Chapter 6

# Critical Pulmonic Stenosis

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**Case Scenario**

A 5-day-old male presents with tachypnea and poor feeding after being discharged home on day 2 of life. His mother had an uneventful pregnancy with routine prenatal care and testing and he was born via spontaneous vaginal delivery without complications. On day 4 of life his mother noted that his breathing was fast and he was not eating well and decided to seek care. At the pediatrician's office the patient was noted to have room air oxygen saturations of 75%.

Transthoracic echocardiography showed the following:

- *Critical valvular pulmonary stenosis, peak gradient 72 mm Hg*
- *Intact ventricular septum*
- *Mild right ventricular hypertrophy*
- *Normal left ventricular function*
- *Mild to moderately depressed right ventricular function*
- *Moderate tricuspid regurgitation*
- *Patent foramen ovale with bidirectional shunting*

He was admitted to the congenital cardiac intensive care unit and was started on prostaglandin $E_1$ and milrinone. He is scheduled for balloon valvuloplasty of the pulmonic valve in the cardiac catheterization laboratory.

## Key Objectives

- Describe the role of the ductus arteriosus during fetal life.
- Discuss echocardiographic findings related to pulmonary stenosis.
- Describe perioperative management of a patient with critical pulmonary stenosis.

## Pathophysiology

### What is the pathophysiology of critical pulmonary stenosis?

During fetal development, little blood flows through the lungs because of high pulmonary vascular resistance (PVR). During fetal life, the majority of pulmonary artery blood flow is directed through the ductus arteriosus (DA) to the aorta, flowing right to left (R-to-L). After birth, PVR is initially elevated and then decreases over the first few days of life. As pulmonary pressures become lower than systemic pressures, flow in the DA reverses, becoming left-to-right (L-to-R) from the aorta to the pulmonary artery.

In normal infants the DA is not needed after birth and begins to functionally close during the first 24–72 hours after birth. It is anatomically closed between the third and fourth week of life. Persistent patency of the DA can be caused by stress, hypoxia, and acidosis.

In this infant, critical PS limited the amount of antegrade pulmonary blood flow (PBF) through the pulmonary valve. Therefore, the major source of source of PBF was provided via the DA, flowing from the aorta to pulmonary artery. (See Figure 6.1.) As the DA began to close during the first few days of life, PBF was reduced. Antegrade blood flow through the critically stenosed pulmonic valve was not sufficient and the infant became hypoxemic, causing tachypnea and poor feeding. (See Figures 6.2 and 6.3.)

### How is critical pulmonary valve stenosis defined?

Intervention is recommended for patients with a gradient across the pulmonary valve >50 mm Hg.

The following echocardiographic classifications are defined:

- **Mild PS**: Peak instantaneous gradient <30 mm Hg
- **Moderate PS**: Peak instantaneous gradient between 30 and 60 mm Hg
- **Severe PS**: Peak instantaneous gradient >60 mm Hg

**Clinical Pearl**

*Echocardiographic and direct catheter measurement of PV gradients vary, as they use different measurement techniques. The peak instantaneous Doppler gradient can overestimate the catheter gradient by 20–30 mm Hg, and the mean echocardiographic gradient may underestimate the gradient compared to catheterization. During cardiac catheterization the pressures on each side of the valve are directly transduced.*



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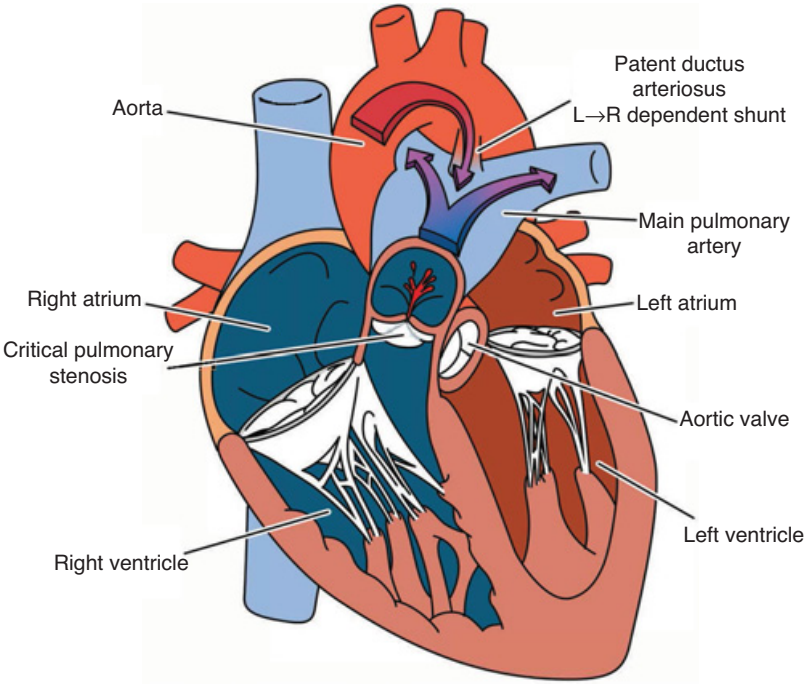


**Figure 6.1 Critical pulmonic stenosis.** Drawing by Ryan Moore, MD, and Matt Nelson.

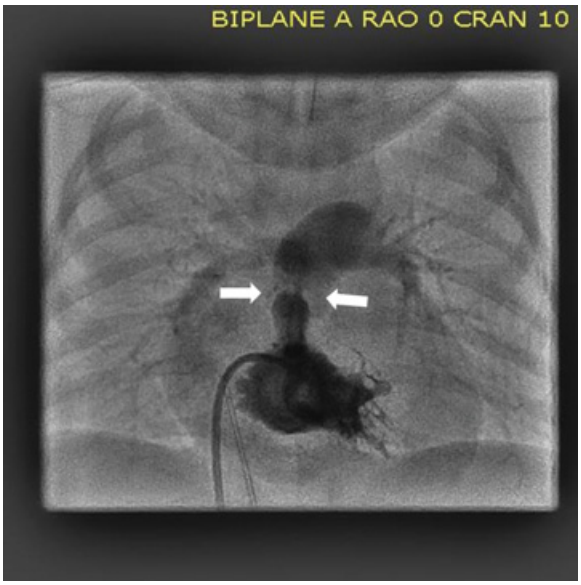


**Figure 6.2 Right ventricle, pre-balloon dilation.** An angiogram is performed in the right ventricle in the AP projection. The pulmonary valve is thickened and doming (arrows). Courtesy of Russel Hirsch, MD.

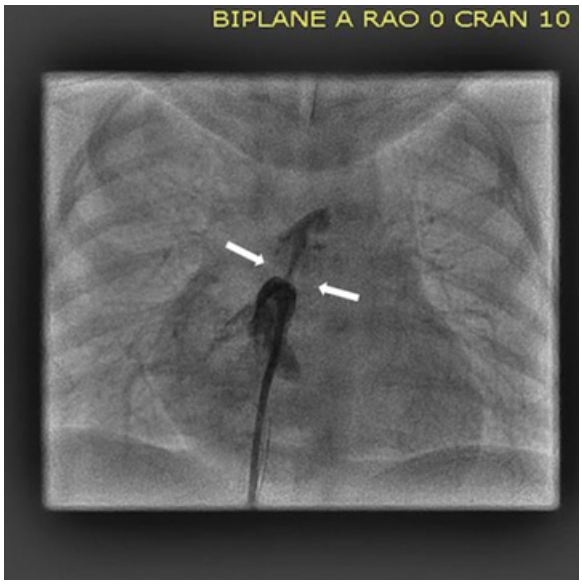


**Figure 6.3 Focused injection under the pulmonary valve.** A hand angiogram is performed under the pulmonary valve in the AP projection. The narrow jet of contrast flow is seen passing through the stenotic pulmonary valve and into the main pulmonary artery (arrows). Courtesy of Russel Hirsch, MD.

## Is critical valvular PS associated with other lesions and syndromes?

Although PS may be an isolated abnormality, it commonly occurs in association with tricuspid atresia, tetralogy of Fallot, dextro (d)- and levo (l)-transposition of the great arteries, and double outlet right ventricle. Critical pulmonary stenosis is associated with Noonan syndrome, Alagille



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syndrome, Williams–Beuren syndrome, and congenital rubella syndrome.

## Why was the patient started on $PGE_1$?

Once the DA is closed or closing, it can usually be reopened therapeutically after initiation of a $PGE_1$ infusion. $PGE_1$ relaxes ductal tissue, improving blood flow through the DA into the pulmonary circulation. The usual starting dose of $PGE_1$ is 0.01–0.05 mcg/kg/minute and it may be titrated up to 0.1 mcg/kg/minute. Adverse effects of $PGE_1$ include apnea, bradycardia, fever, flushing, and less frequently seizures, hypotension, tachycardia, sepsis, and diarrhea. Owing to these adverse effects the dose is titrated down once ductal patency has been reestablished.

**Clinical Pearl**

*Should oxygen desaturation occur in a patient with ductal-dependent PBF, the differential includes both standard pulmonary causes of hypoxia and decreased PBF due to systemic hypotension or hypovolemia, increasing PVR, or inadequate blood flow via the DA.*

## What is an acceptable oxygen saturation for this patient after ductal flow is reestablished?

The effects of $PGE_1$ infusion can be seen within minutes. The $PaO_2$ can often increase as much as 20–30 mm Hg with initiation of the $PGE_1$ infusion. With ductal flow reestablished, the patient's oxygen saturation should be in the mid-80s. If the oxygen saturation falls below 80%, other etiologies for desaturation should be investigated and treated after assuring that the prostaglandins are being delivered. Should the oxygen saturations remain low it may be necessary to increase the $PGE_1$ rate. Due to the presence of the PFO, the oxygen saturation will not approach 100%, as mixing of oxygenated blood with deoxygenated blood is occurring within the heart. As the $PGE_1$ infusion rate is increased, the risk of apnea also increases, and endotracheal intubation may be indicated.

**Clinical Pearl**

*Because of the presence of bidirectional shunting at the level of the PFO, the patient's systemic oxygen saturation will not approach 100%, as mixing of oxygenated blood and deoxygenated blood is occurring within the heart.*

## Why was the patient started on milrinone?

In this case, RV dysfunction exists due to increased right heart pressure secondary to impeded forward flow through the stenotic pulmonic valve, resulting in tricuspid regurgitation. Milrinone is both an inotrope and vasodilator for the systemic and pulmonary vasculature. It improves diastolic function, as well as myocardial contractility. As such, it is often used in patients with poor cardiac function. The usual dose of milrinone is 0.25–0.5 mcg/kg/minute. Milrinone works by decreasing the degradation of cyclic adenosine monophosphate via phosphodiesterase inhibition, with a half-life of approximately 2 hours.

## What does a balloon valvuloplasty procedure entail?

Balloon pulmonary valvuloplasty is the treatment of choice for the typical dome-shaped valve characteristically seen in PS. Balloon valvuloplasty is a catheter-based procedure, performed in the cardiac catheterization suite or hybrid operating room. Vascular access is usually obtained via the femoral veins. Femoral arterial access may also be obtained for arterial pressure monitoring during the procedure. A sheath is placed in the femoral vein, with a catheter entering the right heart via the inferior vena cava, then passing through the stenotic pulmonary valve. A balloon is then inflated at the level of the pulmonic valve. (See Figures 6.4 and 6.5.) All stenosis may not be relieved, as the PV leaflets are often dysmorphic. Additionally, pulmonary insufficiency can result from balloon valvuloplasty as the thickened and calcified pulmonary valve may not coapt well after balloon dilation.

**Clinical Pearl**

*Pulmonary insufficiency can result from balloon valvuloplasty. The thickened and calcified pulmonary valve may not coapt well after balloon dilation, and insufficiency may result.*

# Anesthetic Implications

## What preoperative cardiac evaluation and workup is necessary?

An echocardiogram should be performed, and the following information assessed:

- Biventricular function



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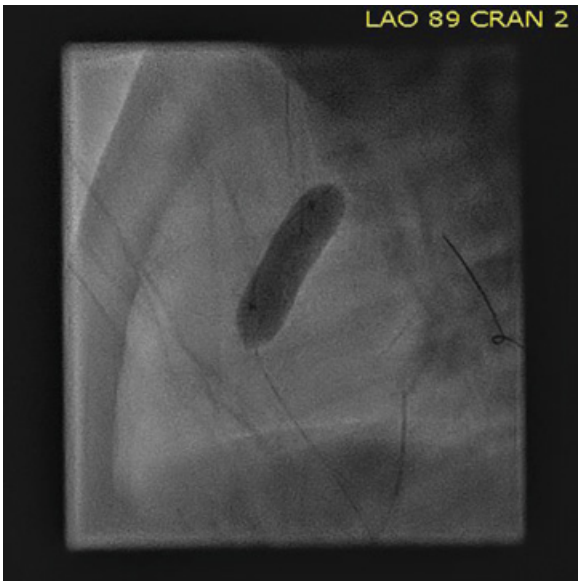


**Figure 6.4 Balloon dilation of the pulmonary valve.** Still frame angiogram in the lateral projection demonstrating balloon dilation across the pulmonary valve. Courtesy of Russel Hirsch, MD.

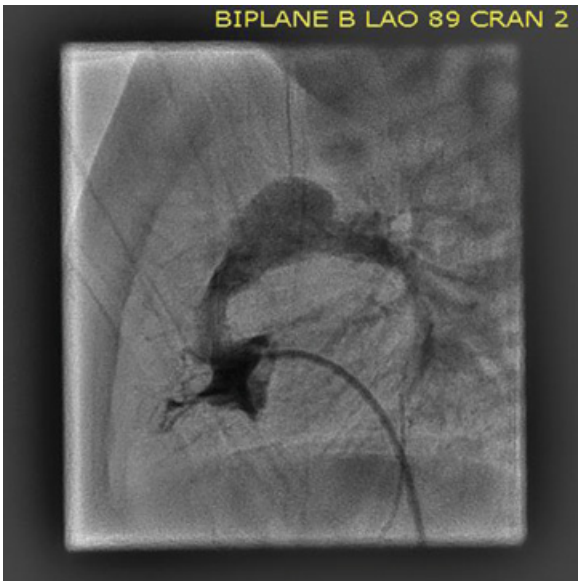


**Figure 6.5 Right ventricle, post-balloon dilation.** An angiogram is performed in the right ventricle in the lateral projection. Improved flow is now noted across the dilated pulmonary valve. Courtesy of Russel Hirsch, MD.

- Presence of tricuspid valve regurgitation, which can be used to estimate RV systolic pressure
- Pulmonary valve gradient to estimate severity of stenosis
- Presence of pulmonary valve insufficiency
- Any other existing valvular pathology

Echocardiographic and direct catheter measurement of PV gradients vary, as they use different measurement techniques. Echocardiography typically visualizes the PV leaflets and annulus. A peak instantaneous Doppler gradient can overestimate the catheter gradient by 20–30 mm Hg, and the mean echocardiographic gradient may underestimate the gradient compared to catheterization data. Continuous wave Doppler through the PV is used to measure the valve gradient. During cardiac catheterization the pressures on each side of the valve are directly transduced. Additionally, the physiologic state (anesthetized or awake, low or high cardiac output) will also impact gradient measurement.

## What are the hemodynamic goals for patients with severe or critical PS?

***RV preload***: RV performance depends on adequate preload. Decreases in central venous pressure will lead to inadequate RV filling and decreased RV stroke volume.

***Heart rate***: Because blood flow across the stenotic pulmonary valve occurs primarily during ventricular systole, heart rate should be maintained within normal limits for age, avoiding significant bradycardia or tachycardia. Arrhythmias are an uncommon but challenging consequence of PS. Maintenance of atrial-ventricular synchrony is key to provide adequate RV filling. In severe PS, tricuspid regurgitation can develop, leading to right atrial distention and increased risk for atrial fibrillation. Additionally, in cases of severe PS severe RVH can increase the risk of subendocardial ischemia. In these cases, a slower heart rate can be considered to improve diastolic time and coronary blood flow.

***Contractility***: With severe or long-standing PS, RVH develops in response to the continued pressure load. Pharmacologic interventions that depress RV function should be avoided as depression of contractile state can lead to RV failure with clinical deterioration. There is particular strain on RV function during induction of anesthesia due to the transition from negative to positive intrathoracic pressure with resultant decreased RV preload and increased afterload.

***Systemic vascular resistance***: Afterload should be maintained to provide adequate coronary perfusion to the hypertrophied RV.

***Pulmonary vascular resistance***: The goals for PVR vary according to the degree of PS and the size and patency of



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the DA. In critical PS, the primary impedance to forward flow is the stenotic pulmonic valve, therefore reducing PVR will do little to increase blood flow to the lungs. However, in patients with mild or moderate PS, major increases in PVR and RV afterload can decrease PBF and lead to RV dysfunction.

In ductal-dependent patients, the ratio of systemic to pulmonary vascular resistance will affect PBF. Therefore, PVR should be kept in the low-to-normal range. Increases in PVR will decrease PBF and decreases in PVR may increase PBF, thereby decreasing both systemic and coronary perfusion. With ductal-dependent PBF, balancing systemic blood flow and PBF by balancing relative resistances requires constant monitoring and management.

**Clinical Pearl**

*In critical PS, the primary impedance to forward flow is the stenotic pulmonic valve; therefore, reducing PVR will do little to increase blood flow to the lungs. However, in patients with mild or moderate pulmonary stenosis, major increases in PVR and RV afterload can decrease PBF and lead to RV dysfunction.*

## What is an appropriate anesthetic induction plan?

Any anesthetic that prioritizes the aforementioned hemodynamic considerations is acceptable. In patients with milder forms of PS an inhalation induction with sevoflurane and oxygen with spontaneous ventilation is an option if the patient does not have intravenous (IV) access. However, in a patient with critical (ductal dependent) PS an IV anesthetic induction is preferable. Crying and agitation during a mask induction may increase PVR, which can negatively impact PBF depending on the degree of PS. Care must be taken to maintain sufficient systemic pressure to ensure coronary perfusion, especially in the setting of RVH. Supplemental oxygen is appropriate during induction for alveolar preoxygenation and to maintain low PVR. Finally, RV workload increases after endotracheal intubation with the transition to positive pressure ventilation.

## How should fluid administration be managed for this patient?

Preload should be maintained in order to optimize RV filling and stroke volume. Care should be taken not to overload the neonatal heart with excess volume.

## What is the most appropriate ventilation strategy for this patient?

Children who have decreased PBF or lower $SpO_2$ as a result of congenital heart disease have poor tolerance for pulmonary vein desaturation due to inadequate ventilation or oxygenation. Ventilation strategies that aim for normocarbia, avoidance of $SpO_2 < 80\%$, and maintenance of functional residual capacity should optimize gas exchange and PVR.

## What is an appropriate anesthetic maintenance plan?

Either inhalation or intravenous medications can be used to maintain anesthesia with attention to hemodynamic goals. Ventricular contractility should be maintained. Tachycardia should be avoided. As there is minimal postprocedural pain, opioids may be used sparingly.

## What potential intraprocedural complications exist?

- ***Arrhythmias*** (although usually transient) can occur due to wire and catheter manipulation.
- ***RV perforation*** can occur, as well as damage to valve leaflets and/or chordae, from catheter manipulation.
- ***Hypothermia*** due to constant flushing of fluid to prevent clot in the sheaths and the cool ambient temperature necessary for the equipment.
- ***Air embolism*** secondary to flushing of the sheaths.
- ***Accumulative blood loss*** from sheath insertion site and continual aspiration of catheters.

**Clinical Pearl**

*Complications from balloon valvuloplasty are not common. However, when they occur, they can progress quickly. Prepare for emergencies by having emergency medications and a defibrillator available.*

## Should this patient be extubated at the end of the procedure?

If the patient is hemodynamically stable after balloon valvuloplasty and does not require inotropic support, extubation in the procedural suite may be considered if all other extubation criteria are met. Since this infant had moderately depressed RV function requiring milrinone



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preprocedure, the length and success of the procedure will guide the decision to proceed toward tracheal extubation.

**Clinical Pearl**

*A rare complication in severely hypertrophied ventricles is "suicide right ventricle." After obstruction is relieved, the now hyperdynamic RV may collapse inward, causing subpulmonary obstruction and decreased RV output. Avoidance of tachycardia is helpful to avoid this complication.*

## What is the postprocedural management for this patient?

Immediately post-procedure, bleeding at the access sites (femoral vessels) and the risk of apnea following general anesthesia in the neonate are the most common complications. Longer term, pulmonary insufficiency or residual pulmonic stenosis are frequently present. It is difficult to have a perfect result with balloon valvuloplasty, and the risk of incurring pulmonary insufficiency must be weighed against the risk of residual PS. A rare complication in severely hypertrophied ventricles is "suicide right ventricle." After obstruction is relieved, the now hyperdynamic RV may collapse inward, causing subpulmonary obstruction and decreased RV output. Avoidance of tachycardia is helpful to avoid this complication.

## What criteria are used to measure the success of the intervention (balloon valvuloplasty)?

Balloon valvuloplasty for PS can either be curative or an initial intervention to improve PBF until a more definitive procedure is planned. Ideal outcomes are little or no residual obstruction in the subvalvular region or valvular level, resolution of RVH over time, decrease in the amount of tricuspid regurgitation, and no R-to-L shunting at the atrial level (PFO). For neonates who were $PGE_1$ dependent prior to the procedure, $PGE_1$ is typically stopped and the infant is monitored in hospital until the DA closes. An acceptable $SpO_2$ indicates that adequate antegrade blood flow through the pulmonary valve is present.

**Clinical Pearl**

*For neonates who were $PGE_1$ dependent prior to the procedure, $PGE_1$ is typically stopped and the infant is monitored in hospital until the DA closes. If $SpO_2$ remains acceptable, adequate antegrade blood flow through the pulmonary valve is present.*

## What other procedures can be done if the catheterization is not successful?

If PBF is insufficient and $SpO_2$ begins to fall despite balloon valvuloplasty, surgical palliation may be indicated. A modified Blalock–Taussig (mBT) shunt can be placed if oxygen saturations cannot be maintained without the DA as a source of PBF. A modified BT shunt utilizes a GORE-TEX® graft (usually 3.5 mm), to connect the right subclavian or innominate artery to the right pulmonary artery, thus providing another source of pulmonary blood flow.

For patients who have sufficient PBF but residual pulmonary valve stenosis or insufficiency, medical management is often employed in anticipation of another catheter-based or surgical procedure.

## Suggested Reading

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