

Hypoplastic Left Heart Syndrome (HLHS)

Srushti Surve

Perfusion Technology Intern, MGM SBS & Hospital, Navi Mumbai

Abstract

Hypoplastic Left Heart Syndrome (HLHS) is a rare but severe congenital heart defect accounting for approximately 2–3% of all congenital cardiac anomalies. It is characterized by underdevelopment of left-sided heart structures, rendering the left ventricle incapable of supporting systemic circulation. Survival depends on early diagnosis and staged surgical palliation or transplantation. This paper summarizes the pathophysiology, clinical presentation, diagnostic modalities, surgical strategies, perfusion management, and long-term prognosis of HLHS.

Introduction

Hypoplastic Left Heart Syndrome (HLHS) is a congenital cardiac anomaly involving severe hypoplasia of the left ventricle, mitral valve, aortic valve, and ascending aorta. In the absence of surgical intervention, it is universally fatal in the neonatal period. Current management involves staged reconstructive procedures (Norwood, Glenn, and Fontan) or cardiac transplantation.

Anatomy and Circulation

Key anatomical features include a hypoplastic or absent left ventricle, mitral valve atresia/stenosis, aortic valve atresia/stenosis, and hypoplastic ascending aorta. Systemic and pulmonary circulation depend on right ventricular output through an atrial septal defect (ASD) and patent ductus arteriosus (PDA). This physiology creates obligatory right-to-left shunting, leading to systemic desaturation.

Causes and Clinical Presentation

Causes include genetic factors (e.g., Turner syndrome, Trisomy 18), maternal diabetes, smoking, infections, and family history of congenital heart disease. Symptoms manifest within the first 24–48 hours of life and include cyanosis, respiratory distress, poor pulses, cold extremities, metabolic acidosis, and cardiogenic shock if untreated.

Diagnosis

Prenatal diagnosis is typically made via fetal echocardiography between 18–22 weeks gestation. Postnatal investigations include echocardiography (confirmatory), chest X-ray (cardiomegaly, pulmonary congestion), ECG (right ventricular hypertrophy), and pulse oximetry (hypoxemia).

Surgical Management

Definitive treatment consists of staged palliative surgeries: 1) Norwood procedure (Stage I, first week of life), 2) Bidirectional Glenn shunt (Stage II, 4–6 months), 3) Fontan completion (Stage III, 18–36 months). A hybrid Stage I approach may be performed in specialized centers. Alternative therapy includes heart

transplantation.

Perfusion Considerations

Perfusion goals during surgery include maintaining systemic oxygen delivery, avoiding pulmonary overcirculation, ensuring cerebral and myocardial protection, and preserving organ function. Pre-bypass management requires prostaglandin E1 infusion to maintain PDA patency, controlled hypoxemia (SaO_2 75–85%), and maintaining systemic vascular resistance above pulmonary vascular resistance. Bypass strategies differ across stages (Norwood, Hybrid, Glenn, Fontan) with variations in cannulation, flow rates, temperature, and cerebral protection techniques.

Prognosis and Outcomes

Without surgical intervention, HLHS is fatal within the first week of life. With staged repair in specialized centers, 10–15 year survival rates reach 60–70%. Long-term complications include arrhythmias, right ventricular dysfunction, thromboembolism, and protein-losing enteropathy. Lifelong cardiology follow-up is essential.

Conclusion

HLHS remains one of the most challenging congenital cardiac defects. Advances in surgical staging, perfusion management, and perioperative care have significantly improved survival. However, morbidity remains substantial, highlighting the need for continued innovation and long-term multidisciplinary follow-up.