Congenital Heart Disease in Adults
A New Cardiovascular Subspecialty

Joseph K. Perloff, MD

Advances in diagnostic methods and in the surgical and medical care of infants and children with congenital heart disease have resulted in one of the most successful rehabilitation programs that medicine has witnessed. In the United States alone, there are presently between 500,000 and 600,000 adults with congenital heart disease. Each year, about 20,000 open operations for congenital heart disease (an approximate minimum) are performed in the United States (personal communication, 1990, Dr. Peter Einstein, Assistant Professor of Surgery and Pediatrics, University of Tennessee). Of an estimated 25,000 infants currently born with congenital malformations of the heart and circulation, over 85% will reach adulthood. The heavy investments in time and resources needed to accomplish these results should now be matched with a similar dedication to the adolescent and adult with congenital heart disease. Potential subspecialty professional resources (1989 figures) include 884 board-certified pediatric cardiologists, too few to cope with the patient load of adults with congenital heart disease even if all 884 were committed to do so, and 12,119 board-certified medical cardiologists, only a small minority of whom have knowledge of, interest in, or responsibility for adults with congenital cardiac malformations. Congenital heart disease in adults is still a largely unrecognized subspecialty but is emerging as a discipline that requires special expertise.

The adult congenital heart disease patient population includes those who have never undergone cardiac surgery, those who have undergone cardiac surgery and require no further operation, those who have had palliation with or without anticipation of reparative surgery, and those who are inoperable apart from organ transplantation. The following remarks deal broadly with what this new area of cardiovascular interest presently comprises: multidisciplinary facilities for comprehensive care, survival patterns (natural and postoperative), medical considerations, surgical considerations, and postoperative residua and sequelae.

The seminal contributions of Maude Abbott, Robert Gross, Helen Taussig and Alfred Blalock, and Clarence Crafoord set the stage. The postwar introduction of cardiac catheterization was a major step forward, and the development of extracorporeal circulation in the early to mid 1950s was destined to make virtually all congenital malformations of the heart accessible to the skills of cardiac surgeons. Survival patterns have been affected, often profoundly. Congenital heart disease should therefore be considered not only in terms of age of onset but also in terms of the age range that survival now permits. Long-term management remains concerned with natural survival but is increasingly concerned with the growing numbers of postoperative patients who, with few exceptions, continue to need medical surveillance. Unoperated adults experience improved longevity and well-being owing to refinements in the medical management of hematologic disorders, renal function, urate metabolism, pulmonary physiology, infective endocarditis, electrophysiological abnormalities, pregnancy, and non-cardiac surgery. The ideal cardiac surgical objective—cure in the literal sense—is rarely achieved. Accordingly, operations leave behind a broad range of residua and sequelae that may require prolonged, if not indefinite, medical attention. Uninterrupted, long-term care is essential if the concerns inherent in this new and increasing patient population are to be addressed.

Multidisciplinary Facilities for the Comprehensive Care of Adults With Congenital Heart Disease

Specialized tertiary medical facilities for the care of adults with congenital heart disease were begun in the 1960s at the Toronto General Hospital (now the Toronto Hospital) and evolved there into the Congenital Heart Disease Centre. An analogous facility was developed in 1975 at the National Heart Hospital London (now the Royal Brompton and National Heart Hospitals). Development of a similar facility at the Mayo Clinic (1988) is indebted to the experience at the National Heart Hospital London. The UCLA Adult Congenital Heart Disease Center began in 1978, and the University of Iowa Adolescent/Adult Congenital Heart Disease Clinic, which began in 1981, is indebted to the UCLA experience. Over 6,000 adults with congenital heart disease are cur-
rently registered in these five specialized centers in Canada, Great Britain, and the United States.² A number of other teaching hospitals are following suit. A first necessity is to recognize the seminal role of pediatric cardiologists and pediatric cardiac surgeons, without whose efforts congenital heart disease in adults would not be a matter of importance. Relevant to this topic are the staff, the laboratories, the criteria for patient entry, the referral sources, the outpatient and inpatient management, the multidisciplinary consultants, and the educational, training, and research commitments of the specialized facilities for adults with congenital heart disease.¹ ²

The staff reflects the current consensus that patients are best managed, at least in the foreseeable future, by collaboration between pediatric and medical cardiologists with the assistance of cardiovascular nurse specialists and physician assistants. Collaborating cardiac surgeons must be equipped to deal with both congenital and acquired cardiovascular diseases and with surgical problems that are often unique to the adult with congenital heart disease. Dedication to task is a collaborative effort, especially in the setting of a university hospital in which intellectual interchange, teaching, and research are as paramount as optimal patient care.

Criteria for patient entry into the specialized facilities are based on age and psychological and physical maturity. Entry may begin at age 12 years, recognizing adolescent medicine as a bridge between the preadolescent patient and the adult. Alternatively, admission may begin at ages 17 or 18 years, depending on maturity.

Referrals reflect the notion that tertiary centers are not designed to compete with practicing physicians or community hospitals but instead to offer services difficult, if not impossible, to duplicate. An important source of referrals is pediatric cardiology at the parent institution, but significant and growing numbers of referrals are from internists, pediatricians, family practitioners, or cardiologists within or outside the immediate geographic area (state, out-of-state, and abroad).

There is a mounting consensus that outpatients are more appropriately seen in clinics for adults (i.e., that adult care is best provided in an adult setting). Advanced planning of diagnostic studies is obligatory for patients who are referred to the outpatient facility from long distances. The number of patients per outpatient clinic largely determines the amount of time available for sharing information between staff and trainees, underscoring the educational mission of the facility including teaching of medical students, house officers (pediatric and medical), and pediatric and medical cardiac fellows.

Inpatients include admissions for cardiac or noncardiac surgery, for labor and delivery, for the medical management of arrhythmias or heart failure, or for the management of noncardiac medical disorders. Inpatients constitute an important part of the educational experience of house staff and fellows. The clinical nurse specialist coordinates the inpatient/outpatient interface.

Noninvasive, catheterization, and angiographic laboratories for adults with congenital heart disease must meet the standards established by pediatric cardiology laboratories. Adults are best studied in laboratories that are designed for adults, but quality should not be compromised for expediency or design.

Noncardiac consultants should be formally incorporated into the adult congenital heart disease facility, including specialists in hematology, nephrology, urate metabolism, pulmonary medicine, electrophysiology, genetics and epidemiology, gynecology and obstetrics, psychiatry, anesthesiology, pathology, social services, insurability, and vocational counseling.¹ ² The objective is to have ready access to consultants who have gained experience in the specific problems associated with congenital heart disease in adults.

Adult congenital heart disease facilities assume a major role in the education and training of fellows, residents, nurse specialists, and physician assistants who will become the next generation of responsible professionals.¹ ² ³ If the medical and pediatric specialty boards placed more emphasis on congenital heart disease in adults, the percent of interested fellows and training program directors would increase accordingly. The cardiology specialty boards can play pivotal roles in encouraging fellowship programs to adjust their curricula. If passing the boards required an appropriate level of knowledge of congenital heart disease in adults, the training programs are likely to take cognizance.² For medical cardiac fellows, formal rotations (inpatient and outpatient) including a rotation in pediatric cardiology should be part of their experience. Anticipated professional requirements dictate the need for specialized fellowship training designed for cardiologists who wish to qualify as career experts in the evolving field of congenital heart disease in adults.² Educational responsibilities extend to community physicians, to visiting physicians from other institutions, and, importantly, to the patients themselves. Stratification of patient care is a major issue.² The base of the pyramid is the primary care physician (general pediatrician, general physician). The next level (stratum) is the cardiologist (pediatric, medical) who provides both consultative and primary care in the community. Tertiary care depends on regional specialized centers that provide experience and expertise even with rare and complex lesions. It is not yet clear how large a population is needed to serve a tertiary care center.

A commitment to research is prompted by a desire to address unresolved questions intrinsic to the adult congenital heart disease population. The investigations generally require collaboration with colleagues in a number of other specialties, thus promoting valuable interdisciplinary interchange. A research base must be provided for cardiac fellows interested in careers in academic medicine.
TABLE 1. Common Defects With Expected Adult Survival

<table>
<thead>
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<th>Defect</th>
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<tbody>
<tr>
<td>Bicuspid aortic valve (functionally normal, stenotic, incompetent)</td>
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<tr>
<td>Coarctation of the aorta</td>
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<tr>
<td>Pulmonary valve stenosis</td>
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<tr>
<td>Atrial septal defect secundum</td>
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<tr>
<td>Patent ductus arteriosus</td>
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Natural Survival Patterns

Natural survival includes malformations that do not require operation, those that remain amenable to operation in adulthood, and those that are inoperable except for organ transplantation. Management of adults with congenital heart disease must take into account acquired disorders of the heart and circulation and noncardiac illnesses that may coexist and modify the physiological expressions of the basic congenital cardiac malformation. Natural survival patterns, in order of clinical importance, include common defects with expected adult survival (Table 1), uncommon defects with expected adult survival (Table 2), and common defects with exceptional adult survival (Table 3).

Survival Patterns After Surgery or Interventional Catheterization

An understanding of prognosis after cardiac surgery or interventional catheterization requires knowledge of the preoperative congenital malformation, the nature and effects of the therapeutic intervention, and the postoperative or postinterventional residua and sequelae. Success is measured not only by the length of survival but by the quality of life and the need for reoperation. Patients who underwent cardiac surgery decades ago benefited from the anatomic repairs but often suffered from the deleterious effects of what would now be considered inadequate myocardial protection. Prosthetic materials—valves, patches, and conduits—that were state of the art decades ago have been superseded by many generations of improved devices and materials. Late survival after cardiac surgery or interventional catheterization relates to repair or replacement of congenitally malformed cardiac valves; intra-atrial repairs including atrial septal defects, Mustard or Senning venous switch operations for complete transposition of the great arteries; intraventricular surgery through a right atrial incision or through a ventriculotomy (Fallot’s tetralogy, for example); central arterial surgery (patent ductus arteriosus, coarctation of the aorta, sinus of Valsalva aneurysms) and partial or complete caval to pulmonary artery communications (Glenn or Fontan procedures).

Medical Considerations

Cyanotic Congenital Heart Disease: Hematologic Management, Renal Function, and Urate Metabolism

Cyanotic patients with erythrocytosis have been classified as “compensated” or “decompensated.” Those with compensated erythrocytosis establish equilibrium hematocrts in iron-replete states. Symptoms attributable to hyperviscosity are usually mild or absent when hematocrit levels are lower than 65% and absent, mild, or moderate even at high hematocrit levels, occasionally 70% or more. Phlebotomy for relief of hyperviscosity symptoms is required rarely, if at all. Patients with decompensated erythrocytosis fail to establish equilibrium conditions and manifest unstable, rising hematocrit levels and recurrent, moderate to severe symptoms attributable to hyperviscosity. Erythrocyte production is not regulated; feedback inhibition fails to occur. Symptomatic hyperviscosity is common, prompting therapeutic phlebotomy that temporarily relieves symptoms but depletes iron stores. Because symptomatic hyperviscosity in an iron-replete state seldom occurs with hematocrit levels of lower than 65%, these symptoms in patients with hematocrit levels lower than 65% are almost always due to iron deficiency; phlebotomy aggravates rather than alleviates the symptoms.

TABLE 2. Uncommon Defects With Expected Adult Survival

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<tr>
<td>Situs solitus with dextrocardia</td>
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<td>Situs inversus with dextrocardia</td>
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<tr>
<td>Congenital complete heart block</td>
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<td>Congenitally corrected transposition of the great arteries</td>
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<tr>
<td>Congenital mitral regurgitation</td>
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<tr>
<td>Ebstein’s anomaly of the tricuspid valve</td>
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<tr>
<td>Uhl’s anomaly</td>
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<tr>
<td>Discrete subaortic stenosis</td>
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<tr>
<td>Congenital pulmonary valve regurgitation</td>
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<tr>
<td>Primary pulmonary hypertension</td>
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<tr>
<td>Atrial septal defect (primum, sinus venosus)</td>
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<tr>
<td>Lutembacher’s syndrome</td>
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<tr>
<td>Aneurysm of a sinus of Valsalva</td>
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<tr>
<td>Coronary arteriovenous fistula</td>
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<tr>
<td>Pulmonary arteriovenous fistula</td>
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<td>Anomalous course of a coronary artery</td>
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TABLE 3. Common Defects With Exceptional Adult Survival

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<th>Defect</th>
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<tr>
<td>Ventricular septal defect</td>
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<td>Fallot’s tetralogy</td>
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TABLE 4. Common Acyanotic and Cyanotic Malformations That Permit Survival to Childbearing Age

Acyanotic
- Atrial septal defect (secundum)
- Patent ductus arteriosus
- Pulmonary valve stenosis
- Coarctation of the aorta
- Aortic valve disease

Cyanotic
- Fallot’s tetralogy

Malformations are listed in descending order of prevalence among women.
In our experience, adults with cyanotic congenital heart disease are at little or no increased risk of stroke, even if the hematocrit level is above 65% and even if the erythrocytosis is decompen-dated (iron deficient).\textsuperscript{1,13,14} Cerebrovascular accidents in cyanotic adults are usually associated with excessive, injuri-ous phlebotomies or with the use of aspirin or anticoagulants that reinforce the intrinsic hemostatic defects (see below) and cause intracranial bleed- ing.\textsuperscript{1,14} Phlebotomy is not recommended for adult patients with compensated erythrocytosis, including those with hematocrit levels in the range of 70%, as long as symptoms attributed to hyperviscosity\textsuperscript{14} are mild or absent. Phlebotomy is recommended in pa-tients with marked to severe hyperviscosity symptoms and with hematocrit levels of 65% or greater, pro-vided dehydration is not the cause. Dehydration is treated by volume replacement, not phlebotomy. A comparatively safe, simple outpatient method for phlebotomy in adults involves removal of 500 ml of blood over 30 to 45 minutes followed by quantitative volume replacement, generally with isotonic saline, less commonly with salt-free dextran in 5% dextrose if patients have congestive heart failure.

When iron is administered in symptomatic iron-deficient patients with inappropriately low hematocrit levels, the dose should be small (325 mg ferrous sulfate per day). Hematocrit levels tend to rise quickly so the erythrocyte response must be moni-tored closely. Patients should be cautioned to avoid over-the-counter preparations that contain iron.

A significant increase in red cell mass can lead to inaccuracies in routine laboratory determinations of hematocrit. Hematocrit levels must be based on automated blood counts, because microhematocrit centrifugation methods result in plasma trapping with falsely elevated levels.\textsuperscript{1,15}

Home oxygen administration has sometimes been advocated, especially during sleep.\textsuperscript{15–19} From the hematologic and respiratory points of view, there is presently insufficient evidence that oxygen is useful in adults with cyanotic congenital heart disease, and the drying effect on nasal mucous membranes tends to increase the risk of epistaxes (see below).

Hemostasis is abnormal in cyanotic congenital heart disease.\textsuperscript{1,14–19} For the most part, bleeding ten-dencies are mild and are characterized by easy bruising, petechial hemorrhages in the skin and mucous membranes, epistaxis, gingival bleeding, and hemoptysis. Occasionally, however, epistaxis and hemoptysis can be copious and infuriatingly recurrent. Platelet counts are generally in the low range of normal, but when the increased blood volume is taken into account, the total circulating platelet mass is closer to normal than platelet concentrations imply. Inherent abnormalities in platelet function\textsuperscript{20} are reinforced by aspirin or nonsteroidal anti-inflammatory agents. In addition, abnormalities of the intrinsic and extrinsic coagulation systems with elevations of prothrombin time and activated partial thromboplastin time, re-spectively, and specific deficiencies of a number of coagulation factors have been reported.\textsuperscript{16} Aspirin, oral anticoagulants, and heparin are ill-advised because the inherent risk of stroke is low, because these drugs have no demonstrated efficacy in reducing that negligible risk, but instead may significantly aggra-vate the existing hemostatic defects and increase the risk of bleeding.

Serious hemorrhage may occur during surgery or accidental trauma.\textsuperscript{21} Phlebotomy has been shown to improve hemostasis temporarily in some erythrocytotic patients. Preoperative phlebotomy is therefore selectively used to reduce the hematocrit level to just below 65%.\textsuperscript{1} Phlebotomized units are reserved for potential autologous transfusions.

Renal function and urate metabolism are often abnormal in adults with cyanotic congenital heart disease.\textsuperscript{22,23} High plasma uric acid levels are secondary to inappropriately low renal fractional uric acid excretion rather than to urate overproduction.\textsuperscript{22} Enhanced urate reabsorption is believed to be caused by renal hyperperfusion reinforced by a high filtration fraction. Accordingly, hyperuricemia serves as a marker of abnormal intrauterine hemodynamics.\textsuperscript{22} Renal histopathology is characterized by enlarged, hypercellular glomeruli, basement membrane thickening, focal interstitial fibrosis, tubular atrophy, and hyalinization of afferent and efferent arterioles.\textsuperscript{24} Arthralgias are relatively common in erythrocytotic adults with cyanotic congenital heart disease, but acute gouty arthritis is relatively infrequent despite elevated uric acid levels, an observation similar to that in other forms of secondary hyperuricemia.\textsuperscript{25} If colchicine is used to treat acute gouty arthritis, care must be taken to avoid the dehydrating effects of vomiting and diarrhea. Nonsteroidal anti-inflammatory agents may then be considered but should be used cautiously because of the intrinsic hemostatic defects. Uricosuric agents are not routinely advised but can be efficacious in patients with hyperuricemia and recurrences of gouty arthritis.\textsuperscript{1}

\textbf{Cyanotic Congenital Heart Disease: Dynamics of Oxygen Uptake and Control of Ventilation}

Exercise may significantly increase the degree of venoarterial mixing and materially influence the dynamics of $O_2$ uptake and ventilation.\textsuperscript{20–28} Patients with cyanotic congenital heart disease exhibit marked abnormalities in achieving a new steady state for $\text{Vo}_2$ after the onset of exercise.\textsuperscript{26} Those with right-to-left shunts have greater increases in ventilation during exercise than do normal subjects; breathlessness on exertion may be a prominent clinical complaint un-associated with heart failure.\textsuperscript{27} Accordingly, assigning a New York Heart Association functional class is misleading (see below).

\textbf{Infective Endocarditis: Risks and Prophylaxis}

With the advent of intracardiac surgery and prosthetic devices, the clinical and bacteriological profile of infective endocarditis changed significantly.\textsuperscript{1,2,12,29} Some operations (for example, ligation of a patent
ductus arteriosus) eliminate the risk altogether, whereas other operations (shunts, prosthetic valves or conduits) materially increase the risk. However, certain general principles still prevail, namely, that two major causes predispose to infective endocarditis: a susceptible cardiac or vascular substrate and a source of bacteremia. Prophylaxis includes both chemotherapeutic (antimicrobial) and nonchemotherapeutic measures. Chemotherapeutic prophylaxis is based on the cardiac lesion, the source of potential bacteremia, and the absence or presence of the history of antibiotic sensitivity. Nonchemotherapeutic prophylaxis includes day-to-day oral hygiene, skin care, nail care, and in females, avoidance of certain contraceptive devices. The spongy, fragile gums of patients with cyanotic congenital heart disease are a special concern. Appointments with dentists or oral hygienists should be made twice yearly for teeth and gum prophylaxes. Meticulous skin care is important, especially in adolescents with acne. Young adults with cyanotic congenital heart disease are prone to develop acne, and pustules may be distributed beyond the face.

Pregnancy and Congenital Heart Disease: The Mother and the Fetus

The common congenital malformations of the heart and circulation in which natural (unoperated) survival of women into childbearing age can be anticipated are listed in Table 4 in approximate order of prevalence among women. However, the postoperative cardiac female now constitutes an important category of pregnancy and heart disease and represents a growing patient population. Successful cardiac surgery improves fertility and stabilizes the pregnancy in patients whose heart disease had reduced their sexual and ovarian functions. Women who were physiologically ill equipped to bear children or previously would not, in all probability, have reached reproductive age are now presenting for obstetric and cardiac care after reparative surgery for congenital heart disease. Although the expectant mother's cardiac reserve is reduced by the hemodynamic burden of pregnancy, the reduction can almost always be counteracted by addressing the factors that encroach on circulatory reserve. Maternal mortality has been said to vary directly with functional class, but in the presence of congenital heart disease, there are two important caveats: 1) cardiac symptoms on which the New York Heart Association functional classes were originally based are more relevant to acquired than to congenital heart disease, and 2) in the presence of certain congenital cardiac malformations, childbearing imposes such a formidable threat to maternal survival that pregnancy is proscribed or should be interrupted irrespective of functional class.

A strong consensus favors spontaneous vaginal delivery. Labor in the lateral decubitus position attenuates the hemodynamic fluctuations associated with major uterine contractions in the supine position. Cesarean section is reserved for cephalopelvic disproportion or for preterm labor in women taking oral anticoagulants. Because the probability of thromboembolism increases during the postpartum period, patients with lesions susceptible to paradoxical embolization (ostium secundum atrial septal defect, for example) require meticulous leg care, use of elastic support stockings, and early ambulation.

The need for antibiotic prophylaxis during routine delivery in pregnant cardiac patients is controversial. It has been argued that prophylaxis is unnecessary in uncomplicated vaginal deliveries because bacteremia is neither a natural nor an inevitable occurrence. However, it can also be argued that it is not prudent to assume that delivery will be uncomplicated, and delivery with an episiotomy is not, strictly speaking, uncomplicated.

Maternal congenital heart disease exposes the fetus to immediate risks that threaten its intrauterine viability and to remote risks that express themselves as congenital and developmental malformations. Immediate risks are chiefly determined by the functional class of the mother (with the qualifications mentioned earlier), by maternal cyanosis, and by anticoagulants. Remote risks to the fetus take the form of genetic parental transmission, teratogenic effects of certain cardiac drugs, and the harmful effects of certain environmental toxins and environmental exposures.

Exercise and Athletics Before and After Surgery or Interventional Catheterization

Consideration must be given to 1) the type, intensity, and duration of exercise; 2) the risk of body collision inherent in a given type of athletic activity, especially in patients receiving anticoagulants; 3) the training program (conditioning) required for a given sport; 4) the emotional response (stress) that the athlete experiences in anticipation of or during a particular sporting event; and 5) the risk of injury either to the athlete or to spectators if the athletic activity provokes loss of consciousness. Two categories of exercise are recognized, isotonic and isometric, but there is often a continuum between the two, with most physical activity incorporating both isotonic and isometric components. Relevant to this discussion are the type and severity of a given congenital malformation and the presence, type, and success of surgery. As a rule, the malformation under consideration is clinically overt (congenital aortic stenosis, for example) but an occult aberrant coronary artery coursing between the aorta and right ventricular outflow tract may first announce itself by exercise-induced angina pectoris, myocardial infarction, or sudden death. Pulmonary vascular disease is an important independent variable epitomized by primary pulmonary hypertension in which syncope on effort is a striking feature that often heralds sudden death. In Fallot’s tetralogy, isotonic exercise augments the right-to-left shunt and results in subjective breathlessness chiefly caused by the response of the respiratory center to the sudden changes in blood gas...
composition and pH, not by heart failure. After a right ventriculotomy, exercise may induce ventricular arrhythmias.\(^3^9\) The Fontan operation results in a circulation in series without a subpulmonary ventricle; exercise performance may improve significantly but cardiac indexes in response to isotonic exercise seldom increase more than twofold.\(^3^0,4^1\)

**Insurability, Employability, and Psychosocial Considerations**

As newer and more successful methods of treatment are applied and as the long-term benefits of these therapeutic advances are realized, patients are likely to enjoy higher access to insurance.\(^1,2,4^2,4^3\) Most young adults who have undergone successful repair of congenital heart lesions are eligible for some form of life insurance but health insurance is more problematic. It has been roughly estimated that in the United States, one third of adults with congenital heart disease have inadequate or no health insurance even though they may be physically capable of working.\(^2\) Constraints imposed by fee-for-service insurance plans (no coverage for preexisting illness) make these plans unattractive, especially for patients who anticipate further surgery or cardiac catheterization or for those who require frequent ambulatory evaluations or diagnostic testing. The future of health maintenance organizations and independent practice associations is uncertain, but there is a trend toward these prepaid medical care providers. The current heightened interest and concern in the funding of health care in the United States\(^4^4\) would do well to include congenital heart disease in adults, a patient population that not only is increasing year by year but also generally has the educational achievements and physical capabilities for productive employment.\(^2\) Innovations such as statewide insurance pools, employer-based health care systems, and national health insurance should be explored so that these deserving and productive members of the community can obtain adequate medical care.\(^2\)

There are advocacy groups for patients with cystic fibrosis and Down’s syndrome; why not for patients with congenital heart disease? If funding for the care and study of adults with congenital heart disease were secure, support for specialized care facilities would improve, more cardiologists would be interested, and a manpower shift might take place.

The opportunities for employment of adults with congenital cardiac defects are influenced by education, type of cardiac lesion, job discrimination, cardiac surgery, and often by the reluctance of an employer to accept the health insurance responsibilities involved in hiring.\(^1,2,4^3\) In certain occupations, the safety of others is in the hands of a single individual (bus drivers and airline pilots, for example). Job discrimination is one of the most important factors affecting employment opportunities for these patients. The smaller the company to which the application is made, the greater is the reluctance of employers to hire someone with a thoracotomy scar or a preexisting cardiac disorder. The National Rehabilitation Act of 1973 prevents job discrimination against the handicapped by almost all employers with 10 or more employees. The Vocational Rehabilitation Act of 1920, strengthened by amendments, offers a wide range of services but these services are significantly underused, especially by cardiac patients.

There are special, if not unique, psychosocial concerns in patients who have experienced dramatic and sometimes traumatic diagnostic and therapeutic interventions during key developmental phases of their lives.\(^1,2,4^5-4^7\) Nevertheless, most patients with congenital heart disease function psychologically within normal range, although low self-esteem, insecurity, and feelings of vulnerability are sometimes matters of concern.\(^4^4\) Parental knowledge, understanding, and attitude largely determine the patients’ and the parents’ perceptions of the congenital heart disease and significantly affect psychological adjustments. The adult with congenital heart disease faces tangible problems in the work force (see above), in dating, marriage, and parenthood. Cyanosis is believed to impair intellectual function, although the degree of impairment is generally mild and may be overestimated by IQ tests that depend on gross motor function at a young age.\(^4^7\)

**Surgical Considerations**

Operation or reoperation in adults with congenital heart disease often involves special surgical concerns peculiar to older patients.\(^1,2\) These concerns must take into account the congenital cardiac malformation per se (previously operated or unoperated) together with acquired cardiac and noncardiac diseases of adulthood. In cyanotic adults confronting their initial operation, hematologic disorders (see above) and aortopulmonary collaterals are matters of concern. In adults who have undergone reparative surgery, the chief concerns at the time of reoperation are prosthetic materials such as conduits and valves. Concerns that apply to both unoperated patients and patients undergoing reoperation include pulmonary vascular disease, ventricular function, myocardial protection (cardioplegia), blood salvage techniques, the risk of infective endocarditis, the residua and sequelae of previous cardiac surgery, and coexisting acquired heart disease, the incidence of which varies with age.

Interventional cardiac catheterization is now the preferred primary treatment or an adjunct to the surgical treatment of increasing numbers of pediatric and adult patients with congenital malformations of the heart and circulation.\(^1,2,4^8,4^9\) Corrective or reparative interventional catheterization procedures address pulmonary valve stenosis, recoarctation of the aorta, patent ductus arteriosus, and in selected patients, ostium secundum atrial septal defect. Palliative interventions can be either in lieu of surgery or as adjuncts to surgery. Therapeutic cardiac catheterization, like cardiac surgery, has three principle objectives: preservation of or improvement in cardiac
function, an increase in longevity, and maintenance of or improvement in the quality of life.

When adults with congenital heart disease require noncardiac surgery, perioperative risks can be reduced, often appreciably, when problems inherent in this patient population are anticipated.\textsuperscript{1,2} Uncomplicated situs inversus with dextrocardia may go unrecognized until an illness requiring surgery brings the patient to attention. Symptoms are likely to be misconstrued and diagnostic conclusions incorrect unless the mirror-image visceral positions are known. In acute appendicitis, the abdominal pain is in the left lower quadrant, and the pain of biliary colic is in the left upper quadrant. Noncardiac surgery in an adult with a functionally normal or mildly stenotic or incompetent bicuspid aortic valve imposes a risk that is essentially that of infective endocarditis. Fibrocalcific obstruction of a congenital bicuspid aortic valve accounts for approximately one half of the cases of surgically important pure aortic stenosis in adults.\textsuperscript{50} The immediate risks of noncardiac surgery are determined by the degree of obstruction, the functional adequacy of the after-loaded left ventricle, and the presence and degree of acquired coronary artery disease. Patients with hemodynamically significant congenital bicuspid aortic regurgitation confront cardiac surgery with risks determined by left ventricular function and susceptibility to infective endocarditis. In patients with primary pulmonary hypertension, even minor noncardiac surgery incurs risks that cause trepidation.\textsuperscript{1,2} The stress of a surgical procedure can trigger circulatory demands that are not adequately met. The corollary to these remarks is that even minor elective surgery in patients with primary pulmonary hypertension is, with few exceptions, to be avoided. Ostium secundum atrial septal defect in asymptomatic young adults with normal pulmonary arterial pressure imposes comparatively little risk during noncardiac surgery but with two qualifications. In response to hemorrhage, systemic resistance rises and venous return diminishes, a combination that sometimes appreciably augments the left-to-right intraventricular shunt. The second concern is the risk of paradoxical emboli from leg veins because thrombi carried by the inferior vena cava tend to stream across the atrial septal defect into the systemic circulation.\textsuperscript{1,3,2} In cyanotic adults with congenital heart disease, the incidence of cholecystitis (calcium bilirubinate stones) is increased. Biliary colic sometimes becomes manifest years after intracardiac surgery has eliminated the cyanosis. Hemostasis in cyanotic adults can be improved by preoperative phlebotomy (see earlier discussion) with the phlebotomized units stored for potential autologous transfusion. Intravenous lines, infusions, and drugs in cyanotic patients must be managed with special care. An air filter should be inserted into the intravenous line. In the adult with uncorrected Fallot's tetralogy who requires noncardiac surgery, meticulous intraoperative and postoperative monitoring of blood pressure is important because a sudden fall in systemic resis-

\textbf{Table 5. Residua After Reparative Surgery for Congenital Heart Disease}

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<tr>
<th>Category</th>
<th>Details</th>
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<tbody>
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<td>Valvular</td>
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<tr>
<td>Ventricular</td>
<td>Chamber morphology, Chamber mass, Chamber function, Myocardial connective tissue</td>
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<td>Vascular</td>
<td>Anatomic (morphological) vascular anomalies or defects, Elevated resistance and/or pressures; systemic, pulmonary</td>
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<tr>
<td>Noncardiovascular</td>
<td>Developmental abnormalities, Somatic defects, Medical disorders</td>
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</table>

\textbf{Postoperative Residua and Sequelae}

Residua after reparative surgery for congenital heart disease (Table 5) are represented by cardiac, vascular, or noncardiovascular abnormalities that are intentionally left behind at the time of reparative surgery.\textsuperscript{1,2,11} With few exceptions, these residua are obligatory, that is, they do not result from surgery having fallen short of its goal, at least in a technical sense.\textsuperscript{51} By contrast, sequelae are alterations or disorders that are intentionally incurred, occasionally or invariably, at the time of reparative surgery and are looked upon as necessary and acceptable consequences of operation (Table 6). Complications are unintentional aftermaths of reparative surgery that range in severity from inconsequential to fatal. The focus here is not on complications, although complications and sequelae may imperceptibly merge.
TABLE 6. Sequelae of Reparative Surgery for Congenital Heart Disease

<table>
<thead>
<tr>
<th>Category</th>
<th>Example</th>
</tr>
</thead>
<tbody>
<tr>
<td>Electrophysiological</td>
<td>Atriotomy</td>
</tr>
<tr>
<td></td>
<td>Intra-atrial repair</td>
</tr>
<tr>
<td></td>
<td>Intraventricular repair</td>
</tr>
<tr>
<td></td>
<td>Ventriculotomy</td>
</tr>
<tr>
<td></td>
<td>Incision site</td>
</tr>
<tr>
<td></td>
<td>Intracardiac repair</td>
</tr>
<tr>
<td>Native valves</td>
<td>Left ventricular or right ventricular outflow repair</td>
</tr>
<tr>
<td></td>
<td>Left ventricular or right ventricular inflow repair</td>
</tr>
<tr>
<td>Prosthetic materials</td>
<td>Patches</td>
</tr>
<tr>
<td></td>
<td>Valves</td>
</tr>
<tr>
<td></td>
<td>Conduits</td>
</tr>
<tr>
<td>Myocardial and endocardial sequela</td>
<td></td>
</tr>
</tbody>
</table>

Electrophysiological residua are represented by abnormalities that are, with some exceptions, inherent components of certain congenital cardiac malformations. These residua persist—sometimes harmlessly, sometimes not so harmlessly—after reparative surgery. Residual abnormalities of cardiac valves after reparative surgery fall into three general categories: 1) congenitally malformed cardiac valves that are functionally normal and do not require attention during reparative surgery, 2) intrinsically normal cardiac valves that are rendered incompetent because of the physiological stress imposed by the congenital malformation that prompted surgical repair, and 3) residually incompetent or stenotic congenitally malformed cardiac valves that do not lend themselves to complete repair. Residual ventricular abnormalities after reparative surgery are represented by disorders that are permanent, such as the intrinsic morphology of the ventricle, or disorders that change with the passage of time, such as alterations in chamber mass and function. The development of increased ventricular mass and its regression after reparative surgery are important properties of the myocardium. Vascular residua after primary repair of congenital cardiac malformations consist of anatomic anomalies or defects or elevated resistance and/or pressure in the systemic or pulmonary circulation. A more than casual relation exists between aortic root disease and bicuspid aortic valves, and on rare occasions the natural history or postoperative history of bicuspid aortic stenosis is dramatically punctuated by dissecting aneurysm. The younger the patient at the time of successful coarctation repair, the more likely is the long-term normalization of postoperative systemic blood pressure. The preoperative status of the pulmonary vascular bed, especially the resistance vessels, is a major determinant of the presence and degree of residual postoperative pulmonary vascular disease. Early operation sets the stage for normal development of the pulmonary vascular bed and reduces the probability that preoperative alterations will be translated into the undesirable postoperative residua of increased muscularity of small pulmonary arteries, intimal hyperplasia, and a reduction in the number of intra-acinar vessels. Noncardiac residua can be important long-term concerns after reparative surgery; these include developmental abnormalities such as mental deficiency, residual somatic defects such as dysmorphism or limb abnormalities, preoperative medical or psychosocial disorders, and healed brain abscess that can serve as a focus of a seizure disorder. Cataracts and deafness persist as residua after division of a patent ductus in children with the rubella syndrome.

Sequelae of reparative surgery for congenital heart disease involve electrophysiological mechanisms, native cardiac valves, prosthetic materials, myocardium, and endocardium (Table 6). Electrophysiological sequelae follow intra-atrial repairs, and intraventricular repairs via a right atrial incision or through a right ventriculotomy. Sequelae involving native cardiac valves occur after left ventricular or right ventricular outflow repairs or left ventricular or right ventricular inflow repairs. Postoperative or post-balloon angioplasty aortic regurgitation as a sequel of repair of congenital bicuspid aortic stenosis is a case in point. Reconstruction of the right ventricular inflow valve (in Ebstein’s anomaly, for example) is necessarily followed by sequelae that are intrinsic to the basic congenital tricuspid malformation, even if competence is established. Prosthetic materials represent a special category of sequelae after reparative surgery for congenital heart disease. Patches are often devoid of concern but prosthetic valves and conduits result in sequelae that vary in significance according to the physical and hemodynamic characteristics of the materials. There is a fine line between sequelae and complications.

Conclusion

Congenital heart disease in adults has emerged as a special area of cardiovascular interest, a new subspecialty. The accrued benefits have been gratifying but uninterrupted, long-term care is essential if the concerns inherent in this increasing patient population are to be addressed. As George P. Elliott wrote in the American Scholar, “In science generally to solve one set of problems may be to create or discover a whole new set, and of no science . . . is this more true than of medicine.”

References

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