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Outcome after Fetal Diagnosis of Hypoplastic Left Heart Syndrome: The Toronto Experience of 267 Patients

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Background. Hypoplastic left heart syndrome (HLHS) is characterized by variable degrees of underdevelopment of the left heart, such that is unable to support the systemic circulation after birth. Available management options offered to parents at our center include termination of pregnancy (TOP), compassionate care at birth (CC), and active management either with staged single ventricle surgery (including two stage 1 options: hybrid procedure vs. Norwood operation) or cardiac transplant. Studies on HLHS outcomes are usually focused on postnatal outcomes and suggested a rather wide range of outcomes among centers and sometimes among surgical options. The aim of this study was to review the overall outcome of a large cohort of HLHS babies from the time of fetal diagnosis to 1-year of life.

Methods. 267 fetal cases diagnosed with HLHS (1990-2011) were identified in our cardiac database. Clinical/echocardiography data were correlated with outcomes of actively managed babies.

Result: Mean age at referral was 23.7 ± 5.4 gestational weeks with mitral atresia/aortic atresia (MA/AA: 49%) being more common than mitral stenosis /aortic atresia (29%) and mitral stenosis/aortic stenosis (22%). Additional major extracardiac abnormalities were detected in 13% of cases, while genetic disorders affected 19% of 119 tested cases, with Turner syndrome as the most common anomaly (7%). 160/267 (60%) resulted in TOP or CC and 13 (4.9%) underwent spontaneous intrauterine demise (IUD). Of 94 live-births with intended active management, 75 (80%) survived to infancy and only 56 (60%) to 1 year. Risk factors for IUD were a highly restricted atrial septum ($p < 0.001$), extracardiac structural abnormalities ($p < 0.001$), at least moderate tricuspid regurgitation ($p = 0.002$) and pericardial effusion ($p < 0.001$). Postnatal mortality was significantly associated with MA/AA ($p = 0.05$) and a highly restrictive atrial septum with a need for urgent neonatal balloon atrioseptostomy ($p = 0.02$). Other variables including the choice of surgery and era of diagnosis did not significantly affect outcomes.

Conclusion: The overall survival to 1 year of life after a fetal diagnosis of HLHS was 21% in this study. Main reasons for the high attrition rate included frequent associations with genetic/extracardiac disorders, parental decision of TOP/CC, and surgery-related complications.