22 Congenital Aortic Stenosis

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Congenital aortic stenosis (AS) is one of the five most common congenital heart lesions and accounts for 5-10% of all congenital heart defects. It can be classified as: valvar (70%), subvalvar (10-20%), supravalvar (5-10%), or mixed (8%) (Figure 22-1). The importance of AS is disproportionate to its incidence because it is either critical and requires emergent intervention or it is progressive and requires frequent follow-up throughout life, regardless of type of intervention. This chapter will focus on valvar AS. Subvalvar and supravalvar AS are described in Chapters 23 and 24, respectively.

Pathophysiology and Clinical Presentation

Only 2% of congenitally abnormal aortic valves develop clinically significant AS or AI by adolescence. Valvar degeneration and dysfunction of congenitally abnormal valves, however, is progressive over time and a higher percentage of patients require intervention later in life. Approximately 20% of patients have associated cardiac lesions including aortic coarctation, VSD, and PDA.

In patients with congenital AS, approximately one third will have tricuspid valves, two thirds will have bicuspid valves (true bicuspid or functionally bicuspid due to fusion of a commissure), 8% will have monocuspid valves, and rare patients will have quadricuspid valves. Long-term outcomes and response to therapy correlates with valvar morphology. The presence of a bicuspid aortic valve (which is thought to be present in as much as 4% of the general population) may also be associated with aortic root dilation and the development of an ascending aortic aneurysm ("bicuspid aortopathy"). The incidence and severity of aortic dilation does not directly correlate with the severity of AS.

Patients with severe AS (approximately 10% of patients with congenital AS) may present in the newborn period or during infancy. Severe AS is well tolerated in utero from the standpoint of cardiac output as the RV maintains adequate cardiac output through the arterial duct. However, in severe cases, LV function may be severely depressed in fetal life. The management of fetal critical AS is highly controversial and beyond the scope of this chapter, other than to note that detection of severe AS and depressed LV function should alert the postnatal management team of the potential for an unstable newborn requiring urgent attention. In postnatal life, as the PDA closes, patients may develop symptoms of low cardiac output and CHF. If the degree of stenosis is severe with depressed LV function, systemic perfusion is compromised, leading to rapidly progressive cardiogenic shock after PDA closure. Pre- and postductal saturations (differential saturations with decreased SaO_2 in the lower extremities) are useful in detecting ductal dependency. In critically ill newborns, PGE infusion to maintain or reestablish ductal patency may be a life-saving temporizing measure.

In older patients, as AS progresses, symptoms including easy fatigability and exertional fatigue occur in about 30% patients. Angina, syncope, and sudden death after exercise are rare. Patients with less severe forms of AS will initially be asymptomatic. However, AS tends to be progressive over time and close follow-up is required.

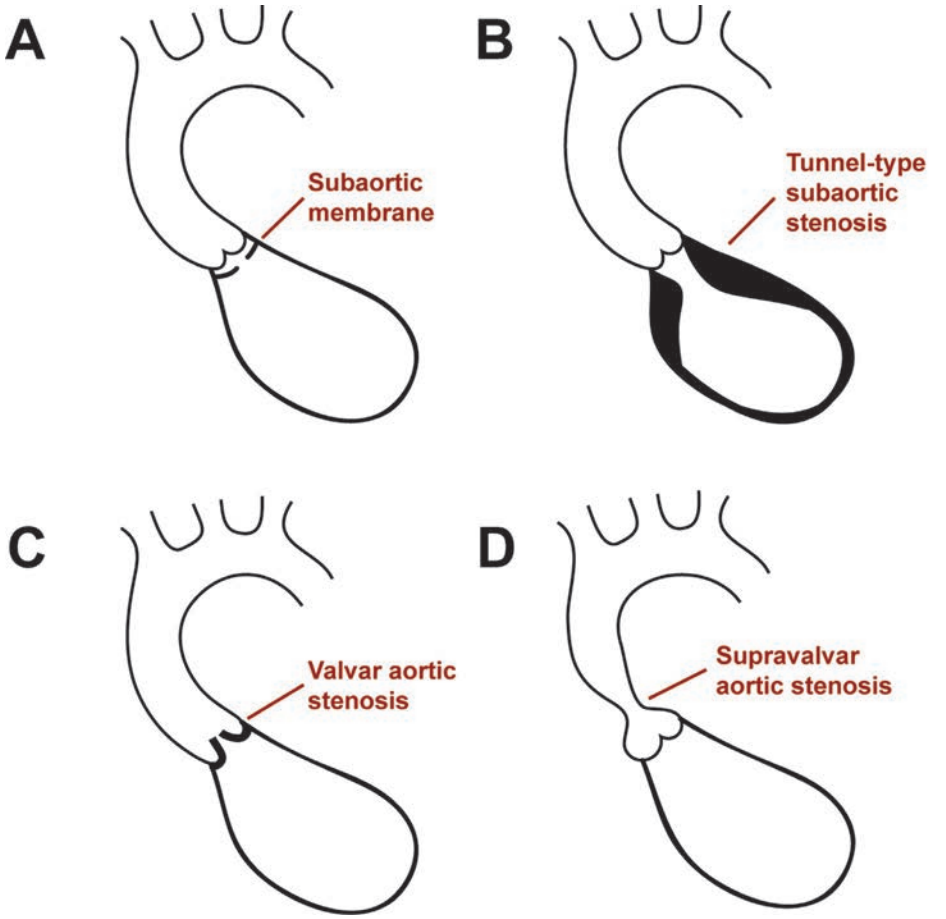

Figure 22-1. Different levels of left ventricular outflow tract obstruction including: A) fixed subaortic obstruction due to a membrane, B) diffuse tunnel-type subaortic stenosis, C) valvar aortic stenosis, and D) supravalvar aortic stenosis.

On physical exam, patients with critical AS may present in cardiogenic shock with weak pulses and mottled appearance. In general, patients with AS will have an increased apical impulse with a harsh systolic ejection murmur at the right upper sternal border radiating into both carotids. A suprasternal thrill with a systolic ejection click is consistent with valvar stenosis. A precordial thrill and an S_4 gallop may be present in severe AS.

Diagnosis

- **ECG.** LVH is often present, although the ECG may be normal even with significant AS. LVH with strain pattern (ST depression and T-wave inversion in lateral leads) at baseline or with exercise is concerning for severe AS.

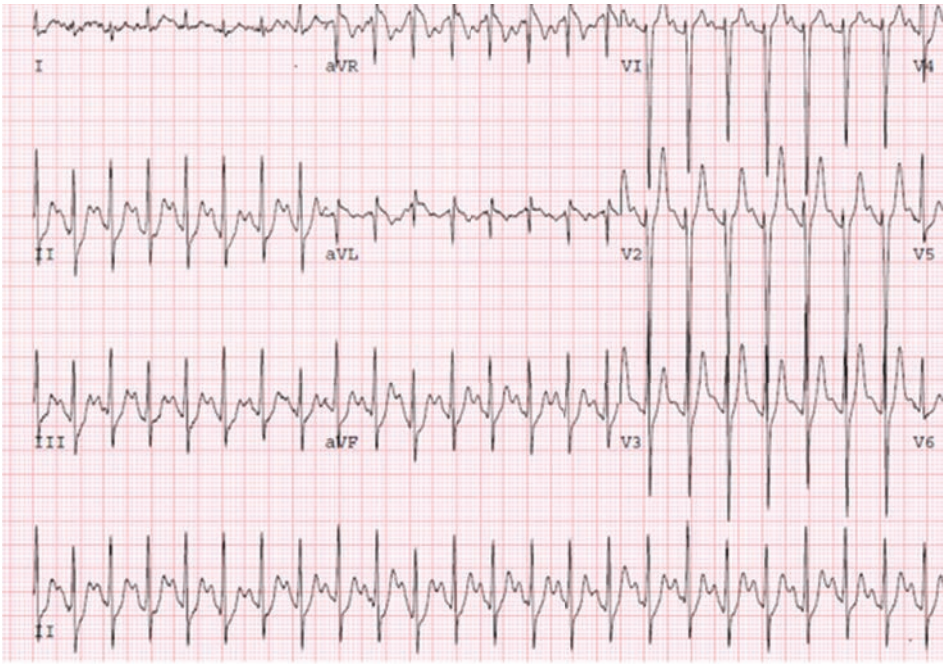

Figure 22-2. ECG showing significant ST-segment changes with exercise.

- **CXR.** Nonspecific. There may be a rounded apex, prominent aortic shadow, LA dilation, and posterior displacement on lateral views.
- **Echocardiogram.** Useful for definitive diagnosis, evaluating valvar morphology, presence of myocardial fibrosis, and associated defects. In utero fetal echocardiography may also be used to diagnose significant AS. Tachycardia, increased contractility, or cardiac output status (including anemia) may falsely increase the observed gradients across the aortic valve. Echocardiography can help define the severity of AS (Table 22-1).
- **Exercise stress test.** Useful to determine exercise capacity in moderate to severe AS. ST-segment changes with exercise are indicative of ischemia (Figure 22-2).

Medical Management

Patients with critical AS presenting as newborns in cardiogenic shock require initiation of PGE (to support the systemic circulation), intubation, inotropic support, and urgent intervention.

Patients with less severe forms of congenital AS may be followed closely. For patients with mild-moderate AS (mean gradient <30 mmHg), follow-up is recommended every 2 years. Patients with moderate-severe AS (mean gradient >30 mmHg), should be followed every year. Life-long follow-up is recommended. Antibiotic prophylaxis is

currently not routinely recommended (Nishimura et al. 2014), although opinions on this issue are highly variable.

Based on ACC/AHA guidelines, patients with AS may need to be exercise-restricted as follows (Bonow et al. 2015):

- Patients with mild AS and normal maximal exercise response should not be exercise-restricted
- Patients with moderate AS can participate in low and moderate static or dynamic competitive sports if normal exercise-tolerance testing
- Asymptomatic patients with severe AS should not participate in competitive sports
- Symptomatic patients with AS should not participate in competitive sports

Indications / Timing for Intervention

AS is progressive over time and valve replacement is the ultimate therapy for most patients with significant AS. Timing for intervention and outcomes correlate with initial gradient and degree of valvar dysplasia. Balloon valvuloplasty and surgical repair may delay timing of valve replacement considerably. Many patients develop progressive AI, in both native and postintervention states.

Newborns and infants that present with critical AS and cardiogenic shock require urgent intervention in the form of aortic balloon valvuloplasty or surgical valvotomy. Asymptomatic patients are put forward for intervention when peak gradient is >50 mmHg. For patients that are symptomatic, show ischemic or repolarization ECG changes at rest or exercise, or are planning to play competitive sports or become pregnant, a peak gradient of >40 mmHg is generally used as an indication for intervention.

Catheter-Based Intervention

In many centers, balloon valvuloplasty (Figure 22-3) is the initial line of therapy for isolated valvar AS, although data are conflicting concerning the question of whether this therapy is as effective and durable as open surgical valvotomy. Freedom from reintervention after balloon dilation is approximately 50% at 10 years. Recovery is typically rapid; most patients with normal ventricular function are eligible for discharge within

Table 22-1. Severity of AS based on echocardiographic and cardiac catheterization findings.

	Peak Velocity[a]	Peak Instantaneous Gradient[a]	Mean Gradient[a]	Peak-to-peak Gradient[b]	Valve Area[c]
Mild	3 m/s	<36 mmHg	<25 mmHg	<30 mmHg	>1.5 cm^2
Moderate	3-4 m/s	36-64 mmHg	25-40 mmHg	30-50 mmHg	1-1.5 cm^2
Severe	>4 m/s	>64 mmHg	>40 mmHg	>50 mmHg	<1 cm^2

[a] By echocardiography.

[b] By cardiac catheterization.

[c] In adolescents and adults, normal valve area is 3-4 cm^2. For children, normal valve area is 2 cm^2/m^2 and severe AS is 0.6 cm^2/m^2.

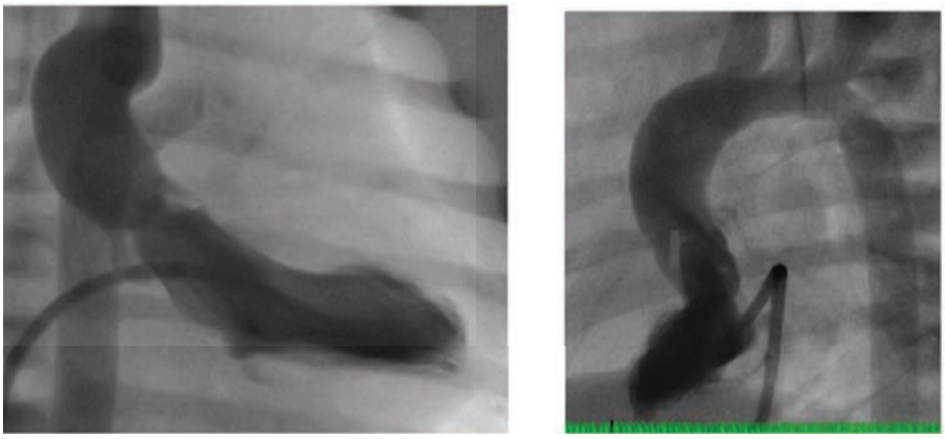

Figure 22-3. Angiogram showing a typically thickened and dysplastic aortic valve with a small effective orifice.

24 hours. Although high risk, balloon valvuloplasty may be better tolerated than surgical valvotomy in neonates with critical AS or with depressed ventricular function, and can delay surgical intervention.

Surgical Repair

Aortic Valve Repair

Open surgical valvotomy is very effective for primary AS and several series suggest this mode of treatment may be associated with superior durability and less AI when compared to balloon valvuloplasty. However, at present, balloon valvuloplasty is more frequently offered in most centers, including TCH.

Primary surgical repair is most frequently offered for patients with associated cardiac lesions, progressive AI, or recurrent AS not adequately treated with balloon valvuloplasty. Techniques for valve repair include accurate commissurotomy with leaflet debulking. In cases where leaflet tissue is limited, cusp extensions may be performed with the use of autologous pericardium or commercially available bovine pericardium. Complex aortic valve repair methodologies have been developed to address even the most malformed aortic valves, including the so-called "monocusp" valves. These techniques essentially amount to an on-table construction of a tissue valve within the patient's native aortic root and represent a formidable geometric and technical challenge. Many units have reported early success with these methods, however, these repairs do not tend to be as durable as replacement, particularly in small children (see aortic valve replacement below).

Secondary valve repair may also be considered after a previous balloon valvuloplasty. In patients undergoing balloon valvuloplasty for primary AS, the valve does not consistently tear along commissural lines; on the contrary, the leaflet is often torn not only in

a radial fashion, but it may be avulsed from the annulus in a circumferential fashion. As such, reparative strategies after previous dilations are typically complex. A central limiting feature in all forms of repair for severe AS is the annular dimension. Neither balloon valvuloplasty or isolated reparative surgery can effectively overcome severe annular hypoplasia. It is imprudent for the surgeon to accept a hypoplastic annulus in performing a primary open valvotomy for AS. In this setting, AS will recur rapidly if the annulus is not dealt with appropriately (see annulus enlarging techniques below).

TCH experience with aortic valve repairs (1995-2011) (Khan et al. 2013)
Number of procedures: 97
Median age 2.6 years (1-18 years)
Median weight 11.6 kg (21-110 kg)
Perioperative mortality: 2%
5-year freedom-from-reintervention or death:
- Simple repair: 84%
- Complex repair: 61%

TCH experience with aortic valve replacements (1995-2011) (Khan et al. 2013)
Number of procedures: 188
Median age 8.3 years (4 days – 18 years), median weight 25.4 kg (2-109 kg):
Types of aortic valve replacements:
- Autograft (Ross): 68 (36%)
- Homograft: 74 (39%)
- Mechanical: 36 (19%)
- Bioprosthetic: 10 (6%)

Perioperative mortality: 3%
5-year freedom-from-reintervention or death:
- Autograft (Ross): 91%
- Homograft: 52%
- Mechanical: 95%

Aortic Valve Replacement (AVR)

The entire topic of AVR in children is complex and highly debated. Our own data (Khan et al. 2013), as well as those from other centers performing large numbers of AVRs in children, appear conclusive in demonstrating that the pulmonary autograft aortic root replacement (Ross operation) is the superior option for most children. Nonetheless, there are instances in which other options can and should be considered. In the setting of profound LV dysfunction, an expedient operation with limited myocardial ischemia may be better tolerated than the more lengthy and technically demanding Ross procedure. As such, viable alternatives for children include homograft aortic root replacement or mechanical AVR. The latter is less attractive in most patients due to the necessity and thereby risk of chronic anticoagulation. Homograft root replacements offer the patient the option of an active lifestyle without anticoagulation, but the implants degenerate at unpredictable rates.

The Ross operation offers the patient the option of a more durable, native semilunar valve in the aortic position. The operation is predicated on a satisfactory pulmonary valve, and we have not offered the Ross operation if we find a bicuspid pulmonary valve or multiple deep-cusp fenestrations (we have seen one case of a quadricusp pulmonary valve). The pulmonary valve complex is harvested as a muscular sleeve of the RV infundibulum. It is important to remember that the pulmonary valve does not have a fibrous "annulus" and it is therefore subject to dilation. As such, many surgeons reinforce the implanted autograft muscular apron with a Dacron® strip or some other material to "fix" the dimension of the annulus. Our opinion is that the full root method, which includes direct reimplantation of the coronary arteries is superior to the sub-coronary technique.

In cases where the aortic annulus is hypoplastic, the Ross-Konno method is preferred to achieve an adequate annular dimension. The root enlarging portion of the operation is actually facilitated by the harvest of the autograft. Once the autograft is procured, the surgeon has a direct view of the ventricular septum, which is then incised for several millimeters to increase the annular dimension. The defect in the septum is repaired with a Dacron® or pericardial patch and the autograft is implanted appropriately.

As noted previously, patients with AS will often have a dilated ascending aorta and in some patients, it is frankly aneurysmal. In the latter setting, the ascending aorta will have to be replaced, frequently including extending the replacement up into the aortic arch ("hemi-arch" replacement) using a Dacron® tube graft. In cases of an ectatic ascending aorta, some surgeons favor an anterior aortoplasty to "tailor" the size of the ascending aorta to match the autograft, while others do not. In either case, the autograft will still have the tendency to dilate at the level of the aortic anastomosis and over time, this will lead to progressive dilation at the level of the autograft valve pillars and subsequent AI. It is therefore prudent to reinforce the anastomosis between the ascending aorta and autograft with a strip of Teflon™ felt or some other durable material.

Postoperative Management

Postoperative management of patients undergoing surgery for AS (either valve repair or replacement) is rather generic in assuring adequate preload (volume), carefully controlled afterload (to control BP in the setting of multiple aortic suturelines and to facilitate LV function), and judicious inotropic and lusotropic support. In many cases, early extubation facilitates management, but in patients who have gone into surgery with profound LV dysfunction, particularly newborns with critical AS, sedation and mechanical ventilation may be useful in reducing overall body oxygen consumption and thereby myocardial demand.

In patients with longstanding AS with associated severe LVH, the LV will often appear "hyperdynamic" after successful intervention or surgery. In these situations, volume resuscitation is paramount. As tachycardia may be poorly tolerated, the judicious addition of a short-acting intravenous beta-blocker (esmolol) may facilitate perioperative management.

Complications

Complications after both balloon valvuloplasty and open surgery for AS should be infrequent. In very small infants, vascular access may be challenging and thereby, compromise of distal perfusion when the femoral approach is used may occur. This typically is responsive to anticoagulation therapy and expectant management, although there have been rare cases of profound extremity compromise that have required surgical revascularization (in very small babies this may require the assistance of surgeons experienced in microvascular methods). In some very small babies, an open carotid artery cutdown has been used for cath access and in these cases, the carotid artery should be repaired primarily following the procedure. While rare, balloon valvuloplasty in small, critically ill newborns has been associated with aortic rupture

and sudden cardiac arrest. It is therefore mandatory that the surgical team be notified and on standby when a critical newborn is taken for an urgent or emergent balloon valvuloplasty, where emergent surgery or ECMO support may be needed.

Complications after surgery should also be infrequent, but may be significant. For the Ross operation, the autograft must be harvested from the RV infundibulum and as such, the left main, LAD, and first septal perforating coronary arteries may be at risk. Furthermore, in performing the root replacement, the coronary ostia must be reimplanted into the autograft root. These considerations all emphasize the need for careful ongoing assessment for the potential of myocardial ischemia in the perioperative period.

Other potential surgical complications include failure of the valvuloplasty or autograft (persistent AS or AI), recurrent AS, complete AV block, myocardial dysfunction, and bleeding, to name a few. All patients face the lifetime risk of need of repeat surgical or catheter intervention and thereby require a consistent and longitudinal follow-up management plan.

Suggested Readings

Bonow RO, Nishimura RA, Thompson PD, et al. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 5: Valvular heart disease. Circulation 2015;132:e292-e297.

Khan MS, Samayoa AX, Chen DW, et al. Contemporary experience with surgical treatment of aortic valve disease in children. J Thorac Cardiovasc Surg 2013;146:512-521.

Morales DL, Carberry KE, Balentine C, et al. Selective application of the pediatric Ross procedure minimizes autograft failure. Congenit Heart Dis 2008;3:404-410.

Nishimura RA, Otto CM, Bonow RO, et al. 2014 AHA/ACC guideline for the management of patients with valvular heart disease. J Thorac Cardiovasc Surg 2014;148:e1-e132.