7

Subaortic Stenosis

7.1 Morphology

The presence of an abnormal tissue beneath the aortic valve or abnormal morphology of the left ventricular outflow tract leads to the left ventricular outflow tract (LVOT) obstruction. The obstruction of subaortic stenosis (SAS) is of four types:

Type 1: Membranous: A discrete membrane consisting of endocardial fold and fibrous tissue. (see Figure 7.1)

Type 2: Muscular: A fibromuscular ridge consisting of a thickened membrane with a muscular base at the crest of the interventricular septum.

The types 1 & 2 account for 70-80% of LVOT obstruction due to the presence of abnormal tissue 0.5 cm to 1.5 cm beneath the aortic valve and involves a variable extent of the LVOT.

Type 3: Tunnel: Diffuse, fibromuscular, and tunnel-like narrowing of the LVOT.

Type 4: Anomalous mitral valve: Obstructing accessory or anomalous mitral valve tissue with an anomalous chordal attachment obstructing the LVOT.

The types 3 and 4 are rare causes of LVOT obstruction.

Certain anatomic features of LVOT, genetically determined abnormal myocardial cell proliferation, and morphologic changes due to abnormal flow patterns predispose the children to develop SAS. Anatomic characteristics that may promote chronic flow alterations in the LVOT include:

a) Long, narrow LVOT.

b) Steep (> 130 ٩) aorto-ventricular septal angle.

c) Increased mitral-aortic separation.

d) Exaggerated aortic override.

A focal myocardial abnormality similar to that found in hypertrophic cardiomyopathy may also play a role.

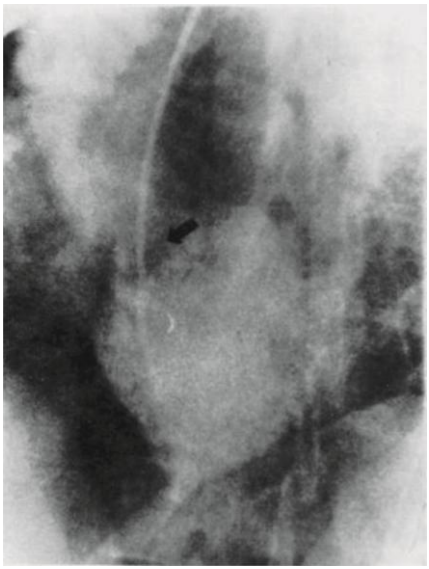

Figure 7.1 *Left ventricular angiogram in left anterior oblique projection demonstrates membranous subaortic obstruction (arrow head) beneath the aortic valve. Reproduced from: Younis G A,, Awdeh M, Benrey J, Price C E, Cooley A D. Discrete Membranous Subaortic Stenosis: Postoperative reversion of Aortic valve Echocardiogram to Normal. Cardiovasc Dis. 1976; 3(4): 424-429. Request for reproduction: Contact: http://www.ncbi.nlm.nih.gov/pmc/about/copyright/.*

7.2 Pathophysiology

During the natural course of SAS, abnormalities of aortic and mitral valve leaflets occur due to tethering effect by encroaching fibroblastic tissue of the membranous and fibromuscular obstruction. Significant obstruction to ejection results in concentric LV (left ventricular) hypertrophy, producing excessive thickness of the ventricular septum. This effect leads to a cycle of further obstruction and localized fibro muscular growth. SAS has variable but unpredictable rates of progression in children whereas the rate of progression in adults is slow.

Heart failure: If it presents early in the course of SAS, it is usually from associated congenital heart defects. The isolated SAS in children produces heart failure, only occasionally, with well-preserved systolic function and maintenance of cardiac output until severe obstruction develops.

Aortic regurgitation: It develops in the majority of patients during the course of SAS due to thickening of valve cusps, asymmetrical post-stenotic dilatation of the ascending aorta, and repetitive trauma from the high-velocity jet of blood flow through the site of stenosis.

The severity of regurgitation increases with an increasing LVOT pressure gradient, reflecting progressive aortic valve damage by the high-velocity jet of blood. Aortic regurgitation persists even after the SAS is corrected. Therefore, significant and progressive aortic regurgitation requires aortic valve repair or replacement at the time of SAS repair. SAS may also recur even after an adequate surgical resection.

Myocardial Ischemia: Aortic regurgitation adds volume overload to an already pressure-overloaded LV (left ventricle). The decrease in aortic diastolic pressure leads to diminished coronary perfusion and in combination with increased myocardial oxygen demand from pressure and volume overloaded ventricle, it predisposes the left ventricular myocardium to severe ischemic injury.

7.3 Treatment

7.3.1 Medical

It has no role as most of the patients are asymptomatic. Surgical intervention is often required, at some point, in the clinical course of the disease to relieve LVOT obstruction, preferably, before heart failure develops. If the disease progresses to the point that heart failure or clinically significant LV dysfunction develops, anti-congestive therapy is indicated until surgery can be performed.

The systemic vasodilators and angiotensin-converting enzyme [ACE] inhibitors are, however, contraindicated.

7.3.2 Surgical Treatment

(I) Surgical Intervention & Indications for Surgery

1. LVOT catheter peak-to-peak gradient /or doppler mean gradients of < 30 mm Hg and without LV hypertrophy:

   Treatment is by medical follow-up only. (The rationale for early surgery in this subset, based on rapid disease progression and eventual aortic valve injury, is outweighed by the problem of high postoperative incidence of recurrence, late reoperation, and development of aortic regurgitation even after successful early relief of the obstruction).

2. LVOT catheter peak-to-peak gradient /or doppler mean gradients of 50 mm Hg or more: Treatment: Surgical intervention.

3. Catheter LVOT catheter peak-to-peak /or doppler mean gradients of 30-50 mm Hg: Treatment: Surgical intervention if:

   - Symptomatic with angina, syncope, or dyspnea on exertion (class I).
   - Asymptomatic but develop ST/T-wave changes on EKG over left precordium at rest or with exercise (class I), or have a case for progressive course, such as an older age at diagnosis and tunnel-like SAS (it should have an early surgical intervention). Prevention of aortic regurgitation is usually not an indication for surgical intervention in those with mild LVOT obstruction.

(II) Catheter Intervention

Percutaneous balloon dilation of a discrete lesion can substantially reduce the LVOT pressure gradient that is relatively brief. Therefore, balloon dilation is not indicated.

7.4 Operative Techniques

7.4.1 Discrete Membranous and Fibromuscular SAS

Complete resection with myotomy, with or without myomectomy is performed through an aortotomy. Significant aortic regurgitation may require aortic valve repair or replacement.

7.4.2 Tunnel-type SAS with Narrow LV-aortic Junction

Aortoventriculoplasty (Konno procedure) may be required. It involves excision and replacement of the aortic valve with a prosthesis, patch augmentation of ventricular septum to enlarge the LVOT, and pericardial patch closure of the right ventriculotomy.

7.4.3 Tunnel-type SAS with Normal LV-aortic Junction and Aortic Valve or Recurrent SAS

Modified Konno procedure without aortic valve replacement may be performed.

7.4.4 Complex SAS or Tunnel Stenosis

Modified Konno with aortic root replacement by using a prosthetic aortic valve, an aortic valve allograft, or a pulmonary valve auto graft (Ross-Konno) is a prescribed procedure (see Chapter 8).

7.5 Postoperative Management

The postoperative course following resection of subaortic stenosis is usually uneventful. The hospital stay averages 5 to 7 days.

7.5.1 Hemodynamic Management

Systemic hypertension may be encountered postoperatively requiring management.

7.5.2 Invasive Monitors

Arterial, CVP (central venous pressure), and LA (left atrial) catheters.

7.5.3 Vasoactive Drug Infusions

Nitroprusside and/or esmolol are often used in the early postoperative period (see Section I Chapters 4 & 16).

Switch to captopril or enalapril to treat hypertension later in the postoperative course.

7.5.4 AV Conduction Abnormalities

AV (atrioventricular) dissociation or complete heart block can occur.

Atrioventricular pacing should be available at the bed side.

7.5.5 Postoperative Bleeding

It is uncommon.