Chapter

58 Transitional Circulation

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A 38-week-old, 3.67 kg boy born to a G15P4 mother had the prenatal diagnosis of a large vein of Galen malformation. The child was found to have high output cardiac failure on a fetal echocardiogram (hydrops faetalis), which required maternal digoxin therapy during pregnancy. Due to significant hydrocephalus, the child was delivered via cesarean section, with Apgar scores of 2 and 7.

The neonatologist intubated the child at birth due to poor respiratory effort, and placed arterial and venous umbilical catheters. The child now requires assisted ventilation with an FiO_2 of 0.50. The chest X-ray reveals cardiomegaly and poorly aerated lungs. The transthoracic echocardiogram reveals a large patent ductus arteriosus and a patent foramen ovale, both with right to left shunting, as well as moderate tricuspid regurgitation with estimated suprasystemic right ventricular systolic pressures, moderately depressed right ventricular function, and normal left ventricular systolic function.

Vital signs include: UAC: 69/30, HR 120, SpO_2 92% (FiO_2: 0.50)

Describe the Pathway of Normal Fetal Circulation

Oxygenated blood leaves the placenta from one umbilical vein. Most of the oxygenated blood bypasses the hepatic circulation via the ductus venosus and enters the inferior vena cava (IVC). At this juncture, oxygenated blood from the ductus venosus mixes with deoxygenated blood from the lower extremities. Blood from the IVC enters the right atrium, a majority of which is directed by the Eustachian valve across the foramen ovale, through the left atrium and into the left ventricle. Here, the left ventricle pumps blood across the aortic valve to supply the coronary vessels and aortic arch, supplying blood to the upper and lower body.

The superior vena cava (SVC) directs deoxygenated blood from the upper body to the right atrium. As opposed to IVC blood, the majority of SVC blood crosses the tricuspid valve and enters the right ventricle. From the right ventricle, the blood enters the pulmonary arteries, and, due to high pulmonary vascular resistance (PVR), the blood crosses the ductus arteriosus to the distal aortic arch. At this point, this blood mixes with blood from the left ventricle as it travels down the descending thoracic aorta towards the internal iliac arteries and back to the placenta. An umbilical artery originates from each of the internal iliac arteries to deliver fetal blood to, and oxygenate, the placenta (Figure 58.1).

What Are the Important Physiological Differences between Fetal and Adult Circulations?

Fetal circulation is characterized by a high PVR state due to fluid filled lungs and a hypoxic environment. In addition, systemic vascular resistance (SVR) is low due to the large surface area and low resistance of the utero-placental interface. There is shunting across the:

- Liver via the ductus venosus (oxygenated blood)
- Right heart across the foramen ovale (oxygenated blood)
- Lungs and upper body across the ductus arteriosus (deoxygenated blood)

The physiologic shunts allow for the more oxygenated blood from the umbilical vein to preferentially perfuse the brain and heart and the less oxygenated blood from the SVC to perfuse the lower body. Taken as a whole, the placenta, rather than the lungs, provides oxygenated blood for systemic oxygen delivery, and as such pulmonary blood flow is diminutive (8–10 fold) compared to that of postnatal life. Shunts carry fully oxygenated blood (ductus venosus) to the heart and blood with mixed oxygenation (foramen ovale and

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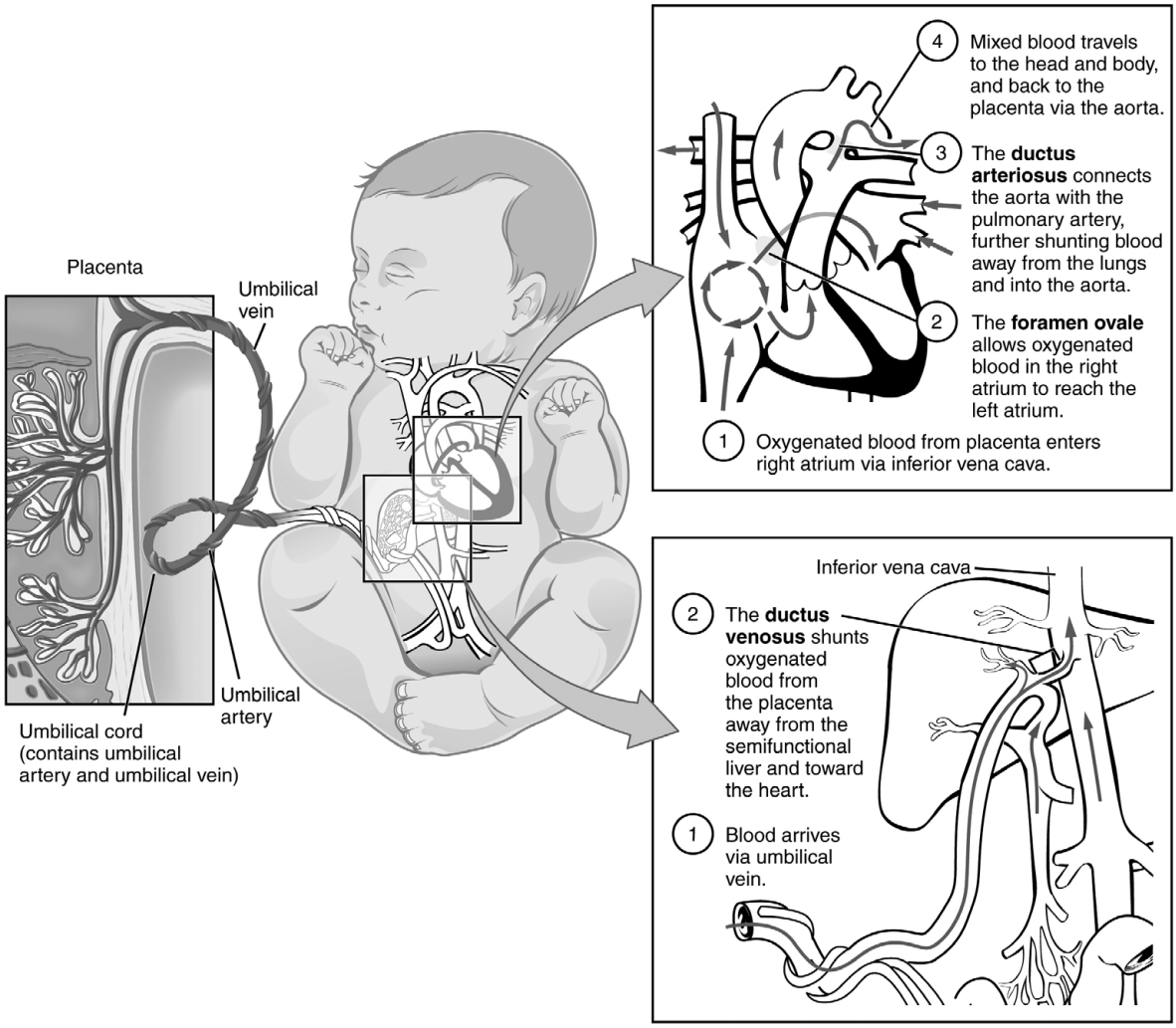

Figure 58.1 Schematic representation of fetal circulation. Image by OpenStax College, reproduced here under CC BY https://upload.wikimedia.org/wikipedia/commons/b/be/2916_Fetal_Circulatory_System-02.jpg

ductus arteriosus) to the body and ultimately back to the placenta via the iliac vessels for oxygenation.

How Is the Fetal Oxygen Carrying Capacity Regulated?

For oxygen transport to occur in the fetal hypoxic environment (normal umbilical vein PaO_2 is 30–35 mmHg), the majority (80%) of hemoglobin (Hgb) is Hgb F, whereas Hgb A accounts for 90% of adult hemoglobin. The different protein structure of fetal hemoglobin confers its greater affinity for oxygen, represented as the PaO_2 at which the hemoglobin molecules are 50% saturated, or the P50 value. Hgb F has a P50 of approximately 19 mmHg whereas Hgb A has a P50 of approximately 26 mmHg (Figure 58.2). This improves fetal oxygen uptake at the placenta. In addition, normal fetal pH (7.25–7.35) is lower compared to adults (7.35–7.45). This acidosis also shifts the oxygen-hemoglobin dissociation curve to the right, facilitating the unloading of oxygen at the fetal tissue level (Figure 58.2).

What Is Transitional Circulation?

Transitional circulation refers to the period of time when the fetal circulation transitions to adult circulation. This period begins immediately after delivery; the

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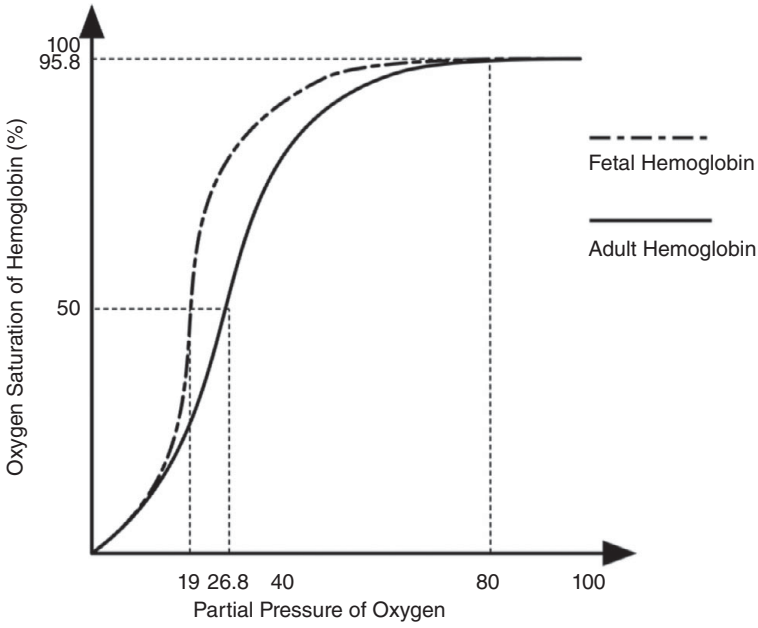

Figure 58.2 Oxyhemoglobin dissociation curve for fetal and adult hemoglobin. Courtesy of Adam C. Adler, MD

umbilical cord is clamped, and the lungs inflate with initial breaths. Umbilical cord clamping increases SVR, and increased PaO_2 reduces PVR. The reduction of PVR increases blood flow to the lungs and left atrium. The increased left atrial pressures functionally close the foramen ovale, eliminating the shunting across the atrial septum. Over the next six to twelve months, the foramen ovale is closed and becomes the fossa ovalis. In approximately 25–35% of adults, closure does not occur, and a probe-patent foramen ovale exists.

While in utero, the ductus arteriosus remains open due to hypoxia, acidosis, and placental prostaglandins. Postnatally, the ductus arteriosus begins to close over the first one to four days of life in the majority of neonates. Functional closure initially occurs as the left atrial and aortic root pressures become greater than the right atrial and pulmonary arterial pressures, and the concomitant increase of PaO_2 and loss of prostaglandin E_2 leads to muscular wall contraction. Over the next one to three months, anatomic closure occurs as proliferation of fibrous tissue occludes the ductus arteriosus lumen. The ductal remnant becomes the ligamentum arteriosum. Indomethacin, a nonsteroidal anti-inflammatory agent, can be used to facilitate closure of a patent ductus arteriosus though inhibition of prostaglandin synthesis. Alternatively, when medical management is unsuccessful, surgical ligation may be required for a patent ductus arteriosus.

When Does Persistent Fetal Circulation Occur?

Persistent fetal circulation (failure in the conversion of fetal to adult circulation) can occur in any condition with severe physiological derangement where there is hypoxia and acidosis. In the vein of Galen malformation (VOGM), one or more branches of arteries of the carotid or vertebral system bypass the cerebral capillary system to feed directly into the Median Prosencephalic Vein of Markowski, an embryologic remnant, creating a left-to-right shunt. In neonates with VOGM, cerebral blood flow may account for up to 80% of cardiac output. Consequently, this lesion can result in high output congestive heart failure, hydrocephalus, and seizures. This high output congestive heart failure due to VOGM may quickly progress to cardiogenic shock during the neonatal period. A persistent transitional circulation, represented here by the presence of a patent ductus arteriosus, patent foramen ovale, and severe pulmonary hypertension, may further complicate the disease process. The increased systolic and diastolic wall stress worsens the cardiogenic shock, which maintains the transitional circulation.

Any condition with pulmonary hypertension (persistent pulmonary hypertension of the newborn) may cause a persistent fetal circulation. The increase in right ventricular afterload leads to right-to-left

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shunting through a patent foramen ovale and/or patent ductus arteriosus, as well as right ventricular dysfunction. Underdevelopment, maldevelopment or maladaptation of the pulmonary vasculature due, for instance, to pulmonary hypoplasia, meconium aspiration, severe respiratory distress syndrome, pneumonia, or episodes of hypoxia can also lead to persistent pulmonary hypertension of the newborn (PPHN).

In congenital heart disease, there are lesions incompatible with postnatal circulation. These could include ductal dependent lesions (such as pulmonary atresia or hypoplastic left heart syndrome) or parallel systemic and pulmonary circulations (such as transposition of the great arteries). In these cases, prostaglandin E_1 is often used to maintain a patent ductus arteriosus until an intervention can occur. Infants receiving prostaglandin E_1 are at risk for apnea, fevers, and generalized edema.

What Are the Anesthetic Considerations in Caring for a Child with a Persistent Transitional Circulation?

A thorough understanding of the disease pathophysiology guides management. For the VOGM, endovascular treatment is often attempted to reduce or eliminate the degree of arteriovenous shunting. Oftentimes, this requires multiple interventional radiology sessions.

Assessment of end-organ function is paramount to the preoperative assessment, not only to tailor anesthetic care, but also to determine whether a patient is a candidate for intervention. Given a wide spectrum of disease, providers must determine the likely long-term outcomes for each patient. A trans-fontanelle ultrasound, brain MRI, head CT, EEG, postnatal TTE, renal and hepatic studies, and a thorough clinical exam can augment decision-making with regard to intervention and anesthetic management. The prenatal ultrasound can help determine the degree of intracranial shunt, as well as the degree of cardiac dysfunction. While conventional angiography is the gold standard for imaging of VOGM, this exam is usually only undertaken if an intervention is planned.

Preoperatively, neonates may require respiratory support with high-flow nasal cannula or endotracheal intubation. Heart failure is managed with diuretics (e.g., furosemide) and inotropic support (e.g., epinephrine, dopamine, milrinone). The patient is closely monitored for multi-organ failure while awaiting endovascular treatment. With very severe heart failure and pulmonary hypertension, inhaled nitric oxide (iNO) may be necessary to reduce pulmonary vascular resistance to maintain oxygenation.

Intraoperative management necessitates balancing competing physiologic goals and mitigating the effects of a persistent transitional circulation. Avoiding intermittent increases in pulmonary arterial pressures (pulmonary hypertensive crises) requires adequate oxygenation, ventilation, analgesia, and iNO as needed. Treatment with sodium bicarbonate of the severe acidosis due to hypoxia and a low cardiac output state may also be necessary.

Ensuring adequate coronary perfusion pressures requires close monitoring for ECG changes indicative of ischemia. Diastolic runoff that compromises coronary perfusion may occur with a patent ductus arteriosus. In this case, blood pressure may need to be augmented or additional oxygen carrying capacity may be required with a transfusion of red blood cells. Excessive reduction in PVR may lead to increased diastolic runoff, which stands in opposition to minimizing the PPHN seen with a persistent transitional circulation. Clinical signs and symptoms (delayed capillary refill, low diastolic blood pressure, worsening end-organ damage) may prompt scaling back of interventions that decrease PVR to reduce diastolic runoff.

In addition to standard ASA monitors, recommended access and monitors include:

- Adequate intravenous access for blood transfusion.
- Arterial line for close hemodynamic monitoring and arterial blood gases.
- Central line for infusion of inotropes.

With regard to patients with VOGM, a large amount of saline may be infused by the interventionalist during the endovascular procedure. It is imperative to monitor for volume overload and consider diuretic administration during the procedure. In addition, consequences of volume overload should be anticipated including: dilutional anemia, dilutional coagulopathy, and electrolyte imbalances.

After the procedure, the patient is left intubated and transported to an intensive care unit for continued critical care and monitoring.

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Suggested Reading

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