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Hypoplastic Left Heart Syndrome & Norwood Procedure

14.1 Morphology & Pathophysiology

The most severe form of hypoplastic left heart syndrome (HLHS) is characterized by atresia of the aortic and mitral valves and hypoplasia of the left ventricle with diminutive ascending aorta. The left ventricle is usually a thick-walled, slit like cavity with endocardial fibroelastosis, and the ascending aorta is often severely hypoplastic, measuring only 2-3 mm in diameter. The left atrium is usually smaller than normal. The right heart (i.e., right atrium, right ventricle, and pulmonary arteries) is markedly enlarged. A patent foramen ovale is common with herniation of the valve of the septum into the right atrium (see Figure 14.1). Ventricular septal defect, though not considered as an integral part of the hypoplastic left heart syndrome, may be present in the syndrome of mitral atresia with a normal aortic root. The most common presentation is a visceroatrial situs solitus with D-ventricular loop and atrioventricular and ventriculoarterial concordance, as well as levocardia.

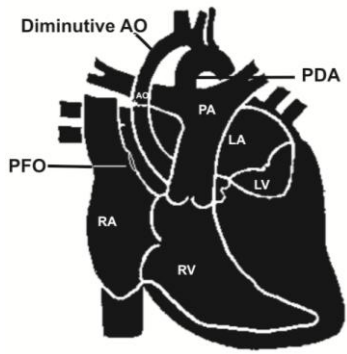

Figure 14.1 *Diagram showing hypolplastic left heart syndrome. LV is hypoplastic with diminutive ascending aorta. The right atrium, the right ventricle, and pulmonary artery are enlarged. The patent foramen ovale allows the blood flow from the LA to RA. The patent ductus arteriosus provides perfusion to the descending aorta and via retrograde to the arch vessels and ascending aorta. The mitral valve is atretic or severely stenotic. AO=ascending aorta, LA=left atrium, LV=left ventricle, PA=pulmonary artery, PDA=patent ductus arteriosus, PFO=patent foramen ovale, RA=right atrium, RV=right ventricle.*

In a newborn, the pulmonary venous blood returning to the left atrium crosses the atrial septum through a patent foramen ovale (partially obstructive in most cases). This blood mixes with desaturated systemic venous blood in the right atrium. The right ventricle pumps this mixed blood to both the pulmonary arteries and the descending aorta, which are connected in parallel by the ductus arteriosus. Coronary artery and cerebral blood flow occurs via retrograde through the aortic arch and ascending aorta. The amount of blood that flows into pulmonary and systemic circulation is based on the resistance of each circuit. Blood flow is inversely proportional to the resistance.

Decreased pulmonary vascular resistance after birth allows a higher percentage of the right ventricular output to go to the lungs instead of the body. The increased pulmonary blood flow results in higher oxygen saturation, but systemic blood flow is decreased with decreased perfusion of the body and development of metabolic acidosis and oliguria. Coronary artery and cerebral perfusion also decrease due to decrease in retrograde flow via the aortic arch and ascending aorta with a risk of myocardial or cerebral ischemia.

If pulmonary vascular resistance is significantly higher than systemic vascular resistance, systemic blood flow is increased at the expense of pulmonary blood flow. This may result in hypoxemia. The circulation in hypoplastic left heart syndrome in a new born infant depends on adequacy of interatrial communication, patency of the ductus arteriosus, and the level of pulmonary vascular resistance. A delicate balance between pulmonary and systemic vascular resistances should be maintained to ensure adequate oxygenation and tissue perfusion.

The infant with HLHS often exhibits signs of cardiogenic shock, including hypothermia, tachycardia, respiratory distress, central cyanosis, pallor, poor peripheral perfusion with weak pulses in all extremities and in the neck, and hepatosplenomegaly.

14.2. Preoperative Management

14.2.1 Open the Ductus Arteriosus

By keeping the patency of the ductus arteriosus with prostaglandin E_1 infusion, immediately establishes ductal patency and ensures adequate systemic perfusion. A larger dose of prostaglandin E_1 is required to reopen the ductus arteriosus in an infant with cardiovascular collapse and shock due to ductal closure. In a relatively asymptomatic infant, a smaller dose of prostaglandin E_1 may be sufficient to keep the ductus arteriosus patent.

(PGE_1 is administered centrally via an umbilical venous catheter).

14.2.2 Correction of Metabolic Acidosis

The infusion of sodium bicarbonate is essential in the early management. This therapy is futile if the ductus arteriosus remains constricted. Metabolic acidosis indicates inadequate cardiac output and adversely affects the myocardium.

14.2.3 Manipulate Pulmonary Vascular Resistance

Note that *pulmonary vascular resistance (PVR*) of a newborn is slightly less than *systemic vascular resistance (*SVR) and begins to fall soon after birth. In HLHS, decreased PVR causes increased pulmonary blood flow and an obligatory decrease in systemic blood flow. Increased alveolar oxygen decreases pulmonary vascular resistance.

1. Oxygen should not be administered unless pulmonary parenchymal disease or pulmonary edema exists. Most infants should remain in the room air with an acceptable oxygen saturation (pulse oximetry in the low 70’s).

   (initially, an umbilical arterial catheter is inserted to obtain frequent blood samples for a serial blood gas analysis).

2. Achieve $PaCO_2$, in the range of 45-50 mm Hg, to increase PVR. This is accomplished by intubation, sedation, mechanical hypoventilation, or the addition of nitrogen or carbon dioxide (with FIO_2 of 15-19%) to the infant's inspired gas via an endotracheal tube or a hood.

Note:

1. Administration of sub-ambient FIO_2 to balance systemic and pulmonary blood flows should be applied only during stabilization of a neonate but should not be pursued for long periods as severe pulmonary hypertension may complicate the postoperative course.

2. Intubation and ventilation along with other measures to balance pulmonary and systemic flows may also improve tricuspid regurgitation.

14.2.4 Inotropic Support

The administration of inotropes may adversely affect the balance between pulmonary and systemic vascular resistances. Therefore, it is indicated only in severely ill neonates with concurrent sepsis or a profound cardiogenic shock and acidosis. The inotropic support should be weaned as soon as the infant is clinically stable.

14.2.5 Diuretics

Diuretics may be used to manage pulmonary overcirculation before surgery.

Agents: Furosemide and spironolactone.

14.2.6 Antibiotics

Antibiotics are indicated if the infant is at a risk of infection. Antibiotics are discontinued after obtaining negative blood cultures.

14.3 Surgical Management of HLHS

The goal of surgery is to separate the pulmonary and systemic circulations. The right ventricle remains as the systemic ventricle while the systemic venous blood passively flows to the lungs through Fontan circulation. The surgical reconstruction is accomplished in three stages:

14.3.1 Norwood Procedure (Stage I)

The goal of Norwood procedure or the stage I procedure is to establish adequate systemic circulation without the ductus arteriosus and provide enough pulmonary blood flow for adequate oxygenation while simultaneously protecting the pulmonary vascular bed in preparation for stages II and III.

14.3.2 Surgical Technique

Operation is usually performed during the first weeks of life after the infant has been stabilized in a neonatal intensive care unit. Surgery is performed on CPB with deep hypothermia and circulatory arrest. The principle surgical steps are:

Creating an anastomosis between the main pulmonary artery and the aorta to provide systemic blood flow by performing the surgical procedures listed below:

1. Ligate the ductus arteriosus. Transect the main pulmonary artery at the level of takeoff of branch pulmonary arteries and close the distal segment of transected main PA with a patch.
2. Perform an anastomosis between the proximal segment of divided main pulmonary artery and the ascending aorta, eliminate coarctation of the aorta, and augment the ascending aorta and arch with a homograft patch to provide adequate sized conduit for systemic blood flow.
3. Perform an atrial septectomy to provide unrestricted blood flow across the atrial septum.

4. Perform the modified Blalock-Taussig shunt to provide pulmonary circulation, using a 5 mm Gore-Tex graft, between RPA (right pulmonary artery) and the innominate artery. Recently, connecting a Gore-Tex graft from the right ventricular outflow tract to the pulmonary artery (Sano operation) has been advocated instead of the conventional modified Blalock-Taussig shunt by some surgeons.

14.4 Postoperative Management

The postoperative course can be variable. The hospital stay may be prolonged as these neonates often require careful monitoring of oral and/or nasogastric feedings. The average stay is two to four weeks.

14.4.1 Hemodynamic Management

Optimal balancing between systemic vascular resistance and pulmonary vascular resistance is mandatory to insure a smooth postoperative course.

Alterations in ventilation, vasoactive infusions, and external stimuli can affect the relative resistances of systemic and pulmonary circulations.

A fall in pulmonary vascular resistance or a rise in systemic vascular resistance will result in overcirculation in the pulmonary circulatory bed (due to increased flow through a shunt), systemic hypoperfusion, and acidosis.

A rise in pulmonary vascular resistance or a fall in systemic vascular resistance results in decreased pulmonary perfusion, hypoxia, and cyanosis.

Postoperative management is directed at achieving the balanced circulation with matched pulmonary and systemic flow. Adequacy of systemic oxygen delivery must be carefully monitored.

Above is accomplished by intubation, sedation, and mechanical ventilation in the immediate postoperative period. The inspired FIO_2 is rapidly weaned to ambient air.

Infusions of a fentanyl and a neuromuscular blocking agent are commonly utilized for sedation.

Arterial oxygen saturation is usually seventy to seventy-five percent on the room air.

Mixed venous oxygen saturation is usually forty-five to fifty-five percent and PaO_2 is between 35 to 40 mm Hg.

14.4.2 Invasive Monitors

Arterial, central venous, and LA (left atrial) catheters.

An oximetry catheter may be used to assess the cardiac output.

14.4.3 Vasoactive Drug Infusions

Dopamine, epinephrine, milrinone, nitroprusside, phenoxybenzamine, and norepinephrine (see Section I Chapters 4 & 16).

14.4.4 Postoperative Bleeding

It is not uncommon complication. Numerous suture lines exposed to systemic level pressure predispose to this complication.

14.4.5 Discharge Medications

Digoxin: To augment the cardiac function.

Diuretics: To manage the right ventricular volume overload.

Aspirin: To prevent thrombosis of the shunt.

Captopril: For the afterload reduction if tricuspid regurgitation is present.

Oxygen saturation is typically 70-80% on the room air.