

# Emergent post-partum atrial septostomy for unanticipated PFO closure following in-utero balloon aortic valvuloplasty for HLHS

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**Introduction:** Hypoplastic Left Heart Syndrome (HLHS) is a constellation of clinical entities that arise from decreased blood flow through the left side of the heart early in fetal development resulting in an underdeveloped left ventricle and ascending aorta. In-utero intervention is aimed at promoting forward flow in an attempt to foster development of these left-sided structures. One of the key anatomical prerequisites for survival is adequate decompression of the pulmonary circulation across the atrial septum or via a decompressing vein from the Left Atrium (LA). Studies of the subset of HLHS patients who are born with an intact septum show extremely poor outcomes. We present the case of a male fetus who after undergoing in-utero aortic valve balloon dilation was born with the physiology of HLHS with an unexpected intact atrial septum. The following report and discussion describe this life threatening scenario.

**Case Report:** Severe congenital aortic valvar stenosis (peak gradient 23 mmHg) was diagnosed by fetal echocardiography at 21 weeks gestation. The 23 wk follow-up demonstrated worsening stenosis (peak gradient 46 mmHg), severe Left Ventricle (LV) dysfunction, and no antegrade flow in the ascending aorta. The parents were counseled regarding HLHS and elected to undergo the in-utero intervention of balloon aortic valvuloplasty. Aortic insufficiency was present after the procedure demonstrating effective valve dilation. The POD#1 echocardiograph demonstrated good forward flow across the Aortic Valve (AV) and antegrade flow around the aortic arch. By 28 weeks little antegrade flow across the AV was noted with once again retrograde flow into the ascending aorta. At this time there was adequate flow across the fossa ovalis. Surveillance was continued every 2 weeks with unchanged findings. After spontaneous labor at 38 weeks and unanticipated delivery at an outside hospital, the baby was hypoxic, hypotensive, and acidotic. Echocardiography demonstrated an intact atrial septum with a dilated LA, LV dilation with severely diminished function and moderate mitral regurgitation (MR). The baby was intubated, umbilical arterial and venous catheters were inserted and medical resuscitation started with alprostadil, dopamine, epinephrine, and dobutamine infusions. The baby was emergently transferred via helicopter and brought directly to the interventional cardiology suite for emergent atrial septostomy. The ABG on presentation to the cath lab was pH 7.223/PaCO<sub>2</sub> 60.1/PaO<sub>2</sub> 27.2/HCO<sub>3</sub><sup>-</sup> 24.2/BE -4.5/SaO<sub>2</sub> 39.3%. The anesthetic management included ongoing resuscitation with continued inotropic support, acid base correction with sodium bicarbonate, appropriate ventilation with associated muscle relaxation and blood transfusion to maintain Hemoglobin at around 15 gm/dl. TEE confirmed an intact atrial septum with LA dilation. The atrial septum was perforated with a radiofrequency ablation catheter with the initial LA pressure measuring 32 mmHg. After balloon dilation and deployment of an intra-atrial stent the LAP decreased to 8 mmHg with no residual LA to Right Atrium (RA) gradient. The dobutamine and epinephrine were weaned to off, and the dopamine was weaned from the presenting 10 µg/kg/min to 3 µg/kg/min. The final ABG prior to exiting the cath lab was pH 7.403/PaCO<sub>2</sub> 39.9/PaO<sub>2</sub> 42.8/HCO<sub>3</sub><sup>-</sup> 24.3/BE -0.3/SaO<sub>2</sub> 78.8%. The follow-up echocardiograph confirmed a well positioned stent across the upper part of the atrial septum with unobstructed left-to-right flow; mild MR/TR/AR/PR, and severe LV dysfunction/dilation.

**Discussion:** The in-utero diagnosis of HLHS affords the opportunity to perform an aortic valvuloplasty to promote forward blood flow to support the development of the left-sided heart structures including

the ascending aorta. The viability of HLHS patients ex-utero requires a pulmonary venous egress pathway (ASD or decompressing veins) to decompress the pulmonary circulation and provide oxygenated blood to the systemic circulation. In a subset of HLHS patients this pathway is absent and they require emergent creation of an intra-atrial communication. This patient is of interest because a fetal intervention attempted to promote development of the left-sided structures. When this failed, the more conventional treatment pathways were pursued given that a pulmonary venous egress pathway existed. At birth, the egress pathway unexpectedly closed necessitating an emergent intervention to create a life-saving intra-atrial communication.