38 Aortopulmonary Shunts and Ductal Stents

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Aortopulmonary shunts and PDA stents are important tools for the palliation of patients with CHD. They are mainly used to augment or maintain a stable source of pulmonary blood flow as part of the palliation strategy in patients with single-ventricle physiology or to postpone a definitive biventricular repair in patients with comorbidities or those in which further somatic growth would allow for a better repair. They can also be used to promote growth of the branch PAs in some patients by augmenting flow (e.g., patients with ductal-origin of the PA or those with pulmonary atresia, VSD, and major aortopulmonary collaterals). Additionally, PDA stents are selectively used as part of a hybrid palliation in patients with hypoplastic left heart syndrome.

The selection between PDA stents and different types of aortopulmonary shunts depends on the particular clinical situation of the patient and is beyond the scope of this chapter.

Aortopulmonary Shunts

Since the original description of the classic shunt by Blalock and Taussig, multiple different types shunts between the systemic and pulmonary circulations have been devised. Some of these shunts include:

- **Classic Blalock-Taussig-Thomas shunt (BTTS).** The subclavian artery is transected and then sutured directly into the PA to increase pulmonary blood flow. This shunt is rarely used currently but may be useful in patients in which growth of the subclavian artery (and the shunt) is necessary (e.g., patients with no plans for future palliation). BP measurement on an extremity that has undergone a classic BTTS will obviously be inaccurate.
- **Modified Blalock-Taussig-Thomas shunt (mBTTS).** See below.
- **Mee shunt (or Melbourne shunt).** Used for patients with pulmonary atresia, VSD, and major aortopulmonary collaterals with diminutive PAs. The main PA is disconnected from the heart and anastomosed on and end-to-side fashion to the ascending aorta in order to promote growth of the branch PAs and allow for future reconstruction. This procedure can be performed through a median sternotomy or a left thoracotomy.
- **Waterston-Cooley shunt.** Anastomosis between the ascending aorta and the right PA. Rarely used currently.
- **Potts shunt.** Anastomosis between the descending aorta and the left PA. Rarely used but has been recently applied to patients with refractory pulmonary hypertension to decompress the pulmonary circulation.

The shunt that is most commonly used currently is the mBTTS. This shunt involves placing a Gore-Tex® tube from the innominate or subclavian artery to one of the branch PAs. The amount of flow through an mBTTS is critical since it determines the degree of overcirculation or cyanosis that the patient will experience. This flow is a complex interplay of multiple factors including the size and length of the shunt, the location

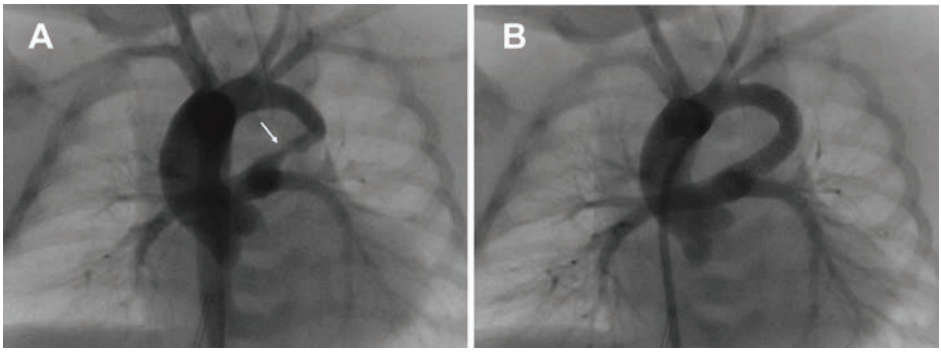

Figure 38-1. Angiogram in a neonate with tetralogy of Fallot, right aortic arch, and cyanosis. A) A PDA can be seen arising from the left innominate artery with constriction (arrow). B) After stent placement, a widely patent PDA is seen.

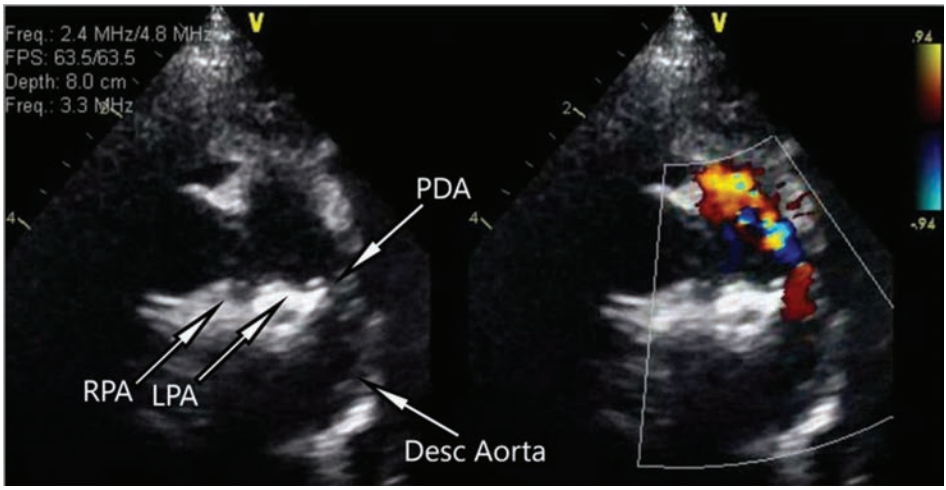

Figure 38-2. High-parasternal color-compare short-axis echocardiographic view ("3-finger view") demonstrating a small PDA.

of takeoff from the systemic circulation, and the difference between systemic and pulmonary vascular resistances.

An mBTTS can be performed either through a median sternotomy or a posterolateral thoracotomy. A clear understanding of the aortic arch anatomy (i.e., arch sidedness, brachiocephalic branching pattern, PDA sidedness) and PA anatomy is critical to plan the operation. On patients with usual anatomy, we have favored performing the procedure via a right posterolateral thoracotomy. Using this technique, the shunt is placed into the distal right subclavian artery (lateral to the recurrent laryngeal nerve), which serves as the restrictor to flow. This allows placement of a larger shunt (usually a 4 mm Gore-Tex® graft in a newborn) that will last longer into infancy as the subclavian artery slowly increases in size. In addition, it decreases the likelihood of overcirculation by

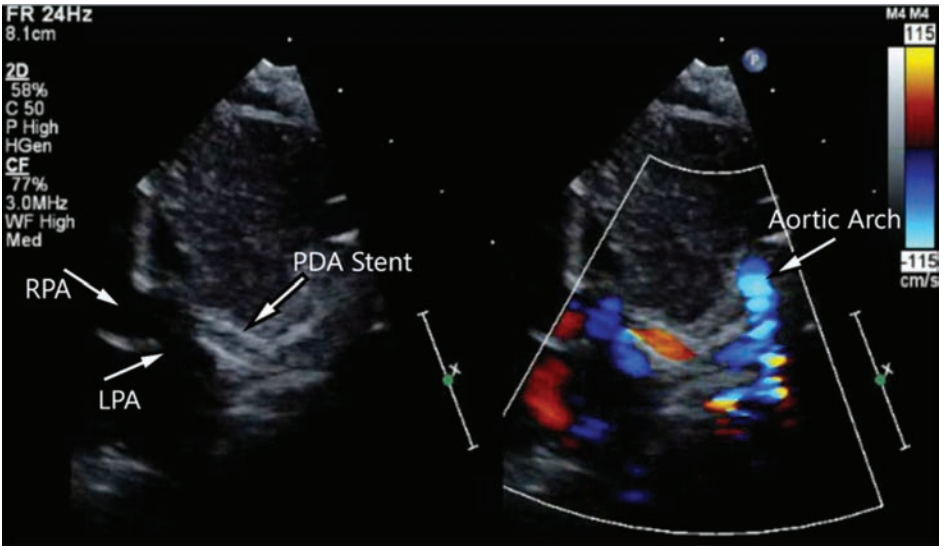

Figure 38-3. Echocardiographic image with color-compare showing a patent PDA stent.

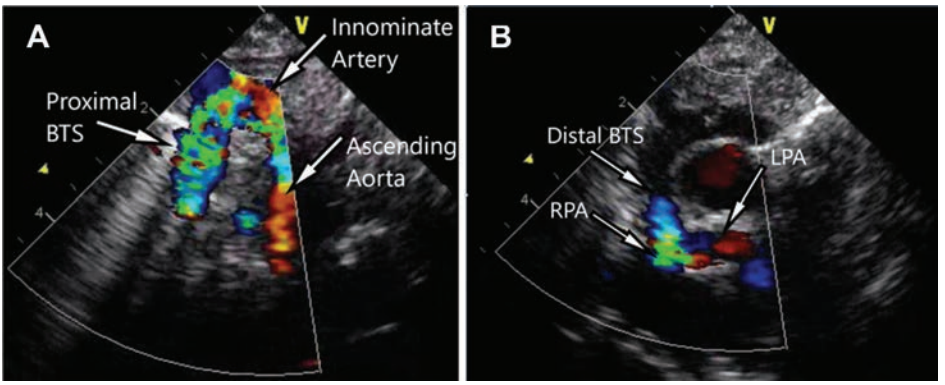

Figure 38-4. Echocardiographic suprasternal notch view showing the proximal (A) and distal (B) ends of a patent mBTTS.

allowing the subclavian artery to modulate the flow. The sidedness of the thoracotomy (right or left) is mainly determined by the location of the PDA (right thoracotomy for a left-sided PDA and left thoracotomy for a right-sided PDA) in order to allow the PDA to provide blood supply to the contralateral lung during the procedure. The PDA is then allowed to close spontaneously after the procedure.

In patients with unusual anatomy or those requiring other procedures, the shunt is created via a median sternotomy. Even when performed through a sternotomy, every attempt is made to place the shunt as far distal into the subclavian artery as possible in order to modulate flow. When performing the procedure via a median sternotomy, a 3.5 mm Gore-Tex® graft is usually used.

Table 38-1. Clinical postoperative scenarios and recommended interventions.

Clinical Scenario	Possible Etiology	Interventions
High SaO_2 (>95%) and early markers of low systemic output (falling NIRS, metabolic acidosis, rising lactate)	Excessive pulmonary blood flow with or without systemic vasoconstriction	Check ABG, avoid alkalosis, reduced FiO_2 if possible, consider increasing sedation to overcome intrinsic tachypnea. If hypertensive (or if systemic BP allows) consider short-acting systemic vasodilator. Optimize hemoglobin.
As (1) with additional markers of organ injury (e.g., elevation of creatinine, oliguria, liver dysfunction, irritability)	Excessive pulmonary blood flow with significant systemic "steal"	In addition to interventions in (1), provide supportive therapy for organ dysfunction, consider neuromuscular blockade. Discuss with cardiology and cardiac surgical team the need for additional interventions to limit pulmonary blood flow, including duct ligation if PDA still present, or potentially downsize the shunt.
Diastolic hypotension with or without signs of ischemia on ECG	Excessive pulmonary blood flow with significant coronary "steal"	This requires a careful balance between managing systemic perfusion without further compromising diastolic pressure. Interventions include optimizing ventilation, infusion of colloid, and addition of a low-dose vasopressin or norepinephrine infusion.
Differential flow apparent on CXR	Normal postoperative findings (plethoric shunted side) *or* obstructed flow to the nonshunted (oligemic) lung	Optimize ventilation. Discuss with cardiology and cardiac surgery. Requires further evaluation with echocardiogram / CTA / catheterization.
Desaturation and arterial hypoxemia without any focal lung pathology to explain it	Pulmonary cause or shunt malfunction (obstruction or occlusion)	Immediate CXR. In the absence of a respiratory etiology, this suggests *shunt malfunction* until proven otherwise. Oligemic lung fields or quiet or even absent shunt murmur will support this, but absence of these does not exclude shunt malfunction. After optimizing ventilation, there should be an immediate discussion with cardiac surgery and cardiology. Urgent echocardiography should be ordered and depending on the findings, further imaging including CTA or cardiac catheterization may be required. Depending upon the clinical condition of the patient, immediate surgical assistance at the bedside should be considered with readiness to provide urgent ECMO support.

PDA Stents

Percutaneous stent implantation can be performed to maintain ductal patency in patients with ductal-dependent pulmonary blood flow. Details of the ductal anatomy are vital to obtain prior to the procedure. Using TTE, the origin and insertion of the ductus, in addition to PA anatomy, should be ascertained. If not possible with TTE, further imaging with a cardiac CT scan may be necessary.

As these infants are maintained on PGE infusion, the size of the PDA is frequently too large to allow placement of an appropriately sized stent (i.e., a stent size that would not result in significant pulmonary overcirculation). PGE is usually stopped the day before the procedure (allowing for a constriction to occur with a safe level of desaturation) to provide an estimate of how long PGE will eventually need to be stopped before the cardiac catheterization procedure. The exceptions to this protocol are patients with ductal-origin of a PA who may not exhibit a change in oxygen saturation if PGE is stopped (the other PA is usually in continuity with the main PA). In this instance, ductal constriction can be gauged by TTE.

The cardiac catheterization procedure is performed under general anesthesia. Based on the origin of the ductus (i.e., from the descending aorta, underside of the aortic arch, innominate artery, subclavian artery, or ascending aorta), an access site is chosen that will result in the straightest trajectory. This may be from a percutaneous femoral artery, carotid artery, axillary artery, umbilical artery, or transvenous route. Patients are heparinized during the procedure to maintain activated clotting times of >250 seconds.

After the stenting procedure (Figure 38-1), patients are started on aspirin and in some instances, heparin therapy is administered until aspirin can be given. In addition to looking for signs of ductal stent patency on physical examination, it is important to interrogate the stented ductus in detail by TTE. Any patient with lower saturations than normal for a given physiology or signs of stent narrowing by echocardiography warrants prompt referral, and immediate catheter/surgical intervention may be indicated.

Imaging

A PDA usually courses from the undersurface of the aortic arch to the main PA. The best and more reliable echocardiographic view is a modified high-parasternal short-axis view with both branch PAs and the descending aorta in the same image ("3-finger view") (Figure 38-2). This view provides the best view of the entire length of the ductus arteriosus including its aortic and pulmonary sides, as well as the direction of blood flow. It is also a reliable view to use in order to "rule out" the presence of a PDA.

If the ideal ductal view cannot be obtained due to poor echocardiographic windows or to unsual anatomy (such as a right aortic arch), one can use a combination of an aortic arch view from the suprasternal notch (with the ascending aorta and the aortic isthmus present) and a high-parasternal short-axis view with both PA branches. It is important to keep in mind that the direction of blood flow might be reversed (right-to-left), in which case a large PDA could be easily be missed if one only focuses on a left-to-right shunt.

A PDA stent is expected to cover the entire length of the ductus without obstruction to blood flow in the proximal descending aorta and proximal left PA. Color and spectral

Doppler should be obtained not only within the PDA, but also in the descending aorta and proximal branch PAs (Figure 38-3).

An mBTTS in a patient with a left aortic arch will be located on the patient's right side, and course from the right subclavian artery to the proximal right PA. A suprasternal notch view of the aortic arch is the best place to start looking for the aortic origin of the shunt (Figure 38-4, A). The ultrasound probe should be angled superiorly towards the right shoulder, following the course of the innominate artery. Once the aortic side of the shunt is found, the course of the blood flow should be followed inferiorly, ideally to its entrance into the proximal right PA (Figure 38-4, B). The echocardiographic image should have the subclavian/innominate artery superiorly and the right PA inferiorly, with the entire length of the mBTTS coursing vertically.

Complete evaluation of an mBTTS includes evidence of any obvious narrowing or blood flow acceleration by color Doppler, as well as spectral Doppler evaluation. Pulse Doppler should be used before and after any level of obstruction. Continuous-wave Doppler should also be obtained, ideally along the entire length of the shunt, or proximally, medial, and distal, if proper alignment of the entire shunt is not possible.

After stenting of an mBTTS, it is important to evaluate for any evidence of obstruction in the innominate artery and proximal right PA. A final velocity inside the stent itself should also be reported.

Perioperative Care

An mBTTS or PDA stent placed during the neonatal period or early infancy should in general, be expected to provide appropriate pulmonary blood flow for adequate oxygenation, systemic oxygen delivery, and somatic growth for at least a period of a few months, if not longer. There is an acceptable mantra that a young infant may have to "grow into" the shunt, as the perfect shunt for a 3 kg newborn, may not remain perfect for very long. Thus, the "ideal shunt" may not appear physiologically ideal in the first instance, meaning that careful intensive care management and exquisite attention to detail are very important during the first days after surgery or PDA stent placement.

The mainstay of preoperative and postoperative management of infants undergoing shunt or PDA stent placement lies in the careful optimization of systemic oxygen delivery, while avoiding excessive pulmonary blood flow. Medical management includes attention to ventilatory management and gas exchange in order to avoid excessive pulmonary blood flow and simultaneously, careful fluid management, titration of vasoactive agents, and optimization of hematocrit to maximize systemic and myocardial oxygen delivery. Finally, avoidance of a hypercoagulable state and appropriate anticoagulation are essential to maintain shunt patency. Table 38-1 illustrates several clinical postoperative scenarios and recommended interventions.

Ventilation

In general, the basic principle that a shunt or stent provides a stable and controlled source of pulmonary blood flow means ventilation should be titrated only to avoid unwanted changes in PVR. The main drivers of PVR are pH and oxygen tension.

All newborns and the majority of young infants remain intubated and mechanically

ventilated immediately after surgery. The duration of ventilation is variable and depends mainly on hemodynamics and systemic oxygen delivery, as well as other factors such as the status of the lungs or other comorbidities. In general, patients should progress to extubation within 48 hours of returning to the ICU.

We typically recommend maintaining a normal pH with the avoidance of alkalosis (respiratory or metabolic) with a normal $PaCO_2$. Inspired oxygen fraction should be titrated to provide a target PaO_2. This will in part depend on the underlying anatomy but would typically be <50 mmHg, though this number should not be overinterpreted in isolation. While ventilation can in theory be used to "control" pulmonary blood flow using hypoventilation or a respiratory acidosis to produce a deliberate elevation of PVR, this is not a recommended approach as acidosis can have unwanted effects on other organs including the brain and myocardium.

Hemodynamic Management

The first few hours after surgery represent a critical period in terms of establishing adequate systemic and myocardial perfusion. During this time, frequent reassessments of the patient should be performed focusing on the clinical assessment of systemic oxygen delivery and organ function. A physiologically generous shunt without careful and preemptive intensive care management, can produce systemic "steal" resulting in secondary organ dysfunction – most commonly acute kidney injury. Similarly, coronary "steal" can lead to myocardial ischemia. Meticulous attention to systemic BP (including diastolic pressure), acid-base balance including lactate, fluid balance, and other non-invasive markers of oxygen delivery including NIRS measurements are recommended at this early stage. It is also important to remember that hemoglobin is an important determinant of systemic oxygen delivery and is typically maintained at ~12-14 g/dL. IV vasoactive medications are often used early after surgery. Considerations include the use of milrinone to optimize systemic afterload, vasopressin, or norepinephrine to optimize diastolic BP and coronary perfusion, and low-dose epinephrine (<0.04 mcg/kg/min), particularly in the presence of any ventricular dysfunction.

Fluid Balance

There are no specific "goals" for early fluid management other than to avoid fluid overload (as this can delay postoperative recovery) and hypovolemia. Hypovolemia can be particularly undesirable early after a shunt – or even at any stage after surgery, as this may compromise shunt flow and can contribute to shunt occlusion. We recommend the careful use of diuretics in order to avoid fluid overload, but not necessarily as a routine.

Shunt Patency

The use of low-dose systemic unfractionated heparin infusion is recommended during the early postoperative period. We typically commence heparin once the coagulation studies are normal, or close to normal, and generally at around 4 hours after returning to the ICU. We typically infuse 10 Units/kg/hour and do not target any coagulation parameters. Indeed, at this low dose, we would not expect much change, if any, of the PTT. Enteral antiplatelet therapy with aspirin (~5 mg/kg/day) is commenced on postoperative day 1, aspirin responsiveness is tested after 3 doses, and if appropriate, heparin is discontinued (see Chapter 59).