Chapter 15

Aortic Stenosis

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Case Scenario

A 13-year-old male with new-onset headaches and right-sided weakness presents for craniotomy and resection of an intracranial mass. He has a past medical history of well-controlled asthma, taking albuterol as needed. His past surgical history includes a Ross procedure performed at age 5 years for congenital aortic stenosis. He reports playing basketball daily with his friends and denies cardiac symptoms of exertional dyspnea, chest pain, or syncope. He currently takes no medications. Preoperative vital signs are heart rate 73 beats/minute, blood pressure 110/69 mm Hg, respiratory rate 18 breaths/minute, and SpO_2 97% on room air. A II/VI high-pitched diastolic murmur can be heard on auscultation of the right upper sternal border. Laboratory values are all within normal limits.

His last cardiology visit was 3 months ago, at which time the cardiologist was pleased with his progress and reported to the boy's mother that his echo looked "fine."

Key Objectives

- Understand the criteria for observation versus intervention on a stenotic aortic valve.
- Discuss potential cardiac surgical interventions for aortic stenosis.
- Understand the difference between the Ross procedure and the Ross–Konno procedure.
- Discuss preoperative assessment of this patient.
- Discuss intraoperative management for this patient.
- Discuss postoperative care and airway management strategies.

Pathophysiology

What is aortic stenosis?

Aortic stenosis (AS) is one of the most common congenital cardiac defects, with a bicuspid aortic valve occurring in approximately 2% of the population. Stenosis can occur anywhere along the left ventricular outflow tract (LVOT) and can be classified as subvalvular, valvular, or supravalvular. (See Figure 15.1.) Nearly two-thirds of stenotic LVOT lesions occur at the level of the aortic valve. Congenital valvular AS occurs in approximately 3%–5% of patients with congenital heart disease (CHD) with a male predominance of 3:1. Evidence suggests that the development of AS is influenced by both genetic and environmental factors [1].

What is the anatomy of valvular AS?

Aortic valve stenosis results from narrowing of the orifice size. While a normal aortic valve is tricuspid, an aberrant aortic valve can be unicuspid, bicuspid, or quadricuspid. The unicuspid valve has a single, thickened leaflet resulting in either an eccentric slit-like orifice or a pinhole central orifice, severely limiting flow through the valve during systole. A bicuspid aortic valve occurs from fusion of two of the three aortic valve cusps. With time these cusps undergo myxomatous changes that result in thickened, fibrous leaflets that further contribute to worsening stenosis. The quadricuspid aortic valve is the most rarely seen and is thought to be due to abnormal division of the developing aortic valve leaflets. This lesion is more often associated with aortic regurgitation than with AS. Symptoms associated with a quadricuspid aortic valve generally do not present until adulthood.

A stenotic aortic valve can be associated with other cardiac lesions including patent ductus arteriosus, coarctation of the aorta, and ventricular septal defects. Aortic root dilation frequently occurs with bicuspid aortic valves, even in the absence of clinical stenosis.

What is the physiology of critical valvular AS in the neonate?

At birth, when the low-resistance placenta is removed from circulation, the left ventricle (LV) experiences an increase in afterload. As ventilation begins and the neonate's lungs expand, pulmonary vascular resistance (PVR) decreases,

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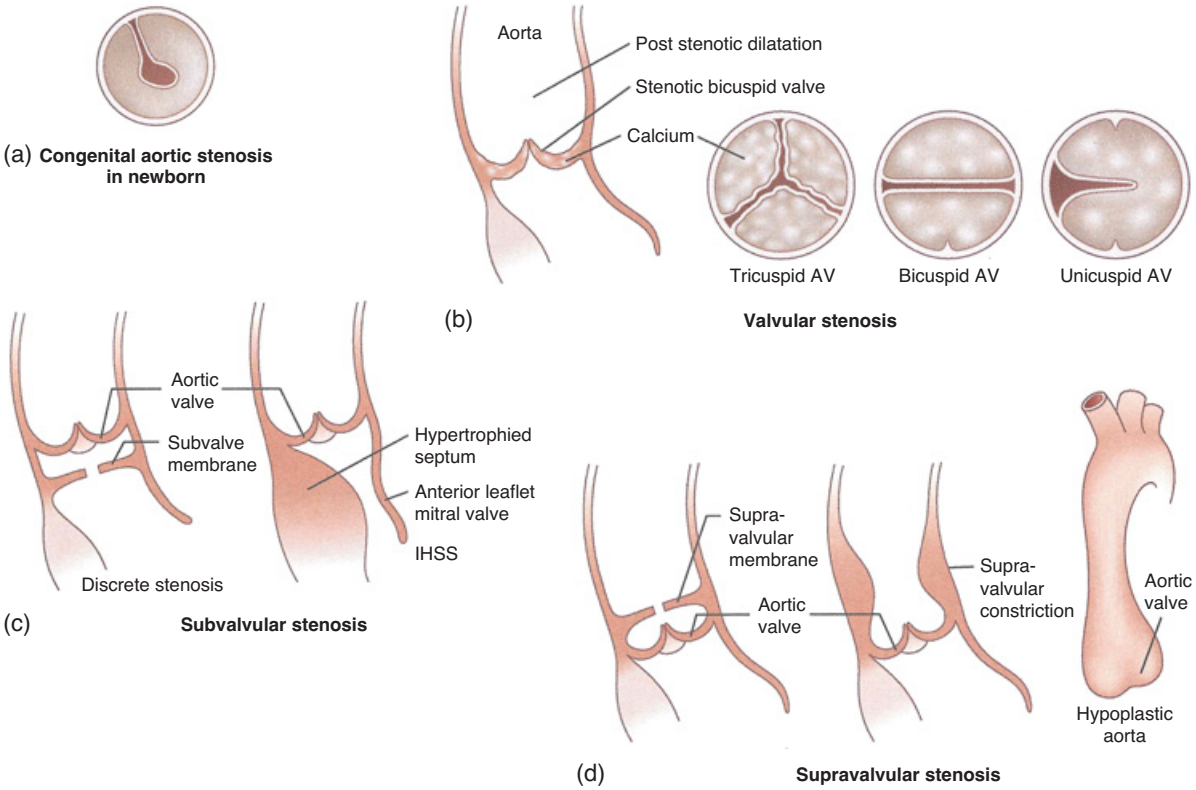

Figure 15.1 Types of aortic stenosis. (**a**) **Congenital aortic stenosis** in the newborn, characterized by hypoplastic aortic valve annulus and malformed unicuspid, unicommissural aortic valve with keyhole appearance to narrow orifice. (**b**) **Valvular aortic stenosis**, characterized by abnormal number of valve cusps or three-cuspid valve. (**c**) **Subvalvular aortic stenosis**, characterized by a thickened ring or collar of dense endocardial fibrous tissue below the level of the valve cusps causing narrowing in the outflow tract from the left ventricle. (**d**) **Supravalvular aortic stenosis**, characterized by narrowing of the ascending aorta just above the aortic valve. From Ottaviani G. and Buja L. M. Congenital heart disease: pathology, natural history, and interventions. In Buja L. M. and Butany J., eds. *Cardiovascular Pathology*, 4th ed. Elsevier, 2015; 611–47. With permission.

resulting in an increase in pulmonary blood flow and increased volume return, or preload, to the left heart. In critical AS, the right ventricle (RV) can generally compensate for compromised LV output in the presence of a patent foramen ovale and patent ductus arteriosus (PDA). Compromised LV output through the stenotic aortic valve is augmented by right to left (R-to-L) shunting through the PDA. As the PDA closes over the first 2–4 days of life the LV is then unable to provide adequate systemic circulation and signs of inadequate systemic circulation and congestive heart failure ensue. The initiation of prostaglandin E_1 (PGE_1) is required for neonates with critical AS to maintain ductal patency, thereby ensuring adequate flow to the systemic circulation. Aortic perfusion distal to the PDA is then largely derived from systemic venous blood, resulting in ***differential cyanosis***, or lower oxygen saturations in the lower extremities compared to the upper extremities. (See Chapter 14.)

Clinical Pearl

While patients with mild AS often remain asymptomatic, newborns with critical AS will become severely ill and present in cardiogenic shock when the PDA closes, requiring immediate intervention. These patients require urgent resuscitation, PGE_1 infusion to maintain the PDA, and intervention to relieve AS.

What is the physiology of valvular AS in children and adolescents?

The obstruction caused by the stenotic aortic valve results in an increase in LV systolic pressure followed by the development of LV wall hypertrophy. This concentric hypertrophy preserves wall stress such that heart rate, stroke volume, and cardiac output are maintained in a normal range. With time, progressive AS will result in

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diastolic dysfunction. The thickened LV myocardium and increased systolic pressure can result in subendocardial ischemia due to a mismatch between oxygen demand and blood supply. In spite of this, children and adolescents with congenital AS often remain asymptomatic. When present, symptoms generally include easy fatigability or dyspnea on exertion. Chest pain and syncope can also be seen in patients with AS and are ominous symptoms of severe stenosis requiring immediate evaluation and intervention.

Clinical Pearl

Congenital aortic valve stenosis is often asymptomatic in infants, children, and adults. When present, symptoms generally include easy fatigability or dyspnea on exertion. Chest pain and syncope associated with AS require immediate evaluation and intervention.

What are the expected echocardiographic findings in patients with AS?

Echocardiography is typically used to evaluate the severity of AS. A bicuspid valve can have a typical "fish mouth" appearance when it opens and closes. Transthoracic echocardiography (TTE) allows visualization of the aortic valve in different planes to assess valve anatomy, annular size, aortic root diameter, and LV function. The anatomy of the ascending aorta, aortic arch, and any other associated cardiac anomalies may also be evaluated.

Measurement of the peak-to-peak transvalvular pressure gradient during cardiac catheterization has historically been used to quantify the degree of aortic valvular stenosis to aid in determining the need for intervention. However, there can be a significant discrepancy between the peak instantaneous Doppler echocardiographic gradient and the peak-to-peak pressure gradient obtained by cardiac catheterization; as peak instantaneous Doppler gradients can overestimate the transvalvular gradient, most clinicians prefer to use continuous wave mean Doppler gradient to guide timing of intervention on a stenotic valve. The mean pressure gradient is relatively consistent between Doppler and catheterization measurements. The following are the guidelines for Stages of Valvular AS from the 2014 American Heart Association/American College of Cardiology Guidelines for the Management of Patients with Valvular Heart Disease [2].

- **Mild stenosis**: Mean gradient <20 mm Hg or aortic V_{max} 2.0–2.9 m/s
- **Moderate stenosis**: Mean gradient 20–39 mm Hg or aortic V_{max} 3.0–3.9 m/s
- **Severe stenosis**: Mean gradient >40 mm Hg or aortic V_{max} >4.0 m/s

What are the criteria for intervention on a stenotic aortic valve?

The decision to intervene on a stenotic aortic valve is based upon the age at presentation, severity of the stenosis, presence of coexisting cardiac lesion(s) and the degree of LV dysfunction. Neonates with critical AS will present with congestive heart failure, respiratory distress, hypoxia, inadequate perfusion, and metabolic acidosis as the PDA closes. Circulatory collapse will ensue if the patient is not aggressively resuscitated and patency of the ductus arteriosus reestablished with PGE_1. Following stabilization, intervention will depend on LV function and size. If LV hypoplasia will not permit a biventricular repair, then the neonate will proceed down the single-ventricle pathway. If the LV is deemed to be of appropriate size and function, most centers will perform a percutaneous balloon valvuloplasty in the cardiac catheterization laboratory, though surgical valvotomy is an option that may be preferred by some.

Patients with mild AS can be monitored with regular echocardiography and require no activity restrictions. Intervention is warranted in those patients who develop either a peak-to-peak gradient >50 mm Hg accompanied by symptoms or electrocardiogram (ECG) changes, or an isolated peak-to-peak gradient >70 mm Hg [3].

Clinical Pearl

Neonates with critical AS can present in cardiogenic shock with metabolic acidosis, respiratory distress, and poor systemic perfusion. These patients require urgent resuscitation, PGE_1 infusion to maintain ductal patency, and intervention to relieve AS.

How is balloon valvuloplasty performed in neonates with critical AS?

The neonate is brought to the cardiac catheterization laboratory and vascular access is obtained. The anterograde approach involves gaining access via the femoral or umbilical veins and then advancing the catheter into the left atrium via the foramen ovale. Alternatively, a retrograde approach can be utilized by cannulating the umbilical, femoral or carotid artery and advancing the catheter to the left side of the heart. The catheter is then positioned across the aortic valve and the balloon inflated, causing disruption of the fused aortic valve leaflets. The

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goal is to relieve stenosis while inflicting minimal aortic insufficiency (AI) from the balloon valvuloplasty. This procedure is palliative, and patients will require intervention later in life for either restenosis of the aortic valve or for AI.

What cardiac surgical options exist for patients with AS?

- ***Repair of the aortic valve*** is the first option. In some centers this is the preferred approach for management of the neonate with critical AS instead of balloon valvuloplasty in the cardiac catheterization laboratory. When possible, surgical repair offers certain advantages. It allows growth, delays aortic valve replacement, and avoids anticoagulation with its associated morbidity. Disadvantages include the need for cardiopulmonary bypass (CPB), potentially at a very early age. Surgical repair may also be complex if associated congenital cardiac abnormalities exist. Studies have demonstrated excellent long-term survival exceeding 95% at 10 years. However, a surgical procedure does not obviate the need for reintervention, with freedom from a second procedure on the aortic valve ranging from 50% to 80% at 10 years [4].
- The ***Ross procedure*** for AS was first described in 1967 by Donald Ross. Described in further detail in the text that follows, the Ross procedure is favored in children and adolescents due to the advantages of potential for growth, freedom from anticoagulation and durability when compared to placement of an aortic homograft. Disadvantages include the fact that the previously healthy right ventricular outflow tract (RVOT) will now have a pulmonary homograft, which is at risk for becoming incompetent and requiring intervention as the patient ages. Additional disadvantages include high early mortality for the Ross procedure when performed on neonates and infants and the high rate of second intervention when performed on young children.
- ***Aortic valve replacement with a mechanical valve*** is the least preferred surgical technique to address AS in pediatric patients. Though this technique carries low early mortality, it is associated with significant morbidity. A mechanical valve requires life-long anticoagulation requiring medication compliance. Patients on anticoagulation cannot participate in contact sports and should avoid becoming pregnant because of teratogenic effects to the fetus. Risks of thromboembolic events and endocarditis exist. Mechanical valves also carry a risk of requiring replacement as children age because the valve does not provide for any growth potential.

Clinical Pearl

The Ross procedure is favored in children and adolescents because of its advantages of potential for growth, freedom from anticoagulation, and durability when compared to placement of an aortic homograft.

What is the Ross procedure?

The Ross procedure involves resecting the stenotic aortic valve and replacing it with the patient's own pulmonary valve (pulmonary autograft). A valved pulmonary homograft is then placed between the RV and pulmonary artery.

After CPB is commenced the diseased aortic valve is removed and coronary buttons dissected. Depending on the comparative sizes of the aortic annulus and the pulmonary autograft the resection can also involve a portion of the aortic valve annulus. This will allow expansion of the annulus to accommodate a larger pulmonary autograft. The pulmonary autograft is then sutured into the LVOT and the coronary buttons are attached. A pulmonary homograft is constructed and placed in the RVOT. (See Figure 15.2.)

What is the Konno procedure?

The Konno operation was described by Konno in 1975 and serves to alleviate LVOT obstruction at the subvalvular, valvular, and supravalvular levels. This is a desirable option when the aortic annulus is very small. Here the incision is made from above the level of the aortic commissures to the aortic annulus and through the RVOT. Another incision is then made across the annulus and extended into the LVOT via the interventricular septum, taking care to avoid the conduction system. A mechanical or bioprosthetic aortic valve is placed in the LVOT, which has been expanded to adequately accommodate the new valve. Both the RV and LV outflow tracts are then augmented with patch closures.

What is the Ross–Konno procedure?

The Konno procedure by itself is rarely utilized but has been combined with the Ross operation to perform a shortened incision through the interventricular septum. The initial dissection of the aortic valve, pulmonary autograft, and coronary buttons are carried out as discussed earlier for the Ross procedure. Again, a portion of the aortic annulus is resected, and an incision made in the interventricular septum. This allows a smaller aortic annulus to accommodate a larger pulmonary autograft. A shorter incision in the interventricular septum than

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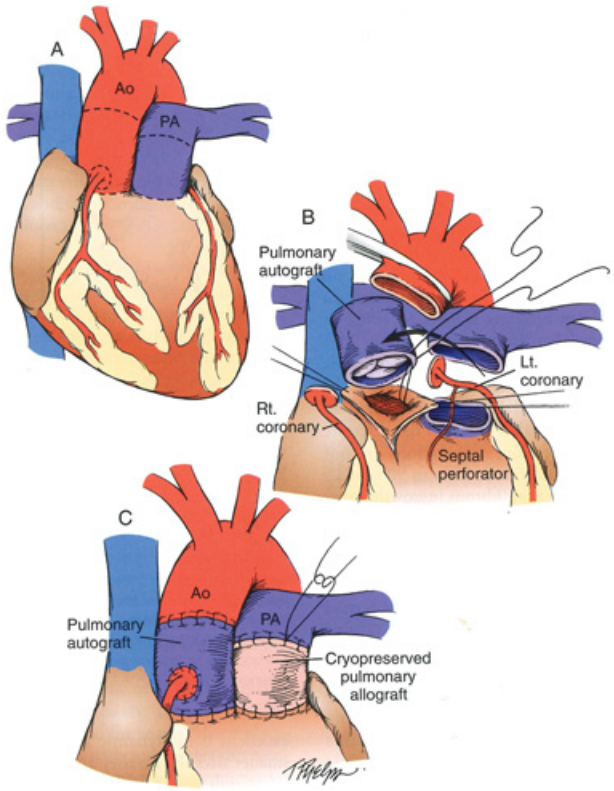

Figure 15.2 **Ross procedure. (A)** Great arteries are transected, aortic sinuses are excised, and coronary arteries are mobilized. **(B)** Pulmonary autograft is excised from the right ventricular outflow tract. The proximal end of the autograft is anastomosed to the annulus. **(C)** The coronary arteries are anastomosed to the pulmonary autograft. Autograft-to-aorta anastomosis is completed and the right ventricular outflow tract is reconstructed usually with a cryopreserved pulmonary allograft. From Alsoufi R., Aljiffry A., and Ungerleider R. M. Left ventricular outflow tract obstruction. In Undgerleider R. M., Meliones J. N., McMillan K. N., et al., eds. *Critical Heart Disease in Infants and Children*, 3rd ed. Elsevier, 2019; 615–31. With permission.

that performed in the classic Konno operation decreases the risk for injury to the conduction system. Coronary buttons are implanted, and a pulmonary homograft placed in the RVOT to complete the Ross–Konno procedure. (See Figure 15.3.)

What are outcomes for patients undergoing the Ross or Ross–Konno procedures?

An analysis of the Society of Thoracic Surgery, Congenital Heart Surgery Database from 2000 to 2009 evaluating outcomes in neonates and infants undergoing aortic valve replacement by the Ross or Ross–Konno procedures found in-hospital mortality to be 40% [5]. This is likely because neonates present with more complex left-sided structural abnormalities and significant LV dysfunction. Long term outcomes in older patients have been favorable, with >75% freedom from re-operation at 10 years. One study also associated the type of surgical reintervention with age at the time of the Ross/Ross–Konno intervention, with infants more likely needing later surgery on the RVOT and children and adolescents needing subsequent reintervention on their LVOT [6].

Anesthetic Implications

What valvular disease might be anticipated in this patient?

After the Ross procedure patients can develop AI. The exact physiology is unclear, but it is thought to be due to either dilation of the neoaortic root or degeneration of the pulmonary cusps of the autograft. Despite concerns about the Ross procedure creating two diseased valves from one dysfunctional valve, observational data do not support this. Pulmonary homograft dysfunction is much less common than AI after the Ross procedure.

What are the physical findings of AS? AI?

Vital signs are generally normal in patients with AS. The classic murmur associated with AS is a systolic crescendo–decrescendo murmur heard best at the right upper sternal border, though in younger children it can be auscultated at the left upper sternal border. The murmur will often radiate to the bilateral carotid arteries.

The murmur of AI is classically a diastolic high-pitched, decrescendo murmur heard best at the right upper sternal

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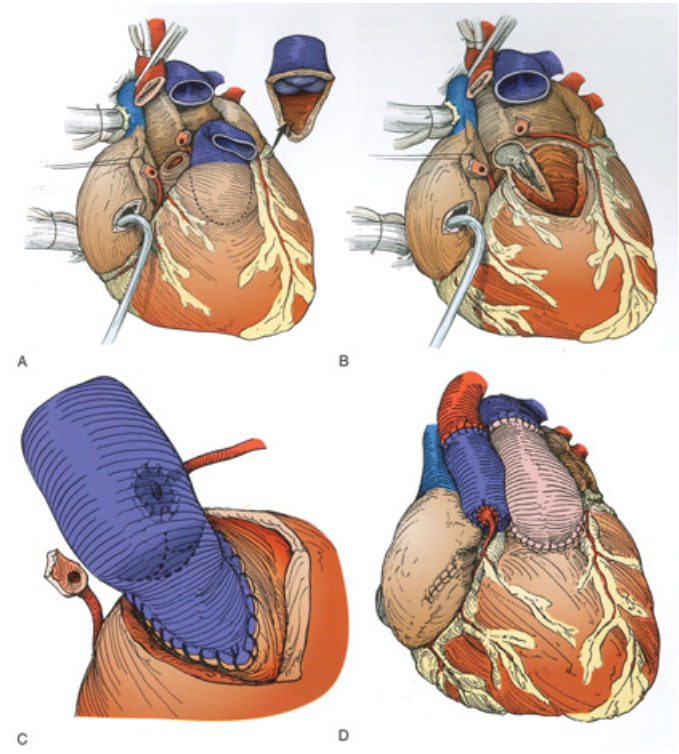

Figure 15.3 **Ross–Konno procedure. (A)** The pulmonary valve is harvested after the aorta has been divided and the coronary arteries removed as buttons. **(B)** An incision is made across the infundibular septum. **(C)** The pulmonary autograft is anastomosed to the aortic root using the infundibular muscle to repair the interventricular septal defect. The coronary arteries are placed into the neoaorta. **(D)** The procedure is completed with a pulmonary homograft to repair the right ventricular outflow tract. From Alsoufi R., Aljiffry A., and Ungerleider R. M. Left ventricular outflow tract obstruction. In Undgerleider R. M., Meliones J. N., McMillan K. N., et al., eds. *Critical Heart Disease in Infants and Children*, 3rd ed. Elsevier, 2019; 615–31. With permission.

border. Severe aortic regurgitation can cause a mid-to-late diastolic rumbling murmur heard best at the apex of the heart. This lesion is also associated with wide pulse pressures and bounding pulses when chronic in nature.

Clinical Pearl

The murmur of AS is a systolic crescendo–decrescendo murmur radiating to the carotids while that of aortic regurgitation is a diastolic high-pitched, decrescendo murmur.

How should this patient be evaluated preoperatively?

A thorough history and physical exam will need to be performed before his craniotomy. While discussing his history, particular attention should be paid to his functional status, activity level, and the presence and/or nature of cardiac symptoms such as shortness of breath, chest pain, or syncope. His medication list should be reviewed and medication compliance confirmed. Current vital signs and laboratory values, particularly his hematocrit, should be assessed.

As part of the preoperative evaluation the most recent pediatric cardiology consultation should be reviewed, and the cardiologist made aware of the impending surgery. A recent ECG and echocardiogram should also be reviewed. When evaluating the echocardiogram, it is important to note the status of the neoaortic valve, noting any significant regurgitation or restenosis. The RVOT should also be evaluated for any pulmonary homograft dysfunction.

The preoperative evaluation should include a discussion about fasting status. If restenosis of the neoaortic valve has occurred, particular care should be taken to avoid prolonged fasting and perioperative hypovolemia. In general patients should be encouraged to eat solid food up until 8 hours before their scheduled procedure and to drink clear liquids up until 2 hours before their procedure. Patients with pre-existing intravenous (IV) access may receive maintenance fluids until the time of surgery.

Clinical Pearl

The most recent pediatric cardiology consultation should be reviewed, and the cardiologist made aware of the impending surgery. Results of the last echocardiographic exam should be thoroughly reviewed, with attention to the status of the RVOT and the pulmonary homograft as well as the neoaortic valve, noting significant regurgitation or restenosis.

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Is there a need for invasive monitoring during the craniotomy?

Craniotomies require close blood pressure monitoring due to the potential for volume shifts with the administration of mannitol. Given this, and the patient's history of palliative cardiac surgery, invasive arterial pressure monitoring would be appropriate during this procedure. A central line allows monitoring of central venous pressure and administration of inotropes if needed. In this patient, the placement of a central line would be best in the subclavian or femoral veins because of the nature of the surgical procedure. However, in the absence of a central line, volume status can also be accurately measured by monitoring urine output, closely following vital signs trends and evaluating arterial blood gas values for base deficits.

What perioperative management concerns exist for this patient?

Placement of a preoperative IV catheter, if not already present, would be appropriate for this patient. In the setting of a neoaortic valve with restenosis, goals of an anesthetic IV induction include avoidance of tachycardia and maintenance of appropriate preload and afterload. Similarly, intraoperative anesthetic goals should include maintenance of appropriate preload, afterload, and contractility while avoiding tachycardia and arrhythmias. In the presence of aortic regurgitation, a higher heart rate is better tolerated. A total IV anesthetic with propofol and narcotic infusions would allow for hemodynamic stability and optimize cerebral oxygen supply and demand. Euvolemia should be maintained and volume replacement should be guided by urine output and arterial blood gas measurements of base deficit. The patient should be monitored closely for anemia; if necessary, packed red blood cells should be available for transfusion.

Clinical Pearl

Anesthetic goals in patients with AS include maintenance of preload, afterload, and contractility with avoidance of tachycardia, arrhythmias, and hypovolemia.

What are the postoperative management considerations for this patient?

In the absence of significant respiratory disease or intraoperative complications, extubation should be considered in the operating room environment. It is important to avoid significant tachycardia on emergence and to ensure adequate analgesia. Postoperative care should take place in an intensive care unit.

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