Congenital Heart Disease in Adults

— Collaboration Between Pediatric and Medical Cardiologists —

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Congenital heart disease in adults has become a special area of cardiovascular interest, but remains a relatively unfamiliar discipline. Advances in diagnostic and surgical techniques have had a striking impact on longevity in infants and children with congenital heart disease. Nevertheless, since true cures are rare, long-term care is obligatory for most if not all patients. In parallel with diagnostic and surgical developments, advances in medical management have had a major impact on life span in both postoperative and unoperated patients. Who will assume responsibility for the long-term care of these adult patients? Relatively few medical cardiologists are equipped to do so and relatively few pediatric cardiologists are sufficiently well versed with the accrued problems of aging. Accordingly, the current approach involves collaboration between pediatric and medical cardiologists. The purpose of this report is to underscore the importance of this collaboration, and to focus on 2 particular issues — the management of cyanotic congenital heart disease and the management of congenital heart disease and pregnancy.

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CONGENITAL heart disease in adults has become a special area of cardiovascular interest, but remains a relatively unfamiliar discipline, especially in departments of medicine. Spectacular advances in diagnostic techniques and in the surgical and medical management of infants and children with congenital heart disease have had a major impact on survival, and this trend promises to continue¹. However, true cures are rare. Most if not all postoperative patients require long-term care because of minor to major residua and sequelae^{1–3}. Over 2 decades ago, Dr. Joseph K. Perloff published his first paper that focused on this subject, arguing that it was simply a matter of time before a population of adults with congenital heart

disease would emerge³. In 1993, the American College of Cardiology recognized the importance of this emerging area of special cardiovascular interest by supporting the 22nd Bethesda Conference², and by supporting an annual course on congenital heart disease in adults in the United States and in Canada. In 1993, the International Society for Adult Congenital Cardiac Disease (ISACCD) was established, with meetings held semiannually during the American College of Cardiology and American Heart Disease Annual Scientific Sessions. In this report, we give an overview of the system used for congenital heart disease in adults in North America, particularly at the Adult Congenital Heart Disease Center at UCLA, and focus on my (HD) experience as a fellow at the Adult Congenital Heart Disease Center at UCLA⁴.

Key words:

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The UCLA Adult Congenital Heart Disease Center

The Center is a combined resource of the Departments of Medicine, Pediatrics and Surgery, and enjoys access to the UCLA Hospital and the UCLA ambulatory care facilities! The Center operates a tertiary care facility.

Patients are referred upon reaching 18 years of age and upon achieving appropriate physical and psychological maturity. Patients include those who have never undergone cardiac surgery, those who have undergone reparative or palliative cardiac surgery, and those who are inoperable apart from lung, heart or heart/lung transplantation.

The Outpatient Clinic is an important aspect of the Center. The scheduled number of clinic follow-up visits and consultations at the weekly clinic are kept at a level that permits time for relatively intense, shared discussion of most if not all patients. A separate "overflow" clinic operates in parallel. Follow-up and consultation reports are dictated only by staff cardiologists because the reports are designed to provide a practical as well as educational experience for the referring physician, and to provide a reliable source of data for the UCLA Adult Congenital Heart Disease Registry. The Outpatient Clinic serves as a teaching facility for fellows, residents, medical students, visiting cardiologists and specialty nurses.

Inpatients are an important part of the educational experience of the staff members and cardiology fellows and include elective admissions for cardiac or noncardiac surgery, for labor and delivery, for medical illnesses, and to the Cardiac Intensive Care Unit, usually for electrophysiologic abnormalities. Hospitalized patients are the responsibility of one of the Adult Congenital Heart Disease Center staff cardiologists in collaboration with a cardiology fellow and the Clinical Nurse Specialist. Preoperative planning and postoperative care are collaborative efforts. Patients are discussed before their operation at biweekly conferences with the cardiac surgeons.

Medical Considerations

Two areas were particularly noteworthy in my (HD) experience as a fellow in the

UCLA Adult Congenital Heart Disease Center: the medical management of patients with cyanotic congenital heart disease^{1,5,6} and the management of pregnancy in congenital heart disease⁷⁻⁹

Cyanotic Congenital Heart Disease

The erythrocytosis of cyanotic congenital heart disease is classified as either "compensated" or "decompensated." Compensated erythrocytosis refers to patients who establish equilibrium hematocrit levels in an iron-replete state, and who experience absent, or mild hyperviscosity symptoms even at high hematocrit levels^{5,6}. Patients with decompensated erythrocytosis fail to establish equilibrium conditions and manifest unstable, rising hematocrit levels and recurrent moderate to severe hyperviscosity symptoms. These symptoms are categorized in detail in each patient, and include headache, faintness, dizziness, visual disturbances, fatigue, lassitude, myalgias, paresthesiae, depressed mentation, and a sense of distance or dissociation. Since studies in the UCLA Adult Congenital Heart Disease Center have not identified a risk of stroke due to cerebral arterial thrombotic occlusion, phlebotomy is confined to patients with decompensated erythrocytosis who manifest sufficiently intrusive hyperviscosity symptoms to warrant temporary relief^{5,6}. Phlebotomy is not recommended in patients with compensated erythrocytosis regardless of the hematocrit level, as long as symptoms attributable to hyperviscosity are no more than mild to moderate.

Pregnancy and Congenital Heart Disease

An important focus of this topic is the intricate interplay between maternal circulatory and respiratory physiology and maternal congenital heart disease, and the effects of this interplay on the fetus, which is exposed to risks that threaten its intrauterine viability and to risks that subsequently express themselves as developmental defects or transmitted congenital malformations of the heart or circulation⁷⁻⁹. There is a uniform consensus that a successful operation prior to gestation is important in reducing maternal risks of congenital heart disease. In women with functionally mild unoperated lesions and in those after successful cardiac surgery, the

management of labor and delivery is the same as for normal gravidas, except for the selective susceptibility to infective endocarditis. For gravidas with functionally important congenital cardiac disease—unoperated or operated—planned management of labor, delivery and the puerperium are crucial if risk is to be minimized.

Certain congenital cardiac diseases are of special interest because of their common occurrence and female prevalence.⁷⁻⁹ Ostium secundum atrial septal defect is important because these patients tend to reach reproductive age, and females predominate. Paradoxical embolization is a risk obviated by closure of the defect. There is an increase in the incidence of atrial tachyarrhythmias when an ostium secundum atrial septal defect is closed after childhood. In pregnant women with severe pulmonary valve stenosis or severe bicuspid aortic stenosis, balloon dilatation late in the third trimester can greatly reduce maternal risk. Fallot's tetralogy is the most common cyanotic malformation that permits unoperated survival into reproductive age, and about 50% of patients are female. A gestational fall in systemic vascular resistance together with augmented venous return to an obstructed right ventricle results in an increase in right-to-left shunt and a fall in systemic arterial oxygen saturation; these changes are harmful to the fetus. Repair of coarctation of the aorta is best accomplished by resection with end-to-end anastomosis, a technique that removes the vulnerable postcoarctation aortic segment. Balloon dilatation of coarctation is not recommended in females. The fetal risk of maternal oral anticoagulants has not been satisfactorily resolved. Accordingly, the use of anticoagulants should be minimized.

Collaboration Between Pediatric and Medical Cardiologists

In the medical system in our country, each department has developed independently. In improving the quality of medical and surgical care for infants, children, adolescents, and adult patients, we often confront new conditions which are difficult to manage using our current system. Even in a general hospital, each independent department of pediatrics, gynecology, psychiatry, and medi-

cine encounters adolescents and adults who have had a disease since their childhood and pregnant women with congenital diseases. Only a few pediatric cardiologists and medical cardiologists have any interest and sufficient knowledge of congenital heart disease in adults. Congenital heart disease in adults is a relatively novel and unrecognized subspecialty. The collaboration of pediatric cardiologists, medical cardiologists, cardiac surgeons, and other specialties is essential for the care of patients in this relatively unfamiliar area.

Conclusions

Congenital heart disease in adults has become a special area of cardiovascular interest, but remains relatively unfamiliar, especially among medical cardiologists. Collaboration between pediatric and medical cardiologists has contributed to the care of these patients, especially in the setting of a formalized facility dedicated to the care of adults with congenital heart disease.

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