

Congenital Heart Disease In Adults: A Historical Perspective

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Abstract

It is uncommon if not rare for a new area of specialized cardiovascular interest to appear on the scene, but such has been the case with congenital heart disease in adults. Until the turn of the 20th century, the Hospital for Sick Children London, the Childrens Hospital Philadelphia and the Boston Childrens Hospital were little more than dim lights of hope in the darkness of pediatric medicine. Congenital heart diseases were considered little more than hopeless futilities, and congenital heart disease in adults was not even a theoretical consideration.

Beginning in the mid 19th century, a series of events prefigured a brighter future. Within the span of four years--1842 to 1846--anesthesia was born in the United States and ranks among America's greatest contributions to medicine. In 1895, Konrad Roentgen discovered "a new kind of rays," and in 1937, Castellanos, Pereias and Garcia in Cuba, and Robb and Steinberg in New York independently developed angiocardiology. "The internal structure of the living heart had been revealed for the first time." Willem Einthoven of Leiden developed the string galvanometer and in 1903, constructed the first electrocardiograph. In 1929 at the Augusta-Viktoria Hospital in Eberswalde near Berlin, Werner Forsmann, only a year after finishing his medical studies, performed the first cardiac catheterization on a living human being---himself. The untiring work of Maude Abbott culminated in her 1936 Atlas of Congenital Heart Disease based on 1000 pathology specimens personally studied at the McGill Medical Museum. The seminal contributions of Robert Gross in Boston (ligation of a patent ductus arteriosus, 1939), Clarence Crafoord in Stockholm (resection of aortic coarctation, 1944), and Alfred Blalock and Helen Taussig in Baltimore (the legendary shunt operation for cyanotic congenital heart disease, 1944) soon followed, and the sense of despair that had surrounded

congenital malformations of the heart---those "hopeless futilities"---began to dissipate. When Helen Taussig began work on her monumental Congenital Malformations of the Heart, surgical treatment of congenital heart disease was not even a faint glimmer on the horizon, but when the book was published ten years later (1947), it included three surgical procedures---ligation of a patent ductus, resection of aortic coarctation, and the Blalock/Taussig shunt. In 1953, John H. Gibbon in Philadelphia performed the first successful operation employing a mechanical heart/lung bypass system for closure of an atrial septal defect in an 18 year old woman, and in 1955, John W. Kirklin at the Mayo Clinic reported eight cases of intracardiac surgery with the aid of a Gibbon type mechanical pump-oxygenator.

Medical ultrasound was introduced in the 1960's, and before long, cardiac ultrasonography was heralded as "the diagnostic pathway to the promised land." Echocardiography soon provided accurate, safe and painless diagnostic information from fetal life to the neonate, child, adolescent and adult with congenital heart disease.

Cardiac catheterization as a therapeutic intervention had its inception in 1966 with Rashkind's creation of an atrial septal defect without thoracotomy as a palliative approach to complete transposition of the great arteries.

Immense technical resources are now at our disposal, permitting remarkably precise anatomic and physiologic diagnoses and astonishing feats of palliative and reparative surgery. Survival patterns have been affected, often profoundly. Approximately 85% of infants with congenital heart disease in developed countries can expect to reach adulthood. In the United States, there are currently approximately 900,000 to a million adults with complex, moderately complex and simple congenital malformations of

the heart, and 20,000 open operations are performed annually. Although cardiac surgery and interventional catheterization have revolutionized survival patterns, cures in the literal sense are few. Postoperative residua and sequelae are the rule, and range in severity from inconsequential to serious. Importantly, even curative cardiac surgery does not preclude noncardiac residua, some of which have major impacts on morbidity and longevity.

The care of adults with congenital heart disease requires knowledge of the basic malformation per se, the type of surgical intervention employed, the postoperative cardiac and noncardiac residua and sequelae, and the acquired cardiac and noncardiac diseases that accrue with age. To address the needs of this complex and growing patient population, regional specialized comprehensive care facilities have evolved. These facilities do not compete with practicing physicians or community hospitals, but instead offer services difficult or impossible to duplicate.

The first publication to characterize the changing population of congenital heart disease was exactly 30 years ago--*Circulation* March 1973, the Pediatric Congenital Cardiac

becomes a Postoperative Adult. In October 1990, the 22nd Bethesda Conference, Congenital Heart Disease after Childhood, provided legitimacy for the new subspecialty, and the 32nd Bethesda Conference a decade later (October 2000), extended that legitimacy. And now, worldwide interest is reflected in the International Society for Adult Congenital Cardiac Disease.

Pediatric cardiology evolved as a specialty after the Second World War as advances in diagnostic methods and in the surgical management of congenital heart disease created an air of optimism in infants who had been considered hopeless futilities. The early years of this fledgling specialty were not easy because traditional biases weighed against innovation. The success of pediatric cardiologists and pediatric cardiac surgeons profoundly changed survival patterns, and as a result congenital heart disease in adults evolved as a specialty. The early years of this fledgling specialty were not easy because traditional biases weighed against innovation. But vigor can be derived from opposition, and remember---there is no avant garde, only those who have been left behind.

References

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