8 Aortic Stenosis and Other Types of Left Ventricular Outflow Tract Obstruction or Anomalies

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- Aortic valve stenosis (valve AS)
- Bicuspid aortic valve
- Subvalve aortic stenosis
- Shone complex
- Supravalve aortic stenosis
- Elastin arteriopathy
- LV-aorta tunnel

Normal Aortic Valve

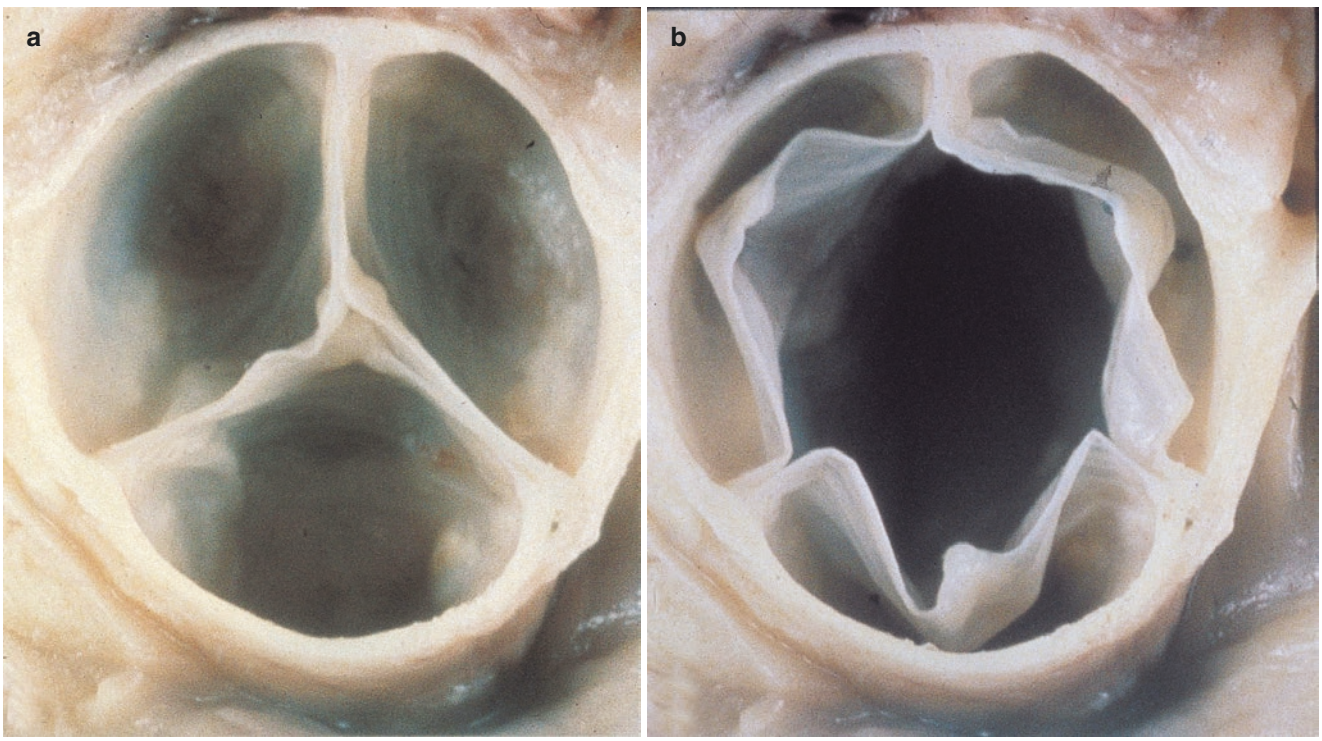

Fig. 8.1 Specimen picture of a normal aortic valve with three leaflets during diastole (**a**) and systole (**b**). (These pictures are provided by Dr. William D. Edwards, Mayo Clinic, USA)

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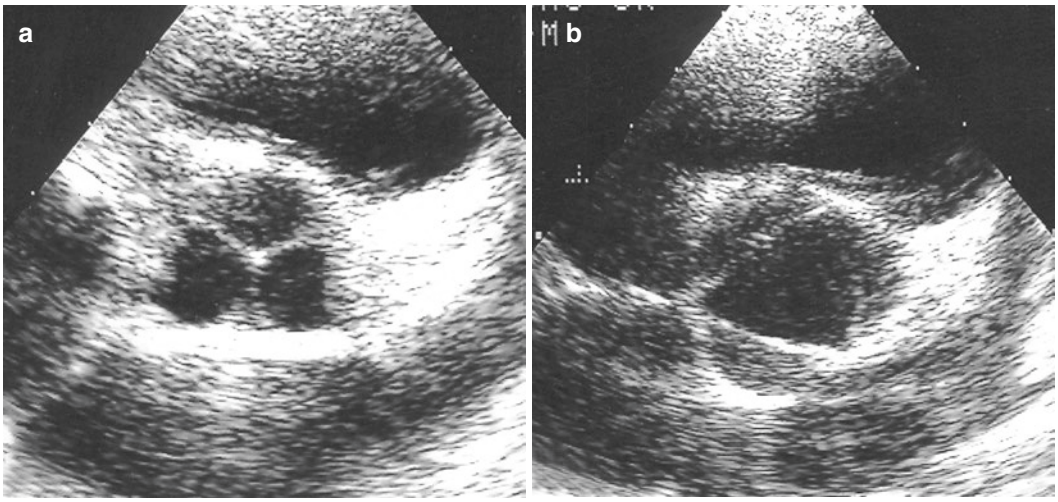

Fig. 8.2 Normal aortic valve with three leaflets (Y-shape) on 2D echocardiogram during diastole (**a**) and systole (**b**)

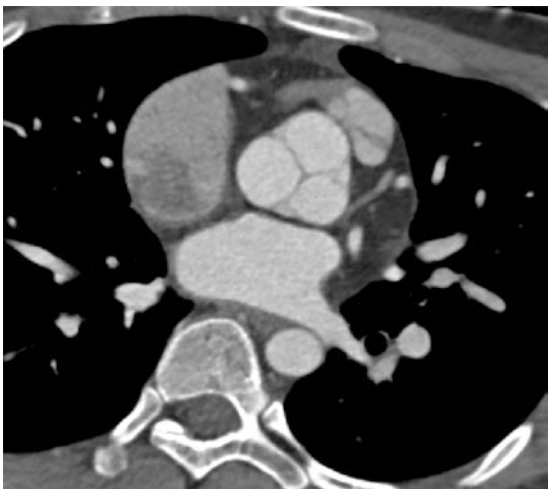

Fig. 8.3 Normal aortic valve on CT image showing three leaflets

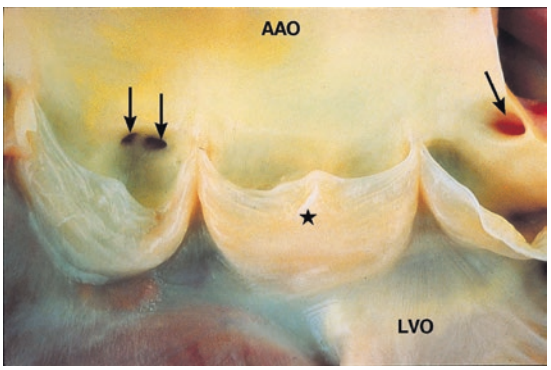

Fig. 8.4 Specimen picture of a normal aortic valve seen with the anterior wall of the ascending aorta open. Arrows indicate coronary orifices from the right sinus (two openings) and left sinus (one orifice). The noncoronary cusp has no coronary opening. (This picture is provided by Dr. William D. Edwards, Mayo Clinic, USA)

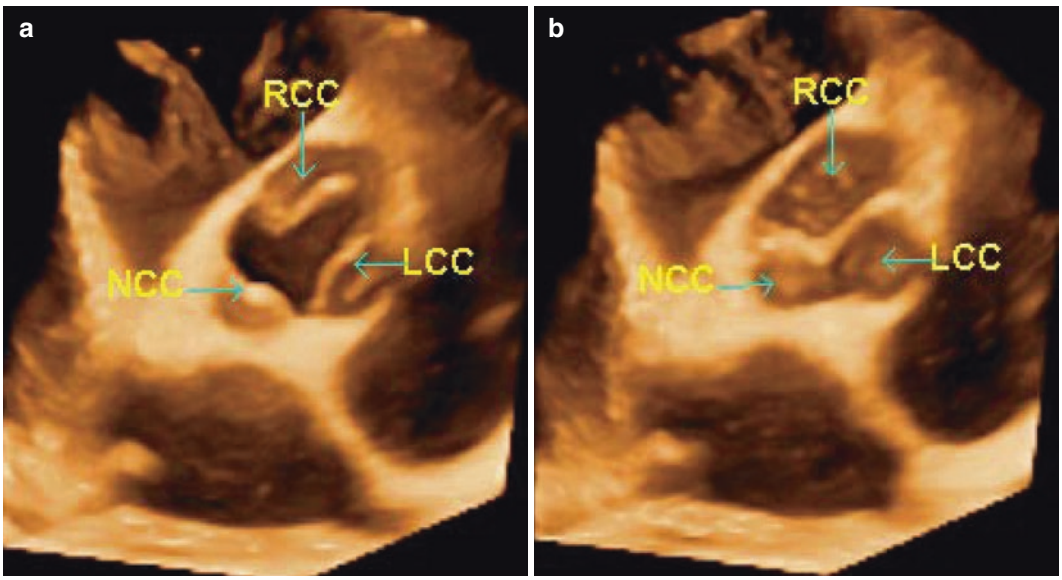

Fig. 8.5 Normal aortic valve on 3D echocardiogram during systole (**a**) and diastole (**b**)

Bicuspid Aortic Valve

The bicuspid aortic valve is the most common congenital heart disease. The incidence is reported to be as high as 1–2% in the (Western) population. It is a highly heritable condition, with transmission likely to be autosomal dominant. Although most of the bicuspid aortic valve without stenosis functions normally during early life, it can progress to valve stenosis and/or regurgitation or aortic root dilatation and dissection later in life.

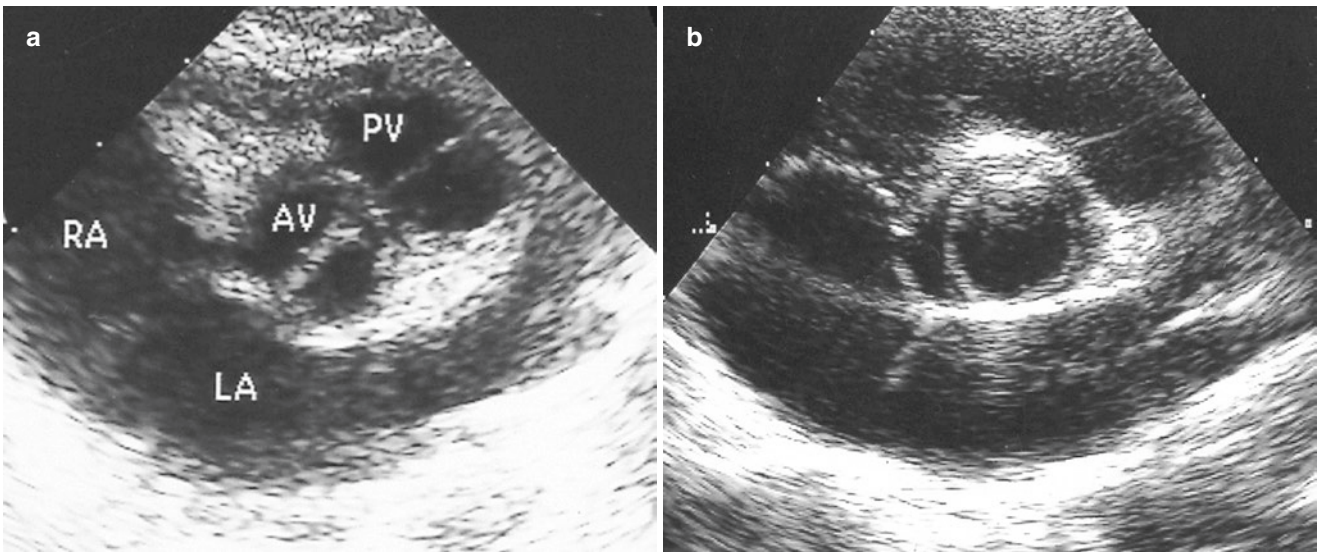

Fig. 8.6 Echocardiographic images (parasternal short axis view) of a bicuspid aortic valve without stenosis during diastole (**a**) and systole (**b**)

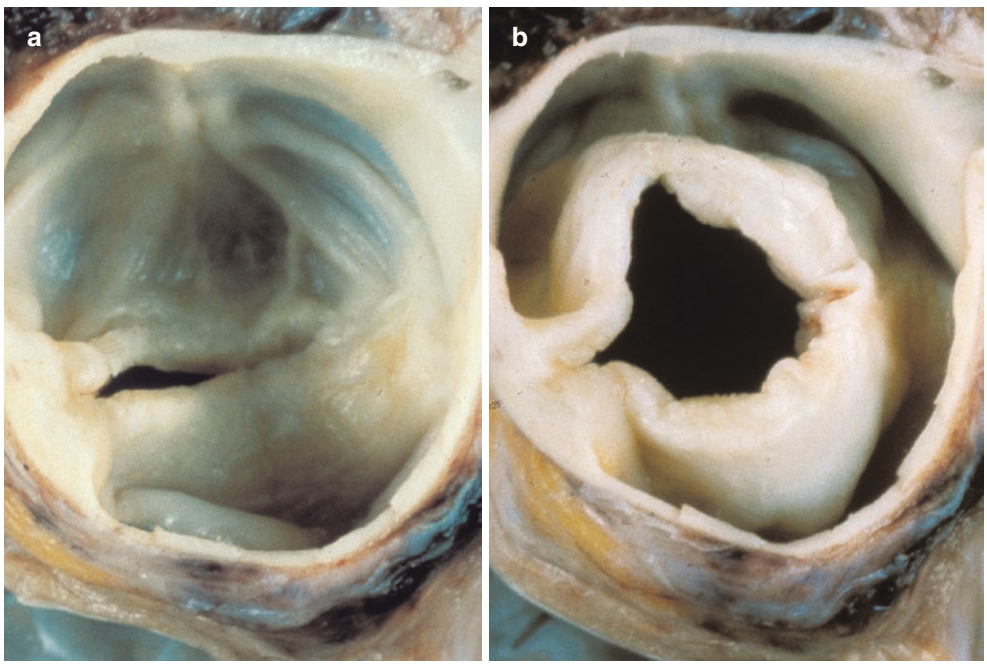

Fig. 8.7 Specimen picture of a bicuspid and very thickened aortic valve during diastole (**a**) and systole (**b**). (This picture is provided by Dr. William D. Edwards, Mayo Clinic, USA)

Aortic Valve Stenosis (Valve AS)

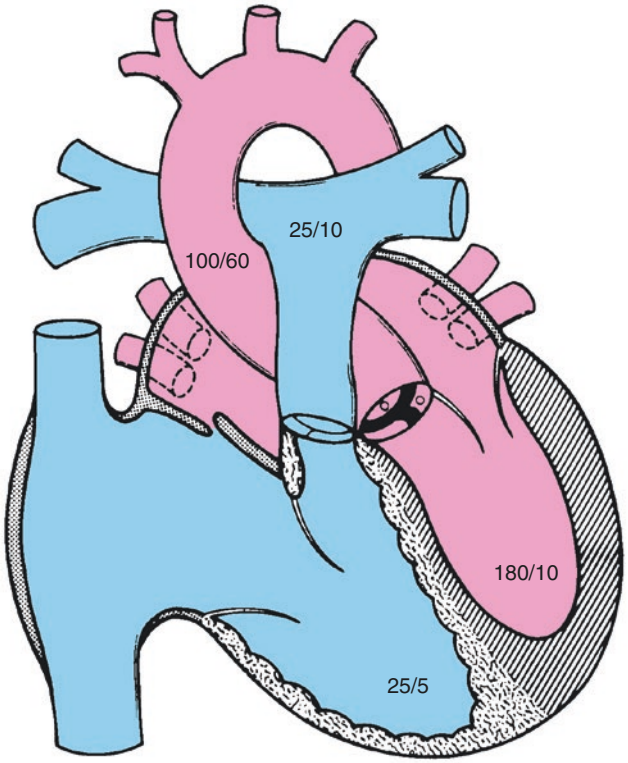

Fig. 8.8 A schematic of aortic valve stenosis (valve AS). Pressure in the LV is elevated, resulting in LV wall hypertrophy and pressure gradient between the LV and ascending aorta. A poststenotic dilatation of the ascending aorta is depicted

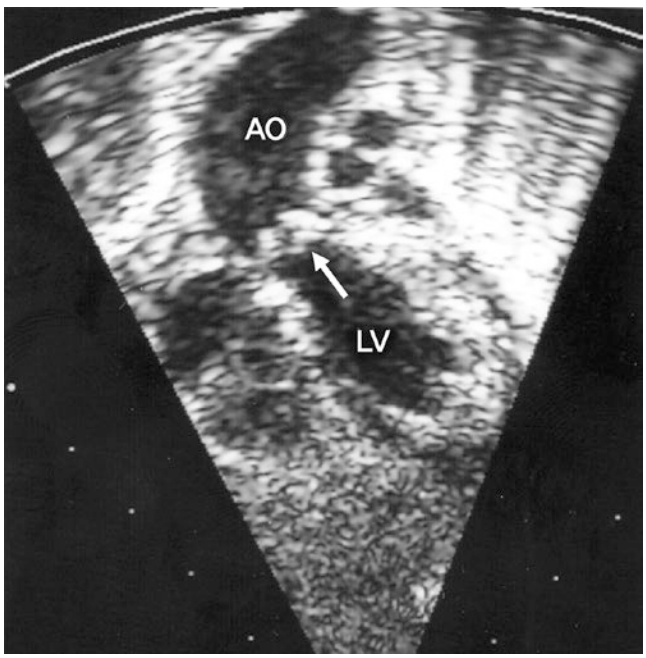

Fig. 8.9 An echocardiographic image (apical 5-chamber view) from a 2-day-old neonate with severe valve AS. The arrow indicates a thickened aortic valve. The ascending aorta (AO) is dilated

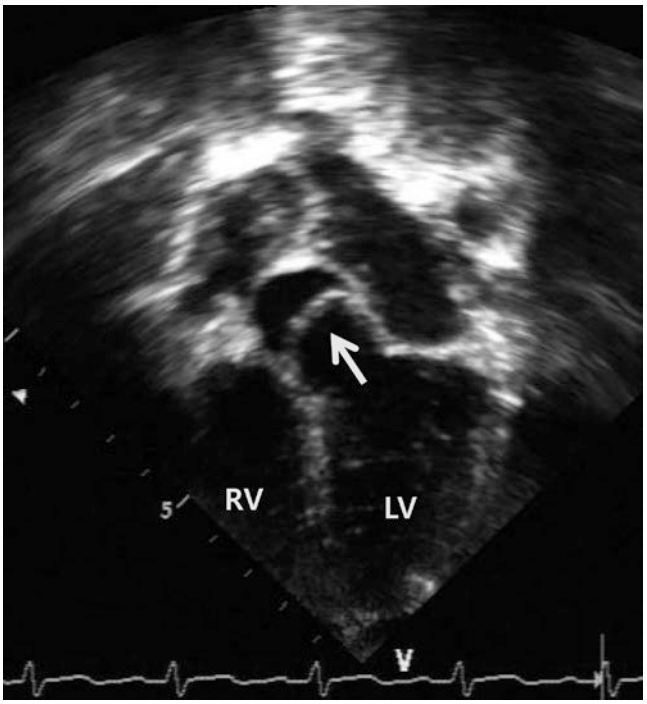

Fig. 8.10 An echocardiographic image (apical 5-chamber view) from another patient with moderate valve AS. The arrow indicates a thickened and doming aortic valve

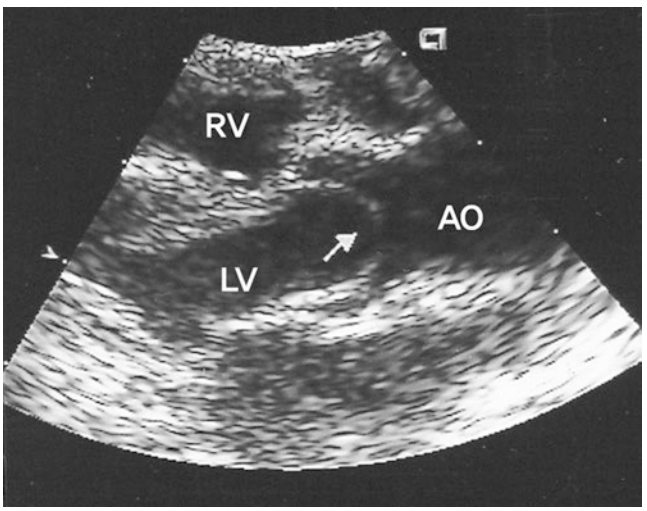

Fig. 8.11 An echocardiographic image (parasternal long-axis view) of mild valve AS showing doming of the aortic valve

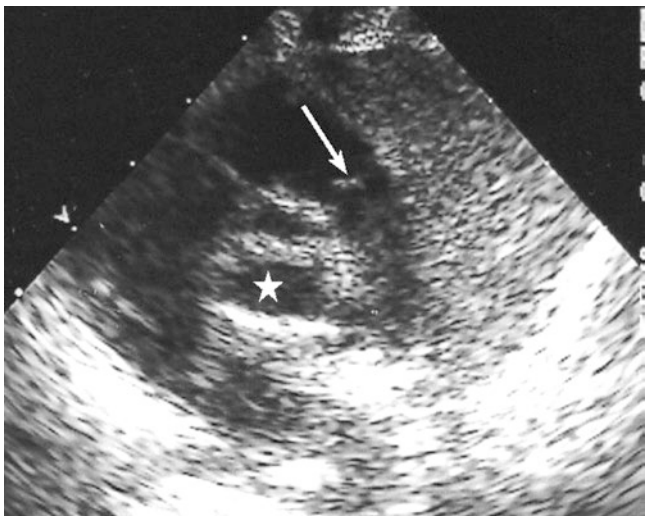

Fig. 8.12 An echocardiographic image (parasternal short axis view) of a thickened and bicuspid aortic valve (★). The arrow indicates a normal pulmonary valve

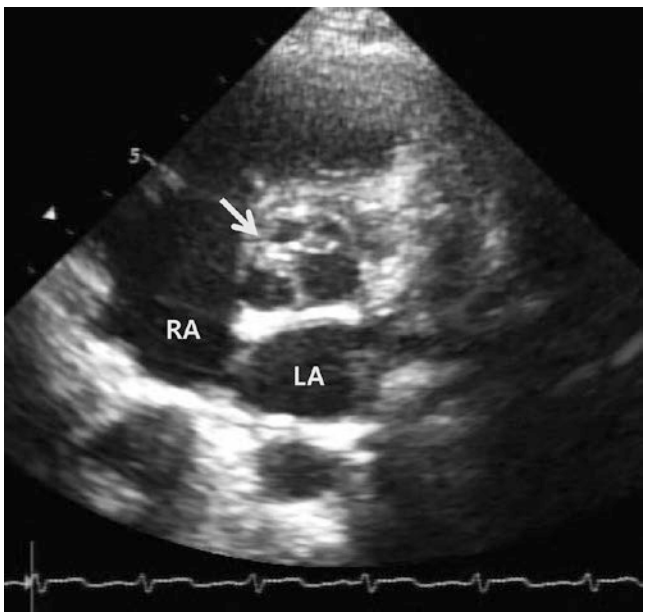

Fig. 8.13 An echocardiographic image (parasternal short axis view) of a very stenotic aortic valve. The arrow indicates a fused commissure between the right and noncoronary cusp

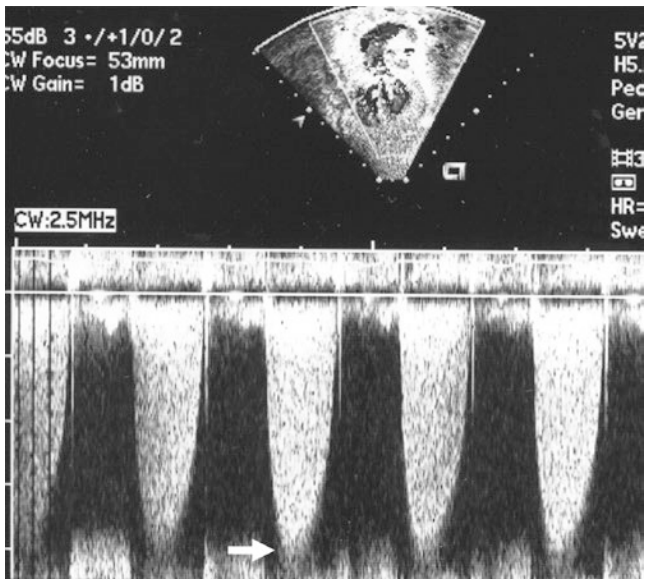

Fig. 8.14 A continuous wave (CW) Doppler measurement of the ascending aorta from a patient with severe valve AS. The maximum velocity is 4 m/s, indicating a peak systolic pressure gradient of 64 mmHg across the stenotic aortic valve

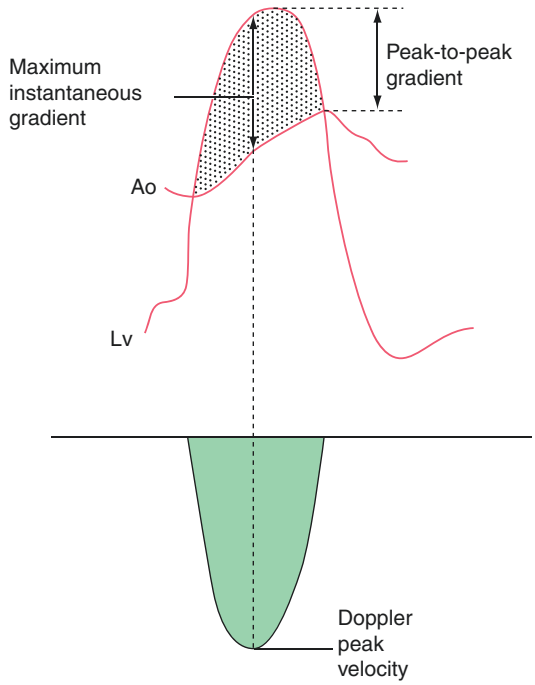

Fig. 8.15 A pressure tracing and CW Doppler recording showing differences among peak-to-peak pressure gradient, maximum instantaneous pressure gradient, and mean gradient. Among these, mean gradient (the average of pressure gradients during the entire period of flow) is the best parameter in estimating the severity of a valve stenosis. Doppler peak velocity correlates with the maximum instantaneous gradient. Peak-to-peak gradient can be measured only by catheterization. Mean gradient can be measured by tracing the time velocity integral and is calculated by a built-in computer program. (Oh JK, Seward JB, Tajik AJ, et al. The Echo Manual. Lipponcott: Williams and Wilkins; 2006. 65, with permission)

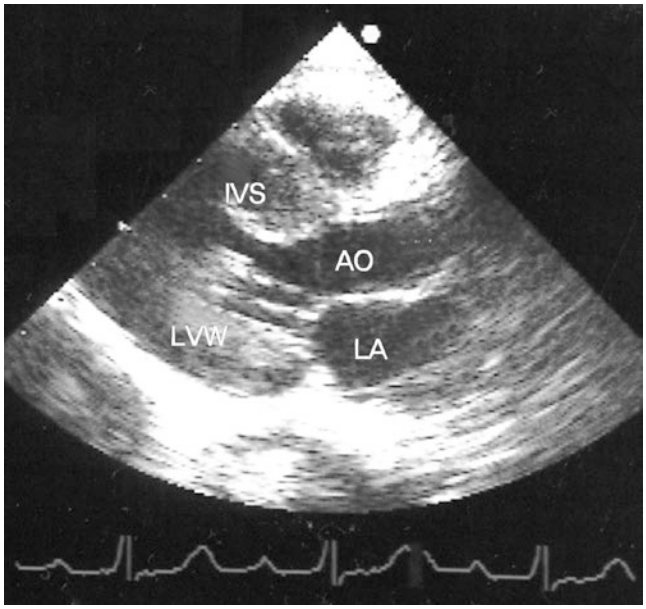

Fig. 8.16 An echocardiographic view (parasternal long axis view) showing severe, concentric LV hypertrophy in a patient with severe valve AS. *IVS* interventricular septum, *LVW* LV wall

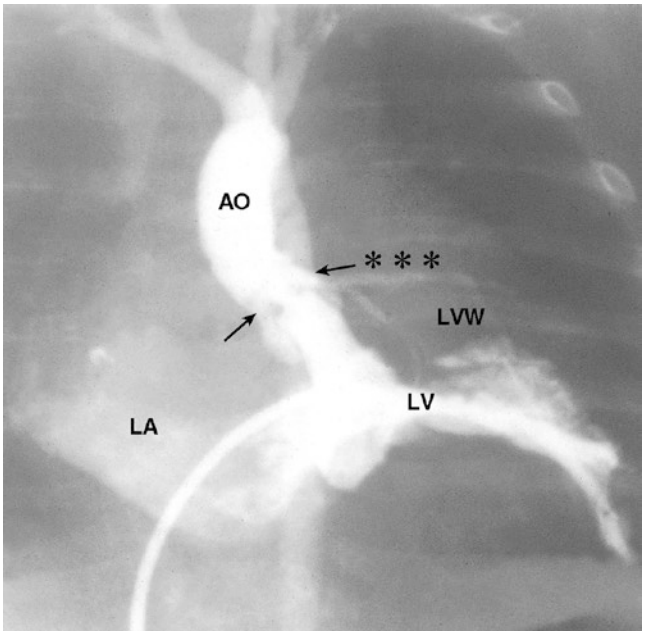

Fig. 8.17 An LV angiogram from a patient with severe valve AS. Severe concentric LV wall hypertrophy is suggested from the mid-cavity obliteration during systole and the long distance between the left coronary-artery (***) and contrast-filled LV cavity. Arrows indicate a thickened aortic valve seen as a radiolucent line

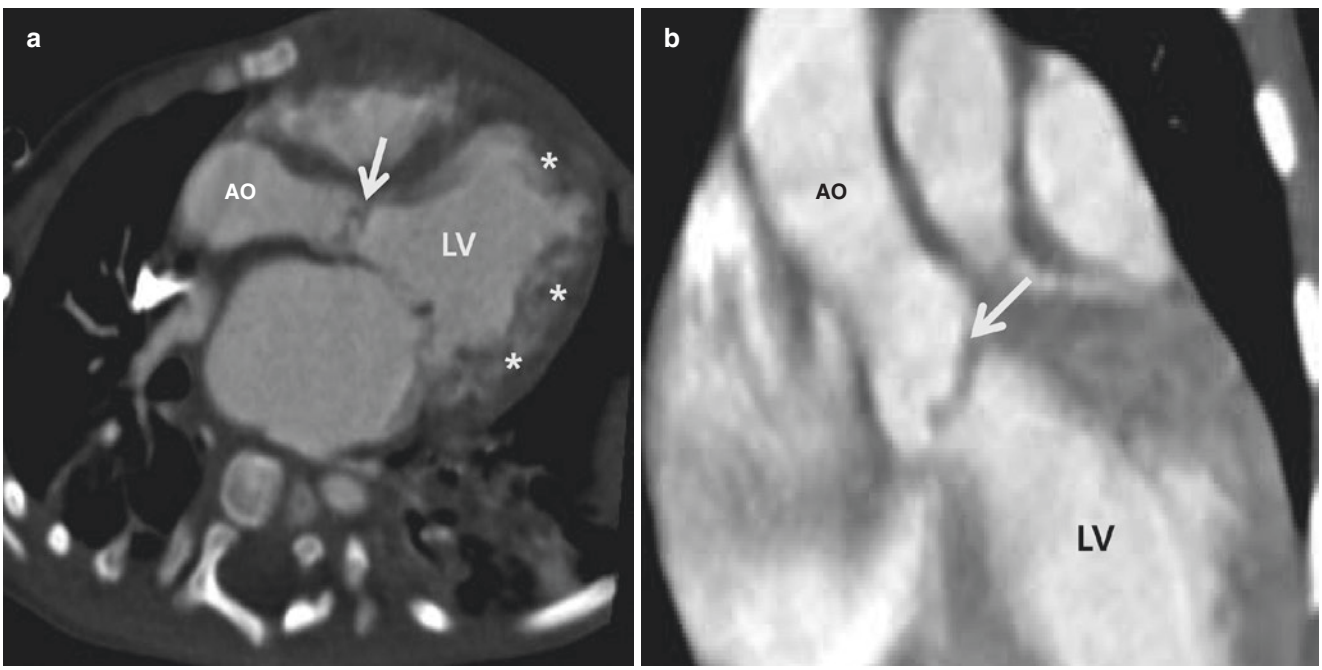

Fig. 8.18 (**a**, **b**) CT images (performed for airway evaluation) from a 2-week-old neonate with severe valve AS showing thickened aortic valve (arrow) and dilated LV. The LV wall has a spongy appearance suggesting noncompaction (***)

Aortic Valve Stenosis

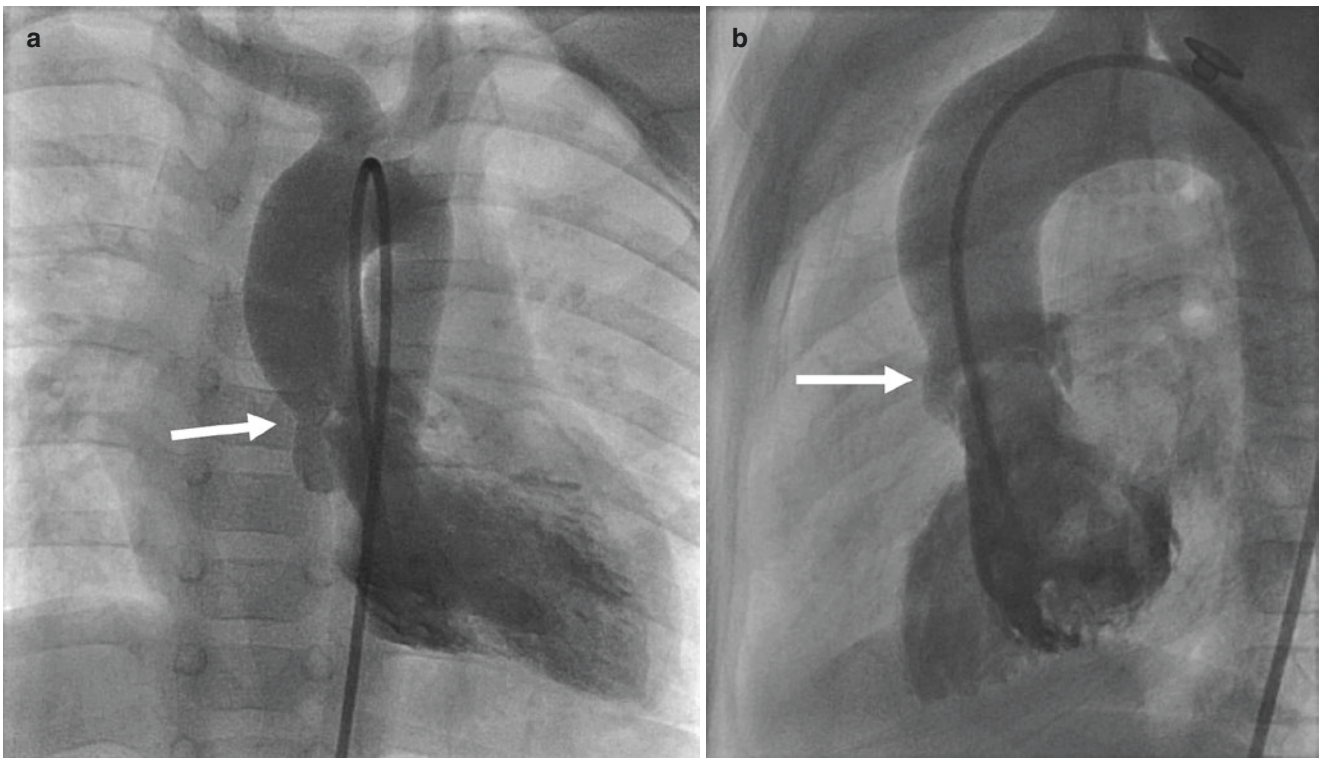

Fig. 8.19 AP (**a**) and lateral views (**b**) of an LV angiogram from a 12-month-old male infant with severe valve AS. Arrows indicate a thickened valve. The systolic pressures were 150 mmHg in the LV and 75 mmHg in the ascending aorta

Balloon Dilatation of an Aortic Valve

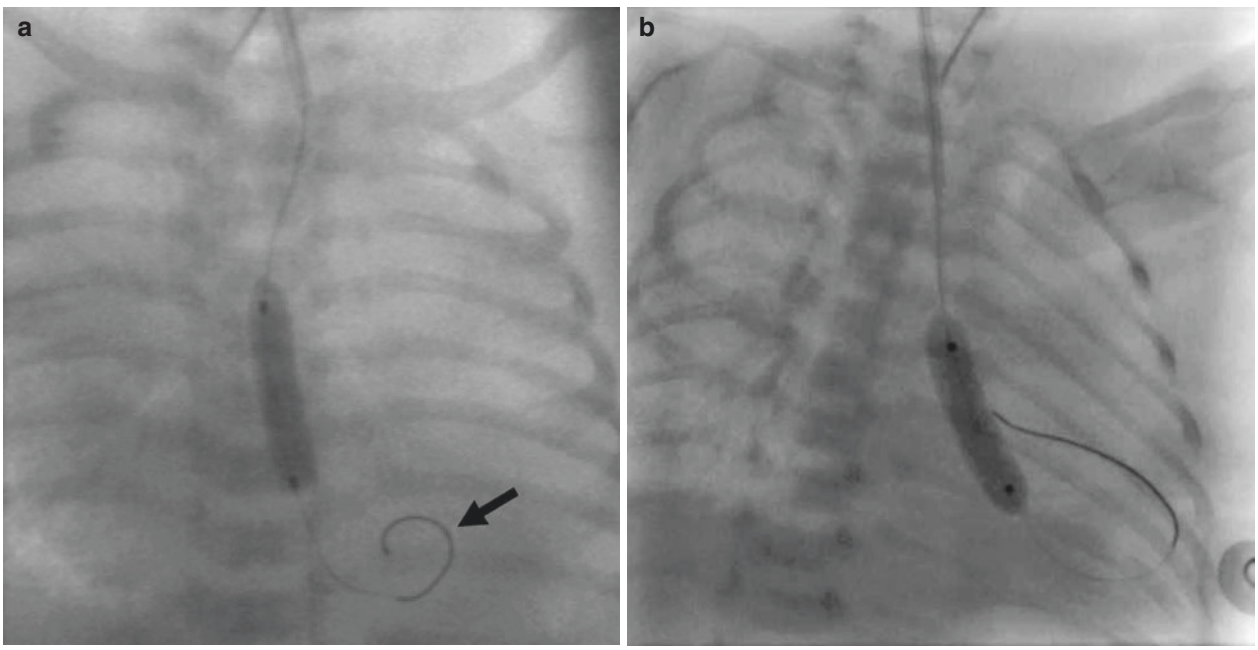

Fig. 8.20 (**a**, **b**) An AP view of a balloon dilatation catheter advanced from a carotid artery from two different newborns with severe valve AS. The soft end of the guide wire (arrow) is seen in the LV. A dilatation balloon is placed across the aortic valve and is fully inflated

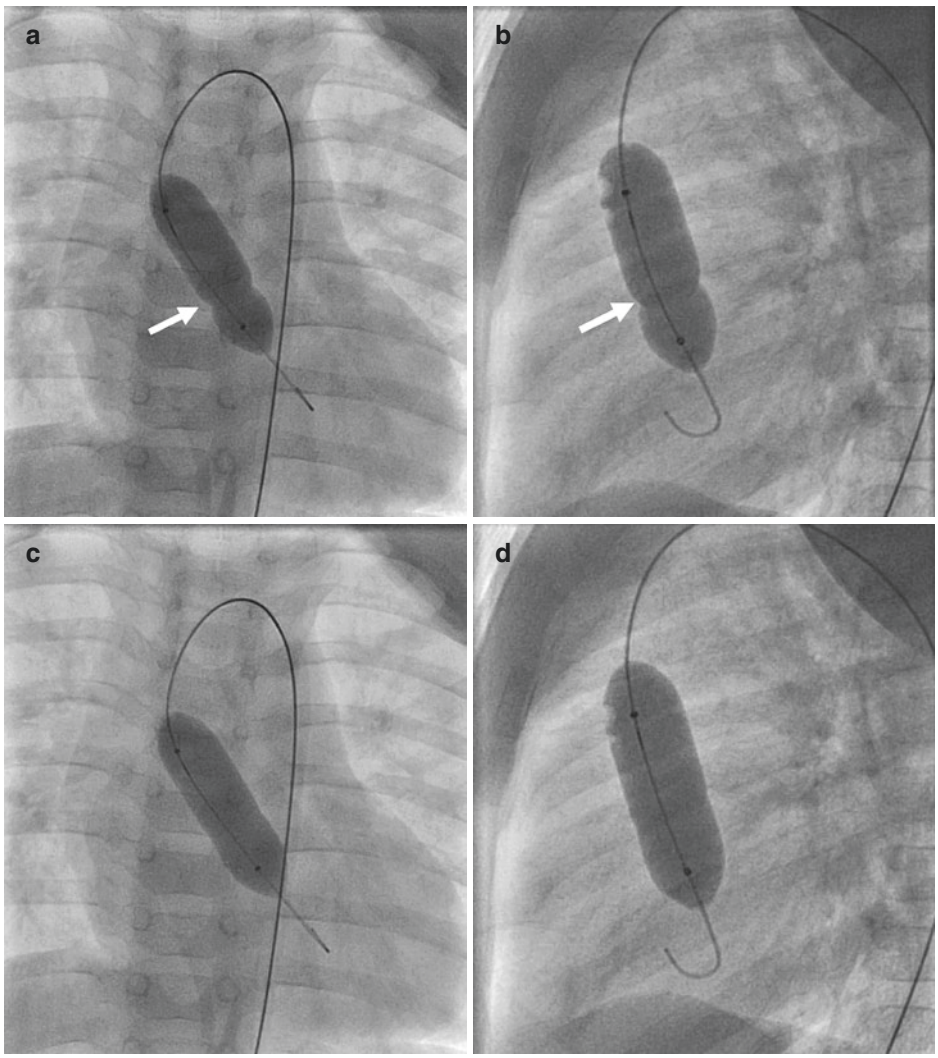

Fig. 8.21 AP (**a**) and lateral views (**b**) of a 12-mm balloon catheter advanced from a femoral artery. The balloon is partially inflated, showing a waist at the level of the stenotic aortic valve. AP (**c**) and lateral views (**d**) of the same balloon catheter fully inflated with no visible waist

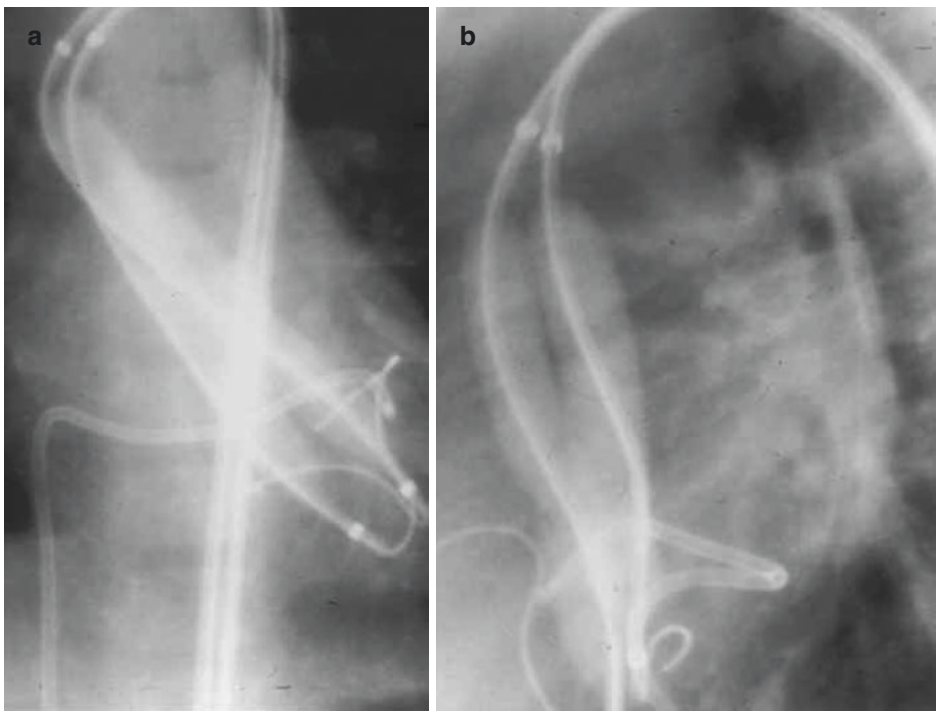

Fig. 8.22 AP (**a**) and lateral views (**b**) of two balloons across the aortic valve. Sometimes it is necessary to use two dilatation balloons instead of one large balloon to avoid damaging the peripheral artery through which the large catheter must pass

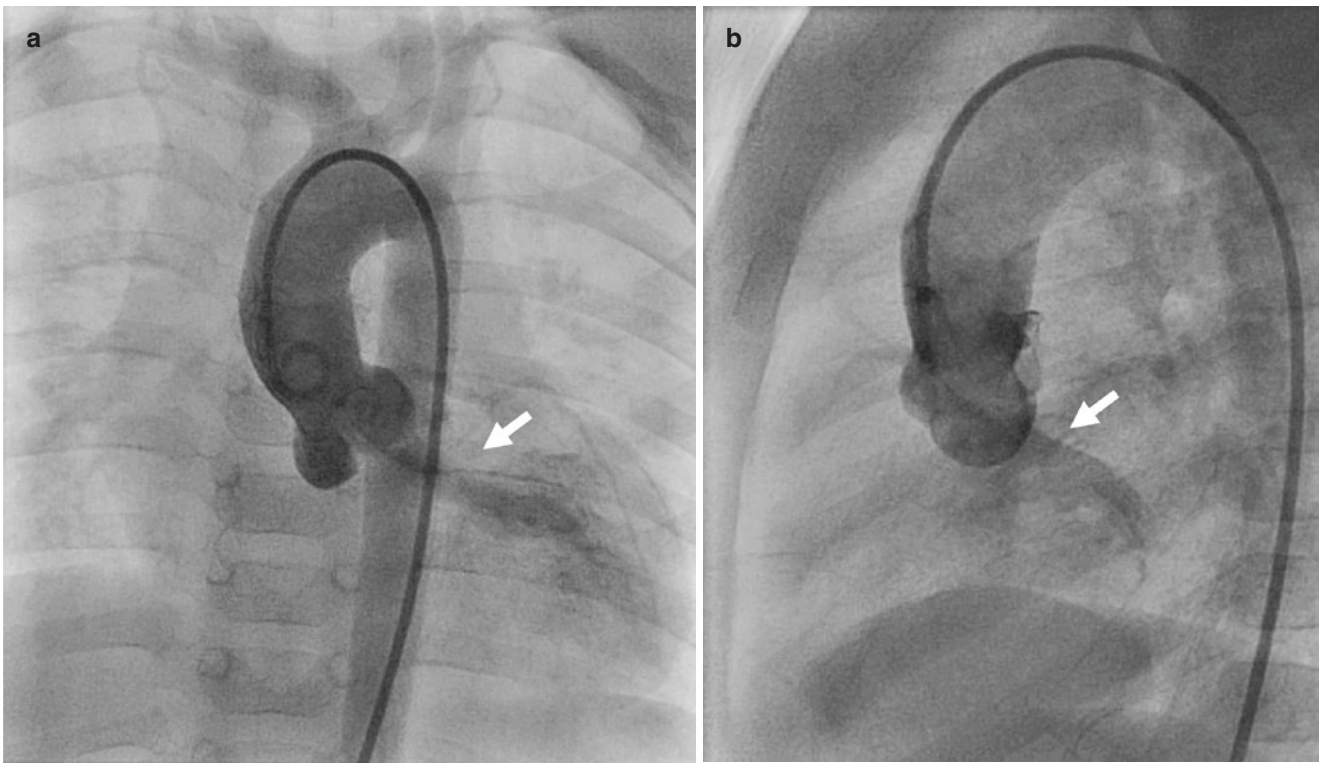

Fig. 8.23 AP (**a**) and lateral views (**b**) of an aortogram performed after balloon dilatation of the aortic valve showing minimal aortic regurgitation (arrows)

EGG of Severe Aortic Valve Stenosis

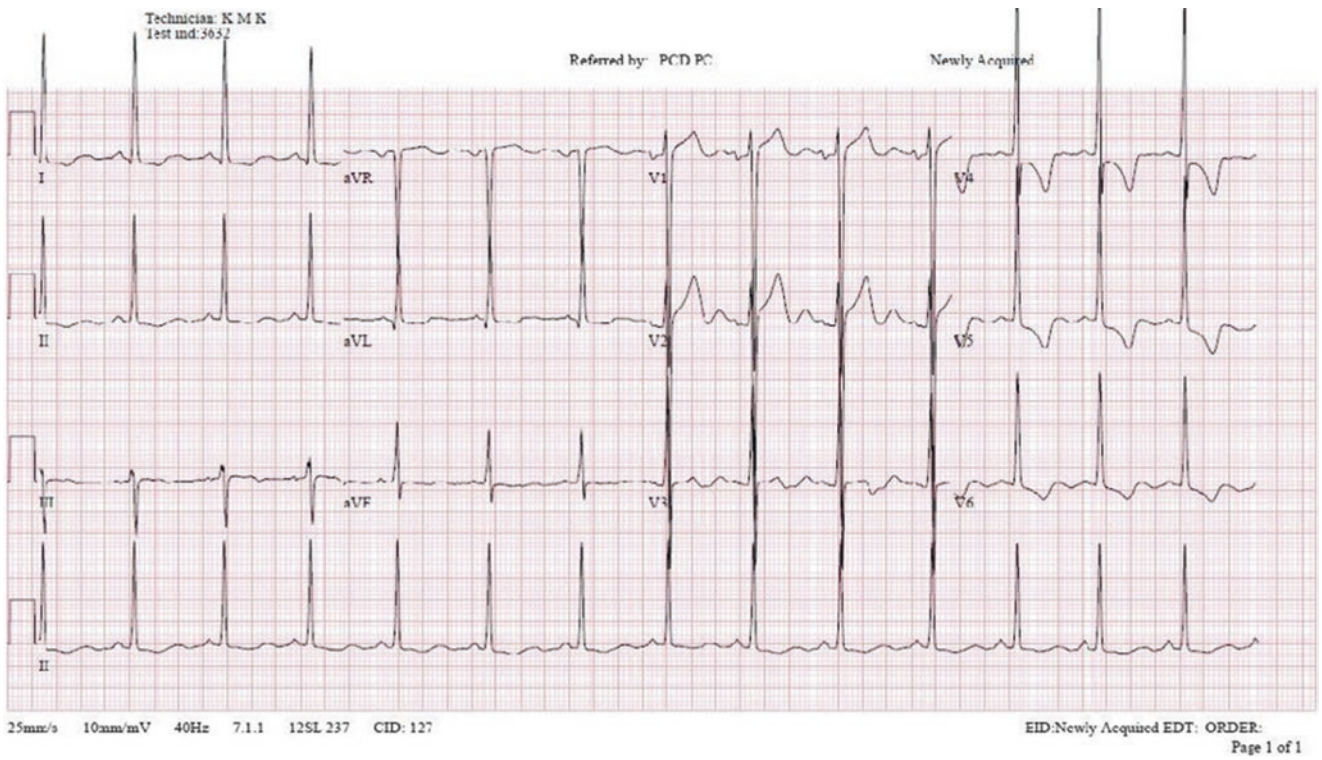

Fig. 8.24 An ECG from a patient with severe valve AS showing severe LVH and strain pattern

Infective Endocarditis in Patients with Aortic Valve Stenosis

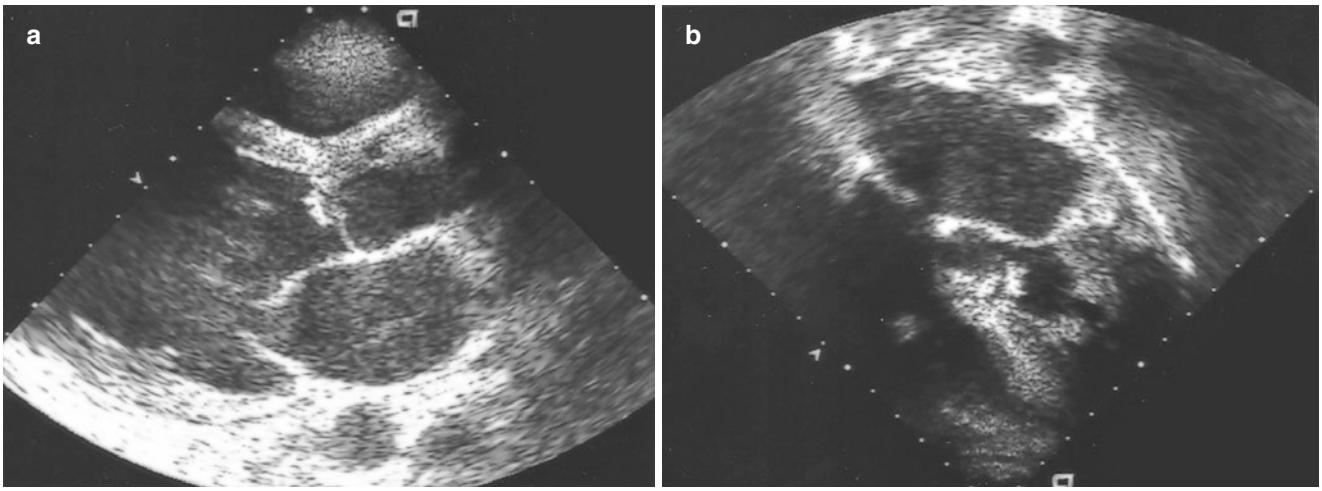

Fig. 8.25 (**a**) A parasternal long-axis view from a 14-year-old boy who presented with a 2-week history of fever of unknown origin. Small vegetation (arrow) is seen immediately under the aortic valve. (**b**) Apical 4-chamber view showing a thick vegetation under the aortic valve. Blood culture grew *Streptococcus sanguis*. He had a history of recurrent ulcer around the mouth and oral cavity, and this is believed to be the source of bacterial entry. After treatment with antibiotics, he underwent Ross operation

Mycotic Aneurysm of the Ascending Aorta

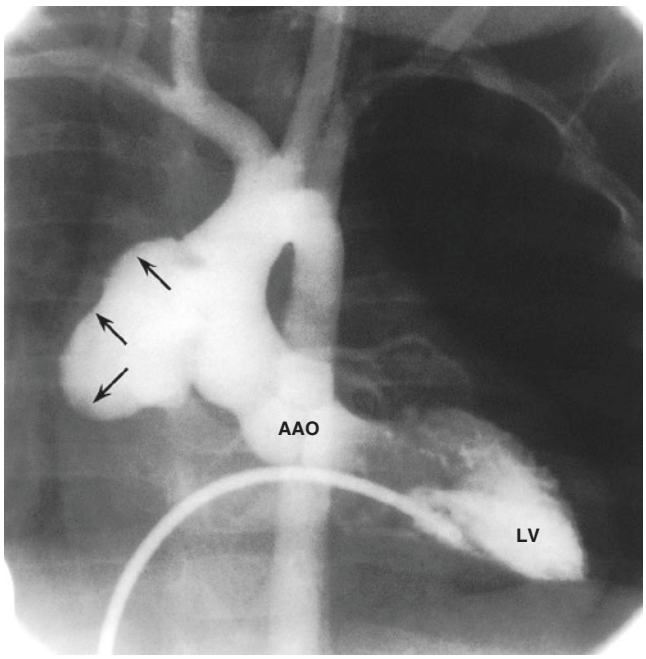

Fig. 8.26 An AP view of an LV angiogram from an 11-month-old infant with bacterial endocarditis. He presented to the hospital with a history of fever of unknown origin. He was found to have a bicuspid aortic valve with mild stenosis. Mycotic aneurysm occurred exactly where the AS jet hits the wall of the ascending aorta. This angiogram was obtained many years ago, when CT and MRI technology was not well developed. Angiography such as this is not only unnecessary but is also dangerous and should not be performed in the modern era when CT and MRI provide superb image

Aortic Valve Stenosis

Incidence

AS accounts for 3–8% of all CHDs.

However, minor abnormalities of the aortic valve could go undetected in many cases early in life.

The incidence of bicuspid aortic valve is reported as high as 1.2% according to studies in Caucasians. Valve AS is four times more common in males than in females.

Hemodynamics

Grading of AS according to peak systolic pressure gradient on echocardiography:

- Mild AS: <50 mmHg
- Moderate AS: 50–75 mmHg
- Severe AS: >70 mmHg
- *These criteria can be applied only when LV function is normal.*
- *In other words, when LV contractility is decreased, measuring pressure gradient is meaningless.*

Due to the increased afterload of the LV, the LV wall becomes hypertrophic.

In severe AS, LV function deteriorates, causing heart failure and myocardial ischemia.

Symptoms

Mild-to-moderate AS causes no subjective symptom in most cases.

Severe AS may cause chest pain, dyspnea with exercise, syncope, and even sudden death.

Physical Signs

- Systolic ejection click (SEC), the opening sound of the stenotic valve, is audible immediately after S_1 (Fig. 8.27).
- Systolic ejection murmur along the left upper sternal border, radiating along the carotid arteries. Loudness of this systolic murmur correlates with the severity of AS.
- S2 becomes split in severe AS.
- To-and-fro murmur (systolic ejection murmur and high-pitch early diastolic murmur, Fig. 8.28) may be audible in patients with AS associated with AR or when the patient develops AR after balloon dilatation of AS.
- Thrill may be palpable with a loud murmur in severe stenosis.
- Peripheral pulses may become weak in severe AS.

Fig. 8.27 A diagram showing a diamond-shaped systolic ejection murmur between S1 and S2. A systolic ejection click (SEC) is audible immediately after S1, a typical auscultatory finding in valve AS

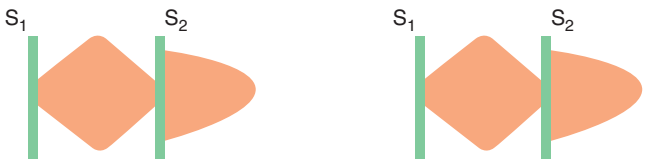

Fig. 8.28 A diagram showing to-and-fro murmur, typically heard in patients with AS and AR. Unlike a continuous murmur, S2 is clearly audible between the systolic and diastolic murmurs

Chest Radiography

The heart size is usually normal until ventricular function deteriorates and dilates.

Electrocardiography

LV hypertrophy and strain pattern in severe AS.

Echocardiography

The following observations should be made during echocardiographic study: morphology of the aortic valve, whether it is bicuspid or tricuspid, Doppler assessment of AS severity, assessment of LV function, aortic root dilatation, function of other valves, and associated lesions.

Cardiac Catheterization and Angiography

This procedure is rarely done for diagnostic purpose only.

It is necessary when attempting catheter intervention.

CT and MRI

Not necessary for uncomplicated simple AS.

Natural Course

- Sudden death in 1–2% of patients with severe AS.
- AS usually progresses with advancing age.
- Aortic valve regurgitation can occur with time, particularly in the bicuspid aortic valve.
- Bacterial endocarditis is a serious complication of AS and is reported in approximately 4% of patients with valve AS. Therefore, lifelong prophylaxis against bacterial endocarditis is important.

Catheter Treatment (Balloon Dilatation of the Aortic Valve)

- Treatment is indicated when the pressure gradient is greater than 50 mmHg (only when the LV contractility is normal).
- When the LV function is impaired, pressure gradient is meaningless.
- Balloon dilatation should be attempted as the first-line treatment when valve morphology is feasible.
- This procedure is particularly valuable in patients with LV dysfunction, when open heart surgery carries a high risk.
- Minimal aortic regurgitation is inevitable after balloon dilatation. However, care should be taken to avoid severe AR after balloon dilatation. Actually, it is better to ignore mild AS than causing severe AR.
- The diameter of a dilatation balloon should not exceed the diameter of the aortic valve annulus (different from PS dilatation).

Surgery

- Surgery is necessary when balloon dilatation is ineffective or is not feasible.
- Surgical procedures include (1) open commissurotomy, (2) Ross operation (pulmonary autograft aortic valve replacement along with replacement of the pulmonary outflow tract by using a homograft or mechanical valve), and (3) aortic valve replacement with a mechanical valve.
- Choosing the treatment modality in individual patients depends on valve characteristics, age, and gender of the patient, previous surgical procedure, and surgeon's experience.

Follow-Up

- Lifelong follow-up is necessary in patients with AS, regardless of mode of treatment and severity of AS, with regular ECG and echocardiography.
- Stringent prophylaxis against bacterial endocarditis is needed.
- Individual guidelines for participation in sports should be given.

Critical Aortic Valve Stenosis in Infancy

- Critical AS in neonates and infants carries significant morbidity and mortality because of frequent association with endocardial fibroelastosis (EFE) of the LV, LV dysfunction, and small LV cavity.
- They present with severe congestive heart failure and low cardiac output.
- Heart murmur may be faint or not audible at all. Peripheral pulses are typically weak.
- The most important determinant of mode of treatment and outcome is the size of the LV.
- When the LV size is adequate, opening of a stenotic valve, regardless of LV function, preferably by percutaneous balloon dilatation, is the treatment of choice.
- However, when the LV size is inadequate, PGE_1 infusion is necessary to keep systemic blood flow through the PDA and they should be treated as a variant of hypoplastic left heart syndrome.
- The following criteria of the size of the LV, and mitral and aortic valves adequate for biventricular repair, have been suggested as a treatment guideline. (Leung MP, McKay R, Smith A, Anderson RH, Arnold R. Critical aortic stenosis in early infancy. Anatomic and echocardiographic substrates of successful open valvotomy. J Thorac Cardiovasc Surg. 1991b;101:526–35.):

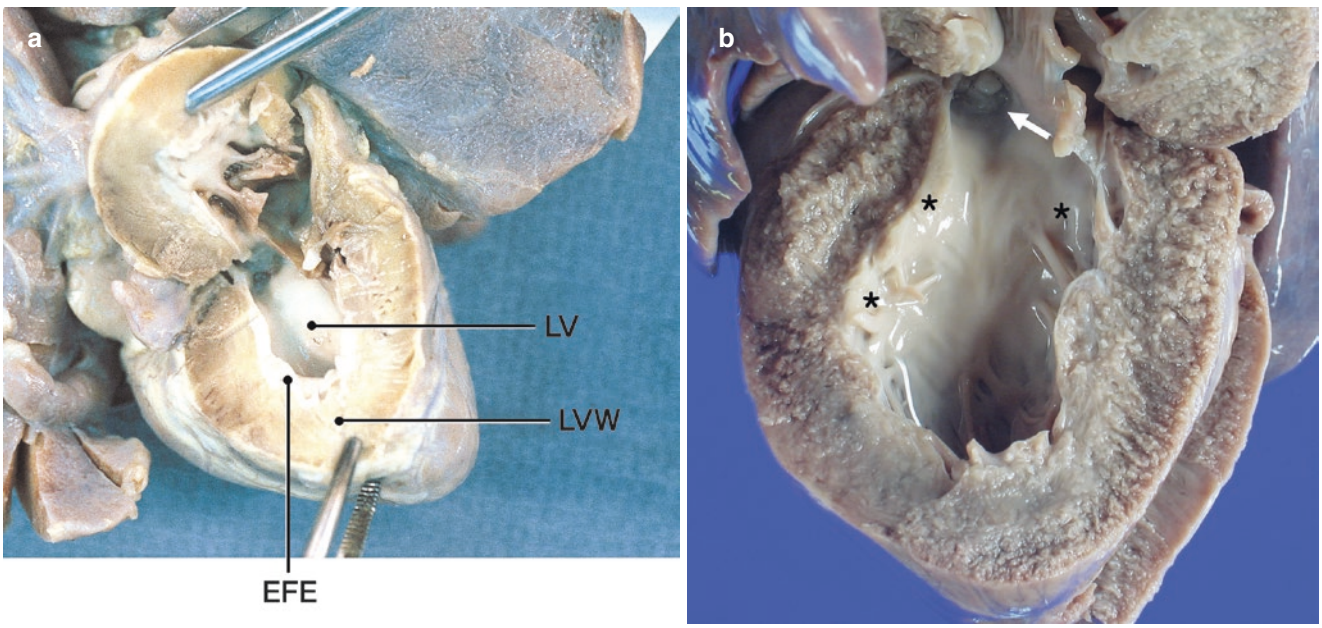

Fig. 8.29 Pathological specimens from two neonates who died of severe heart failure and low cardiac output due to valve AS and severe endocardial fibroelastosis (EFE) at 1 month (**a**) and 2 weeks of age (**b**), respectively. The LV endocardium (*) appears whitish and is very thick due to EFE. Biventricular repair would have not been possible owing to the small LV cavity, particularly in case A. The arrow in case B indicates the aortic valve

  - LV end-diastolic volume >20 mL/m^2 body surface area
  - Aortic valve annulus >5 or 6 mm
  - LV inflow length >25 mm
  - MV orifice or annulus >9 or 11 mm
  - Cardiac apex formed by LV
- Successful resection of EFE of the LV along with open commissurotomy of the aortic valve has been reported and is worth considering in selected cases.

Aortic Valvuloplasty in a Fetus

Aortic Valve Stenosis in a Fetus

Diagnostic clues of valve AS in a fetus are (1) turbulent flow in the ascending aorta and (2) poststenotic dilatation of the ascending aorta.

Prenatal diagnosis of valve AS is not easy because flow velocity across the aortic valve may not represent the true severity of the stenosis during fetal life.

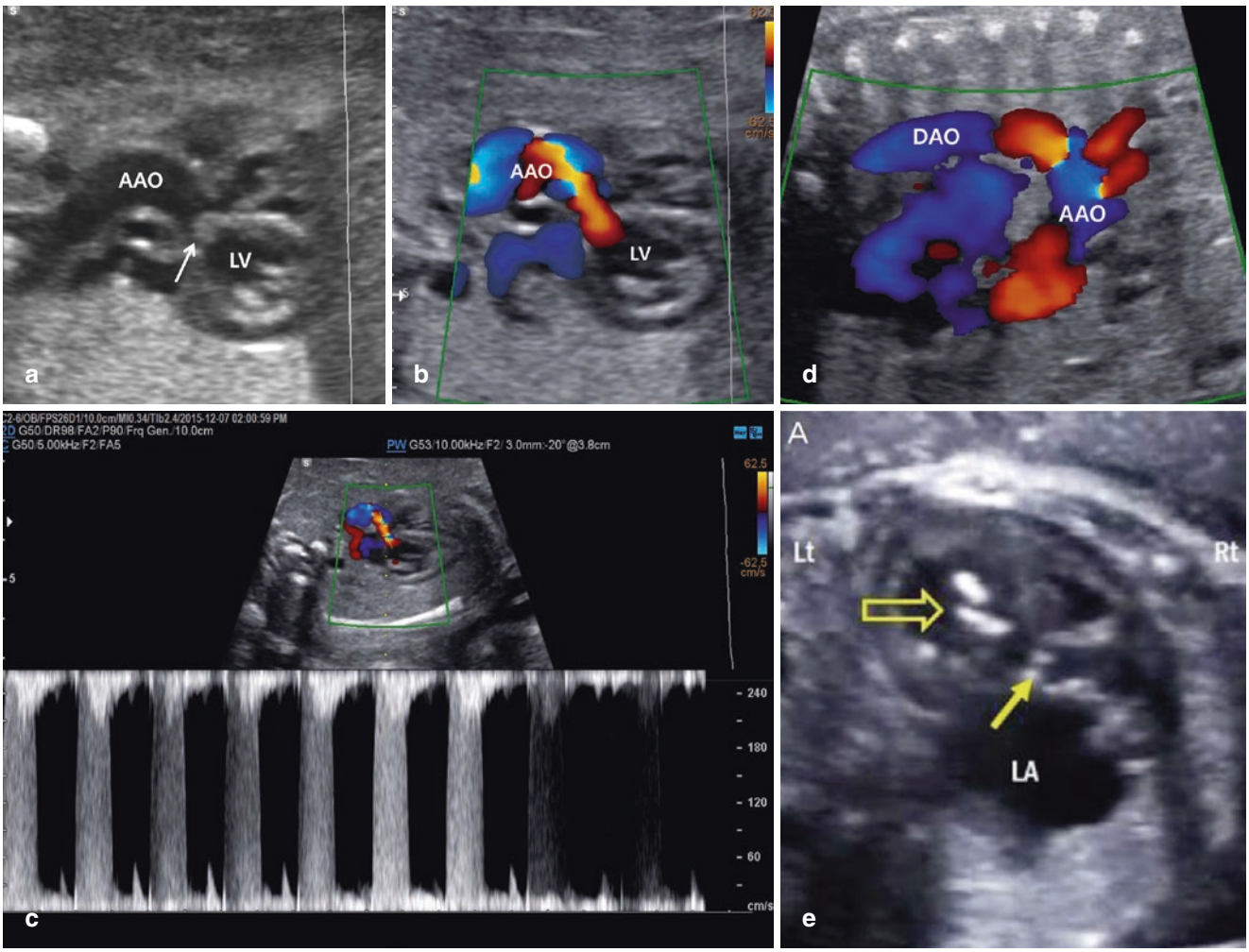

Fig. 8.30 (**a**) The left ventricular outflow tract view of the fetal heart at 24.1 weeks of gestation showing the thickening of the aortic valve (arrow in **a**), with turbulent flow across the aortic valve annulus on a color Doppler image (**b**, **c**) The pulsed wave Doppler showed an increased velocity of >3 m/sec. (**d**) Follow-up evaluation at 26.1 weeks showing a retrograde flow in the ascending aorta (AAO) and the aortic arch seen in blue color, indicating a very severe stenosis. Therefore fetal aortic valvuloplasty was performed under maternal spinal anesthesia at 29.2 weeks of gestation and was a technical success. However, fetal bradycardia sustained, and an emergency cesarean delivery was performed. Postnatal echocardiography indicated mild residual AS with aortic regurgitation and a moderate amount of mitral regurgitation. (**e**) The open arrow indicates the cannula located in the LV and the thickened aortic valve is demonstrated (solid arrow in **e**). *AAO* ascending aorta, *LV* left ventricle, *DOA* descending aorta, *AA* aortic arch. (These figures are provided by Dr. Hye-Sung Won. Asan Medical Center, Seoul, Korea. (Published in part in Obstet Gynecol Sci. 2017 Jan;60 (1):106–109))

Subaortic Stenosis

- Subaortic stenosis can be due to a membrane ridge or fibromuscular tunnel.
- Unlike valve AS, occurrence of subvalve AS is equal among males and females.
- If untreated for a long time, subvalve AS causes aortic valve damage, resulting in valve stenosis or regurgitation.
- Because of the progressive nature of subvalve AS, surgical resection is indicated in patients with less severe stenosis as compared with the treatment criteria for valve AS.
- Balloon dilatation had been performed and reported, but is not recommended because of lack of efficacy.
- Prophylaxis against bacterial endocarditis is equally important in subvalve AS as in valve AS.

Subaortic Stenosis Due to a Membrane or a Fibrous Ridge

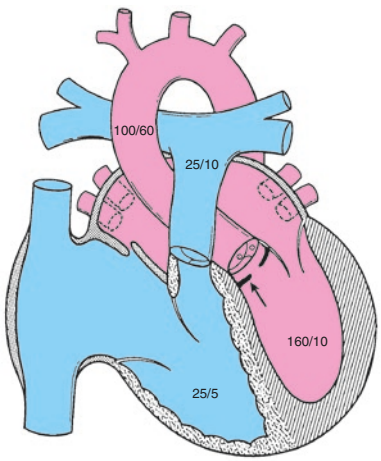

Fig. 8.31 A schematic of subaortic stenosis due to a membrane or a fibrous ridge (arrow). LV pressure is elevated

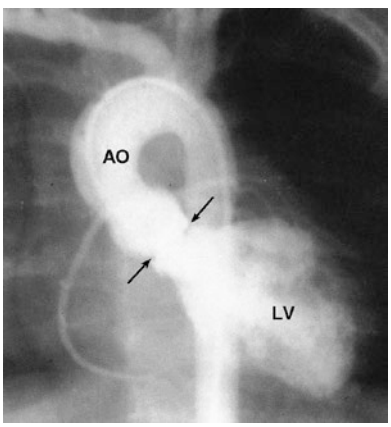

Fig. 8.32 AP view of a LV angiogram showing a membrane seen as a radiolucent line (arrows), under the aortic valve. The LV wall appears thickened

Subaortic Membrane

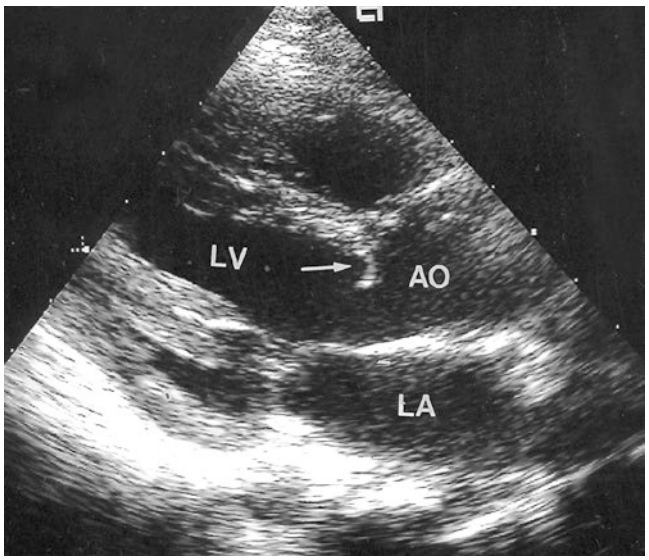

Fig. 8.33 An echocardiographic (parasternal long-axis) view showing a relatively thin membrane under the aortic valve causing turbulence in the ascending aorta

Subaortic Fibrous Ridge

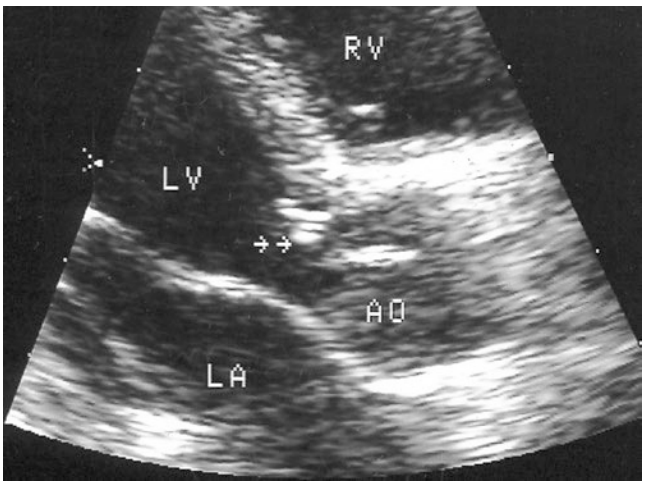

Fig. 8.34 An echocardiographic (parasternal long-axis) view showing a relatively thick ridge under the aortic valve

Subaortic Stenosis Due to a Fibromuscular Tunnel

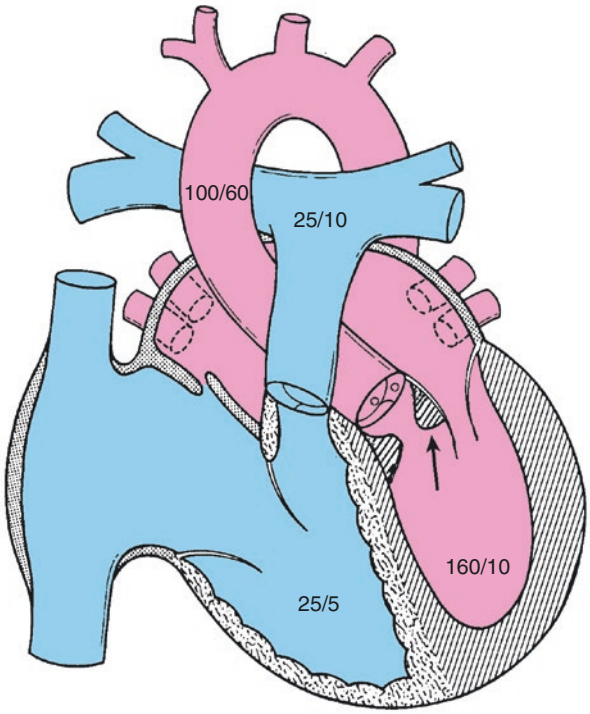

Fig. 8.35 A schematic of subaortic stenosis due to a fibromuscular tunnel (arrow)

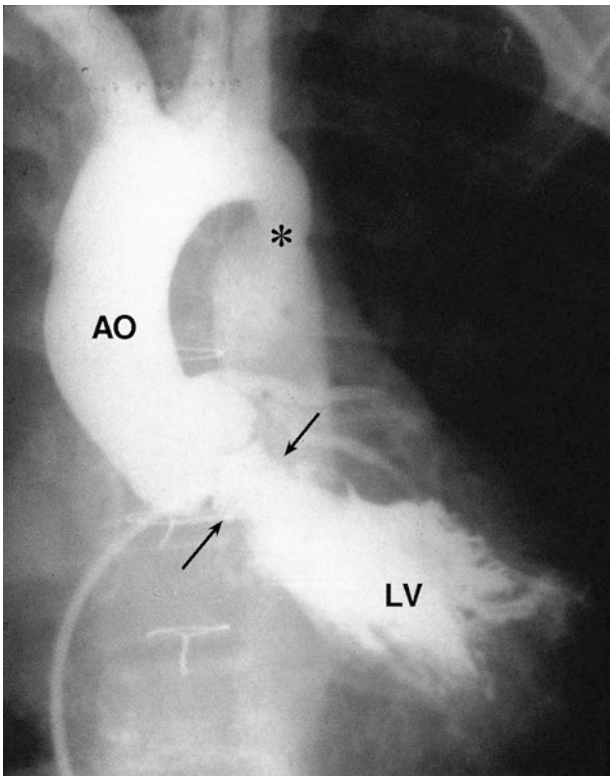

Fig. 8.36 An LV angiogram from a patient with severe subvalve AS due to a fibromuscular tunnel (arrow). The LV wall is hypertrophic. Mild degree of coarctation of the aorta is also present (*)

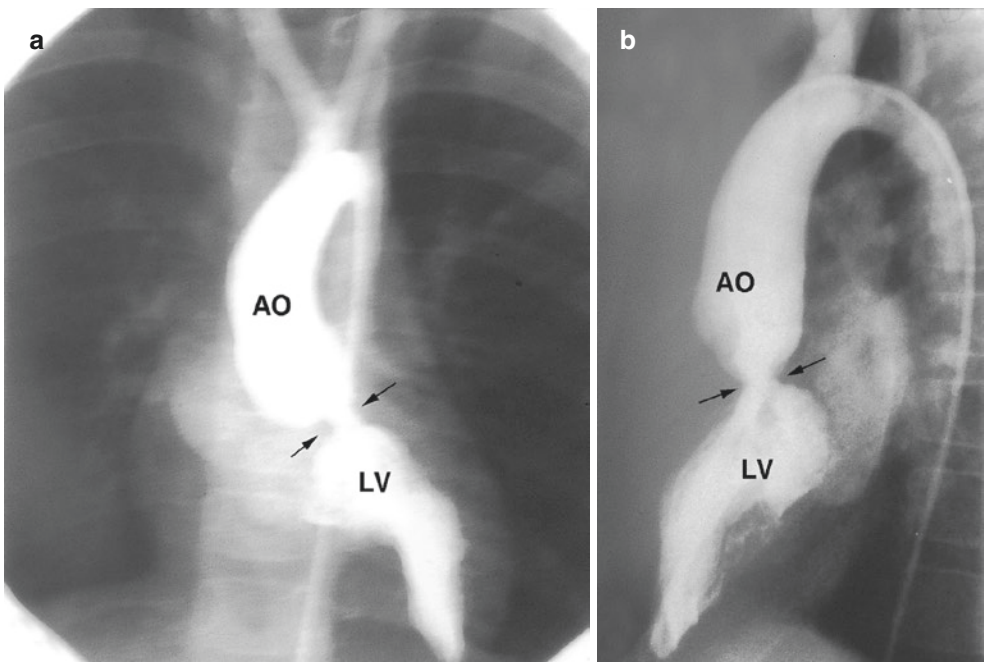

Fig. 8.37 AP (**a**) and lateral views (**b**) of a LV angiogram from a patient with severe subaortic stenosis due to severe circumferential muscle hypertrophy (arrow) under the aortic valve

Multiple Left Heart Obstructive Lesions (Shone Complex)

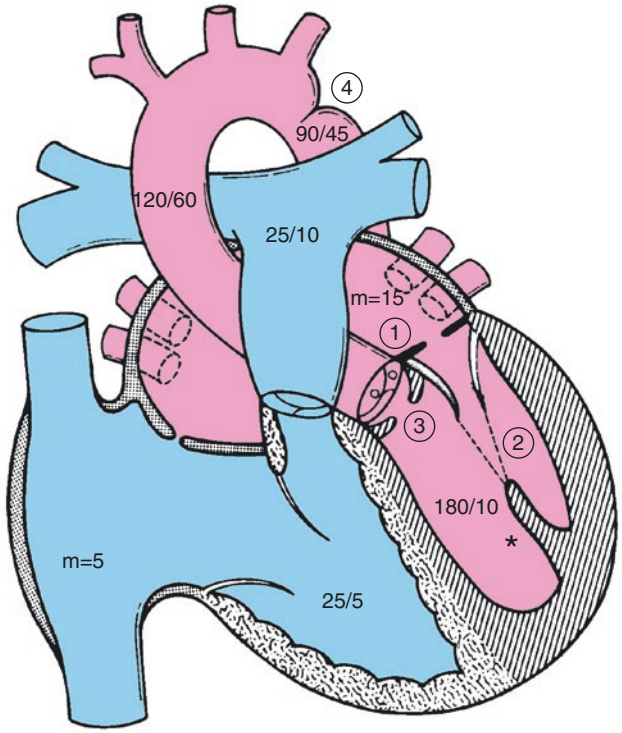

Fig. 8.38 A schematic of Shone complex, multiple obstructions at the inflow and outflow tracts of the LV. Each patient has two or more of the following components. Rarely all of the following lesions are seen in a single patient. This complex consists of the following lesions. VSD can be associated. ① Supravalve mitral ring: A membrane-like ring above the mitral valve, causing obstruction to LV inflow. ② Parachute mitral valve: The chordae of the mitral valve are attached to a single papillary muscle, making parachute-like appearance. ③ Subaortic stenosis due to a membrane or fibromuscular ridge. ④ Coarctation of the aorta

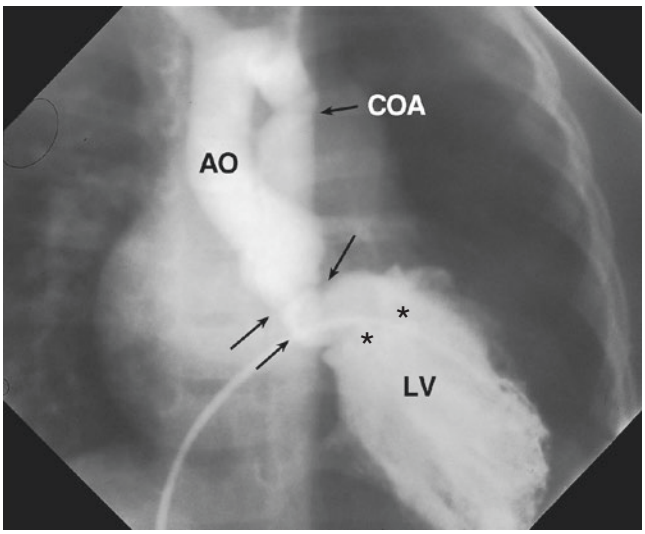

Fig. 8.39 AP view of an LV angiogram from a patient with Shone complex, showing a small mitral valve annulus (**), subaortic stenosis (arrows), and coarctation of the aorta

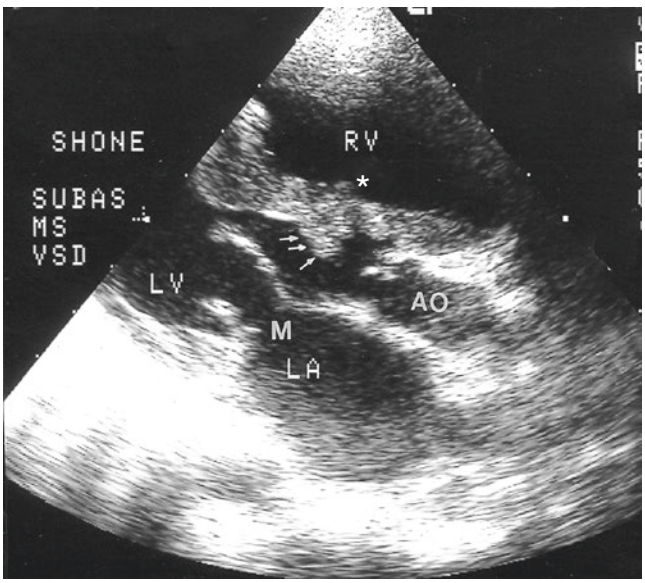

Fig. 8.40 An echo image (parasternal long-axis view) from a patient with Shone complex, showing a thick subaortic ridge (arrows), mild mitral valve stenosis (M), and perimembranous VSD, which is covered with aneurysm (*)

Supravalve Aortic Stenosis in Elastin Arteriopathy

Williams syndrome (Williams-Beuren syndrome, 7q11.23 deletion) and nonsyndromic supravalve AS are collectively called elastin arteriopathy as these syndromes are caused by mutations within the elastin gene on chromosome 7q11.23 (Merla G, Brunetti-Pierri N, Piccolo P, Micale L, Loviglio MN. Supravalvular aortic stenosis: elastin arteriopathy. Circ Cardiovasc Genet. 2012;5:692–6.):

- Approximately 75% (55–80%) of patients with Williams syndrome have CHD.
- In these syndromes, walls of peripheral arteries and pulmonary arteries become thickened and rigid because of abnormal collection of collagen and smooth muscle cells.
- Supravalve AS tends to progress with time, while PA branch stenosis tends to improve as patients get old.
- When supravalve AS is severe, surgery (aortoplasty) is needed to relieve the obstruction.

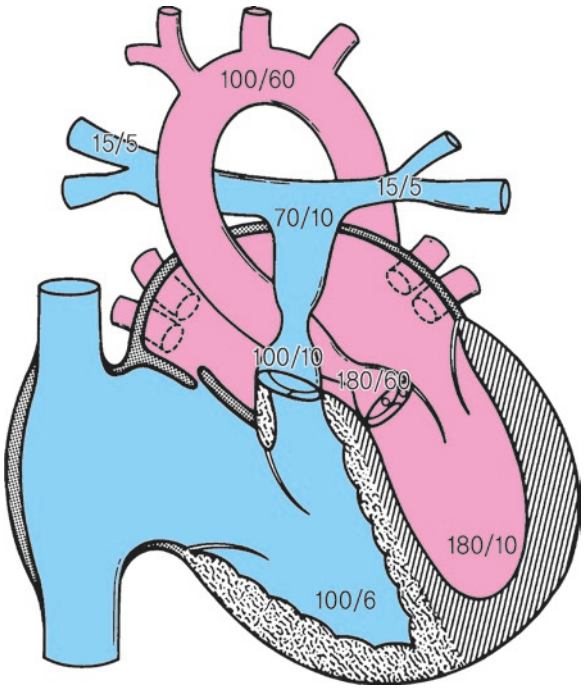

Fig. 8.41 Typical cardiac lesions in Williams syndrome, including supravalve PS, bilateral multiple pulmonary artery branch stenosis, and supravalve AS. Pressures are elevated in both RV and LV

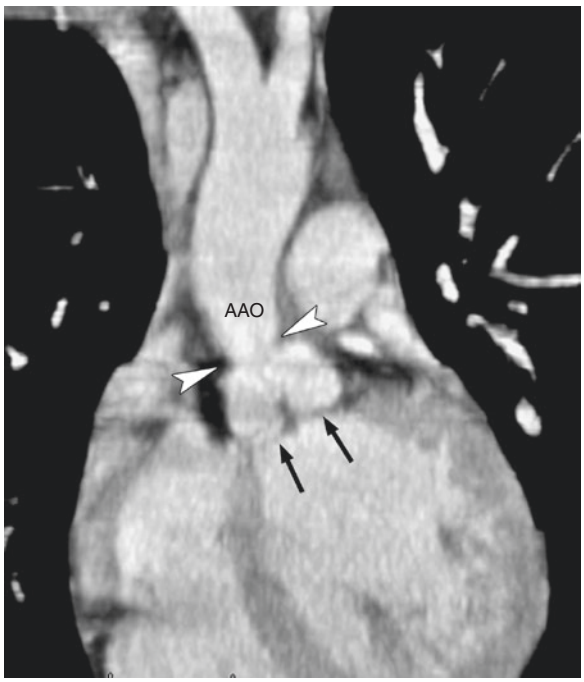

Fig. 8.43 A CT image showing supravalve AS (white arrowhead) and valve AS (black arrowhead)

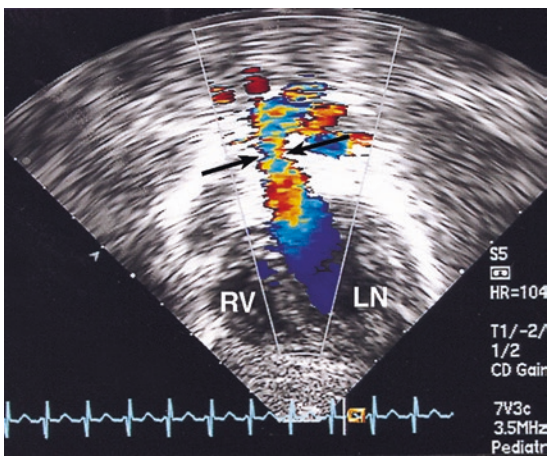

Fig. 8.42 A color Doppler image from a five-chamber view showing turbulent flow in the ascending aorta immediately above the aortic valve. Arrows indicate the site of supravalve AS

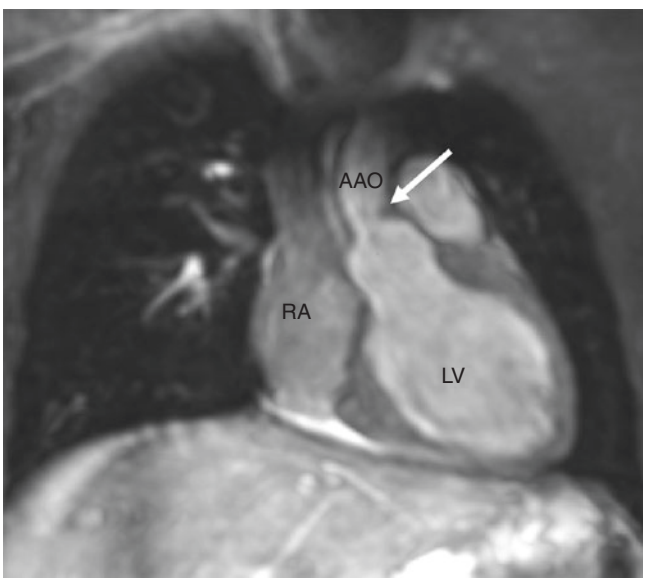

Fig. 8.44 A cardiac MR angiogram from a 3-year-old boy with Williams syndrome, showing supravalve AS (arrow)

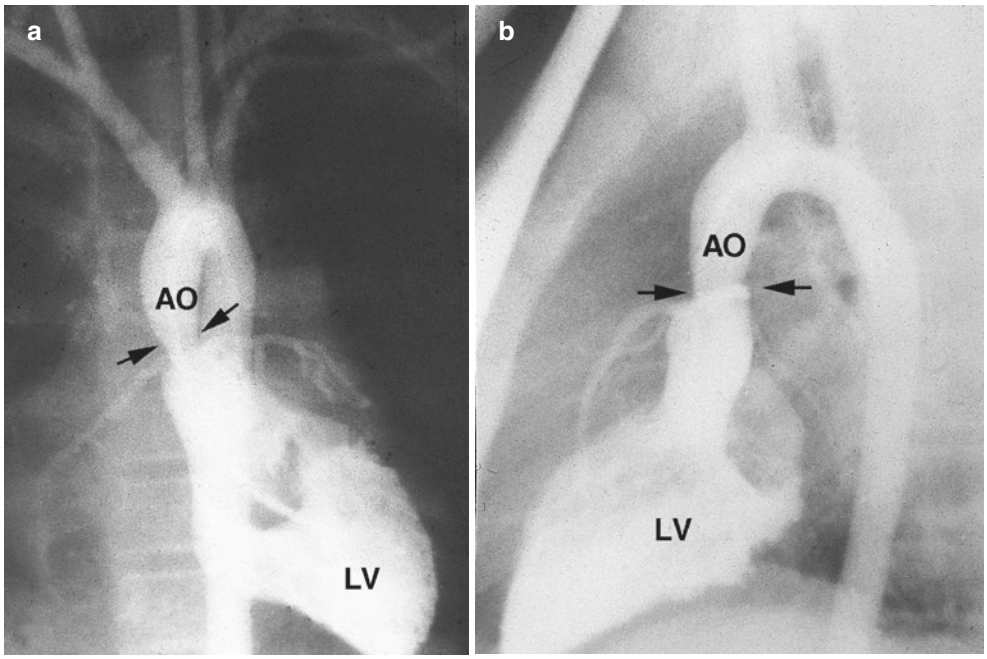

Fig. 8.45 AP (**a**) and lateral views (**b**) of a LV angiogram from a patient with Williams syndrome, showing supravalve AS (arrow). The ascending aorta appears slightly narrow

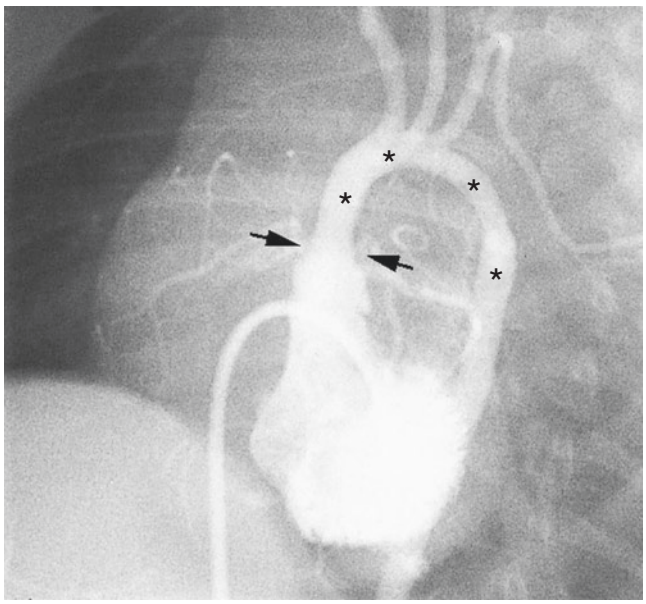

Fig. 8.46 LAO view of a LV angiogram, showing supravalve AS (arrows). The ascending aorta, transverse arch, and thoracic aorta all appear narrow (*), typically seen in patients with Williams syndrome

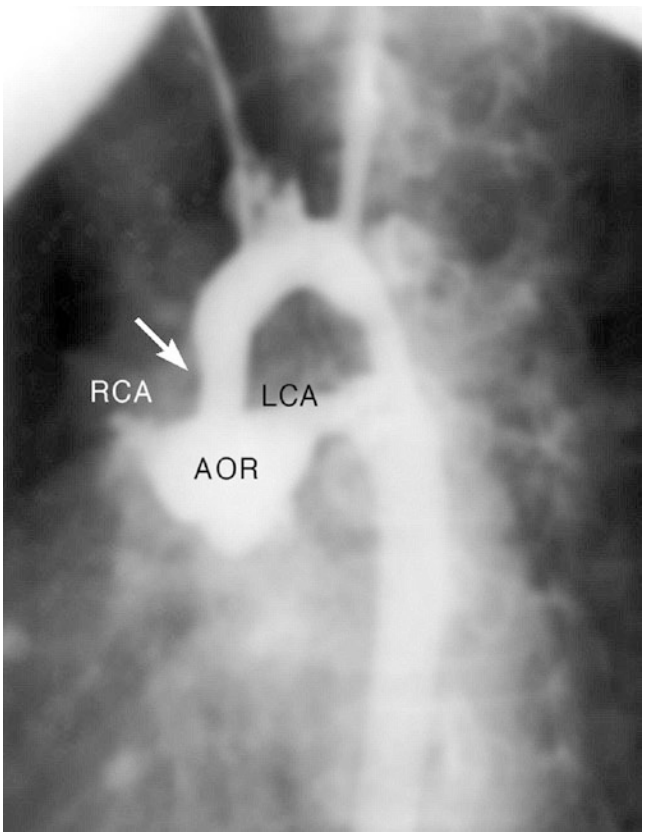

Fig. 8.47 An aortogram from a patient with familial (nonsyndromic) supravalve AS, showing a markedly dilated aortic root (AOR), severe supravalve AS (arrow), and very narrow ascending aorta, transverse arch and thoracic aorta. Both coronary arteries (RCA and LCA) appear very dilated, indicating a high probability of coronary lesions due to a high pressure

LV-Aorta Tunnel

- LV-aorta tunnel, an abnormal communication through a long tunnel between the ascending aorta and the LV myocardium, is a rare congenital cardiac anomaly.
- Patients usually present with a diastolic murmur, similar to aortic regurgitation.
- Surgery is necessary to close the tunnel.

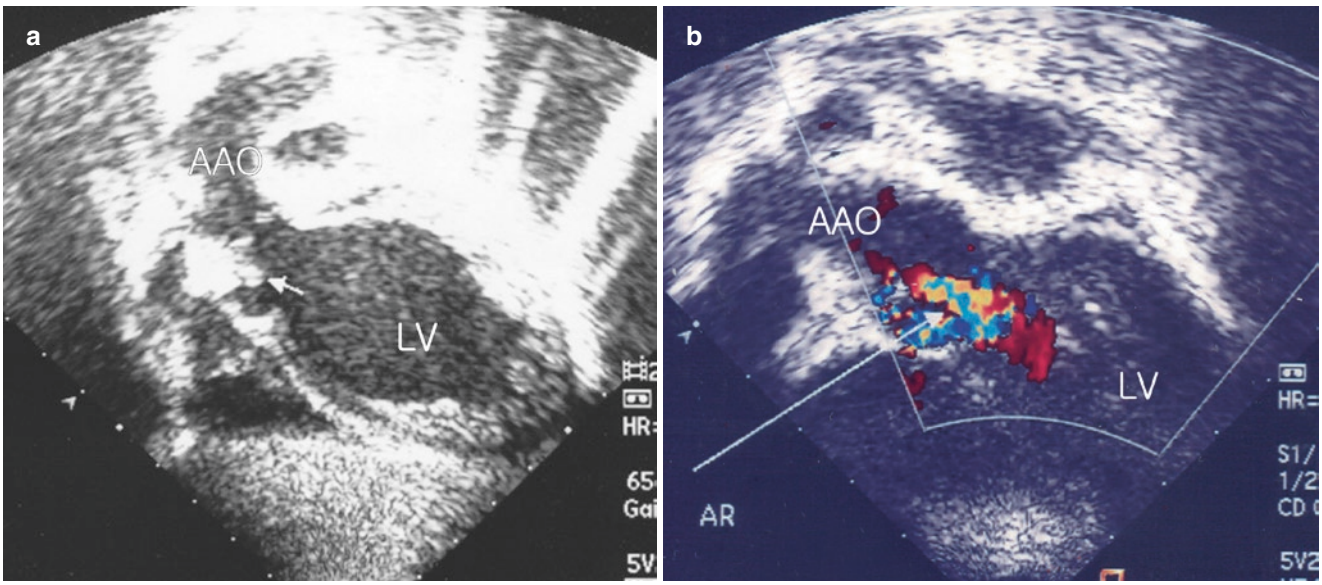

Fig. 8.48 A subcostal LVOT view from a 20-month-old boy who was referred with a diagnosis of "subarterial VSD and aortic regurgitation." (**a**) The arrow indicates the opening of the LV-aorta tunnel on the LV side, which could be misinterpreted as "VSD." (**b**) Color Doppler from the same view showing a diastolic jet coming into the LV cavity, similar to AR jet. However, unlike AR jet, this diastolic jet originates from the "hole" in the LV wall, instead of the aorta

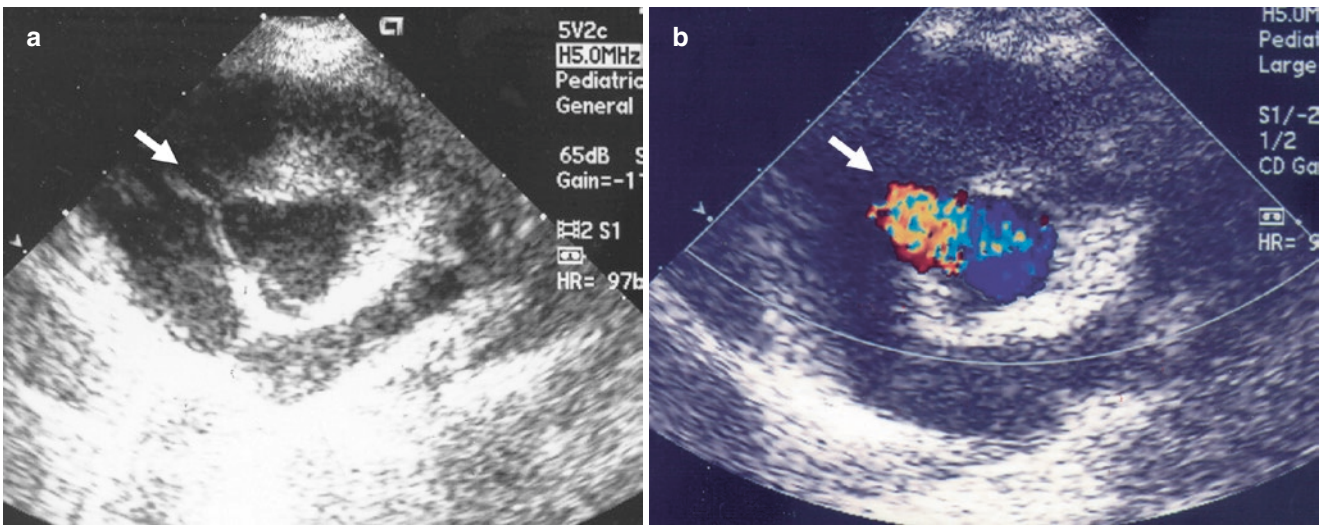

Fig. 8.49 A parasternal short-axis view (**a**) from the same patient showing an aortic opening of the LV-aorta tunnel (arrow). Color Doppler image (**b**) from the same view showing color jet passing through this opening, clearly different from AR jet

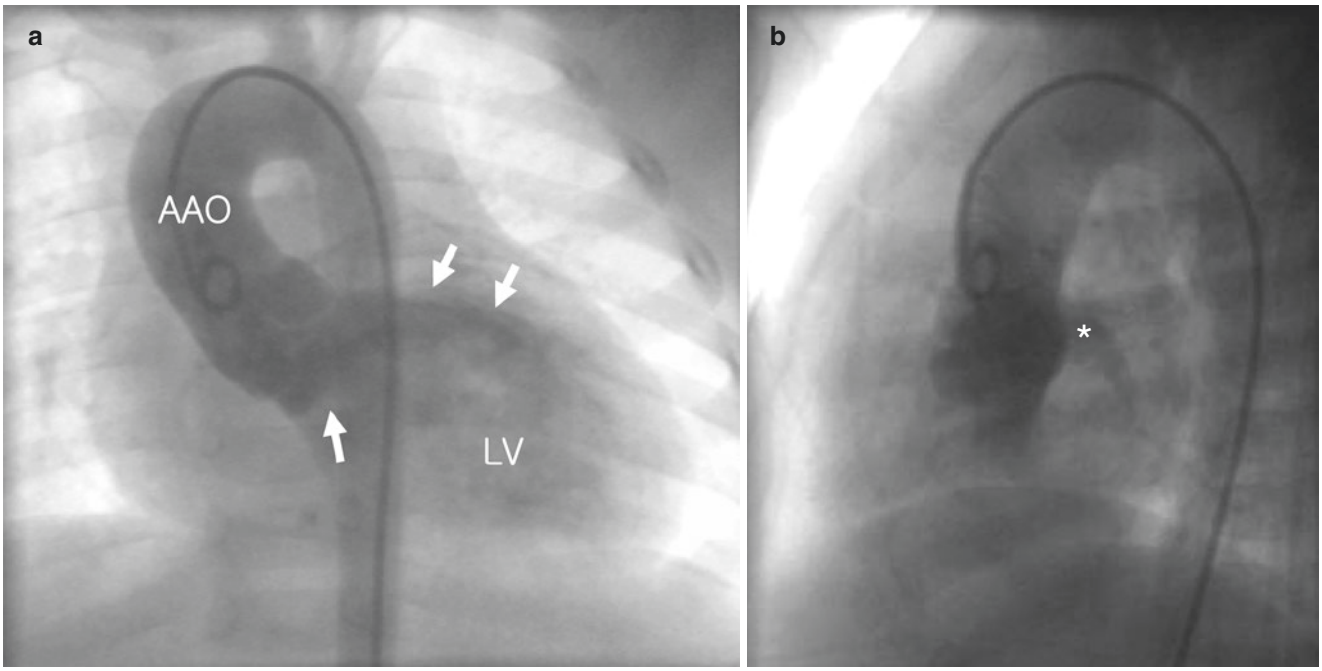

Fig. 8.50 AP (**a**) and lateral views (**b**) of an LV angiogram from the same patient showing flow through the LV-aorta tunnel (*). The lower arrow indicates the aortic opening of the tunnel, and upper arrows indicate the course within the LV myocardium and LV side opening of the tunnel

Pearls and Pitfalls

- The quality of the heart murmur of AS is similar to that of PS. However, they can be differentiated by the location of maximum intensity. PS murmur is loudest at the left upper sternal border and radiates to both lung fields, while AS murmur is loudest at the right upper sternal border and suprasternal notch, and radiates along the carotid arteries.
- The severity of stenosis correlates with the loudness of the heart murmur in AS and PS as long as ventricular function is normal.
- Diagnosis of critical AS in neonates and infants can be missed because heart murmur is typically not loud when LV function is decreased. The maximum flow velocity by CW Doppler across the aortic valve may not be high when LV function is impaired.
- Unlike valve PS, valve AS tends to worsen with time.
- Strenuous exercise should be avoided in patients with moderate-severe AS. Isometric exercise, such as weight lifting, is particularly more dangerous than dynamic exercise.
- When a patient is found to have supravalve AS, it is necessary to perform a chromosome study, mutation analysis on elastin gene, and echocardiography on family members.
- In Williams syndrome, supravalve AS tends to worsen, while peripheral PS tends to improve with time.
- The treatment of choice for valve AS is controversial, and the mode of treatment should be determined on an individual basis, as no treatment modality is problem-free.

Suggested Reading

1. Chartrand CC, Saro-Servando E, Vobecky JS. Long-term results of surgical valvuloplasty for congenital valvar aortic stenosis in children. Ann Thorac Surg. 1999;68:1356–60.
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4. Leung MP, McKay R, Smith A, et al. Critical aortic stenosis in early infancy. Anatomic and echocardiographic substrates of successful open valvotomy. J Thorac Cardiovasc Surg. 1991a;101:526–35.
5. McElhinney DB, Petrossian E, Tworetzky W, et al. Issues and outcomes in the management of supravalvar aortic stenosis. Ann Thorac Surg. 2000;69:562–7.
6. Merla G, Brunetti-Pierri N, Piccolo P, et al. Supravalvular aortic stenosis: elastin arteriopathy. Circ Cardiovasc Genet. 2012;5:692–6.
7. Parsons MK, Moreau GA, Graham TP Jr, et al. Echocardiographic estimation of critical left ventricular size in infants with isolated aortic valve stenosis. J Am Coll Cardiol. 1991;18:1049–55.
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