Case Report

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# Antenatal Diagnosis of Hypoplastic Left Heart Syndrome at 22 Weeks of Gestational Age: A Case Report

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## Abstract

Hypoplastic left heart syndrome (HLHS) is a severe congenital heart defect characterized by the underdevelopment of the left-sided heart structures, leading to systemic blood flow obstruction. This case report details the antenatal diagnosis of HLHS at 22 weeks of gestation in a 29-year-old woman. Fetal echocardiography confirmed severe hypoplasia of the left ventricle, mitral valve atresia, aortic valve stenosis, and a hypoplastic ascending aorta. The patient was counselled on the prognosis and management options, including staged surgical palliation. At 39 weeks, a male infant was delivered via planned cesarean section and underwent the Norwood procedure at 5 days old. The infant's postoperative course was uneventful, and he was discharged with plans for further surgical interventions. Early antenatal diagnosis and coordinated care significantly improve outcomes for HLHS, highlighting the importance of detailed fetal echocardiography and multidisciplinary management.

**Keywords:- Hypoplastic Left Heart Syndrome, Fetal echocardiography, Congenital Heart Defects, Prenatal Diagnosis**



## INTRODUCTION

Hypoplastic left heart syndrome (HLHS) is a complex congenital heart defect characterized by underdevelopment of the left-sided cardiac structures, leading to severe systemic blood flow obstruction.[1] This condition is fatal without surgical intervention and is often detected during routine prenatal ultrasound examinations. The early identification of HLHS is crucial for perinatal management and planning for postnatal care, which typically involves a series of staged surgical procedures to reconstruct the heart's anatomy and function. [2]

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HLHS occurs in approximately 2-3 per 10,000 live births and accounts for about 2-3% of all congenital heart defects. The pathophysiology involves malformation of the left ventricle, mitral valve, aortic valve, and ascending aorta. This leads to hypoperfusion of the systemic circulation and reliance on the right ventricle and ductus arteriosus to maintain systemic blood flow. Without intervention, HLHS is uniformly fatal within the first few days or weeks of life due to heart failure or inadequate systemic perfusion.[3]

The clinical presentation of HLHS in the fetus is typically asymptomatic, making prenatal screening crucial. Diagnosis is generally made via detailed fetal echocardiography, which can visualize the underdeveloped left heart structures. Associated findings may include a small or absent left ventricle, mitral and aortic valve atresia or stenosis, and a hypoplastic ascending aorta. These anatomical findings necessitate a multidisciplinary approach for management, involving obstetricians, cardiologists, and pediatric cardiac surgeons.[4]

An important aspect of prenatal diagnosis of HLHS is the ability to counsel parents about the prognosis, potential interventions, and long-term outcomes. Early diagnosis allows for planned delivery at tertiary care centers equipped with the necessary neonatal and cardiac surgical facilities. This case report highlights the significance of early antenatal detection and multidisciplinary planning in managing HLHS.[5]

# CASE REPORT

A 29-year-old G2P1 woman at 22 weeks of gestation presented for a routine prenatal ultrasound. The patient had an unremarkable medical history, and her first pregnancy resulted in the birth of a healthy child. During the current pregnancy, a detailed anomaly scan was performed, revealing concerns regarding the fetal heart.

The initial ultrasound indicated a small left ventricle with a disproportionately large right ventricle, raising suspicion for a congenital heart defect. The patient was referred for a detailed fetal echocardiogram, which confirmed the diagnosis of hypoplastic left heart syndrome. The echocardiogram showed a severely hypoplastic left ventricle, mitral valve atresia, aortic valve stenosis, and a hypoplastic ascending aorta. Doppler studies indicated normal flow through the ductus arteriosus and foramen ovale, essential for maintaining fetal circulation.

Following the diagnosis, the patient was counseled extensively regarding the nature of HLHS, potential treatment options, and the prognosis. The options included staged surgical palliation beginning with the Norwood procedure shortly after birth, or the possibility of heart transplantation. The importance of delivery at a specialized center equipped for neonatal cardiac care was emphasized.

Subsequent prenatal visits involved close monitoring of fetal growth and well-being, with regular fetal echocardiograms to assess cardiac function and amniotic fluid levels. No additional anomalies were detected, and the pregnancy progressed without complications.

At 39 weeks of gestation, the patient was admitted to a tertiary care center and delivered a male infant via planned cesarean section. The newborn weighed 3.2 kg and had Apgar scores of 8 and 9 at 1 and 5 minutes, respectively. Postnatal echocardiography confirmed the prenatal diagnosis of HLHS with the same anatomical findings.

The neonate was admitted to the neonatal intensive care unit (NICU) and started on a prostaglandin E1 infusion to maintain ductal patency. At 5 days old, the infant underwent the first stage of surgical palliation, the Norwood procedure, which was performed successfully. The postoperative course was uneventful, and the infant was extubated and weaned off inotropic support over the next week. The baby was discharged home at 4 weeks of age with plans for close follow-up and the subsequent stages of surgical palliation.

| Parameter | Value |
|---|---|
| **Gestational Age at Diagnosis** | 22 weeks |
| **Left Ventricle** | Severely hypoplastic |
| **Mitral Valve** | Atresia |
| **Aortic Valve** | Stenosis |
| **Ascending Aorta** | Hypoplastic |
| **Birth Weight** | 3.2 kg |

**Table 1: Haematological Profile of the case.**

# DISCUSSION

Hypoplastic left heart syndrome is a life-threatening congenital heart defect requiring early

diagnosis and a coordinated approach to management. Similar cases in the literature underscore the importance of detailed fetal echocardiography in the prenatal detection of HLHS. For instance, a study by Tworetzky et al. demonstrated that early fetal diagnosis and timely intervention significantly improved the survival rates and outcomes for infants with HLHS.[6]

The case also highlights the critical role of multidisciplinary counseling and the psychological impact on the parents. Comprehensive counseling includes discussions about the natural history of HLHS, the surgical interventions required, and the long-term outcomes. Studies also have shown that early parental counseling and planned delivery at specialized centers improve postnatal outcomes and parental satisfaction.[7]

The management of HLHS involves staged surgical palliation, with the Norwood procedure being the first stage, followed by the Glenn and Fontan procedures. The Norwood procedure, performed within the first week of life, is essential for establishing a stable systemic circulation. The success of the initial surgery and subsequent stages is critical for survival and quality of life. According to Murtuza et al, outcomes of the Norwood procedure have improved significantly over the years due to advancements in surgical techniques and perioperative care.[8]

This case adds to the body of evidence demonstrating the feasibility and importance of antenatal diagnosis of HLHS. Early detection allows for optimal perinatal management and surgical planning, ultimately improving the survival and quality of life for affected infants. Ongoing research and advancements in fetal surgery may further enhance outcomes for these patients in the future.[9,10]

## .CONCLUSION

The antenatal diagnosis of hypoplastic left heart syndrome at 22 weeks of gestation is critical for the effective management of this congenital heart defect. Early detection through fetal echocardiography, multidisciplinary counseling, and planned delivery at specialized centers significantly improve postnatal outcomes. This case underscores the importance of comprehensive prenatal care and coordinated management in improving the prognosis for infants with HLHS.

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