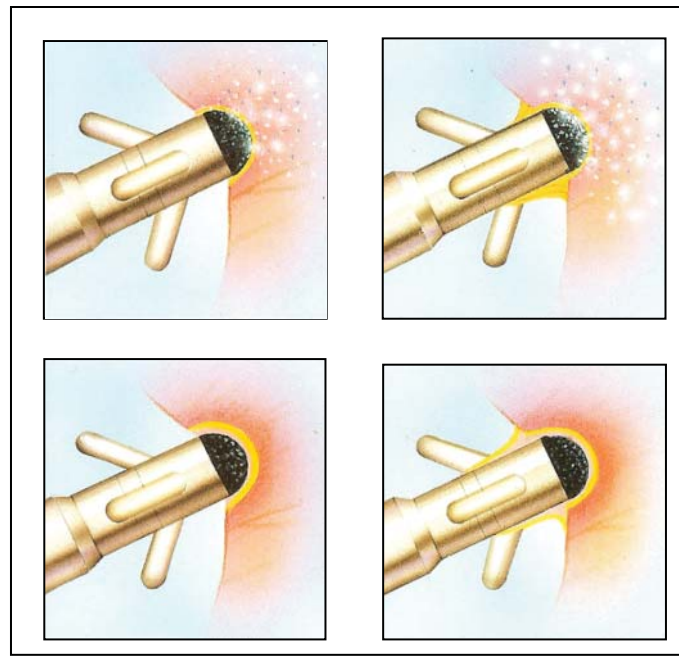

Figure 2.2 *The illustration compares a steroid eluting lead (top views) to a lead without steroid (bottom views) from the day of implant into the chronic phase. The steroid eluting from the tip of the lead suppresses each stage of the inflammatory process. The result is less inflammation, and a thinner capsule surrounding the lead tip. (Source: Reproduced with permission of Medtronic, inc.)*