Congenital Heart Disease: A Clinical Challenge from Infancy through Adulthood

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Congenital heart (CH) disease is the presence of a gross structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional significance [Mitchell SC et al. *Circulation* 1971]. It remains a major contributor to infant morbidity and mortality worldwide, despite advances in medical and surgical treatment, and it was the second leading cause of death in children aged <5 years in Jamaica in 2003. Early intervention is critical, but even adults with CH disease who have been previously treated are at greater risk for premature mortality, complications, and repeat interventional procedures, as repair does not necessarily constitute a cure. The complex management of CH disease during childhood and throughout adulthood was the subject of several sessions at the Caribbean Cardiac Society Meeting in July 2010.

Bovette Butler, MD, Bustamante Hospital for Children, Kingston, Jamaica, presented findings from a Jamaican study of a neonate population that focused on identifying patterns within clinical presentations of CH disease and evaluating outcome determinants. This 5-year retrospective study evaluated infants (aged 0 to 28 days; n=327) who were diagnosed with congenital cardiac lesions at Bustamante Hospital for Children between July 2002 and June 2007. The lesions were classified into 6 groups, including left-to-right shunt (75%), right ventricular outflow tract obstruction (10%), common mixing (6%), complete transposition (4%), left ventricular outflow tract obstruction (4%), and miscellaneous (1%), according to the dominant lesion. The mean age at presentation was 10.6±8.1 days, with initial presentation occurring within the first week of life in 43% of patients (n=139).

Of the 327 neonates evaluated, 100 (31%) were symptomatic at presentation, most of whom presented with symptoms that were related to heart failure (67%). Twenty-eight percent was cyanotic at presentation, owing to tricuspid artresia (TA) and transposition of great arteries (TGAs), with a single ventricle occurring in 54% (n=15) of these cases. Asymptomatic presentation occurred in 123 patients, 93 of whom had evidence of cardiac murmur (76%).

Although lesions that resulted in left-to-right shunting were the most common, lesions that resulted in outflow tract obstruction and complete transposition caused the highest rate of mortality (Table 1). Thirty-six neonates died of CH disease-related causes, and 69% of those who died presented within the first 7 days of life (p=0.001). Symptomatic presentation was associated with a higher mortality rate than asymptomatic presentation (p<0.001).

Table 1. Mortality Rate Associated with Diagnosis.

Diagnosis	Lesion Classification	Number (%) of Deaths
Coarctation of the Aorta (n=4)	Left ventricular outflow tract obstruction	3 (75%)
Hypoplastic Left Heart Syndrome (n=7)	Left ventricular outflow tract obstruction	5 (71%)
Transposition of Great Arteries (n=10)	Complete transposition	5 (50%)
Pulmonary Atresia (n=3)	Right ventricular outflow tract obstruction	2 (67%)

Highlights from the

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CH disease is a serious illness that is associated with a high rate of infant mortality. This study by Butler and colleagues demonstrates the severity of the disease and identifies lesions that are associated with the highest frequency and mortality rates. It is important to note that this analysis may have regional limitations due to the structure of the health care and referral system, as well as the type of CH disease that is found in that area.

Richard Perryman, MD, Memorial Healthcare System, Hollywood, Florida, USA, discussed CH disease in the context of adulthood. Due in part to steadily improving pediatric outcomes, there has been a marked increase in adult congenital heart disease (ACHD) in the United States since 1998, indicating a pressing need for specialized care centers to manage the broad spectrum of clinical complications that are associated with ACHD.

Dr. Perryman emphasized the clinical advantages of regional ACHD centers and what these centers should entail. ACHD centers should offer continuous access to treatment, with 24/7 availability to ensure consistency of care. Facilities should also include specialized services, such as clinicians with expertise in the area of congenital cardiology and advanced diagnostic imaging modalities. Multidisciplinary teams (ie, neurology, nephrology, cardiac pathology, social services, rehabilitation services, high-risk obstetrics, transplant specialists, and geneticists) may help reduce disease burden and provide more sustainable treatment solutions. The concept of the ACHD center may also resolve the issue of optimal care for adults who suffer with a pediatric-based affliction.

There has been some uncertainty as to the best facility and specialist for adults with CH disease (ie, adult-versus-pediatric facility and adult-versus-pediatric surgeon) due to the disease etiology and complexity of this population. In a study by Kogan and colleagues, between 2000 and 2007, 303 surgical procedures for ACHD were evaluated. Of the 303 procedures, 185 were performed in an adult hospital (versus 118 in a pediatric hospital), and 46 procedures were performed by an adult cardiac surgeon (versus 257 by a pediatric cardiac surgeon). A multivariate analysis revealed that surgery that was performed by an adult cardiac surgeon (p=0.0004), surgery that was performed at a children's hospital (p=0.013), and older age at the time of surgery (p=0.028) were significant risk factors for 30-day mortality. The best surgical outcomes for ACHD patients were associated with procedures that were performed by a congenital heart surgeon in an adult hospital setting [Kogan BE et al. *Ann Thorac Surg* 2009]. These data support the idea that comprehensive care in a multidisciplinary setting, such as that found in an ACHD center, would improve the management of ACHD and foster risk reduction and optimal therapeutic approaches.

Regional programs and early detection and intervention are crucial to improving the morbidity and mortality rates of children and adults with CH disease. Despite advances in CH disease treatment, the disease burden remains high in both developed and developing countries. The management of CH disease relies on specialized care and organized regional programs to broaden the scope of treatment for this clinically complex population.