**FETAL DIAGNOSIS AND NEONATAL PALLIATION OF ABSENT AORTIC VALVE WITH HYPOPLASTIC LEFT HEART SYNDROME**

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**Introduction:** Congenital absence of the aortic valve leaflets is a rare association with hypoplastic left heart syndrome (HLHS). Most of the cases that were previously reported resulted in fetal or neonatal demise.

**Case:** A 37-year-old pregnant woman was referred for fetal evaluation of possible HLHS at 22 weeks gestation. The fetal ECHO was remarkable for a hypoplastic left atrium, a nearly atretic mitral valve, a small hypertrophic left ventricle with poor contractility, and a hypoplastic left ventricular outflow tract with severe aortic insufficiency. A female infant was born at term and the postnatal ECHO confirmed the above findings (figure). In addition, there was a complete absence of the aortic valve leaflets. The patient underwent Norwood procedure on day 5 of life with atrial septectomy, PDA ligation, over-sewing of the aortic valve, and a placement of a 4 mm Sano shunt between the right ventricle and the main pulmonary artery. She tolerated the procedure well and is currently maintaining her oxygen saturation in the mid-80s on room air. The plan is to complete her Fontan procedure in the future.

**Conclusion:** Prenatal diagnosis of absent aortic valve should be suspected in the presence of severe aortic insufficiency in the fetal ECHO. Early postnatal intervention is critical as those patients are likely to deteriorate quickly. The over-sewing of the aortic valve may be important to prevent coronary steal and myocardial hypoperfusion, which could potentially be detrimental.

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