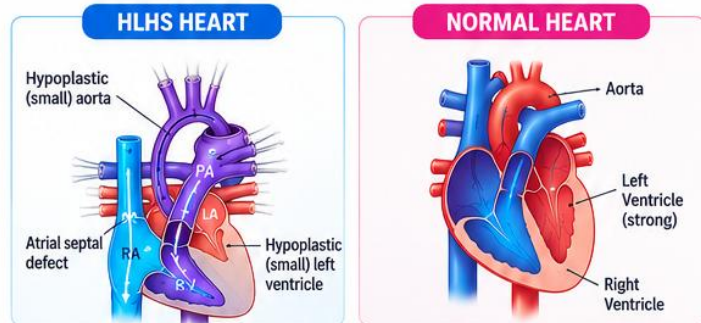


Hypoplastic Left Heart Syndrome (HLHS)

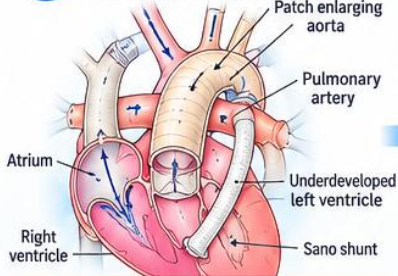
Management, Long-Term Follow-Up, and Activity Recommendations

HLHS is a serious congenital heart defect in which the left side of the heart is underdeveloped and cannot pump blood effectively to the body.



STAGED SINGLE-VENTRICLE PALLIATION (MOST COMMON APPROACH)

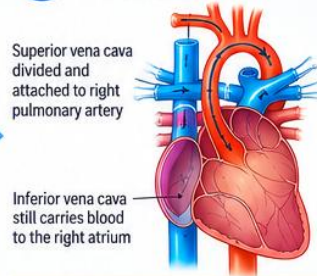
1 Norwood Operation (First Week of Life)



GOALS:

- Create a new aorta
- Provide controlled pulmonary blood flow (using a shunt such as a Sano shunt or BT shunt)
- Establish systemic circulation using the right ventricle

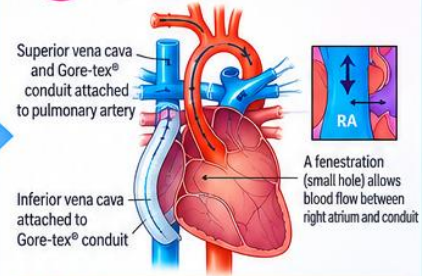
2 Bidirectional Glenn (4–6 Months)



GOALS:

- Connect superior vena cava directly to pulmonary arteries
- Reduce workload on the right ventricle
- Improve oxygen delivery

3 Fontan Operation (2–4 Years)



GOALS:

- Connect inferior vena cava to pulmonary arteries
- Complete Fontan circulation
- Separate systemic and pulmonary venous return

INITIAL MANAGEMENT IN THE NEWBORN

- Prostaglandin E1 infusion to maintain ductus arteriosus patency
- Oxygen and ventilation support when needed
- Management of metabolic acidosis
- Nutritional support
- Evaluation for associated genetic syndromes and extracardiac abnormalities

OTHER TREATMENT OPTION Heart Transplantation

Considered when:

- Anatomy is unfavorable for staged palliation
- Severe ventricular dysfunction
- Significant valve abnormalities

Limitations:

- Donor shortage
- Lifelong immunosuppression
- Risk of rejection

LONG-TERM FOLLOW-UP: LIFELONG CARE IS ESSENTIAL

- **Clinical assessment:** growth, development, exercise tolerance, oxygen saturation
- **Echocardiogram:** ventricular function, valve function, Fontan pathway, aortic arch
- **ECG:** monitor for arrhythmias and conduction abnormalities
- **Holter/Event monitoring:** when palpitations or syncope
- **Cardiac MRI:** ventricular function and blood flow
- **Cardiac catheterization:** for selected cases
- **Liver surveillance:** monitor for liver fibrosis and complications
- **Neurodevelopmental assessment:** learning and attention issues

POTENTIAL LONG-TERM COMPLICATIONS

Cardiac Complications	Fontan-Specific Complications	Neurological & Developmental	Hepatic Complications
• Ventricular dysfunction • Heart failure • Tricuspid valve regurgitation • Arrhythmias • Sudden cardiac death (rare) • Fontan pathway obstruction	• Protein-losing enteropathy (PLE) • Plastic bronchitis • Fontan-associated liver disease • Ascites • Edema • Venous thrombosis • Lymphatic dysfunction	• Learning difficulties • Executive function deficits • ADHD • Developmental delays • Anxiety and depression	All Fontan patients should undergo periodic surveillance for: • Liver fibrosis • Cirrhosis • Hepatocellular carcinoma (rare)

PROGNOSIS

- Outcomes for HLHS have improved dramatically over the past three decades.
- Survival after Norwood surgery exceeds 80–90% at experienced centers.
- Many patients now survive into adulthood.
- Increasing numbers transition successfully to adult congenital heart disease programs.
- Lifelong surveillance remains essential because Fontan circulation is a palliative—not curative—strategy.

ACTIVITY AND EXERCISE RECOMMENDATIONS

Activities Usually Encouraged	Activities Requiring Individual Assessment	Exercise Testing	Red Flags: Seek Immediate Evaluation
✓ Walking ✓ Hiking ✓ Swimming ✓ Cycling ✓ Recreational sports ✓ Golf ✓ Yoga ✓ Light-to-moderate aerobic exercise ✓ School physical education (when appropriate)	• Competitive sports • High-intensity endurance events • Wrestling • Football • Hockey • Martial arts • Heavy weightlifting Eligibility depends on: ventricular function, oxygen saturation, arrhythmia burden, Fontan pressures, and exercise testing results.	Recommended for most older children and adolescents: • Cardiopulmonary exercise testing (CPET) • Exercise ECG testing These help determine: • Safe participation levels • Conditioning programs • Competitive sports eligibility	• Increasing cyanosis • Unexplained fatigue • Reduced exercise tolerance • Palpitations • Syncope • Progressive edema or ascites • Chronic diarrhea (possible PLE) • Hemoptysis or respiratory symptoms suggestive of plastic bronchitis



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