

MEDICAL CENTER EXPERIENCES

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Background

Specialized 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 have evolved into the Congenital Cardiac Centre under the direction of Drs. Gary D. Webb, Peter McLaughlin and Peter Liu. An analogous facility was developed at the National Heart Hospital, London in 1975 under the auspices of Dr. Jane Somerville. What evolved into the University of California, Los Angeles (UCLA) Adult Congenital Heart Disease Center began in 1978 and is currently under the direction of Drs. Joseph K. Perloff, John S. Child and Samuel Kaplan. Development of the University of Iowa Adolescent/Adult Congenital Heart Disease Clinic in 1981 profited from Dr. David J. Skorton's experience at UCLA. Development of an analogous facility at the Mayo Clinic in 1988 profited from Dr. Carole A. Warnes' experience at the National Heart Hospital, London. The driving force behind the development of the facilities in Canada, Great Britain and the United States was the expanding number of adults with congenital heart disease. Advances in diagnostic methods and in the surgical and medical care of infants and children with congenital heart disease have profoundly affected survival patterns. A therapeutic era that brought together cardiologists and cardiac surgeons proved to be one of the most successful rehabilitation programs that medicine has witnessed. The heavy investments in time and resources that were needed to accomplish these results in the young patient must be matched with a similar dedication to the adolescent, postadolescent and adult with congenital heart disease. In the United States alone, there are currently well over one-half million patients with congenital heart disease who have reached adulthood and each year 20,000 open heart operations (an approximate minimum) are performed for congenital malformations of the heart and circulation (personal communication, Dr. Peter Einstein, Assistant Professor of Surgery and Pediatrics, University of Tennessee, Memphis). In 1989, there were 884 board-certified pediatric cardiologists in the United States, too few to cope with the

patient load of adults with congenital heart disease, even if all 884 were committed to do so. In the same year, there were 12,119 board-certified medical cardiologists. It is likely that only a small minority have knowledge of, interest in or responsibility for adults with congenital heart disease. Congenital heart disease in adults is still a largely unrecognized subspecialty, but is emerging as a discipline that requires special expertise. The collaboration of pediatric cardiologists, medical cardiologists, cardiac surgeons and hospital administrators is central to the success of this development. A first necessity is to recognize the seminal role and prerogatives of pediatric cardiologists and pediatric cardiac surgeons, without whose efforts congenital heart disease in adults would not be a present matter of pride and interest.

The number of patients registered in The Toronto Congenital Cardiac Centre for adults is nearly 1,600; in the "Grown-Up" Congenital Heart Unit of the Royal Brompton and National Heart Hospital, London, 3,000; in the UCLA Adult Congenital Heart Disease Center, 700; in the University of Iowa Adolescent/Adult Congenital Heart Disease Clinic, 600; and in the Mayo Clinic Adult Congenital Heart Disease Facility, nearly 300 patients. Accordingly, >6,000 adults with congenital heart disease are currently registered in five specialized centers in Canada, Great Britain and the United States. Approximately 50% of these patients have had cardiac surgery.

Characteristics of the Special Care Facilities for Adults With Congenital Heart Disease

Staffing and interdisciplinary approach. The special care facilities necessarily differ among the five centers, but the differences are more matters of detail than of principle. A

The term "medical cardiologists" is used here and throughout rather than the more conventional term "adult" cardiologist to avoid the implication that it is the cardiologist who is the adult. In Departments of Pediatrics, there are pediatric cardiologists. In Departments of Medicine, there are medical cardiologists. Cardiac surgeons, including pediatric cardiac surgeons, are clearly so designated.

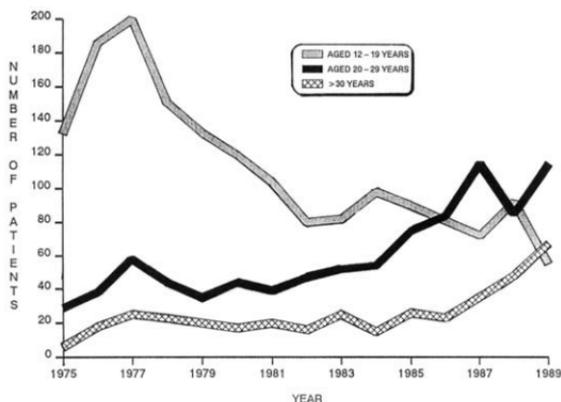


Figure 1. Admissions of patients with congenital heart disease to the National Heart Hospital, London, 1975 to 1989.

sense of direction can therefore be extracted. There is a consensus that patients are best managed, at least in the foreseeable future, by close interdisciplinary collaboration between pediatric and medical cardiologists. The collaborating cardiac surgeon must be equipped to deal with both congenital and acquired cardiovascular diseases and with problems that are special and often unique to these patients. Surgeons who deal largely if not exclusively with acquired heart disease are not likely to be comfortable with this task. Pediatric cardiac surgeons who have knowledge of and technical skills in congenital heart diseases more readily apply those skills to coexisting acquired cardiac diseases and obviate the need for operation by two cardiac surgeons. The fourth staff person of importance is the cardiovascular nurse specialist. It is necessary to underscore the importance of this assignment.

Each of the five centers has formally incorporated into its facility consultants who have gained experience in the specific problems associated with congenital heart disease in adults. The types and numbers of these consultants differ among the centers, but in general include electrophysiologists, high risk pregnancy obstetricians, gynecologists, hematologists, nephrologists, rheumatologists, pulmonologists, psychiatrists, social service personnel (insurance, vocational counseling), cardiac anesthesiologists and cardiac pathologists. It is desirable to anticipate consultative needs rather than seek ad hoc opinions. Some of the centers have support groups under the auspices of the consulting psychiatrist, social services and clinical nurse specialist.

The nonphysician staff plays a significant role, a point emphasized by each center. Secretarial staff members work in administration, coordination, appointments and patient affairs. In addition to the clinical nurse specialist, it is necessary to have outpatient nurses, preferably the same

personnel, assigned to each clinic. The University of Iowa clinic includes two physician assistants in addition to the nurse practitioner.

Criteria for entry into the specialized facilities. Criteria are based on age and appropriate psychologic and physical maturity. Three of the centers accept only postadolescents with an age at entry ≥ 17 or 18 years; the other two centers (Royal Brompton and National Heart Hospital and University of Iowa) accept adolescents with an age at entry of 12 years (Fig. 1). Adolescent medicine is a section of pediatrics that serves as a useful bridge between the preadolescent patient and the adult. All of the centers, by definition, include patients with congenital heart disease, but four (The Toronto Hospital, University of Iowa, The National Heart Hospital and UCLA) include patients with the Marfan syndrome.

Referral patterns and stratification of patient care. These are major issues. Broadly speaking, the base of the pyramid is the primary care physician in the community (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 facilities represented by the five centers contributing to this document. Regionalization to specialized centers assures adequate experience even with rare complex lesions. It is not yet clear how large a population is needed to serve a tertiary care center, but some information can be derived from experiences in Canada, Great Britain and the United States. Ontario is unique in North America in having centralized the care of infants and children with congenital heart disease (Hospital for Sick Children). Policy permits patient care up to the 18th birthday, after which responsibility must be delegated to The Toronto Hospital Congenital Cardiology Centre for adults or

to community hospital physicians. The "Grown-Up" Congenital Heart Disease Unit of the Royal Brompton and National Heart Hospitals in London has an extensive follow-up system. The Hospital for Sick Children in London is a particular source of referrals, but patients are referred from throughout Great Britain. The Adolescent/Adult Congenital Heart Disease Clinic of the University of Iowa is situated in a largely rural state with a population of approximately 3 million. The University of Iowa is the only medical school in the state and the Medical Center is the largest university-owned health center in the United States. At the Mayo Clinic, surgery for congenital heart disease has been performed since 1955. By the early 1970s, approximately 10,000 open heart operations had been performed, most of which were for congenital heart disease. The UCLA Hospital is in a large, medically competitive population area, but referrals of adults with congenital heart disease accelerated as soon as proper referral facilities were developed. It has been gratifying to witness the expansion of referrals from outside of the immediate geographic area to include the state of California, the United States and abroad. There was a consensus among the centers that an important stimulus is from the patients themselves, once they learn that such referral options are available to them. The tertiary referral centers do not compete with practicing physicians or community hospitals, but instead offer services difficult if not impossible to duplicate. The community rapidly becomes aware of the needed resource.

Funding for special care facilities. A concern frequently expressed was how the special care facilities are to be funded, especially how patient care should be supported. The essence of the concern, at least in the United States, is the problem of obtaining health insurance for adult patients with congenital heart disease (see Insurability and Employability, by Drs. Mahoney and Skorton elsewhere in this conference report). Approximately one-third of patients do not have private insurance, federal, state or county insurance or insurance through health maintenance organizations or analogous carriers. Health care delivery systems in Canada and Great Britain differ from those in the United States, but even so there was a strong feeling that health care planners and finance officers in Canada and Great Britain have yet to understand the size and complexity of the services required for adults with congenital heart disease. The sentiment was expressed that if funding for the care and study of these patients were more secure and attractive and if these programs were recognized academically, more cardiologists would be interested in the problem and a manpower shift would take place. Attention was called to the success in securing health insurance for adults with *noncardiac* diseases that begin in childhood (namely, cystic fibrosis and diabetes). There are advocacy groups for patients with cystic fibrosis and Down's syndrome—why not for patients with congenital heart disease? Funding is also needed for trainees and faculty, especially junior faculty, who select

congenital heart disease in adults as an area of special interest.

Tertiary care facilities. Such facilities for these patients can be sufficiently attractive institutional resources that university hospitals are encouraged to confront and resolve the fiscal and administrative constraints. There is reason to believe that response to the resource grows quickly once the facility is developed. Institutional obstacles were matters of concern in all five adult congenital heart disease centers, though to varying degrees and with different emphasis. The first step, indeed the obligatory step, is a relation of mutual respect, trust and collaboration between pediatric and medical cardiologists, who form the core of the necessary interdisciplinary collaboration. The closer and more cordial the working relation, the greater the success of the venture. Overcoming potential and real bureaucratic barriers in developing a jointly administered adult congenital heart disease facility with the requisite number of consulting services is no mean feat, but experience at the five centers contributing to this report encourages the belief that bureaucratic obstacles can be overcome. Access to satisfactory outpatient facilities has not been a significant problem, but access to inpatient facilities may pose formidable institutional obstacles. Logistic problems are posed when medical and pediatric cardiologists do not share the same hospital facilities. In any case, admission privileges must be resolved. Hospital administrations should grant the necessary privileges to qualified pediatric cardiologists who wish to extend their expertise to adults with congenital heart disease.

Outpatients. With one exception, outpatients in the five centers were seen in clinics for adults. There was a feeling, but not a consensus, that adult care is best provided in an adult setting, whether outpatient or inpatient. For patients who come to the outpatient facility from long distances, advance planning of anticipated diagnostic studies is obligatory. The centers all accepted their educational mission, including the teaching of medical students, house officers (pediatric and medicine) and pediatric and medical cardiac fellows. The size of the clinic determines the amount of time available for sharing information between the staff and trainees. A common problem was the length of time a patient had to spend during a given visit in a teaching setting.

Inpatients. Admissions can be cumbersome unless appropriate admission privileges are secured, especially for the pediatric cardiologist with an interest in adults with congenital heart disease. It is undesirable, both in a practical sense and symbolically, for a talented pediatric cardiologist to be denied access to an adult inpatient facility without the permission of a physician on the medical staff. The inpatient problem is compounded when admission is for cardiac surgery. An alternative suggestion was to establish an adult inpatient unit in a children's hospital, but there was a general feeling that this was a less than attractive solution. Inpatients constitute an important part of the educational experience of housestaff and fellows. Admissions can be for cardiac or noncardiac surgery, labor and delivery, cardiac intensive

care (generally arrhythmias), heart failure, hematologic disorders, renal disorders, disorders of acute metabolism or unrelated medical illnesses. Personnel caring for inpatients must be aware of certain specific concerns, such as the need for filters in intravenous lines of cyanotic patients.

Medical records. There was a uniform consensus that records should serve multiple purposes, including providing practical information to the referring or primary care physician and acquainting community physicians about the referral facilities. The quality of the records should be sufficient to serve as research information. There was a tendency to recommend dual records (one set for the hospital files and one for the congenital heart disease center files). It was recommended that copies be given to patients who can then carry their own records when moving to another area. Patients have proved to be responsible emissaries.

Diagnostic studies. Noninvasive, exercise, catheterization and angiographic laboratories must provide the same high quality for adults with congenital heart disease as that provided by the pediatric laboratories for infants and children. The five centers confronted the problem by having the diagnostic study performed in a pediatric laboratory or in an adult laboratory with a medical and pediatric cardiologist collaborating or by training technologists and medical cardiologists in the requisite techniques. It goes without saying that quality should not be compromised for expediency or design. Nuclear magnetic resonance imaging can be performed in collaboration with a specialist in the division of cardiac radiology or by a medical or pediatric cardiologist with the necessary interests and expertise.

Education and training. Training and education extend to community physicians, medical students, housestaff and fellows, visiting physicians from other institutions, nurses (especially specialty nurses) and last, but with emphasis, the patients themselves. Tertiary care centers are responsible for informing practicing physicians in the community that adults with congenital heart disease require special expertise not usually available in local hospital (cardiac) facilities. Consultation and follow-up reports should take this obligation into account. Telephone calls, with few exceptions, are gratefully received by referring physicians. The level of awareness of primary care cardiologists in the community is important in channeling patients to tertiary care centers and in improving primary care.

Medical students and housestaff (medical and pediatric) benefit from outpatient rotations that are designed to acquaint them with congenital heart disease in adults and the educational process is also served by the inpatients. All five contributing centers accommodate visiting physicians from their own countries and abroad. Nurses, especially graduate cardiovascular nurse specialists, are easily and usefully accommodated in the educational and training process. There was considerable interest and discussion regarding the

training of medical and pediatric cardiology fellows, but less than a consensus on how best to proceed. Three categories of fellows were recognized: those who are not interested, those who are interested but not committed and those who are dedicated. The feeling was expressed that if the medical and pediatric specialty boards placed more emphasis on congenital heart disease in adults, the percent of fellows and training program directors interested in this field would increase significantly. 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. There was general agreement that for medical cardiac fellows, formal rotations (inpatient and outpatient), including a rotation in pediatric cardiology, should be part of the fellowship experience. These experiences are designed to inform the fellows about congenital heart disease in adults without the intention of developing expertise, but with the avowed hope that if the seeds are planted they may germinate. Unsettled is how best to provide formal training for fellows who wish to become experts. In an evolving field, how can we provide sufficient intensity of exposure? Academic cardiologists with expertise serve as models to attract fellows in following suit. Still another practical issue is whether the skills and expertise accrued are marketable. Can a major interest in congenital heart disease in adults serve as a focus for an academic or practice career? The problem may be resolved if and when congenital heart disease in adults is recognized as a legitimate area of special interest within cardiovascular medicine.

Research. All five contributing adult congenital heart disease centers reported a commitment to research prompted by a desire to address unresolved questions posed by this patient population. A rich harvest is in store through collaboration with colleagues in a number of other disciplines and a research base must be provided for the fellows interested in careers in academic medicine.

Conclusions

The last 3 decades have witnessed the maturity of diagnostic, surgical and medical skills in the management of congenital heart disease. Advances have been impressive. Survival patterns have been affected, often profoundly. A happy result is the adult with congenital heart disease, an expanding patient population that has emerged as a special area of cardiovascular interest to which the 22nd Bethesda Conference was devoted. Recognition of our responsibilities to these patients is essential if the concerns inherent in this postpediatric population are to be addressed. Unresolved problems abound, but there is an air of optimism.