Mechanism of Diagnosis and Outcome in Critical Congenital Heart Disease

Block, Joseph R.; and Berger, Stuart

Children's Hospital of Wisconsin-Herma Heart Center, Medical College of Wisconsin, Milwaukee, WI

Purpose: Critical Congenital Heart Disease (CCHD) is defined as a congenital lesion in which treatment is required within the first month of life. Outcomes in CCHD may be compromised if the diagnosis is unrecognized prior to onset of symptoms. We evaluated the rate and effect of prenatal diagnosis of CCHD on morbidity and mortality on the patients undergoing surgery for one of twelve lesions at the Children's Hospital of Wisconsin (CHW) or American Family Children's Hospital (AFCH) from 2007 to 2012. Those twelve Lesions {Hypoplastic Left Heart Syndrome (HLHS), Pulmonary Atresia (PA), Tetralogy of Fallot (TOF), Total Anomalous Pulmonary Venous Return (TAPVR), Transposition of the Great Arteries (TGA), Tricuspid Atresia (TA), Truncus Arteriosus (Truncus), Coarctation of the Aorta (COA), Double Outlet Right Ventricle (DORV), Ebstein's Anomaly (EA), Single Ventricle (SV), and Interrupted Aortic Arch (IAA)} which have been identified as those potentially detected by routine pulse oximetry screening were assessed.

Methods: A retrospective chart review was performed on Wisconsin-born patients with an operation for one of these twelve CCHDs at CHW or AFCH. Prenatal vs. postnatal diagnosis, date of birth, surgery, death, diagnosis, presence of a genetic syndrome, length of intubation, birth weight, gender, Aristotle score, and length of stay were determined.

Results: 658 patients were identified and with the following distribution by diagnoses: HLHS 93 patients (14%), PA 51 (8%), TOF 124 (19%), TAPVR 32 (5%), TGA 88 (13%), TA 25 (4%), COA 88 (13%), DORV 57 (9%), EA 5 (1%), IAA 17 (3%), SV 50 (8%). Of our cohort there was a prenatal diagnosis made in 343 patients (52%) and the prenatal detection rate increased from 32% to 66% from 2006 to 2012 ($p = 0.0001$). The hospital mortality rate was 5%. Prenatally diagnosed patients had a higher Aristotle mortality score ($p= 0.0052$), longer hospital stay ($p< 0.001$), lower birth weight ($p<0.001$), presence of a genetic syndrome ($p= 0.0139$) and longer intubation ($p<0.001$). 34/47 patients who died and 12/17 who went on to transplant had a prenatal diagnosis.

Conclusion: The prenatally diagnosed cohort had a higher Aristotle mortality score, longer intubation time, longer length of stay and mortality, suggesting that the most complex forms of CCHD are being discovered prenatally. As this study was limited to those patients surviving to reach CHW and AFCH, deaths due to a missed diagnosis of CCHD cannot be fully assessed yet.