Definition

- **Epidemiology**
  Seven to nine percent of all congenital cardiac anomalies • More common in males than in females (2:1) • Increased incidence in Turner syndrome.
- **Etiology, pathophysiology, pathogenesis**
  Underdevelopment of the left heart • No impairment of fetal circulation • Severe obstruction of the LV and LVOT • Oxygenated blood from pulmonary veins enters the right atrium through the foramen ovale • Dilatation of the right heart and pulmonary arteries • Systemic perfusion occurs through the PDA.

Imaging Signs

- **Modality of choice**
  Echocardiography • MRI and invasive studies for postoperative care (e.g., after the Fontan procedure).
- **Chest radiograph findings**
  Cardiomegaly • Increased pulmonary venous markings • Possible congestion with interstitial edema • Narrow mediastinum.
- **Echocardiographic findings**
  Decreased aortic diameter (< 5 mm) • Small LV • Dilatation of the right heart and pulmonary arteries • PDA • Duplex scan shows left-to-right shunt through the PFO • Echocardiography can be used to assess pressure relationships.
- **CT and MRI findings**
  Used mainly for postoperative evaluations • Patency of aortopulmonary (Blalock–Taussig) shunt and cavopulmonary (Glenn) shunt • Pulmonary arterial anatomy • MR flowmetry for evaluation of cardiac and shunt function.
- **Invasive diagnostic procedures**
  Can be used to detect coronary anomalies • Used mainly in postoperative follow-up • Flow visualization in the hypoplastic ascending aorta • Visualization of the connection of the pulmonary arteries through the PDA • Determination of postoperative pressure relationships.

Clinical Aspects

- **Typical presentation**
  No clinical symptoms immediately after birth • Rapid deterioration after closure of the ductus arteriosus • Heart failure • Volume overload on the pulmonary circulation • Cardiogenic shock • Cyanosis.
- **Treatment options**
  Prostaglandin E_1 to maintain ductal patency • With an interatrial defect small enough to maintain a pressure gradient, the ASD can be expanded by balloon dilatation (Rashkind atrioseptostomy) • Palliative surgical intervention by the Norwood procedure • Heart transplantation is advocated at some centers.
- **Course and prognosis**
  Untreated newborns will die in a matter of days or weeks • Otherwise the prognosis depends on the course and complications of the Fontan procedure.

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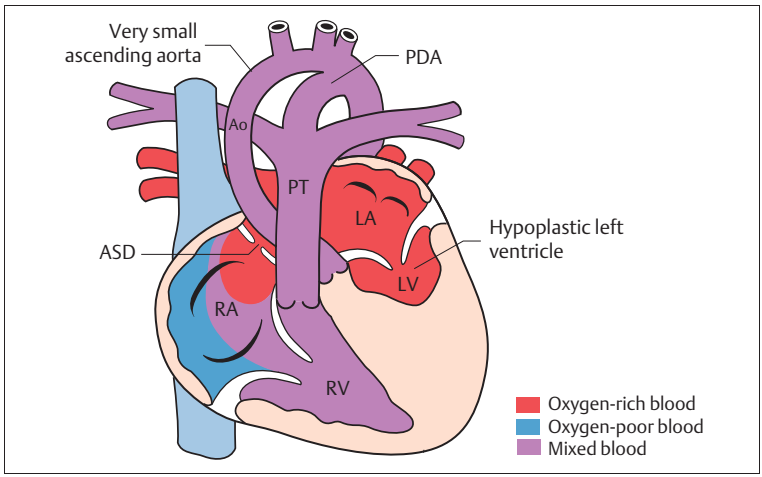

Fig. 9.30 Diagrammatic illustration of HLHS, characterized by hypoplasia of the LA, LV, aortic valve, and ascending aorta (type I). Blood is oxygenated by mixing in the RA with a left-to-right shunt through the ASD. Systemic blood flow relies on a PDA.

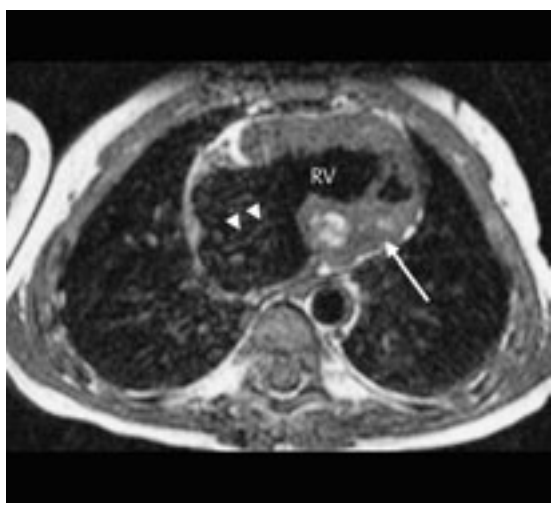

Fig. 9.31 MR image in a 6-month-old boy with HLHS. T1-weighted TSE sequence in an axial plane demonstrates a hypoplastic LV (arrow), an ASD (arrowheads), and a single, hypertrophic RV.

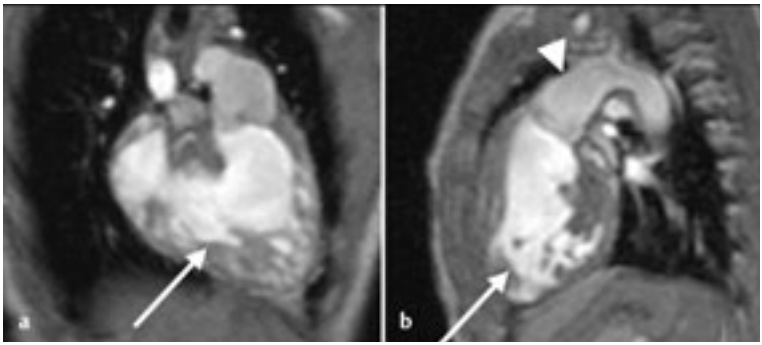

Fig. 9.32 a, b Same patient as in Fig. 9.**31**. Cine GRE sequence in an oblique coronal plane (**a**) and oblique sagittal plane (**b**). Note the hypertrophic single RV (arrow) and the functionally arterialized pulmonary artery (arrowhead) with a hypoplastic aorta. Blood is directed to the lungs through a Glenn or Fontan shunt (not shown).

▸ **What does the clinician want to know?**
Degree of LV and aortic hypoplasia • Size of the PDA and ASD • Ventricular function • Tricuspid insufficiency • Coronary anomalies.

Differential Diagnosis

Aortic stenosis, coarctation of the aorta, interrupted aortic arch	– LV pressure overload in a normally developed heart
Cardiomyopathy	– Generally enlarged heart with normal morphology – Myocardial dysfunction
Arteriovenous malformation	– Morphologically normal heart with volume overload of all chambers

Tips and Pitfalls

If a diagnosis is not made immediately after birth, HLHS should be suspected in infants who show progressive cyanosis and rapid clinical deterioration • Echocardiography should be scheduled as soon as possible.

Selected References

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