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Prenatal Diagnosis of Hypoplastic Left Heart Syndrome with In-Utero Aortic Valvuloplasty: A Case Report with Novel Management Insights

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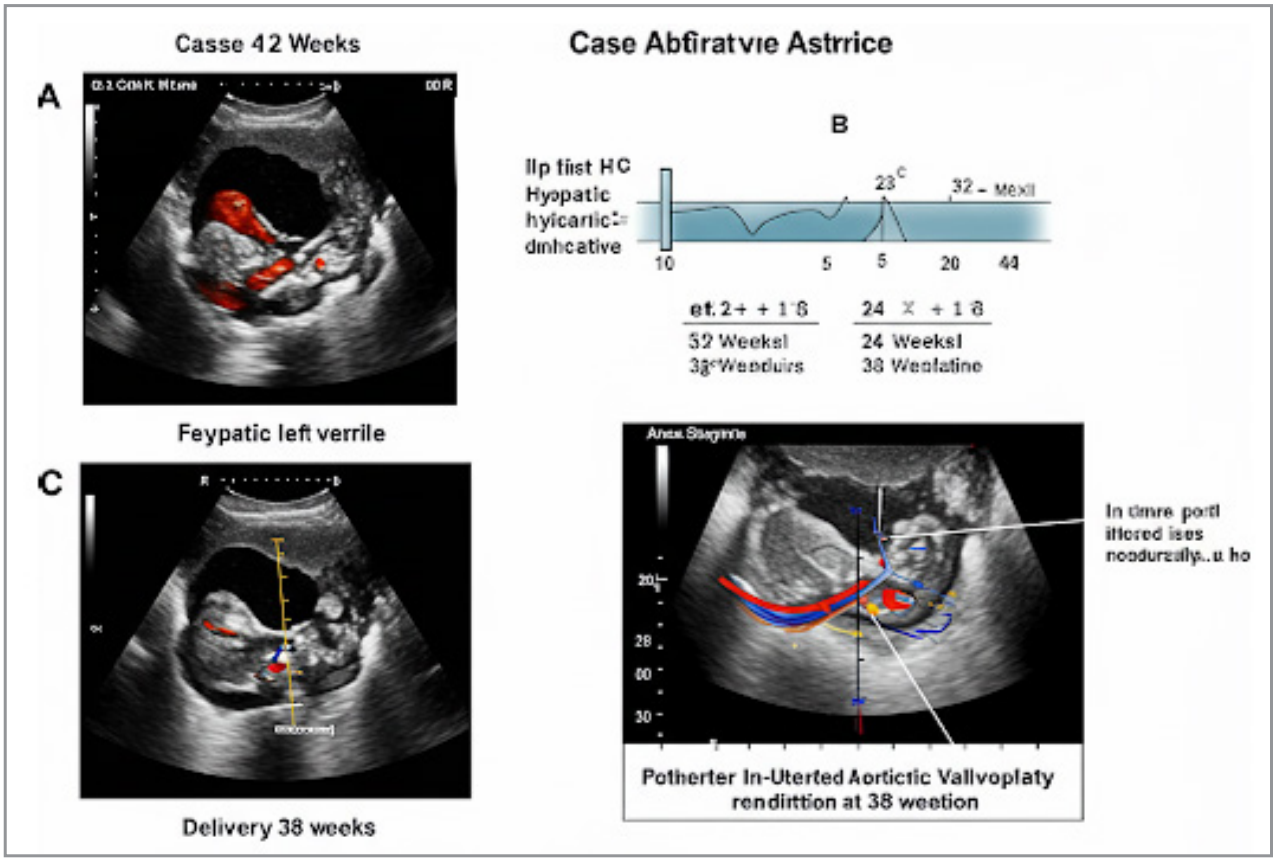
Introduction

Hypoplastic Left Heart Syndrome (HLHS) is a severe congenital heart defect characterized by underdevelopment of the left-sided heart structures, necessitating staged surgical interventions postnatally. Prenatal diagnosis via fetal echocardiography allows for early planning, but outcomes remain challenging due to the complexity of the condition. This case report describes a rare presentation of HLHS diagnosed prenatally, with a novel in-utero intervention—aortic valvuloplasty—performed to mitigate progression of left ventricular hypoplasia. We discuss the diagnostic approach, intervention, and outcomes in the context of European Society of Cardiology (ESC) guidelines, highlighting a potential new strategy for managing severe HLHS [1, 2].



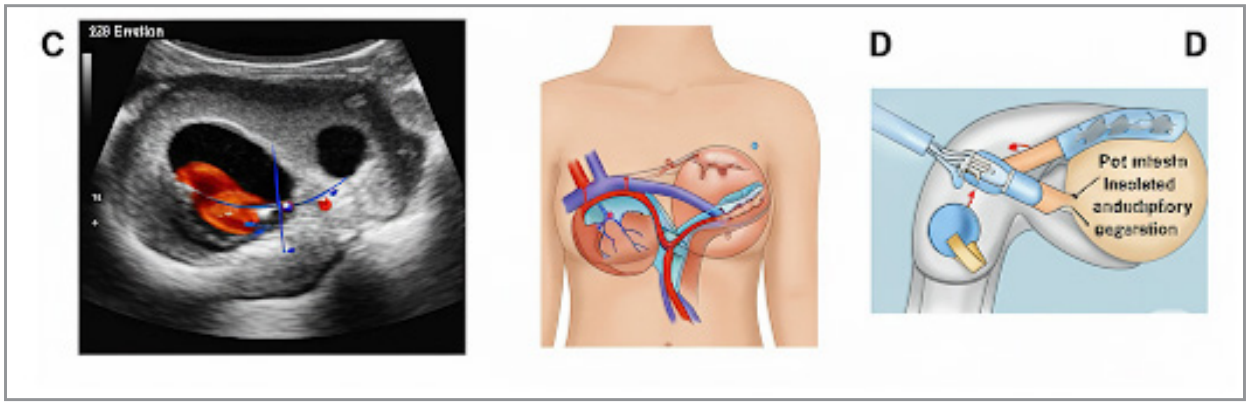


Figure 1: A graphical abstract illustrating the case timeline and key findings.

- **Panel A:** Fetal echocardiogram at 22 weeks showing a hypoplastic left ventricle (LV) and a small aortic root.
- **Panel B:** Timeline of events: diagnosis at 22 weeks, in-utero aortic valvuloplasty at 24 weeks, and delivery at 38 weeks.
- **Panel C:** Postnatal echocardiogram showing improved LV size and flow across the aortic valve.
- **Panel D:** Diagram of the in-utero procedure with a catheter inserted via the maternal abdomen to dilate the aortic valve.

Case Presentation

A 28-year-old primigravida woman was referred to our tertiary care center at 22 weeks gestation following an abnormal routine obstetric ultrasound. The patient had no significant medical history, and there was no family history of congenital heart disease. A fetal echocardiogram was performed using a Philips X5-1 transducer, revealing a 4-chamber view with a severely hypoplastic left ventricle (LV), a small aortic root (Z-score -3.5), and restricted flow across the aortic valve (peak velocity 1.2 m/s). The right ventricle (RV) was dominant, and the atrial septum showed a restrictive foramen ovale. The fetal heart rate was 74 bpm, which, while on the lower side of normal, was not concerning at this stage. These findings were consistent with Hypoplastic Left Heart Syndrome (HLHS) with critical aortic stenosis [3].

Given the severity of the left ventricular hypoplasia and the risk of progression to end-stage HLHS, the multidisciplinary team, including pediatric cardiologists, fetal medicine specialists, and cardiac surgeons, discussed the potential for an in-utero intervention. At 24 weeks gestation, an in-utero aortic valvuloplasty was performed under ultrasound guidance. A 22-gauge needle was inserted through the maternal abdomen, and a balloon catheter was advanced into the fetal left ventricle to dilate the aortic valve. The procedure was successful, with post-intervention Doppler showing improved flow across the aortic valve (peak velocity reduced to 0.8 m/s).

The pregnancy was closely monitored with serial echocardiograms. At 28 weeks, the left ventricle showed mild growth (Z-score improved to -2.0), and the aortic root size increased slightly (Z-score -2.5). The fetus remained stable, and spontaneous vaginal delivery occurred at 38 weeks gestation. The newborn, a male weighing 3.1 kg, was admitted to the neonatal intensive care unit (NICU). A postnatal echocardiogram confirmed HLHS but noted improved left ventricular size and function compared to typical HLHS cases, with an ejection fraction of 45% in the left ventricle. Prostaglandin E1 was initiated to maintain ductal patency, and the infant underwent the Norwood procedure at 7 days of life. At G months follow-up, the infant was stable, with plans for the Glenn procedure at 1 year of age.

Discussion

This case highlights the potential of in-utero aortic valvuloplasty as a novel strategy to alter the natural history of HLHS. Typically, HLHS leads to a severely underdeveloped left ventricle, necessitating a single-ventricle palliation pathway (Norwood-Glenn-Fontan). However, in this case, the in-utero intervention promoted left ventricular growth, potentially allowing for a biventricular repair in the future—an outcome rarely achievable in HLHS. The ESC guidelines on the management of grown-up congenital heart disease (2020) emphasize the importance of early diagnosis and multidisciplinary care in HLHS, but they do not yet address in-utero interventions due to their experimental nature. This case suggests that such interventions may improve outcomes by preserving left ventricular function, aligning with the ESC's focus on optimizing long-term prognosis in congenital heart disease [4, 5].

The technical success of the in-utero procedure, performed at 24 weeks, underscores the importance of early diagnosis and intervention. However, risks such as fetal demise, preterm labor, or maternal complications must be carefully weighed. Larger studies are needed to establish the efficacy and safety of this approach, but this case provides a proof-of-concept for a novel management strategy in severe HLHS.

Patient Perspective

The mother expressed initial anxiety upon learning of the diagnosis but felt reassured by the multidisciplinary team's thorough explanation of the in-utero procedure and postnatal plan. She appreciated the opportunity to potentially improve her child's outcome through the intervention and remains hopeful for his future.

Statement of Consent

Written informed consent was obtained from the patient's mother for the publication of this case report, including the use of de-identified images.

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