Twenty-Five Years of Progress in the Medical Treatment of Pediatric and Congenital Heart Disease

DAN G. McNAMARA, MD, FACC

Houston, Texas

In the past 25 years there has been a many-fold increase in the prospect that with early recognition and modern treatment the newborn with critical congenital heart disease will reach adult life in a healthy condition, prepared to earn a living and to function as a spouse and as a parent.

Advancements in the medical treatment of congenital heart disease may create less public acclaim than may surgical treatment but many purely medical developments provide the basis for achieving ultimate surgical success and, by judicious use of some forms of medical treatment, operation can be avoided altogether.

The eight major contributions to patient care that are discussed in this review and the 35 that are simply listed are merely examples of the many developments that have occurred in the past 25 years. These include: 1) the organization of pediatric cardiology and the contribution of volunteer health organizations, 2) continuing medical education aimed at promoting early diagnosis of congenital heart disease and prompt referral to a cardiac center, 3) advances in the technology of cardiac catheterization, 4) Rashkind's balloon atrial septostomy and other catheter manipulative procedures, 5) pharmacologic manipulation of the ductus, 6) beta-adrenergic blockade for control of a variety of problems, including paroxysmal hypoxemic attacks, certain arrhythmias and relief of symptoms in hypertrophic cardiomyopathy, 7) echocardiography, and 8) advances in arrhythmias, electrophysiologic studies and use of pacemakers.

The 25 year period from January 1958 to January 1983 has seen many improvements in pediatric cardiology for the benefit of the infant, child or adolescent who has cardiovascular disease and the potential for developing it. Some advancements have been developed by pediatric cardiologists; others have been developed by basic scientists or adult cardiologists and then adapted to the needs of the pediatric patient. Many important developments were preceded by years of basic research in order to produce clinically useful methods of treatment. In other instances investigators promptly applied new techniques and concepts to meet an urgent clinical need, conducting little or no preceding fundamental research.

This review will include only a few of the important developments since 1958 and it is clearly weighted toward developments with which I am most familiar. Important contributions that are not listed have been omitted only to conserve space. This limited review may prompt others to prepare an all-inclusive syllabus of historical advances in pediatric cardiology in which appropriate attention is given to all investigators and their research products.

The rationale for a 25 year review of developments that have contributed to patient care is to encourage a new generation of physicians to see what can be accomplished through research in such a comparatively short period of time, and to strive to improve on and surpass these achievements in answering the unmet needs of patients with pediatric and congenital heart disease.

Developments in the Organization of the Discipline of Pediatric Cardiology

Certification Board

In January 1958, approximately 30 pediatricians in the United States were devoting all or most of their time to heart disease in children. Probably the greatest boon to the improved diagnosis and treatment of the infant and child with heart disease at that time was the establishment of the subspecialty of pediatric cardiology. Adams (1) described the process as follows:

"In August 1954 the Committee on Rheumatic Fever and Congenital Cardiac Disease of the American Academy of Pediatrics recommended the creation of a Section on Cardiology within the..."
Academy and the establishment of a certification board in pediatric cardiology. The Section on Cardiology, however, did not become formally organized until 1957, the Sub-Board of Pediatric Cardiology until 1961. Hugh McCulloch and James W. DuShane were primarily instrumental in forming the Section on Cardiology. The first members of the Sub-Board of Pediatric Cardiology were James W. DuShane, Forrest H. Adams, Edward C. Lambert, Alexander S. Nadas, Saul J. Robinson and Helen B. Taussig. As with other specialty boards in medicine, the purposes of the Sub-Board of Pediatric Cardiology were two-fold: to set minimum standards for training centers and to certify those physicians possessing the professional qualifications necessary to practice pediatric cardiology."

Growth of the Sub-Board

In 1967 the Board was awarded a contract with the National Heart and Lung Institute to study the examination procedure and the activities of Board-certified pediatric cardiologists. In this study the professional activity of the 207 Board-certified pediatric cardiologists was sought. Among the 192 who responded, 73% were engaged full-time in pediatric cardiology with a medical school, another 20% were based full-time in a hospital but had no medical school affiliation, and only 12 (7%) of the 192 were engaged in the private practice of pediatric cardiology in an independent office. It was clear that the field of pediatric cardiology was not conducive to independent private practice and required full-time hospital and academic support for success.

Although the diplomates of the Sub-Board of Pediatric Cardiology were mostly academicians, the study uncovered the surprising fact that the average time spent in research for the entire group was only 7%; 5% for junior full-time faculty and 20% for senior full-time faculty. Patient care had been changing so rapidly and was so time consuming that full-time junior and senior academicians were occupied with patient care an average of 62% of their time. Recognizing the small amount of time devoted to research, training program directors began to increase the amount of time devoted to instruction in investigative techniques and to allow junior faculty more opportunity for investigative studies.

Contributions to the Growth of Pediatric Cardiology by Professional Societies

The Cardiology Section of the American Academy of Pediatrics. This Section was organized in 1957 to provide a forum for presentation of scientific papers and the exchange of ideas among pediatric cardiologists. In addition, the Section has been continuously concerned with the establishment of standards of care for cardiac patients (2).

Council on Cardiovascular Disease of the Young of the American Heart Association. The American Heart Association’s Council on Cardiovascular Disease of the Young, formerly known as the Council on Rheumatic Fever and Congenital Heart Disease, has, since its inception, addressed topics important to treatment of pediatric patients with heart disease, including the establishment of standards for care (3,4). This Committee prepares and continually updates statements for physicians, for example, on prophylaxis against infective endocarditis. Council members met with major life insurance companies and assisted in the development of reasonable criteria for insurability of patients with congenital heart disease, both preoperative and postoperative (5). Until then, life insurance companies seldom covered the patient with any form of congenital heart disease.

The Committee on Pediatric Cardiology of the American College of Cardiology. This Committee is now concerned with continuing medical education programs in pediatric and congenital heart disease. Undoubtedly, these education programs have an influence on patient care throughout the United States.

Today there are not enough young, recently trained pediatric cardiologists with both strong clinical skills and a research background to fill the academic positions that are available. a problem that will increase during the next 10 years because of attrition by age. The Committee is studying these manpower needs for pediatric cardiology in this country (personal communication: Standing Committee on Pediatric Cardiology of the American College of Cardiology: Graham TP [chairman], Engle MA, Gutgesell HP, Manning JA, Nadas AS, Rosenthal A, 1982).

Contributions to Patient Care Through Programs Supported by the Federal Government

The Inter-Society Commission for Heart Disease Resources. This Commission was established in 1969 to set optimal standards for patient care; the report for pediatric cardiology was published along with the reports for other disciplines in cardiovascular disease in 1971. The paper “Prevention, Acute Care and Long Term Care for Congenital Heart Disease and for Pediatric Cardiology” (6) has set standards adopted by many institutions throughout the country.

The Crippled Children’s Services. This agency, developed in the 1960s, was designed to support the diagnosis and definitive surgical treatment of infants and children with congenital heart disease. Its service has provided the means by which many cardiac programs for children have been able to function. In some states the Crippled Children’s Services Advisory Committee is charged with quality control by periodic review of the conduct of approved programs of the Crippled Children’s Services.

Training programs. The National Heart, Lung, and Blood Institute supported clinical cardiology training for approximately 20 years at a time when the diagnosis and treatment of each patient with congenital heart disease was virtually a research effort. Many of the current pediatric
cardiologists in cardiac centers were trained under this program. In the past 10 years the Institute's training support has been confined to research training, thus potentiating our expectation for further improvement in treatment of heart disease in the pediatric patient.

Grants-in-aid. The research grants of the National Heart, Lung, and Blood Institute have provided the means for most of the developments discussed or listed in this review. Future improvements in patient care of infants and children with heart disease will rely on a continuation of this support.

Early Diagnosis and Referral
Changes in Attitude Concerning the Infant With Congenital Heart Disease

In 1958 cardiac centers were just beginning to take a close look at the problem of the infant with a morbid form of congenital heart disease. Before that time many centers put higher priority on the patient beyond 2 years of age, simply because the diagnosis and treatment of the older child was associated with a more favorable outcome. In several reports in the late 1950s operations for congenital heart defects in infants who were expected to die proved to be successful, with early survival surgery in about 70% of them (7). Ochsner et al. (7) pointed out that despite a concerted effort to treat critically ill infants, during a 6 year period (1954 to 1960) 300 operations were performed on infants under 1 year of age for critical congenital heart disease (with a 74% survival rate), but 115 infants with congenital heart disease under our care died without an attempt at surgical treatment, although half of them had defects amenable to surgery. This study prompted a more aggressive approach to definitive diagnosis and treatment in our hospital and probably in others around the country.

Late Diagnosis of Congenital Cardiac Defects: Reasons for the Problem and Steps to Correct It

At this time it became clear that the diagnosis of heart disease was being made rather late in the course of an infant's illness (8). This late diagnosis resulted sometimes because the heart murmur was late in appearing and in other cases because the symptoms of heart failure in the infant were nonspecific; these symptoms often suggested disease of some system other than the cardiovascular, especially when no heart murmur could be heard (9). Tachypnea in the infant, despite the absence of other traditional signs of heart disease, was emphasized as a manifestation of pulmonary edema and elevated left atrial and pulmonary venous pressures. Between the years 1969 and 1981, during which the reasons for late diagnosis and referral were made known to primary care physicians and nurses responsible for care of the newborn, the percent of infants referred among the total number of new patients per year rose from 24 to 46% and the percent of patients referred on the first day of life increased from 3 to 13% (10).

Contributions of the New England Regional Infant Cardiac Program: A Model of Patient Care for the Nation

This program provided further stimulus for early referral and served as the standard for the entire country for ideal management of the infant with critical congenital heart disease (11). The program was established in 1968 and by 1974, case finding increased by 20% and the number of deaths in patients not reaching the hospital decreased by 50%. In 1968, 20% of the patients had been referred to the New England hospitals by less than 2 days of age; by 1977, 34% of the patients referred had reached the hospital by this age. The New England program did result in a decrease in the number of hospitals offering infant cardiac services. The data clearly pointed out what many had suspected. Results in hospitals with pediatric cardiac services that managed only a small number of patients per year were inferior to results in hospitals treating a large number of patients per year. Patient care was best served by reducing the number of cardiac centers with a small case load.

Another contribution of the New England Regional Infant Cardiac Program was that the investigators tabulated the number of extracardiac anomalies in a large number of patients (2,251) and found that only 11% of all patients had a severe uncorrectable extracardiac defect, that is, one that interfered with the life or well-being of the patient (for example, Down's syndrome). Another 8% of patients with a cardiac defect had a severe, but correctable, noncardiac defect and in yet another 8% the extracardiac defect was mild and required no treatment. In anomalies such as transposition of the great arteries, severe noncardiac anomalies were extremely rare (0.4%), whereas in anomalies such as endocardial cushion defect, 45% of the patients had severe extracardiac anomalies (especially Down's syndrome). If babies with Down's syndrome are excluded, the patients with endocardial cushion defect had a 19% incidence rate of severe extracardiac anomalies.

Some physicians had assumed that patients with congenital heart disease had such a large incidence of extracardiac anomalies that major efforts to correct the cardiac defect might not always be rewarding. Thus, the knowledge that the number of uncorrectable extracardiac anomalies is much lower than many physicians thought has been a contribution to the enthusiasm with which pediatricians are likely to refer and treat these patients and to the willingness of the state Crippled Children's Services to continue to support them.
Cardiac Catheterization

By January 1958, cardiac catheterization and angiographic techniques had been so well developed during the preceding 10 years that few cardiologists at the time would have expected much change. However, the developments in the 25 years after 1958 have been so extensive as to revolutionize the capabilities of the catheterization laboratory.

Contribution of the Cooperative Study

The cooperative study on cardiac catheterization published in 1968 by Braunwald and Swan (12) emphasized the high mortality rate (6.2%) of cardiac catheterization in infants less than 1 month of age and the high complication rate (15.3%); subsequently steps were taken in virtually every center to improve the situation.

The report of the New England Regional Infant Cardiac Program (11) revealed that 2,535 cardiac catheterizations were performed between 1969 and 1976 and there was no death within 24 hours of catheterization among infants more than 4 months old. Braunwald and Swan (12) reported that there had been five deaths in infants more than 4 months of age among the 1,161 infants less than 1 year of age who underwent cardiac catheterization.

Contribution of Technical Advances for Studying Pediatric Cases

Miniaturization of equipment and supplies. Miniaturization of equipment and supplies appropriate for use in the infant brought about a great deal of improvement in the safety and the information gained from the procedure.

Control of body temperature. Control of body temperature during the procedure added to the safety of cardiac catheterization. It had not been customary in most centers to measure body temperature continuously or even frequently during cardiac catheterization. Recognizing that increased metabolic activity was harmful to the patient with congestive heart failure, many clinicians assumed that a low body temperature might even be somewhat protective and were inclined to ignore body temperature at less than physiologic range. But the harmful effect of reduced body temperature on cardiovascular function was pointed out in a study by Silverman et al. (13).

Monitoring of blood gases. Further improvement came with the frequent monitoring of the patient’s status by blood gases, pH levels and the metabolic state, all of which could become quickly altered in the critically ill infant with congestive heart failure or cyanosis, or both: it was learned that measurable biochemical changes preceded the clinically obvious manifestations.

Use of image intensifier. The image intensifier was devised to reduce radiation exposure to the patient and personnel, but it also allowed viewing of the fluoroscopic image with enough light in the room to permit observation of the clinical appearance of the patient.

Other technical advances. Other advances in the catheterization laboratory include the technique for percutaneous catheter entry (14,15), the use of disposable catheters, plastic needles for atraumatic arterial cannulation and the development of less toxic radiopaque contrast media.

Improved angiographic techniques. The availability of biplane cineangiography improved diagnostic sensitivity and accuracy and, by reducing the number of injections of contrast medium needed and avoiding turning the patient on the side for a lateral projection, greatly increased the safety of the procedure. Biplane fluoroscopy has added to the safety of catheter manipulation, by allowing a lateral view of the course of the catheter, and has further increased the chance of pulmonary artery entry (16).

The angle views of Bargeron et al. (17) first made it possible to pinpoint the exact location of the ventricular septal defect as well as the different types of atrial septal defect. Likewise, these angle positions have allowed an unobstructed view of the pulmonary artery branches and have improved ability to detect pulmonary artery branch stenosis before operation. The use of videotape for immediate replay and review of the angiogram while the patient is still in the catheterization laboratory has improved diagnostic ability in small infants and allows an immediate decision for treatment while the catheterization is still in progress.

Umbilical vessel catheterization. Umbilical vessel catheterization techniques have made it possible to perform studies of complicated malformations in the first few hours of life without resorting to percutaneous entry or cut-down (18,19).

Transseptal left heart catheterization. The transseptal left atrial catheterization was first reported in 1959 (20), but the procedure was largely abandoned because of the limitation of equipment and the danger of inadvertent extracardiac entry. The use of this technique was revived by Duff and Mullins (21) and proved feasible in small infants as a result of improvement of the equipment and the use of the lateral fluoroscope to more accurately guide the course of the needle (1).

Sedation for cardiac catheterization. In 1958, Smith et al. (22) reported on their experience with an ataractic “cocktail” composed of meperidine hydrochloride, chlorpromazine and promethazine for use in infants and children undergoing cardiac catheterization. This method was adopted by the majority of catheterization laboratories in this country and abroad that were concerned with studying infants and children. Israel et al. (23) studied the effect of this mixture on the blood gas tension in subjects without cardiopulmonary disease and found that in only 3 of the 58 children studied was there an effect on ventilatory function. They concluded that the mixture was safe for use in infants and children without profound effect on ventilatory function. They did not, however, study patients with cyanotic heart disease or...
those with pulmonary edema in whom heavy sedation can profoundly interfere with gas exchange in the lung and depress cardiovascular function. Their study did point out the advisability of monitoring the arterial blood gases and pH by the new polarographic and potentiometric techniques for measuring oxygen and carbon dioxide tension. It may have been their report on the effect of the catheterization "cocktail" that led to the widespread use of monitoring of blood gas tension and pH during catheterization in order for the clinician to detect and manage early preclinical changes.

**Balloon Atrial Septostomy**

William Rashkind’s balloon atrial septostomy stands as one of the most important therapeutic procedures developed in the past 25 years for the treatment of certain types of congenital heart anomalies (24). Also, the concept of balloon catheter manipulation has had a profound influence on the entire field of interventional catheterization.

The Rashkind septostomy:

1. Provided a nonsurgical means of creating an atrial septal defect and thus virtually replaced the Blalock-Hanlon operation, a procedure that was technically difficult and carried a mortality rate of 20 to 50%.

2. Increased the safety of cardiac catheterization in the critically ill neonate with transposition of the great arteries by providing immediate clinical improvement, a rise in arterial oxygen saturation when it was severely depressed and a reduction in left atrial pressure when it was abnormally elevated (25).

3. Led to a broadened indication for creation of atrial septal defect in congenital heart disease, including: (a) pulmonary valve atresia with intact ventricular septum and tricuspid atresia with pulmonary stenosis or atresia, so that, if a later systemic to pulmonary artery anastomosis must be performed, the large atrial septal defect provides better tolerance of the increased pulmonary venous return to left atrium (26); (b) total anomaly of pulmonary venous return and other anomalies in which there is either volume or pressure load on the right or the left atrium. The procedure allowed more infants to survive the first few weeks and months of life and to reach an age and size reasonable for definitive repair.

4. Probably stimulated pediatric cardiologists to use the Swan-Ganz gas-inflated balloon catheter to flow guide the catheter tip for difficult entry into certain cardiac chambers. Because the report of Rashkind and Miller preceded that of Swan and Ganz (29) by about 4 years, some have wondered if Rashkind’s balloon catheter stimulated the original concept and development of the Swan-Ganz flow-directed balloon-tipped catheter. However, I learned recently that this was not the case. Swan explains how the idea occurred to him: "One bright fall afternoon in 1967, while observing the motion of sailboats on Santa Monica Bay, I wondered if the attachment of a sail to a cardiac catheter would allow progression to the pulmonary artery and pulmonary artery wedge position. . . . ." (30). Swan discussed his idea with engineers. It evolved that a balloon would be less cumbersome than a sail and would avoid the chance of thromboembolic complications.

5. The Rashkind procedure made cardiologists more conscious of the dynamics involved in transposition of the great arteries, total anomaly of pulmonary venous return and other anomalies in which there is either volume or pressure load on the right or the left atrium. The procedure allowed more infants to survive the first few weeks and months of life and to reach an age and size reasonable for definitive repair.

The dexterous Rashkind had not appreciated that the procedure might be less successful in other cardiologists’ hands. Rashkind had taught that rapid withdrawal across the foramen ovale of the fully inflated balloon was important, but he insisted that the procedure is actually very simple: "All it requires is a big jerk on the end of the catheter. . . . ." Rashkind would quip whenever he was invited to speak on his method. It was partly Rashkind’s humorous and entertaining manner of speaking, his enthusiasm for teaching and his interest in travel that took him to virtually every major city in most countries of the world to tell about the balloon catheter and the whole subject of transposition of the great arteries. Here, for the first time, was a lifesaving procedure for the patient with transposition which could be carried out in communities with limited surgical facilities.

However, the technical skill required for successful septostomy may have discouraged a few cardiologists from persisting in the method and this limitation of balloon atrial septostomy may have led some to search for the means to achieve early definitive repair in anomalies such as transposition. Today, in some cardiac centers, these defects can be surgically corrected in small infants even in the first few days of life if septostomy is unsuccessful; thus the need for long-term palliation by creation of a large atrial septal defect may become less critical. Nonetheless, balloon atrial septostomy remains useful in a large number of critically ill infants with congenital heart disease, allowing the patient to arrive in the operating room in better hemodynamic condition and increasing the chance of long-term survival.
Prostaglandin E₁

Coceani and Olley (34) demonstrated dilation of the ductus by infusion of prostaglandin E₁ in fetal lambs. Others confirmed the observation in different fetal animals. The drug was beneficial in opening the ductus and raising the systemic arterial oxygen saturation in neonatal patients with ductus-dependent congenital heart defects (35–37). Continuous administration of prostaglandin E₁ for hours, days or weeks, if necessary, may allow either spontaneous improvement sufficient to stop the treatment or time to prepare for surgical creation of a systemic to pulmonary artery anastomosis.

Prompted by the finding of Clyman et al. (38) that ductus relaxation could occur with prostaglandin E₁ despite a normal arterial oxygen tension, Heymann et al. (39) used the drug successfully in opening the ductus in patients with interruption of the aortic arch and in those with coarctation of aorta with closing ductus; Lang et al. (40) reported similar success.

Prostaglandin E₁ has been used in the patient with interrupted aortic arch to obtain temporary clinical improvement in the patient’s cardiovascular status until a bypass graft could be placed surgically across the interruption. Surgical success with this anomaly, which is invariably lethal if untreated in the first few days of life, is now possible in certain cases because of the improvement obtained with prostaglandin E₁.

In a study involving 56 centers (41), prostaglandin E₁ was used in 492 infants with ductus-dependent anomalies. Prostaglandin E₁ provided excellent palliation for the 385 infants with ductus-dependent cyanotic congenital heart disease. Among the 107 acyanotic patients with either coarctation of the aorta or interrupted aortic arch, dilation of the ductus produced clinical improvement in 80% of each group and reduced the pressure gradient from 45 to 9 mm Hg.

Indomethacin

It was not until the mid 1960s that investigators of neonatal cardiopulmonary problems recognized that a large ductus arteriosus coexisting with respiratory distress syndrome in the premature infant could be so silent clinically yet aggressive, if not cause, the pulmonary disease in some 15% of premature infants weighing less than 1,750 g (42,43). Despite small size and poor condition, many such premature infants who had been unresponsive to digitalis and ventilatory support recovered after surgical ligation of the ductus (44–46).

On the basis of knowledge that prostaglandins E₁ and E₂ dilate the naturally constricted ductus in fetal animals and in neonatal human patients (with a ductus-dependent cardiac defect), Friedman et al. (47) theorized that administration of a prostaglandin inhibitor might constrict the ductus. Flower and Vane (48) had shown that indomethacin is a potent inhibitor of prostaglandin synthetase. The report of Friedman et al. concerned the success with ductus closure after administration of indomethacin to six consecutive premature infants for whom surgical ligation of the ductus would ordinarily have been performed.

Heymann and Rudolph (49) had inhibited prostaglandin synthesis in fetal lambs by administration of aspirin and gave the drug to 3 premature infants with respiratory distress syndrome with success in 1, but confirmed Friedman’s results in ductal constriction with indomethacin in 14 of 15 premature infants.

The studies of Heymann et al. (49,50) demonstrated the usefulness, and the necessity, of the echocardiogram in supporting the diagnosis of ductus and in following the course of the infant with critical cardiopulmonary disease during pharmacologic manipulation of the ductus. Because of the atypical murmur of ductus in the premature infant, pharmacologic closure of the ductus often could be confirmed only by a decrease in the echo-derived ratio of diameter of left atrium to aortic root. As a result, echocardiographs were made available to many neonatal intensive care units, aiding the diagnosis and follow-up of infants with all forms of cardiopulmonary dysfunction (51,52).

In a large, multicenter, prospective, randomized study indomethacin has proved to be of value. Nadas, the principal investigator, at first characteristically cautious about indomethacin (53), now has this to say at the end of the study (which awaits publication): “Indomethacin ought to be used in small (less than 1,750 g) premature when he has a hemodynamically significant ductus and who within a short period of time do not respond to anticongestives.” Nadas and his co-investigators found that in about 75% of cases the ductus would close with indomethacin. (Nadas AS, personal communication, 1982.)

Beta-Adrenergic Blockade

Until the mid 1970s the occurrence of a spell of profound hypoxemia in the patient with tetralogy of Fallot (or other malformations with ventricular septal defect and severe pulmonary stenosis) was one of the emergency situations that called for immediate operation. In many cardiac centers today, such spells appearing in an infant who is below the ideal age and size for intracardiac repair can usually be handled with the use of propranolol. The drug offers the best medical means currently available to prevent recurrences and allows a reasonable delay in definitive surgical repair.

In 1964, Honey et al. (54) predicted that beta-receptor blockade would have a place in the treatment of cyanotic attacks in tetralogy of Fallot. Singh and Gotsman (55) used the beta-receptor adrenergic blocking agent pronethalol for the successful treatment of an acute attack.

The study carried out at the Medical University of South Carolina and reported by Ponce et al. (56) was the first to
show that propranolol was effective in abolishing spells of paroxysmal hypoxemia in 17 of their 18 patients, allowing a delay in operation ranging from 3 to 27 months (55). The drug was used with 80% success by Garson et al. (57), allowing a safe delay in operation averaging 13 months. Propranolol treatment was not associated with low cardiac output after intracardiac repair. It was once rumored that propranolol treatment is associated with a high risk of myocardial depression after operation. This widespread notion was dispelled by the study of Garson et al.

Beta-adrenergic blockade has also found wide use for: 1) relief of symptoms in left ventricular hypertrophic cardiomyopathy, 2) control of supraventricular tachycardia, especially in patients who have Wolff-Parkinson-White syndrome with short refractory period of the accessory connection, and 3) control of systemic hypertension.

Echocardiography

Diagnostic application. One of the first texts on echocardiography (58) concerned pediatric application of the method. Echocardiography has become indispensable in the management of patients with known or suspected congenital cardiac disease, especially of the seriously ill newborn. In one of the earliest applications of M-mode echocardiography to congenital heart disease, Meyer and Kaplan (59) demonstrated both left and right hypoplastic heart anomalies. Others demonstrated the utility of echocardiography in the diagnosis of transposition of the great arteries (60), endocardial cushion defect (61), Ebstein’s anomaly (62), tetralogy of Fallot (63) and total anomalous pulmonary venous connection (64). With the rapid improvement of two-dimensional echocardiographic equipment in the mid and late 1970s, it has become possible to make a precise anatomic assessment of virtually all of the major congenital cardiac defects (65–67). Perhaps as important as the ability to establish the diagnosis of congenital heart disease in the neonate is the ability to exclude structural cardiac deformities as a cause of cyanosis or respiratory distress; today cardiac catheterization usually can be avoided in the infant with cyanosis due to lung disease or persistent fetal circulation.

Serial studies. Because it is safe and painless, echocardiography is also valuable in the serial assessment of children with known cardiac disease. Measurement of systolic time intervals from pulmonary valve motion provides an estimate of pulmonary artery pressure with normally related great arteries (68) and in transposition of the great arteries (69). Measurement of left ventricular wall thickness and cavity diameter provides an estimate of left ventricular pressure in children with aortic valve stenosis (70). Serial measurement of left ventricular diameter and shortening fraction may be used to follow changes in ventricular function in children with myocardial disease (71).

Evaluation of murmurs. In addition to its value in management of children with serious forms of cardiac disease, echocardiography has provided an explanation of some cardiac murmurs in less severe but far more common disorders. The most notable examples are mitral valve prolapse and bicuspid aortic valve. Although these lesions are usually of no hemodynamic significance, noninvasive documentation provides a rational basis for parental reassurance and for recommendation of bacterial endocarditis prophylaxis.

Improved Diagnosis and Management of Cardiac Arrhythmias in Pediatric Patients

Electrophysiologic testing. Our understanding of cardiac arrhythmias in children began a “growth spurt” in 1969 when Scherlag et al. (72) published their paper on His bundle recording in human beings. Watson et al. (73) had published an example of a His bundle recording in 1967, but the usefulness of the technique was not apparent at that time. Brodsky et al. (74), Gillette et al. (75) and several other groups have shown that the technique is feasible in children.

The usefulness of electrophysiologic testing in children was amplified by the addition of programmed stimulation as demonstrated first in the evaluation of arrhythmias after the Mustard baffle operation for transposition of the great arteries (76,77). Probably the most important use of electrophysiologic techniques in children has been in the evaluation of supraventricular tachycardia (78), particularly that associated with the preexcitation syndromes (79).

Studies on the mechanisms and potential consequences of arrhythmias have resulted in a more systematic and successful use of antiarrhythmic drugs. Two that have proved useful in the pediatric patient are verapamil (80) for supraventricular reentrant tachycardias and phenytoin (81) for ventricular arrhythmias.

Intraoperative applications. Electrophysiologic techniques have been found useful in the cardiovascular operating room in patients who have tachyarrhythmias resistant to medical treatment. The Wolff-Parkinson-White syndrome may be treated with success by operation in more than 90% of the patients by locating and dividing the accessory connection. Patients without overt Wolff-Parkinson-White syndrome may have concealed accessory connections and thus are candidates for surgery as well (82). Intraoperative mapping has also led to the successful surgical treatment of automatic focus atrial and junctional tachycardias and of ventricular tachycardia in children (83).

Pacemakers. Technologic advances in pacemakers and leads have allowed the increased use of pacemakers in children (84). The size of both pacemakers and leads has decreased so that transvenous techniques are applicable to children as young as 5 years of age. New pacemakers allow the restoration of atrioventricular synchrony, thus improving cardiac performance.
Conclusion

It is difficult to select only eight areas of advancement in pediatric cardiology that have contributed to the medical care of the patient since 1958. In preparing for this review I asked the 11 members of our faculty in pediatric cardiology to list the advances of the past 25 years, other than those that I covered, that they would consider important to the medical care of the patient. The following list includes only some of their suggestions, not listed chronologically or in order of importance:

- Afterload reducing agents
- Neonatal transport
- Neonatal intensive care units
- Better utilization of nursing talent in care of infants with heart disease
- Microtechniques for blood gases
- Transcutaneous oxygen monitor
- Respirators for ventilatory support
- Twenty-four hour coverage in tertiary care centers for cardiac catheterization and cardiovascular surgery
- Open radiant warmer for neonatal care
- Doppler blood pressure device
- Nuclear cardiology for quantification of left to right shunt and for ventilation-perfusion studies
- Telephone electrocardiographic monitor
- Holter 24 hour monitor
- Telemeterized electrocardiographic monitor
- Exercise stress testing
- Antihypertensive treatment
- Potent oral diuretic agents
- Behavioral science in the care of patients with heart disease
- Potent antihypertensive agents
- Anticoagulant therapy
- Deep vein thrombosis
- Monitoring of right and left heart filling pressures
- Van Praagh's segmental approach to diagnosis of complex congenital heart disease
- Understanding of oxygen toxicity in the neonate and with prolonged ventilatory support
- Pulmonary vein wedge angiography for distal pulmonary artery visualization in pulmonary trunk atresia (85).

It is the task of investigators, both current and future, to explore methods to solve the unmet needs of the pediatric patient with heart disease or the potential for heart disease. It is the task of the clinician, who is involved daily with the diagnosis and treatment of patients with a cardiovascular problem, to identify these needs, to lay them on the workbenches of the research laboratories in our academic institutions and to call for public support of the studies that are required to respond to these needs.

References


