Hypoplastic Left Heart Syndrome – A Rare Case Report

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Abstract: The term Hypoplastic left heart is used to describe a related group of anomalies that include under development of the left side of the heart (atresia of the aortic or mitral orifice) and hypoplasia of the ascending aorta. The left ventricle may be moderately hypoplastic, very small and non functional, or totally atretic, in the immediate neonatal period. The right ventricle maintain the both upper pulmonary circulation and the systemic circulation via the ductus arteriosus. Hypoplastic left heart syndrome (HLHS) has been report to occur in approximately 0.016 to 0.036% of all live births.. Here we present a rare case report- Hypoplastic left heart Syndrome.

Keywords: Hypoplastic left heart syndrome, Hypoplastic arch, Hypoplastic LV, Single-ventricle, Norwood procedure, Sano procedure

I. Introduction

Hypoplastic left heart syndrome (HLHS) refers to the abnormal development of the left-sided cardiac structures, resulting in obstruction to blood flow from the left ventricular outflow tract 1. In addition, the syndrome includes underdevelopment of the left ventricle, aorta, and aortic arch, as well as mitral atresia or stenosis. The Hypoplastic left heart syndrome may be a progressive lesion, beginning with simple valvar aortic stenosis in mid gestation period. The decrease flow through the stenotic aortic valve reduce the flow through the left ventricle during development, resulting in gradual ventricular chamber hypoplasia 2. HLHS is potentially detectable on prenatal sonography between 18 and 22 weeks’ gestation with a 4-chamber view of the fetal heart and it is one of the most common cardiac malformation detected in fetal life 3.

II. Case Report

Baby boy delivered normally at term weighing 2530 gms, length 49 cms, presented on 3rd day of life with respiratory distress. On Examination there was central cyanosis and pallor with cold extremities, weak peripheral pulses, poor perfusion in shock. Auscultation revealed loud S2. Due to progressive deterioration, mechanical ventilation was provided with inotropic support. However, baby could not be revived. On Ultrasonography 29 weeks single, live intrauterine pregnancy with dilated fetal aorta. Fetal echocardiography showed heart more in left hemithorax, two chambered heart – morphological RA and RV. LA severely hypoplastic to atretic and LV atretic. Large RA and large RV, single pulmonary vein draining into almost atretic LA. Large pulmonary trunk arising from the RV, dividing into RPA and LPA; ductus arch large and continuing into descending thoracic aorta. Ascending aortic atretic. Postnatal echocardiography done on 1 day of life which revealed Mitral atresia, aortic atresia, Rudimentary and Hypoplastic LV. Small, restrictive PFO measuring 3.2 mm, left to right shunt with severe PH. Hypoplastic arch, Pulmonary artery largely continuing as descending aorta – HLHS.
III. Discussion

Hypoplastic left heart syndrome with an intact atrial septum is a rare finding, reported in only 1% of pathologic specimens with hypoplasia of the aortic tract complex. With prenatal restriction, or complete premature closure of the foramen ovale (i.e., intact atrial septum), flow is diverted away from the left atrium and left ventricle. Once separation from placental circulation takes place at birth, pulmonary blood flow increases substantially, as does pulmonary venous return to the left atrium. If left ventricular hypoplasia and an intact atrial septum are present, effective egress from the left atrium is impossible, resulting in marked elevation of pulmonary venous pressure. The two surgical procedures most commonly utilized are the Norwood procedure or the Sano procedure; an alternative therapy is cardiac transplantation. The combination of single-ventricle physiology and impediment to pulmonary venous egress can result in postnatal hypoxemia to a degree which may be incompatible with life; similar to our case.
References