HYPOPLASTIC LEFT HEART SYNDROME WITH COMPLETE HEART BLOCK: CASE PRESENTATION AND LITERATURE REVIEW

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Objectives: To describe an unusual case of hypoplastic left heart syndrome (HLHS) associated with complete heart block diagnosed in-utero. The available medical literature of this extremely rare association including diagnosis, management, and short-term outcome is also provided.

Background: Congenital complete atrioventricular block (CCAVB) is a rare condition occurring in about 1 in 20,000 live births, whereas HLHS occurs 1 in 5,000 live births. HLHS associated with CCAVB is exceptionally rare.

Case Report: HLHS with mitral and aortic atresia and unrestrictive atrial level communication was diagnosed on 23-week gestation fetus of a 30-year-old G3P1 mother. At 35 week gestation, fetal cardiac monitoring demonstrated frequent premature atrial contractions (PACs), some of them were blocked, with an average fetal heart rate of 152 BPM. Subsequent weekly follow up until delivery continued to show frequent PACs, mainly in bigeminy and blocked beats. Average fetal heart rate was mostly in the 60s BPM. CCAVB was suspected. The fetus continued to do well with no evidence of fetal hydrops.

Postnatal transthoracic echocardiogram and 12 lead ECG confirmed prenatal diagnosis of HLHS and CCAVB. The first 4 days of life the infant was started on prostaglandin and was intubated for repeated apnea episodes. At 4 days of age a palliative hybrid approach was implemented. Through a small median sternotomy the pulmonary arterial branches were banded, a 9x19mm Genesis stent was deployed in the ductus arteriosus. Atrial septal defect was adequate and as such no intervention was needed to enlarge atrial septal communication. In addition an epicardial DDD pacemaker was placed. At 14 days of life an atrial septostomy was performed due to the increasing pressure gradient across the once non-restricted defect. Progressive worsening of right ventricular systolic function and desaturation developed in the subsequent weeks despite providing optimal cardiac support and lack of clear etiology for increasing ventricular contractility and increasing AV valve regurgitation. Inability to improve ventricular function and tricuspid regurgitation led to listing for cardiac transplantation.

Conclusions: HLHS and CCAVB is an extremely rare lesion with uniformly lethal outcome without intervention. CCAVB despite early detection and treatment may impose a deleterious effect on the single ventricle function leading to unfavorable outcome of HLHS patients. Even though cardiac transplantation is a final resort for failing single ventricle hearts it is most probably the only option available for these patients.