Chapter 14

Critical Aortic Stenosis

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Case Scenario

A 2-day-old male neonate, diagnosed with critical aortic stenosis, is scheduled for emergent balloon valvuloplasty in the cardiac catherization laboratory. He was born at 38 weeks gestational age via spontaneous vaginal delivery. His Apgar scores were 9 at 1 and 5 minutes, and he was admitted to the neonatal intensive care unit due to mild tachypnea and a murmur on auscultation. No obvious dysmorphic features were noted. On day 2 of life he was noted to be more tachypneic and tachycardic, with diminished pulses, cool extremities, and poor capillary refill. A chest radiograph revealed cardiomegaly and pulmonary edema.

Venous lactate is 4 with a blood pH of 7.1. A prostaglandin E_1 infusion and low-dose inotropic support were started.

Transthoracic echocardiogram was remarkable for the following:

- *A dysplastic aortic valve with a mean gradient of 45 mm Hg*
- *A dilated, thick left ventricle with qualitatively moderately reduced function*
- *No patent ductus arteriosus*

Key Objectives

- Define critical aortic stenosis.
- Discuss the options for treatment of a patient with ductal-dependent aortic stenosis.
- Describe the preoperative assessment and intraoperative management of this patient.
- Discuss the potential complications and expected outcomes of balloon valvuloplasty.
- Discuss the postprocedural management and disposition.

Pathophysiology

What is critical aortic stenosis?

Critical aortic stenosis (AS) is defined as the presence of severe aortic valve stenosis with systemic perfusion that is dependent on right ventricular (RV) output through a patent ductus arteriosus (PDA). Mean gradients of >40 mm Hg or peak-to-peak gradients >50 mm Hg are usually reported. As the PDA begins to close, left ventricular function deteriorates, with signs of decreased perfusion to end organs such as the kidneys, gastrointestinal tract, and brain, manifesting as renal failure, necrotizing enterocolitis, and intracerebral bleeds, respectively. Critical AS is not defined by an absolute valve area or gradient because in patients with ventricular dysfunction (either systolic or diastolic) critical AS may exist with larger valve areas and gradients may be underestimated. (See Figure 14.1.)

Clinical Pearl

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What is the pathophysiology of AS in a neonate?

Aortic stenosis in older infants, children and adults leads to left ventricular (LV) pressure overload and progressive LV concentric hypertrophy and failure, with resultant elevation in end-diastolic pressure and pulmonary edema. The hypertrophied LV thus becomes susceptible to subendocardial hypoperfusion and ischemia.

In neonates with critical AS, a PDA provides systemic cardiac output via flow from the RV to the descending aorta via right-to-left (R-to-L) shunting, and thus systemic perfusion is preserved. The degree of LV hypertrophy in these neonates, though not as severe, still exists, and the ventricle may be dilated and poorly contractile. This heart failure is associated with increases in heart rate and LV end-diastolic pressure which can lead to inadequate coronary blood flow and the risk of ischemia. The elevated left atrial (LA) pressure leads to pulmonary edema and pulmonary hypertension with dilation of the RV; in addition,

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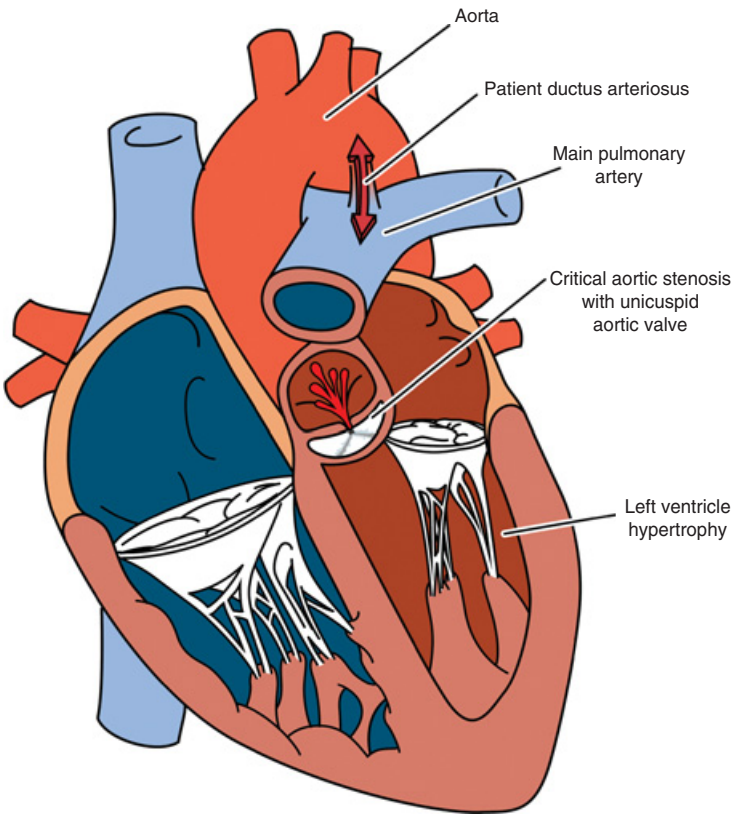

Figure 14.1 Critical aortic stenosis. Drawing by Ryan Moore, MD, and Matt Nelson.

LA hypertension leads to stretching of the foramen ovale and left-to-right (L-to-R) shunting at the atrial level.

What are the options for treatment of a patient with ductal-dependent AS?

The immediate treatment of patients presenting with critical AS is to ensure systemic output and perfusion by maintaining a PDA or reopening a closed ductus. A prostaglandin E_1 (PGE_1) infusion is immediately initiated at birth in patients with a prenatal diagnosis or when critical AS is suspected postnatally. Additional details for medical management are discussed later in this chapter.

Once PGE_1 is initiated and the patient stabilized, then the intervention to relieve the outflow obstruction can be planned. This intervention can be either catheter or surgically based. The decision is based on other associated cardiac factors such as hypoplasia of the LV, mitral valve, or aortic arch.

First, it must be determined whether a biventricular repair is feasible; the question of whether or not the left heart structures are of adequate size to sustain the systemic circulation needs to be answered.

Echocardiography is used to assess all the left-sided structures, namely:

- The size and function of the LV
- The extent of endocardial fibroelastosis
- The size of the aortic annulus and the morphology of the aortic valve
- The size and function of the mitral valve
- The presence of coarctation of the aorta

Once it is determined that a biventricular repair can be undertaken, the next step involves deciding what intervention will be the most beneficial for the individual neonate: surgical aortic valvotomy (SAV) versus balloon aortic valvuloplasty (BAV) in the cardiac catheterization laboratory. Results from the most recent meta-analysis showed that although the rate of reintervention following BAV is higher than following SAV, the survival rates, need for aortic valve replacement, and development of late aortic insufficiency (AI) are equivalent [1]. The final decision will often depend on the patient's weight and clinical condition, local experience, the skills and preference of the surgical and interventional cardiology teams, and overall center outcomes.

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How is SAV performed?

Surgical aortic valvotomy can be done either using a transventricular approach without cardiopulmonary bypass (CPB), or more commonly, via open valvotomy utilizing CPB. In the former approach, after a thoracotomy or sternotomy, the valve is accessed through the LV apex and is serially dilated with balloons or Hegar dilators. Open valvotomy is achieved via sternotomy and may involve just detaching commissures from the aortic wall to reduce doming of the valve (commissurotomy), or a two-stage process. The two-stage process involves thinning the leaflets to improve mobility, followed by commissurotomy.

How is BAV performed?

Balloon aortic valvuloplasty can be approached either from a retrograde or antegrade direction. (See Figure 14.2A and B.) Using a retrograde approach, the balloon sheath is most commonly advanced from the femoral artery, and less commonly from the umbilical artery or carotid artery (usually after surgical cutdown). For the antegrade approach the femoral vein or umbilical vein is cannulated and the catheter advanced through the foramen ovale into the left atrium and ventricle; this approach is easier if the LV is somewhat dilated.

Critical information to be obtained includes:

- Mixed venous saturation
- Aortic valve gradient
- Left ventricular end diastolic pressure (LVEDP)
- Angiograms demonstrating the annulus size; valve anatomy; degree of AI; and LV anatomy, size, and function

The aim of BAV is to adequately relieve the LV outflow tract obstruction (as demonstrated by a reduction in the gradient and/or LA pressure) without causing significant AI, allowing recovery of LV function. The intention is not to dilate the valve annulus but rather to tear the valve leaflets, which are often partially fused, ideally along the fused commissure.

With poor LV function and stable wire position in the aorta, balloon dilatation can be undertaken with minimal chance of the balloon being ejected from the aortic position by a forceful ventricular contraction against the balloon-occluded outflow. If there is adequate LV function, either rapid overdrive pacing or a bolus of adenosine or esmolol, followed by an esmolol infusion, may be administered prior to dilating the valve, to transiently lower cardiac output.

The valve is dilated until the largest reasonable balloon has been inflated across the valve with minimal AI; that is, a balloon usually not exceeding the diameter of the aortic root or a balloon to annulus ratio <0.9. A reduction in the gradient by approximately 50% or to <30 mm Hg is deemed a success. (See Figure 14.3.) If LV function is poor and the gradient is low at baseline, the gradient may

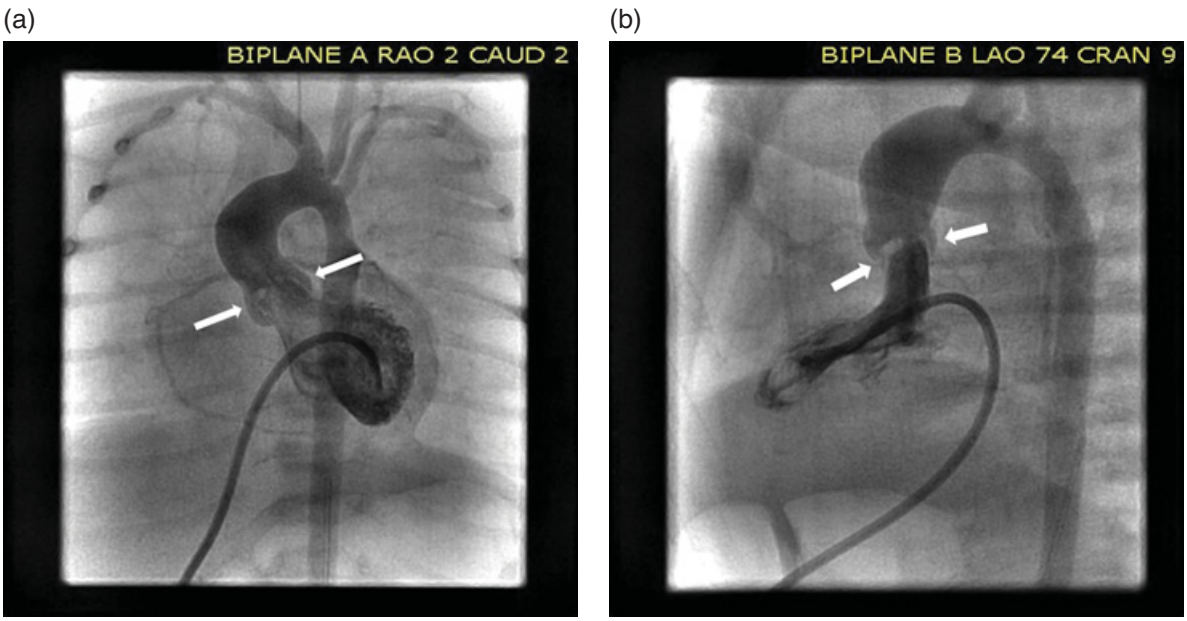

Figure 14.2 Doming aortic valve. An angiogram is performed in the left ventricle in the AP (A) and lateral (B) projections. The thickened and doming aortic valve is noted (arrows). Courtesy of Russel Hirsch, MD.

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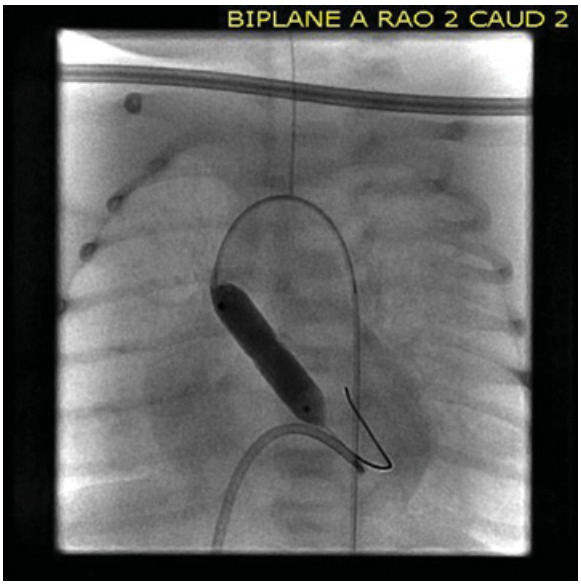

Figure 14.3 Balloon inflation across the aortic valve. Still frame angiogram in the AP projection demonstrates the balloon inflated across the aortic valve. Courtesy of Russel Hirsch, MD.

remain the same after a successful dilation, and it may actually increase days after the procedure once the LV function begins to improve.

Clinical Pearl

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What are the advantages of SAV versus BAV?

Open valvotomy carries the advantage that the surgeon directly visualizes and probes the valve before performing a defined, anatomically sound valvotomy with potentially less resultant AI. However, because this approach requires CPB it carries a greater potential risk to the neonate. Balloon aortic valvuloplasty avoids the need for CPB and sternotomy but may lead to potentially higher rates of cusp disruption with resulting AI that may progress over time, ultimately requiring a surgical repair. However, it is interesting to note that while the development of AI often occurs secondary to BAV, Ewert et al. reported a 49% incidence of AI in patients with untreated AS, suggesting a natural course toward insufficiency [2].

Anesthetic Implications

What key information should be obtained in the preoperative cardiac assessment of a neonate with critical AS?

The aim is to establish whether the neonate has been optimized for the catheterization laboratory: determining what treatment has been implemented, its effectiveness, and any need for further interventions prior to transfer to the catheterization laboratory. A neonate with critical AS and a closed PDA is a medical emergency. Medical management involves establishing intravenous (IV) access and administering PGE_1 to reopen the ductus arteriosus, and utilizing inotropic therapy if necessary to improve LV dysfunction. There should be signs of improvement in systemic perfusion usually within 4 hours. These would include improvements in tachycardia, capillary refill and pulses, decreasing lactate levels, resolution of acidosis, and increased urine output. A preductal/postductal oxygen saturation gradient (SpO_2) suggests R-to-L shunting is occurring through the PDA and is also reassuring. All existing lines should be evaluated for functionality and to confirm that infusions are being delivered to the patient. Dextrose-containing IV fluids should be infusing for this 2-day-old neonate; some centers add calcium gluconate as well. In critically ill infants in whom the duct has closed and remains closed despite PGE_1, surgical and extracorporeal membrane oxygenation (ECMO) backup should be readily available.

Respiratory status should be assessed to decide whether tracheal intubation and ventilatory support, which would decrease the work of breathing and contribute to the overall stabilization of the patient, is warranted prior to transfer to the lab. Often patients on PGE_1 are intubated due to the occurrence of frequent apneic episodes caused by administration of prostaglandins, rather than because of deteriorating pulmonary function. In addition, findings from the chest radiograph, electrocardiogram (ECG), echocardiography, and the most recent lab results should be evaluated. It is also important to ensure the availability of packed red blood cells during procedures in the catheterization laboratory.

Clinical Pearl

A preductal/postductal SpO_2 gradient suggests an open duct; R-to-L shunting through the duct is reassuring.

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What useful information can be ascertained from the chest radiograph, ECG, and echocardiogram?

Chest Radiograph Heart size, the presence or absence of pulmonary edema, appropriate endotracheal tube position, and, if in situ, appropriate positioning of umbilical vein catheter (UVC) and umbilical artery catheter (UAC) should be evaluated. The UVC and UAC should both be above the level of the diaphragm; the UAC should be between thoracic vertebrae 6 and 9 and UVC should be in the inferior vena cava as it enters the right atrium.

Electrocardiogram Right axis deviation and RV hypertrophy (RVH) should be within normal limits for age. In some infants, significant RVH is noted, with tall right precordial R waves and upright T waves in the right precordial leads.

Echocardiogram The following should be assessed: location and nature of the left ventricular outflow tract obstruction (valvular, subvalvular, supravalvular); assessment of the severity of obstruction (peak/mean gradients (PG/MGs); LV wall thickness and chamber size; LV function; patency of the ductus arteriosus and flow through it, atrial level L-to-R shunting through the foramen ovale; and the presence of associated cardiac lesions.

When is an echocardiographic gradient not accurate?

These conditions will result in an inaccurate estimation of the gradient across the aortic valve:

- Presence of a PDA that provides flow from the RV to the descending aorta
- Presence of other obstructive lesions, particularly mitral valve stenosis and coarctation of the aorta
- Depressed LV function

Clinical Pearl

Depressed LV function results in underestimation of the degree of severity of AS.

What should parents be told about anesthetic risk?

Though several scoring systems have been developed to predict the risk of serious adverse events during cardiac catheterization, none specifically address the anesthetic risk. An adverse event is defined as any event leading to mortality, permanent morbidity, need for further interventions, or extended length of hospital stay. The Catheterization Risk Score for Pediatrics (CRISP), created from information obtained from 20 international centers [3], allows points to be assigned according to the patient's age, weight, level of inotropic support/need for extracorporeal membrane oxygenation, systemic illness/organ failure, physiologic status, pre-catheterization diagnosis, procedure type, and the procedure performed. Patients are then placed into one of five CRISP categories and assigned their individual risk for serious adverse events. This risk ranges from a 1% risk for serious adverse events in a CRISP 1 patient to a 36.8% risk for a CRISP 5 patient. Based on a CRISP risk category of 4, this patient would be quoted an incidence of serious adverse events of 14.4%. In considering the risks of general anesthesia, younger age, higher American Society of Anesthesiologists (ASA) status, and emergency procedures have all been reported as risk factors for cardiac arrest during pediatric procedures [4]. Specific to catheterization procedures, infants requiring interventional procedures are at a higher risk for adverse events and hence this patient would fall into the high-risk category. Recent expert consensus statement recommendations have stated that these patients should ideally be managed by a pediatric cardiac anesthesiologist [5]; at a minimum, care should be provided by a pediatric practitioner familiar with the cardiac anatomy and physiology along with the catheterization laboratory environment.

What are the special considerations involved in providing care in the cardiac catheterization laboratory?

The following considerations make the cardiac catheterization laboratory a challenging environment:

- Satellite location, often located away from operating room and intensive care environments
- Suboptimal lighting
- Limited space due to equipment, leading to limited patient access
- Ionizing radiation exposure
- Difficulty with direct communication with other team members
- Increased patient risk for pressure injuries or nerve traction injuries (such as brachial plexus injuries) due to the requirement for arms to be above the head in order to facilitate imaging; meticulous positioning and padding are important
- Difficulty maintaining normothermia in neonates due to both low room temperature (to prevent overheating of cameras) and frequent flushing of catheters and sheaths

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- Volume overload, also due to frequent flushing of catheters and sheaths

What are the anesthetic management goals for this patient?

Anesthetic management is directed toward meeting the increased oxygen requirement of the hypertrophied LV myocardium and optimizing cardiac output.

The goals of management include:

- ***Maintain preload***: A thick LV requires adequate volume loading. However, in the setting of reduced systolic function +/− LV dilation, coupled with preexisting diastolic dysfunction, fluid administration should be judicious.
- ***Maintain afterload***: A thickened LV requires adequate perfusion pressures to minimize sub/endocardial ischemia; vasoconstrictors such as phenylephrine will help with a fall in systemic vascular resistance.
- ***Maintain contractility***: If systolic function becomes reduced, inotropes may become necessary, but should be used judiciously given their propensity to increase heart rate and thus myocardial oxygen requirements.
- ***Maintain heart rate and sinus rhythm***: Avoid bradycardia as neonatal cardiac output is dependent on heart rate given their fixed stroke volume, even in the absence of AS. It should be noted that most infants are tachycardic at baseline, but increases in tachycardia should be avoided, as these will increase oxygen demand and increase ischemic risk in a thick, failing LV.

What are the monitoring requirements for this case?

Standard ASA monitoring is required, but with a few special considerations. As for all catheterization cases where the groin is being accessed, monitoring and vascular access should be on the upper limbs where possible. A preductal SpO_2 monitor will accurately reflect changes in ventilation and respiratory status. Noninvasive blood pressure measurements from a cuff on an arm elevated above the head may underestimate the actual reading and should be correlated with the direct femoral artery pressures that the interventionist can intermittently transduce. End-tidal CO_2 ($ETCO_2$) is a useful surrogate for pulmonary artery blood flow and cardiac output; an abrupt decrease in $ETCO_2$ may be the first indication of a decrease in cardiac output and/or pending disaster.

Clinical Pearl

End-tidal CO_2 is a useful surrogate for pulmonary artery blood flow and cardiac output; it may be the first indication of a decrease in cardiac output and/or pending disaster.

What are special steps in preparation for the case?

In addition to induction drugs and intubation equipment, the following are necessary:

- ***Upper limb IV access*** for volume resuscitation; if the groin is accessed, the interventionist's catheter may obstruct the vessel or the IV may be distal to potential vessel injury.
- ***Blood product*** availability.
- ***Resuscitation drugs*** such as epinephrine, phenylephrine, and calcium gluconate/chloride. Start with low doses as boluses, for example 1 mcg/kg of epinephrine or 1–2 mcg/kg of phenylephrine and repeat as necessary. Calcium gluconate may be used in a bolus of 25 mg/kg. A norepinephrine infusion may become necessary, commencing at 0.02 mcg/kg/minute and titrated to effect.
- ***Defibrillator*** with appropriate size pads/paddle.

What are the most critical points during this case?

As some life-threatening events are preventable, the importance of preparation, anticipation, prevention, and communication has to be emphasized at every step.

Anesthetic induction: An opioid-based anesthetic plus the use of neuromuscular blockade with intubation and mechanical ventilation is generally well tolerated. Regardless of the choice of drug, a slow titration of the induction agent is important given the depressed LV function.

As the catheter courses through the heart, dysrhythmias can frequently occur, including ventricular fibrillation.

Balloon inflation obstructs antegrade blood flow and leads to a drop in cardiac output and blood pressure with a fall in $ETCO_2$. These effects may be less significant if some cardiac output or pulmonary blood flow is maintained via a PDA. However, significant hypotension and bradycardia may be commonly seen at this time. The patient may or may not require pharmacologic intervention to recover.

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What are the possible procedural complications?

- Blood loss during vessel cannulation by the interventional team
- Dysrhythmias
- Valve or heart perforation
- Cardiac arrest from decreased cardiac output during balloon dilatation
- Stroke
- Air embolism
- Acute severe AI (leaflet avulsion, cusp prolapse, disruption of the annulus) requiring urgent surgical valve repair or replacement
- Femoral vein or artery thrombus; heparin should be considered early for lower extremities that show signs of compromised perfusion
- Endocarditis

What are the results of BAV?

Balloon aortic valvuloplasty has been shown to decrease the gradient of AS by 50% but at the expense of a 15% incidence of AI. In the meta-analysis recently published [1], mortality rates following BAV were 11% (95% CI: 8–14). Reintervention following an initial BAV procedure for treatment of AS was 37% (95% CI: 30%–44%) with a mean time to reintervention of 2.7 years (95% CI: 1.4–4.1). This is probably a reflection of the growing preference for a graduated approach to relief of AS, and a trend toward leaving a higher degree of residual stenosis to reduce the risk of significant AI. The incidence of aortic valve replacement following BAV was 20% (95% CI: 17–23). Long-term and mid-term follow-up showed moderate to severe AI was present in 28% of patients (95% CI: 20–37).

Where should the patient be transferred following the procedure?

The patient should be transferred to the appropriate intensive care unit (ICU) for postprocedural monitoring. If stable post-dilatation with normal lactate levels and mixed venous saturations, extubation may be considered. Frequently, however, the patient is transferred to the ICU sedated, intubated, and ventilated. The cardiologist usually decides if PGE_1 can be discontinued in the catheterization laboratory or later during recovery in the ICU, with the decision usually based on how confident he or she is that there will be adequate forward flow through the balloon dilated aortic valve. Post-catheterization chest radiograph and echocardiogram are routinely obtained. Groin access is observed, with a low threshold to start a heparin infusion if no distal pulses are palpable even after the bandage is loosened.

In conclusion, the neonate with critical AS is dependent on a PDA for maintenance of systemic cardiac output, and PGE_1 should be initiated while the management decision regarding catheter versus surgical intervention is being discussed.

References

1. M. T. Saung, C. McCracken, R. Sachdeva, et al. Outcomes following balloon aortic valvuloplasty vs surgical valvotomy in congenital aortic valve stenosis: a meta-analysis. *J Invasive Cardiol* 2019; **31**: E133–44.
2. P. Ewert, H. Bertram, J. Breuer, et al. Balloon valvuloplasty in the treatment of congenital aortic valve stenosis: a retrospective multicenter survey of more than 1000 patients. *Int J Cardiol* 2011; **149**: 182–185.
3. D. G. Nykanen, T. J. Forbes, W. Du, et al. CRISP: catheterization risk score for pediatrics: a report from the congenital cardiac interventional study consortium (CCISC). *Catheter Cardio Inte* 2016; **87**: 302–9.
4. C. Ramamoorthy, C. M. Haberkern, S. M. Bhananker, et al. Anesthesia-related cardiac arrest in children with heart disease: data from the pediatric perioperative cardiac arrest (POCA) registry. *Anesth Analg* 2010; **110**: 1376–82.
5. K. C. Odegard, R. Vincent, R. Baijal, et al. SCAI/CCAS/SPA expert consensus statement for anesthesia and sedation practice: recommendations for patients undergoing diagnostic and therapeutic procedures in the pediatric and congenital cardiac catheterization laboratory. *Catheter Cardio Inte* 2016; **88**: 912–22.

Suggested Reading

Daaboul D. G., Dinardo J. A., and Nasr V. G. Anesthesia for high-risk procedures in the catheterization laboratory. *Pediatric Anesthesia* 2019; **29**: 491–8.

Odegard K. C., Bergersen L., Thiagarajan R., et al. The frequency of cardiac arrests in patients with congenital heart disease undergoing cardiac catheterization. *Anesth Analg* 2014; **118**: 175–82.

Vergnat M., Asfour B., Arenz C., et al. Aortic stenosis of the neonate: a single-center experience. *J Thorac and Cardiovasc Surg* 2019; **157**: 318–26.

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