

Subaortic Stenosis

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Subaortic stenosis (SAS) makes up a relatively small portion of all CHD but it can be one of the most vexing problems faced by providers because no consensus exists to guide management in a large subset of these patients. While SAS can occur in association with other cardiac lesions (e.g., malalignment VSD, hypertrophic cardiomyopathy), this chapter focuses on SAS in an otherwise normal heart.

Pathophysiology and Clinical Presentation

SAS is usually classified into discrete (i.e., “subaortic membrane”) and tunnel-type SAS. The former is much more common and is characterized by a partially or completely circumferential fibrous ridge in the LVOT. Tunnel-type SAS describes a long (1-3 cm) segment of circumferential fibromuscular narrowing of the LVOT that may be associated with hypoplasia of the aortic valve annulus.

The pathophysiology of SAS is unclear. However, it is believed that the turbulence and shear stress caused by abnormal flow patterns incite an inflammatory reaction that is felt to be responsible for occurrence and progression of disease. An acute angulation of the LVOT or associated anomalies (e.g., VSD) may be partly responsible for the turbulent flow. It is also possible that some patients may be more prone to develop fibrosis and scarring, increasing the incidence and recurrence of SAS. The fibrous membrane that develops as a consequence of the fibrotic reaction tends to grow (worsening the stenosis) and attach to surrounding structures including the mitral and aortic valves (causing AI).

In children, progression is associated with higher gradient at diagnosis (mean gradient >30 mmHg), aortic valve thickening at diagnosis, shorter distance from the membrane to the aortic valve, and membrane attachment to the mitral valve. In addition to stenosis, some degree of AI may be present, likely the result of the membrane encroaching on the valve or the effect of a high-velocity turbulent jet damaging the valve over time. Long-term preservation of the aortic valve is an important consideration when determining management. LVH, initially a compensatory response to decrease wall stress, becomes pathologic over time and can lead to heart failure.

Patients with SAS are usually young (<10 years old) and asymptomatic. A murmur is the most common reason for patients to seek medical attention. It is typically a harsh, high-frequency, crescendo-decrescendo murmur (without a click) at the left midsternal border that radiates to the carotids. An early diastolic decrescendo murmur indicates AI. An S_4 gallop would suggest severe obstruction and diastolic dysfunction. Even with severe obstruction, symptoms are rare in children. They include decreased endurance, chest pain, and presyncope or syncope.

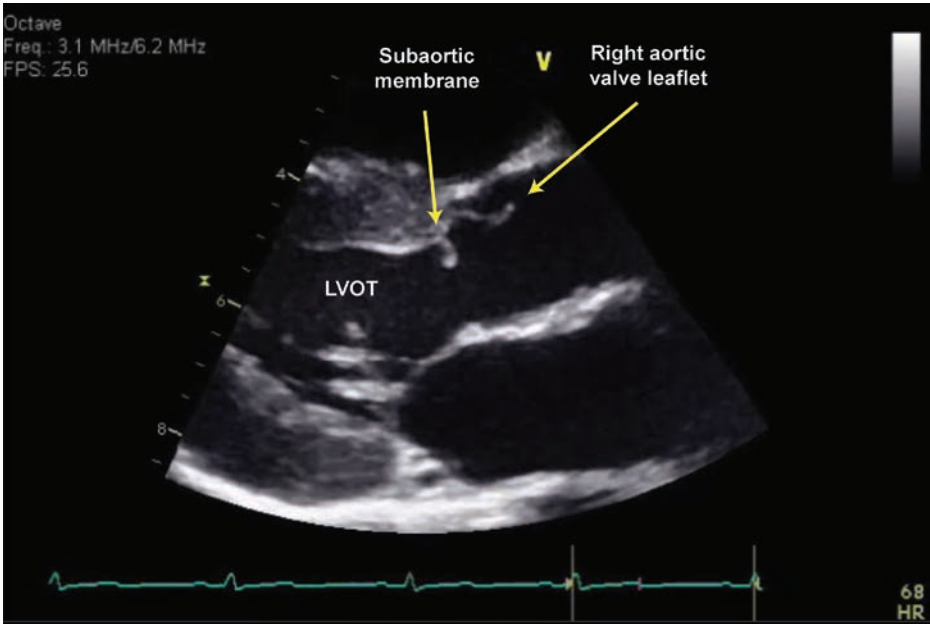


Figure 23-1. Parasternal long-axis echocardiographic image of a discrete subaortic membrane. Image courtesy of Dr. Josh Kailin, www.pedecho.org.

Diagnosis

- **ECG.** The degree of LVH correlates poorly with the degree of obstruction. ST depression in lateral leads represents significant myocardial strain or ischemia regardless of the gradient, and is a potential indication for surgery.
- **CXR.** Usually normal.
- **Echocardiography (Figure 23-1).** In discrete SAS, parasternal long-axis demonstrates a ridge extending from the septum into the LVOT. One should look closely for a corresponding tissue ledge arising from the anterior leaflet of the mitral valve. In tunnel-type SAS, the LVOT will appear diffusely narrow. Aliasing of the color Doppler signal in this view reveals the level of obstruction and can demonstrate AI. Parasternal short-axis 2D view demonstrates the aortic valve architecture and leaflet mobility (sometimes limited by an encroaching membrane), and color will localize the insufficiency. Anterior images in the apical view disclose the location and extent of the obstruction, and the angle of interrogation with color Doppler from this view is favorable in estimating the degree of obstruction and insufficiency. The subcostal view adds to the findings from the apical view and often provides even better resolution in both 2D and color. The importance of slow, complete sweeps through the LVOT cannot be overstated with careful characterization of the LVOT, membrane, and mitral and aortic valves with 2D, color, and spectral Doppler. The suprasternal view often produces the highest measured velocities and should be performed routinely. When choosing an echocardiographic probe, a PEDOF (Pulsed

Echo Doppler Flow-velocity meter) probe will provide the most accurate velocity. When determining the severity of obstruction, one must be careful to consider the angle of interrogation as well as confounding lesions (VSD, contamination from MR). The mean gradient is felt to most closely approximate the catheter gradient in this lesion, and this value should be trended over time.

- **Exercise stress testing (EST).** Indicated when considering participation in competitive sports.
- **Cardiac catheterization.** Can measure the gradient with great accuracy and is sometimes used to confirm or supplant echocardiographic estimations, particularly when imaging is poor or intervention is being considered. However, most patients do not require catheterization prior to intervention.

Indications / Timing of Intervention

SAS is often progressive in children (less so in adults), but the rate of progression is highly variable. At least annual visits and echocardiograms are warranted to monitor for progression; more often when progression occurs. Patients with symptoms (ischemic chest pain or decreased exercise performance), severe obstruction, or LV dysfunction should be referred for surgical intervention. At TCH, patients with AI, especially if progressive, are usually considered for intervention in order to avoid further injury of the aortic valve. The optimal timing of surgery for asymptomatic patients without AI is controversial. It has been suggested that a mean gradient of 30 mmHg or peak gradient of 40 mmHg is a reasonable indication for intervention. However, no consensus recommendation exists, and one should carefully consider the decision on a case-by-case basis.

When an athlete is considering highly static or dynamic competitive sports participation, eligibility guidelines will factor into the decision-making process, as would EST results. Desire for pregnancy influences management in females of childbearing age. It is the recurrence rate of these membranes (up to 20%) and need for reoperation (15%) that necessitate a thoughtful approach.

Surgical Repair

The procedure is performed via median sternotomy and under CPB with aorto-bicaval cannulation. After cardioplegic arrest and venting of the left heart via the atrial septum or the right upper pulmonary vein, an oblique aortotomy is performed. The aortic valve is carefully retracted to examine the LVOT. The extent of involvement of the subaortic membrane is usually more extensive than appreciated on echocardiography (Booth et al. 2010), and it is common for the membrane to involve one or more leaflets of the aortic valve.

The resection is started by incising the membrane in the LVOT, at a level below the nadir of the right coronary cusp. The area rightward of this site contains the conduction tissue and should be addressed carefully. Depending on the thickness of the ventricular septum and the LVOT angle, the incision may extend for a few millimeters into the muscular septum as there is a suggestion that performing a myectomy at the time of SAS resection may reduce recurrence by rectifying the LVOT angle. Resection of the membrane

is then continued leftward towards the mitral valve. Additional muscular septum may be excised just anterior to the mitral valve. The membrane is then peeled off the mitral valve and, carefully, off the area near the conduction system. When the membrane involves the aortic valve, it is peeled

off with care not to injure the valve. Some membranes are intimately attached to the valve and peeling the membrane may be challenging.

If the SAS is more diffuse or tunnel-like, a more extensive resection into the body of the ventricle or an alternative surgical strategy (e.g., modified Konno operation or Ross-Konno operation if the aortic valve annulus is small) may be required.

TCH experience on subaortic stenosis (1995-2018)

(Binsalamah et al. 2019)

Number of patients: 84

Median age: 6.6 years

5-year freedom from reintervention: 87%

Postoperative Management

Patients are usually extubated in the OR or shortly thereafter. Postoperative management of patients with SAS is focused on limiting the hyperdynamic response after SAS resection. Patients may have significant LVH and diastolic dysfunction. As such, an esmolol infusion may be beneficial to reduce heart rate in some patients. A left-bundle-branch block may be present if the subaortic resection involved an extensive septal myectomy. If present, there may be some additional systolic dysfunction.

Complications

SAS resection is usually a low-morbidity operation. However, potential complications include the creation of a VSD from an extensive myectomy (which should be identified and repaired at the time of surgery) and the presence of complete heart block from injury to the AV conduction system. Heart block may be temporary but if normal conduction is not recovered by postoperative day 10, a pacemaker may be indicated (see Chapter 75).

Long-Term Follow-Up

The incidence of reintervention after SAS resection at TCH is approximately 15%. As such, relatively close follow-up after surgical intervention is mandatory. Risk factors for membrane recurrence are an increased peak gradient at the time of diagnosis (>60 mmHg), membrane involvement of the aortic valve, a short distance between the membrane and the valve, and early age at diagnosis. Performing a myectomy at the time of SAS resection seems to be protective for development of recurrence. Data are mixed as to whether early intervention curbs the progression of AI, in particular for patients with more advanced degrees of insufficiency.

Suggested Readings

Binsalamah ZM, Spigel ZA, Zhu H, et al. Risk factors for reoperation after isolated subaortic membrane resection. Presented at the 55th Annual Meeting of the Society of Thoracic Surgeons, San Diego, CA, 2019.

Booth JH, Bryant R, Powers SC, et al. Transthoracic echocardiography does not reliably predict involvement of the aortic valve in patients with a discrete subaortic shelf. *Cardiol Young* 2010;20:284-289.

Ezon DS. Fixed subaortic stenosis: a clinical dilemma for clinicians and patients. *Congenit Heart Dis* 2013;8:450-456.