

2

Left Ventricular Inflow Obstruction

2.1. Cor Triatriatum

Cor triatriatum is a rare congenital cardiac anomaly in which a membranous structure divides the left atrium into two chambers. The distal chamber consists of pulmonary veins and the proximal chamber includes true left atrium and left atrial appendage. The communication between the pulmonary venous chamber and the rest of the atrium is usually restrictive; therefore, it commonly results in pulmonary venous congestion. The most common associated lesion with cor triatriatum is atrial septal defect. The atrial septal defect can either communicate with the pulmonary venous chamber or with the proximal chamber. A large secundum atrial septal defect or even two separate defects can communicate with the two chambers.

Adult patients with cor triatriatum who have no or mild symptoms may have very mild obstruction at rest. Obstruction may become obvious during stress and an increase in heart rate. When there is significant obstruction, patients usually present with secondary pulmonary hypertension. Associated mitral valve regurgitation can also be present. The mechanism for this is not well understood.

2.1.1. Echocardiographic Examination

Parasternal long-axis view shows a linear structure in the left atrium. Apical four chamber view demonstrates a membranous like structure in the left atrium that usually originates from the upper portion of the left lateral atrial wall. The insertion point on the septum is variable. The opening of the membrane can often be seen on two-dimensional images and color flow Doppler confirms its location and diameter. The degree of inflow narrowing can accurately be assessed by pulsed wave Doppler velocities. A low velocity of less than 1.2 m/s that demonstrates early and late diastolic components suggests no obstruction. In contrast, high early diastolic velocity that has lost its late component together with a dilated pulmonary venous chamber or large left-to-right shunt between pulmonary venous chamber and right atrium indicates significant obstruction.

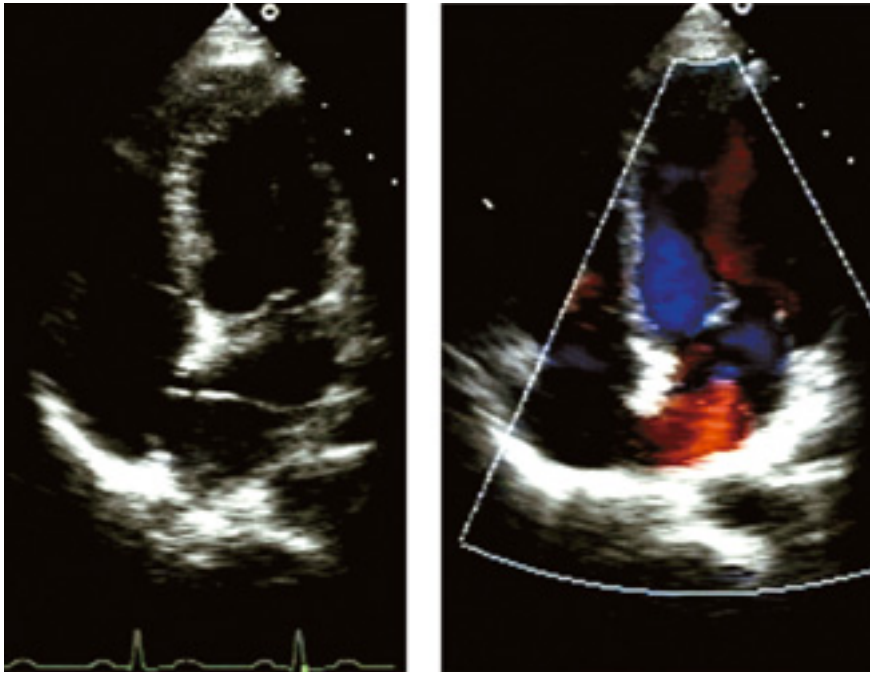


FIGURE 2.1. Apical four chamber view from a patient with a membranous structure in the left atrium.

Increased pulmonary artery pressure, which can be assessed from the peak tricuspid regurgitation velocity, is an indirect sign of significant obstruction. Patients with mild obstruction but dilated pulmonary venous chamber may develop significant obstruction during exercise. This can be clearly demonstrated by pharmacological stress echocardiography (see [Figures 2.1](#) and [2.2](#)).

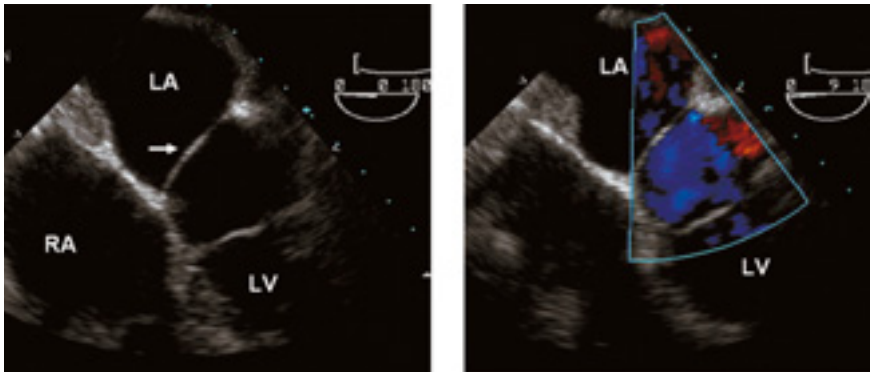


FIGURE 2.2. Transesophageal echocardiogram from a patient with cor triatriatum demonstrating an intra-atrial membrane that divides the left atrium into two chambers.

2.2. Supramitral Valve Membrane

This is a very rare condition in which an abnormally growing membrane between the left atrial appendage and the mitral valve leaflets is present. The level of the high velocity on color flow Doppler is usually shown above the mitral valve orifice level in the inflow tract. In patients with supramitral valve membrane, the mitral valve leaflets themselves are usually normal in anatomy and function. The parasternal long-axis view and apical four chamber view are ideal to demonstrate the lesion.

2.2.1. Management

Management of cor triatriatum and intra-atrial membrane is usually by surgical removal of the membrane as well as repair of the atrial septal defect.

2.3. Mitral Stenosis

Isolated mitral valve stenosis is one of the rarest forms of congenital heart disease. In adults, a variety of abnormal valve anatomy may be present and may contribute to the obstruction of blood flow from left atrium to left ventricle.

2.3.1. Double Orifice Mitral Valve

Double orifice mitral valve is a rare condition, often associated with atrioventricular septal defect. It is quite common for one of the two orifices to be either stenotic or regurgitant. If the diagnosis is suspected, the anatomical and physiological function of both orifices should be assessed. Double orifice mitral valve could remain a silent anomaly for years until valve incompetence becomes severe. At this stage valve repair may be considered in order to protect the left ventricle from functional deterioration.

Parasternal long and short-axis views of the mitral valve leaflets demonstrate the two orifices. In some cases, it may be difficult to demonstrate the two orifices in one echo plane (see [Figure 2.3](#)).

2.3.2. Parachute Mitral Valve

While normal mitral valve leaflets are supported by the anteromedial and posterolateral papillary muscles, parachute mitral valve is supported by either one papillary muscle, two fused papillary muscles, or three chordae attached to one head of papillary muscle. Although parachute mitral valve leaflets are anatomically abnormal, their function may be completely normal or only mildly stenosed.

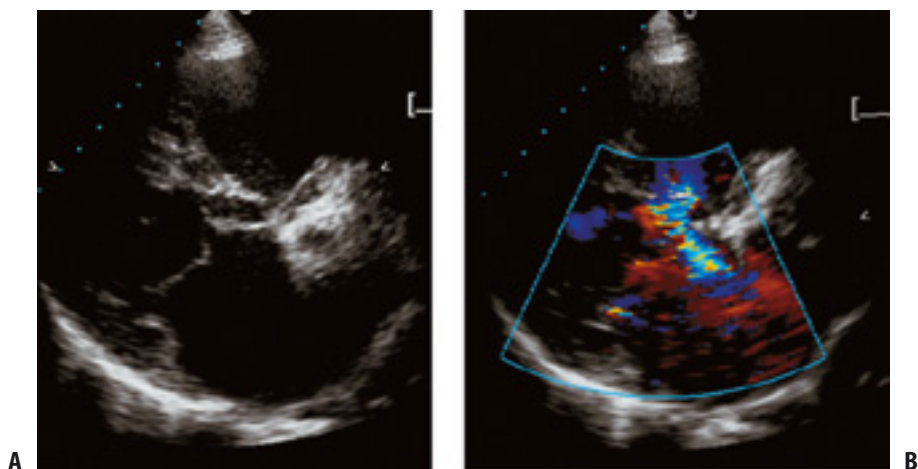


FIGURE 2.3. (A) Parasternal long-axis and short-axis views from a patient with double orifice mitral valve. (B) Note the presence of an incompetent orifice.

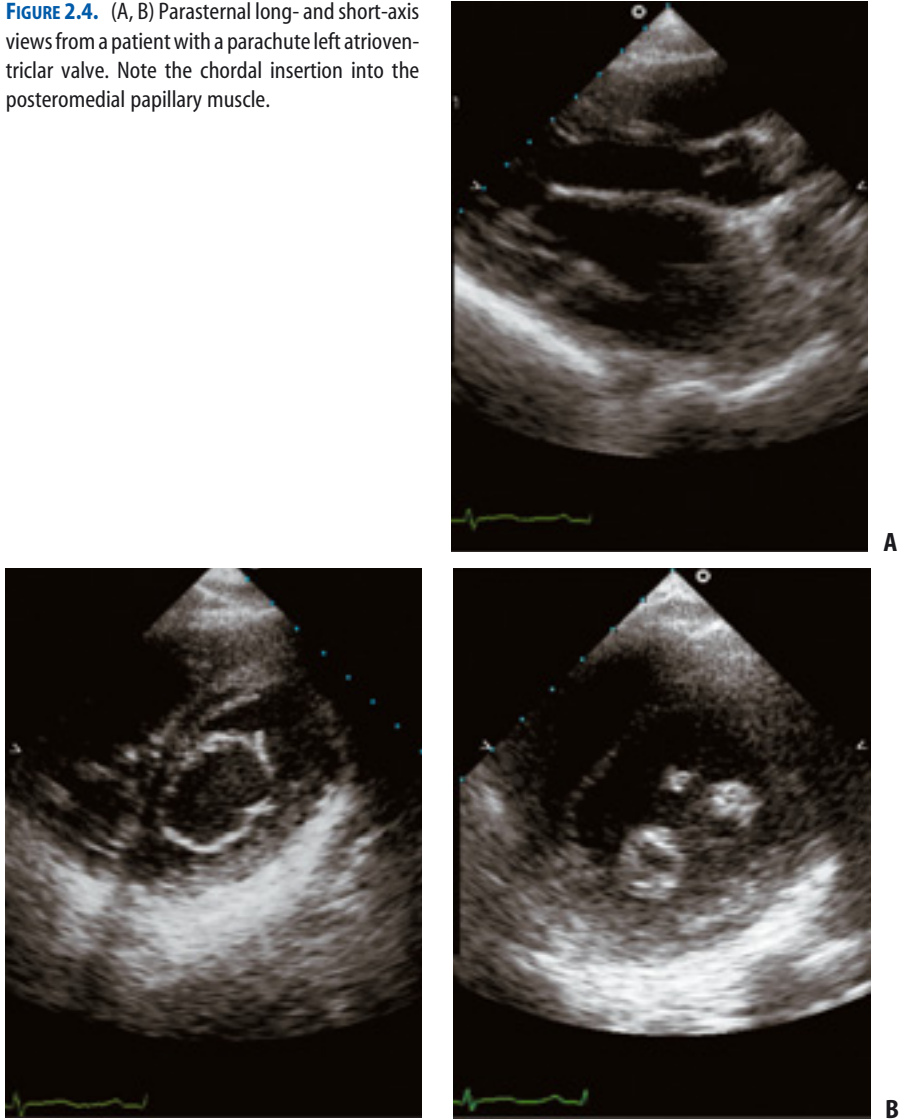
Parasternal long-axis view and apical four chamber view demonstrate the limitation of valve leaflets opening and subvalve narrowing. Short-axis view can demonstrate the chordal insertion into a single papillary muscle. Color Doppler shows the level of increase of flow velocity, not only at leaflets level, but also at subvalvar level (see [Figures 2.4 and 2.5](#)).

2.4. Mitral Valve Prolapse

Mitral valve prolapse is defined as the systolic billowing of one or both mitral valve leaflets into the left atrium. Congenital mitral valve prolapse is the most common cause of mitral regurgitation in adults. This lesion can be seen in isolation or in association with other anomalies, such as atrial septal defect or Marfan syndrome. Mitral valve prolapse and resulting regurgitation can be of variable degrees.

In patients with mild mitral valve prolapse, a midsystolic click is usually heard. It is caused by the backward mitral leaflet displacement in midsystole. This is often associated with late systolic mitral regurgitation, which is almost always mild in severity. Severity of mitral leaflet prolapse does not determine severity of mitral regurgitation. With severe regurgitation the murmur is usually short and peaks in midsystole (see [Figure 2.6](#)).

FIGURE 2.4. (A, B) Parasternal long- and short-axis views from a patient with a parachute left atrioventricular valve. Note the chordal insertion into the posteromedial papillary muscle.



2.4.1. Assessment of Mitral Valve Regurgitation

2.4.1.1. Anatomical Diagnosis

This is easily achieved from transthoracic echocardiographic images and identification of the part of the leaflet that is prolapsing, in particular on the parasternal long-axis view. Good short-axis images may assist in determining the prolapsing scallop and its progressive dysfunction during follow up.

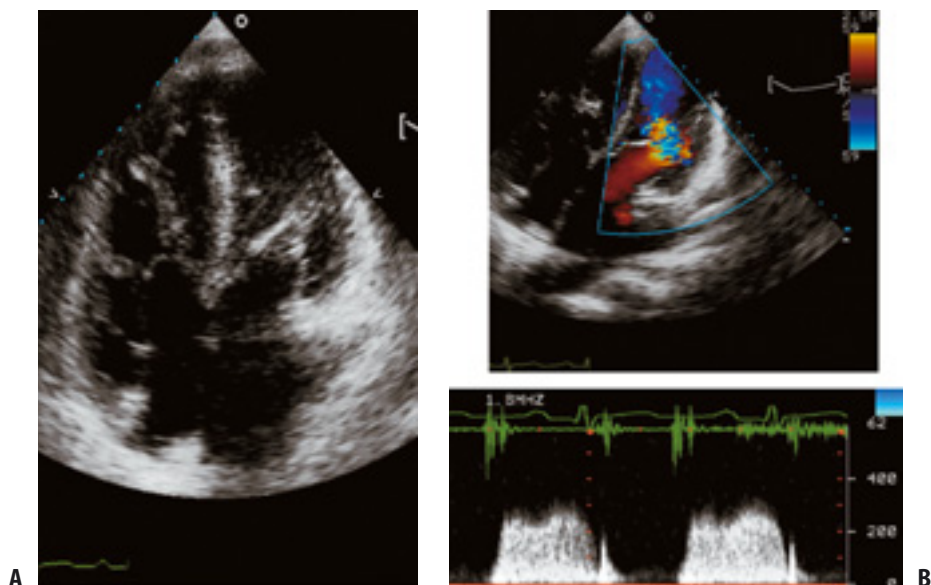


FIGURE 2.5. (A, B) Apical four chamber view of the same patient showing limited opening of the left atrioventricular valve. Color flow map showing turbulent flow starting at subvalve level and continuous wave Doppler demonstrating significant pressure gradient and important stenosis.

Transesophageal echocardiographic images may also help in delineating clearer images and accurate assessment of the exact prolapsing portion of the leaflet. Detailed assessment of the valve structure and function is crucial particularly when considering valve repair (see [Figures 2.7](#) and [2.8](#)).

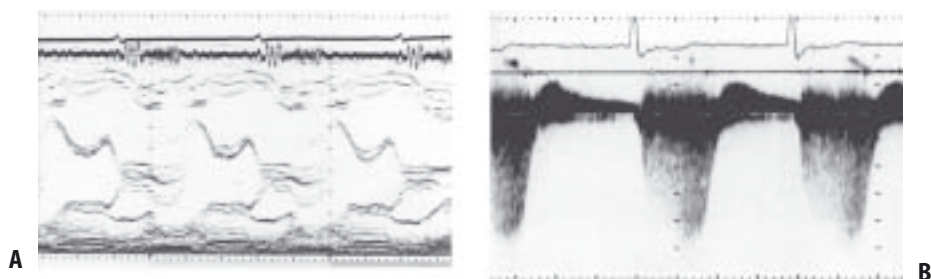


FIGURE 2.6. (A) M-mode echogram of the mitral valve prolapsing and (B) continuous wave Doppler showing late systolic mitral regurgitation, suggesting mild degree of regurgitation.

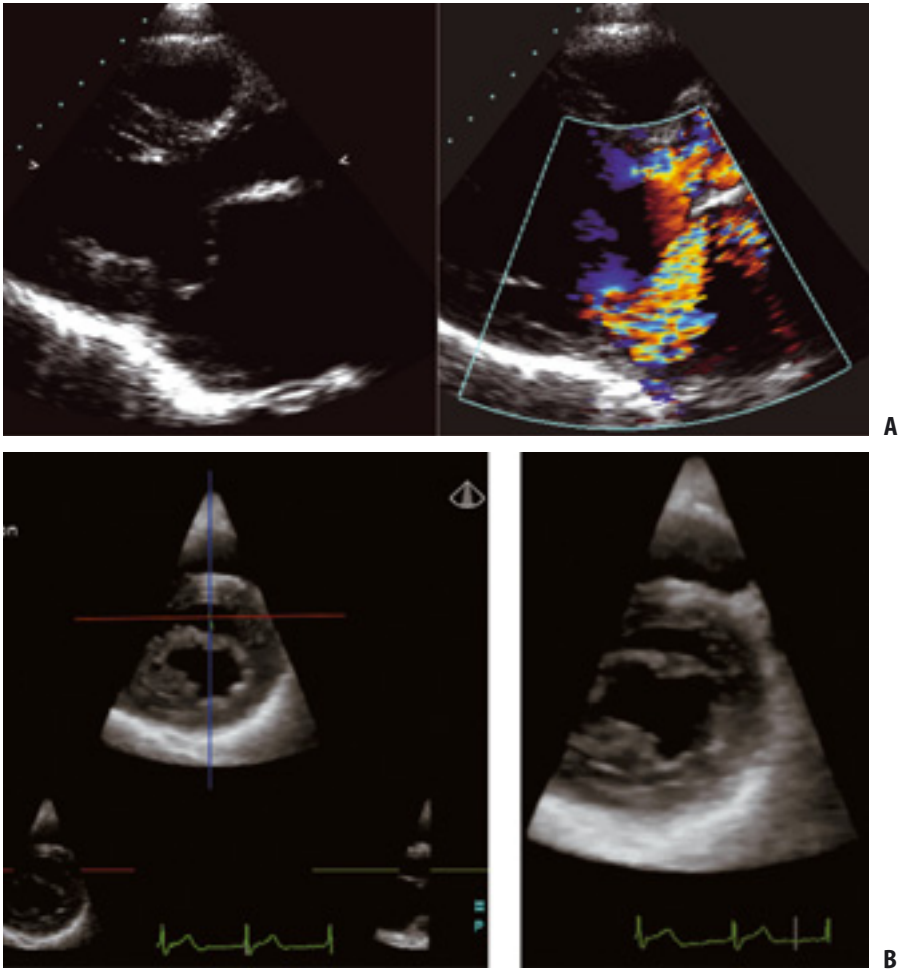


FIGURE 2.7. (A, B) Parasternal long-axis and three-dimensional short-axis view of the mitral valve from a patient with anterior mitral valve leaflet prolapse. Note the prolapsing mid-third of the anterior leaflet and resulting mitral regurgitation.

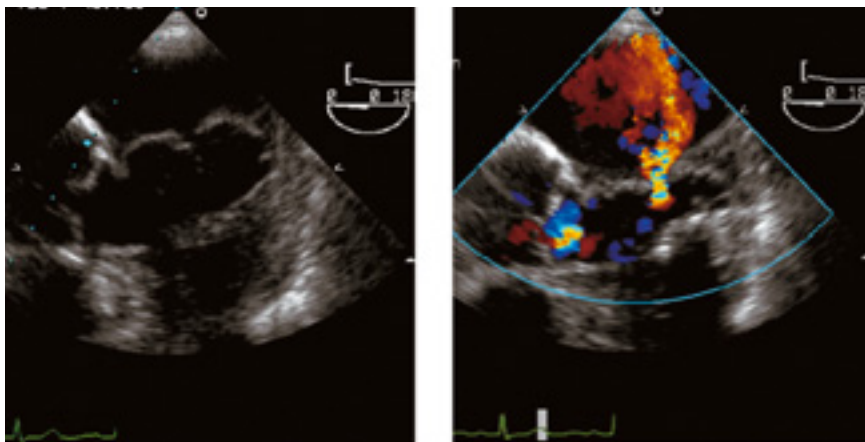


FIGURE 2.8. Transesophageal echocardiogram demonstrating posterior mitral valve leaflet prolapse causing mitral regurgitation.

2.4.1.2. Physiological Diagnosis

2.4.1.2.1. Color Flow Doppler

The degree of mitral regurgitation can be assessed by color flow Doppler based on:

1. **Jet Diameter.** A jet diameter at the leaflet tip level (vena contracta) broader than 5 mm suggests significant mitral regurgitation.
2. **Jet Area.** A mitral regurgitation jet area of more than 35% of the left atrial area is consistent with significant mitral regurgitation.
3. **Proximal Isovelocity Convergence Area (PISA).** This physiological principle estimates the degree of mitral regurgitation by measuring the diameter of the proximal velocity convergence at the leaflet tip level. This technique has its limitations, patients with displastic leaflets and those with distorted orifice may fail to demonstrate a clear spherical velocity convergence zone.

A common case for this is an eccentric mitral regurgitation jet, which is a common finding in patients with mitral valve prolapse. Therefore, color flow Doppler assessment of mitral regurgitation severity if taken in isolation may be misleading in these patients (see [Figures 2.9](#) and [2.10](#)).

2.4.1.2.2. Continuous Wave Doppler

Continuous wave Doppler is a very reliable technique for assessing severity of mitral regurgitation. Mild mitral regurgitation recordings display a decelerating

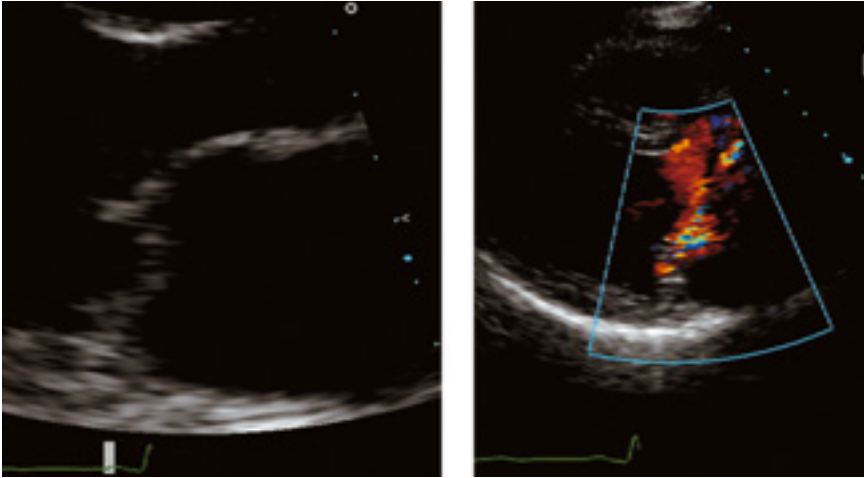


FIGURE 2.9. Parasternal long-axis view from a patient with posterior mitral valve leaflet prolapse that causes mild regurgitation with an anteriorly directed jet.

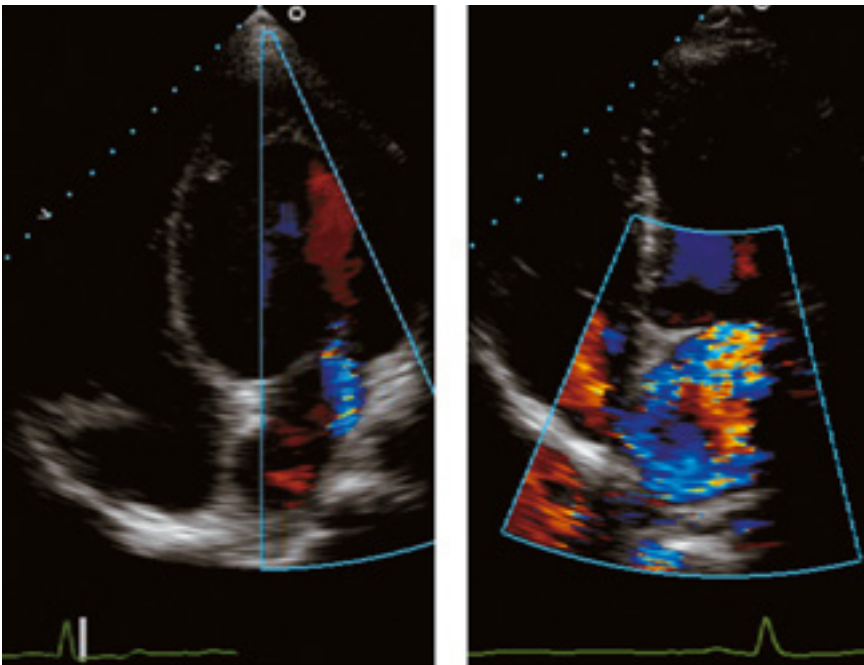


FIGURE 2.10. Apical four4 chamber view from two patients with mitral regurgitation on color Doppler, mild regurgitation (*left*) and severe regurgitation (*right*). Note the significant difference in jet diameter and area with respect to that of the left atrium.

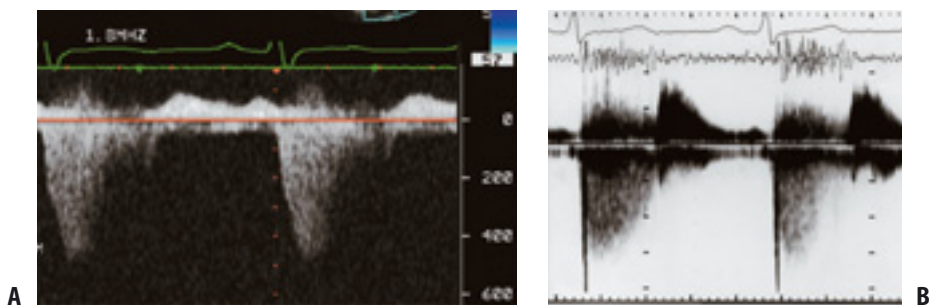


FIGURE 2.11. Continuous wave Doppler from a patient with (A) mild mitral regurgitation and another patient with (B) severe regurgitation. Note the difference in mitral regurgitation continuous wave Doppler signal, demonstrating early equalization of left atrial/left ventricular pressure, at end ejection in the patient with severe regurgitation.

pressure that extends well beyond end ejection (approximately 80 ms), whereas severe regurgitation demonstrates early equalization of left atrial and left ventricular pressures at end ejection (at the time of the second heart). Severe mitral regurgitation also results in suppression of systolic pulmonary venous flow component or even complete flow reversal. This sign should not be taken in isolation, particularly in patients with additional severe left ventricular systolic dysfunction. In them, mitral regurgitation peak pressure drop underestimates the driving left ventricular systolic pressure because of the raised left atrial pressure. Left atrial pressure in these circumstances can be estimated by subtracting the peak mitral regurgitation pressure drop from that of the systolic blood pressure (see [Figure 2.11](#)).

2.4.1.2.3. Left Ventricular Activity

Chronic overload of mitral regurgitation results in progressive increase in left ventricular dimensions, particularly in diastole. End systolic diameter always falls due to the large stroke volume, thus making conventional measurements of left ventricular systolic function from fractional shortening or ejection fraction erroneous. In patients who develop significant left ventricular disease and increase of end systolic volume, assessment of mitral regurgitation based only on left ventricular activity may underestimate severity of ventricular dysfunction (see [Figure 2.12](#)).

2.4.1.2.4. Management of Mitral Regurgitation

Mild mitral regurgitation is generally well tolerated by patients as long as left ventricular function is maintained. Management of severe mitral regurgitation

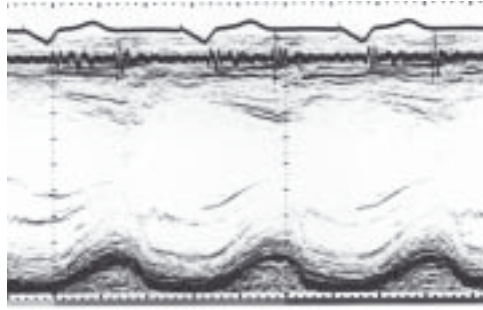


FIGURE 2.12. M-mode images of left ventricle cavity from a patient with severe mitral regurgitation. Note dilated left ventricle cavity size and active wall motion.

due to mitral valve prolapse is almost always by surgical leaflet repair with or without ring insertion, particularly in patients with posterior leaflet prolapse. Results of anterior leaflet repair is now very satisfactory in well-selected cases. In patients with chronic atrial fibrillation and severely dysmorphic mitral leaflets, valve replacement may be a better option. Simultaneous ablation of pulmonary veins has become a regular practice in these patients as an attempt to provide simultaneous treatment for the atrial arrhythmia.

2.4.1.2.5. Intraoperative Echocardiography

Intraoperative echocardiographic monitoring of mitral valve surgery has become a routine practice in most cardiac centres. Perioperative assessment of valve anatomy by transesophageal approach provides detailed anatomical imaging. Furthermore, any additional pathology that might have been missed preoperatively can be studied and assessed. A final careful examination of the valve function before closing the chest is crucial for excluding any residual lesion that should not be left uncorrected (see [Figure 2.13](#)).

2.4.1.2.6. Postoperative Follow Up

Transthoracic echocardiography provides an ideal means for postoperative follow up of patients after mitral valve surgery. The degree of residual or progressive valve regurgitation is assessed by different echocardiographic techniques, as mentioned above. Also, left ventricular dimensions, function, and activity are all assessed and quantified. In the absence of recurrent mitral regurgitation, significant increase in left ventricular end systolic volume or left atrial diameter represent increased ventricular stiffness and raised left atrial pressure as a possible cause for patient's breathlessness.

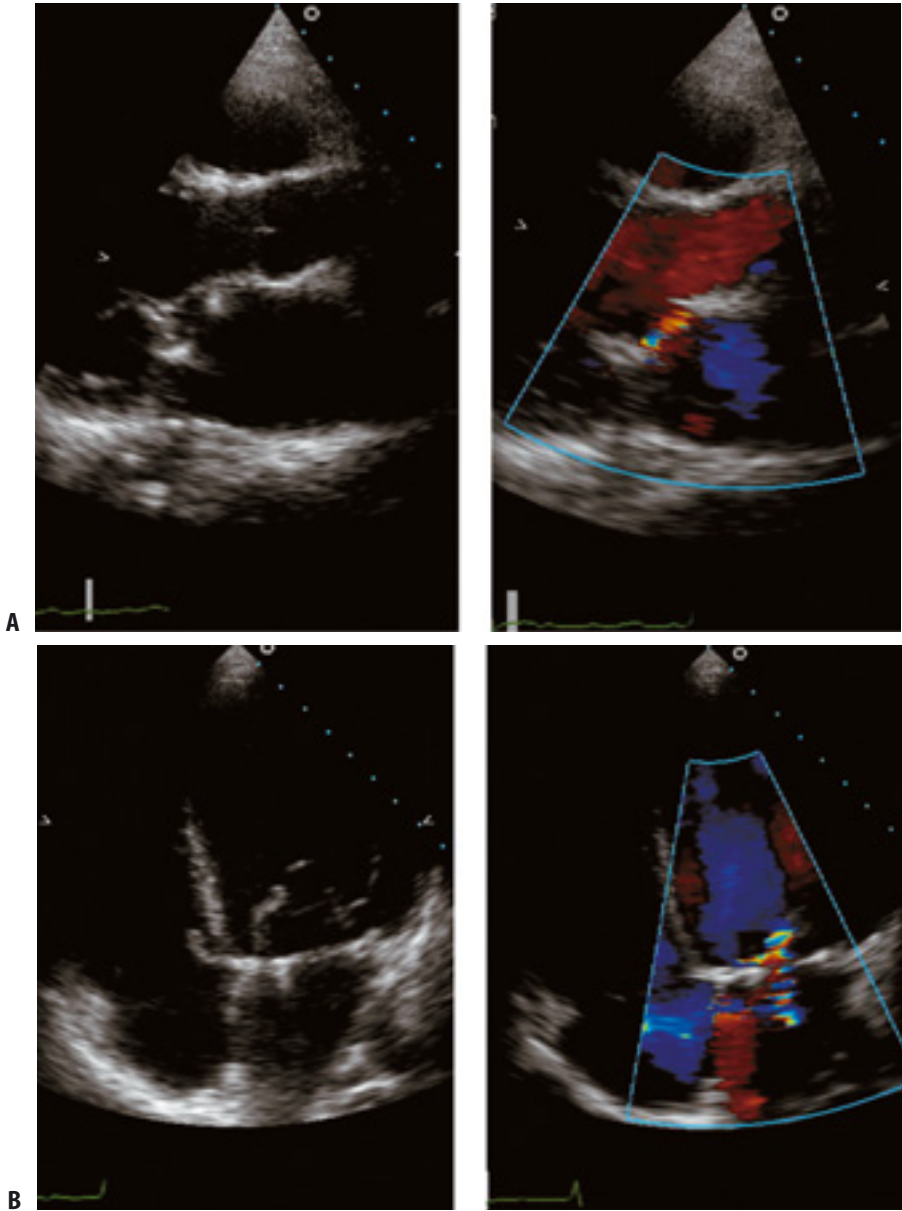


FIGURE 2.13. (A) Parasternal long-axis view and (B) apical four chamber view of the mitral valve from a patient after valve repair and insertion of a ring. Note the optimum repair and the valve competence postoperatively.

2.5. Partial Anomalous Pulmonary Venous Connections

Partial anomalous pulmonary venous connection is defined as the connection of one or more pulmonary veins to a site other than the morphological left atrium. It can occur as an isolated anomaly or in association with other intra-cardiac malformations. Here, we mainly discuss the anomalous connection in the setting of usual atrial arrangement. The connection can be described as totally or partially anomalous and unilateral or bilateral. When it is total, all four veins drain to the systemic circulation; when partial, some of the veins drain to the pulmonary circulation. The sites of anomalous connection are conventionally described as supracardiac, cardiac, and infracardiac. The most common type of supracardiac connection is for the pulmonary veins to join a confluence that drains through a common ascending vein to the brachiocephalic vein or directly to the right superior caval vein. The connection is described as cardiac when the veins insert directly into the right atrium or into the coronary sinus. In the infracardiac connection, the pulmonary veins usually converge to a horizontal confluence from which a vertical vein descends alongside the esophagus. This is also the type of connection most prone to stenosis and most challenging to repair.

The display of pulmonary venous anatomy may be complex, requiring interrogation from a number of transducer locations and planes. In the normal heart, the pulmonary veins may be seen from the subcostal, parasternal short-axis, suprasternal coronal, and apical four chamber views. In infants and small children, the pulmonary veins can be identified most readily from the subcostal location in the coronal view.

2.5.1. Subcostal Imaging

When the drainage site is below the diaphragm, the confluence tends to lie just below the diaphragm and is usually imaged with caudal angulation. The descending vein through the diaphragm usually drains through the esophageal hiatus. This route is anterior to and slightly rightward to the descending aorta, and posterior to and slightly leftward of the inferior vena cava. Thus, in this form of anomaly, three vascular structures drain through the diaphragm. When pulmonary venous drainage is intracardiac, either via the coronary sinus or directly into the right atrium, the draining confluence and the veins draining into it can be seen with more cranial angulation. With supracardiac pulmonary venous drainage, one must direct the subcostal transducer plane in the most cranial angulation. In addition to the confluence, it is important to identify all pulmonary veins draining into it. This can be aided by color flow Doppler and mapping of the region. When the drainage is to the superior caval system or directly to the right atrium, the plane must be directed in a most cranial direction from this transducer location.

2.5.2. Parasternal Imaging

The high parasternal short-axis view provides an alternative approach to image the pulmonary veins and the left and the right atria. The pulmonary veins lie between the pulmonary artery and left atrium and can be identified with cranial–caudal rocking of the transducer. Parasternal long-axis imaging is helpful for identifying the large coronary sinus. Color flow Doppler assists in identifying the exact location of the veins.

2.5.3. Suprasternal Imaging

In the suprasternal approach to imaging the anomalous pulmonary veins, their connection to a confluence and the site of drainage can also be achieved from the suprasternal notch. This may provide an excellent opportunity to image the vertical vein as it runs cranially or even to see its descent through the diaphragm.

2.6. Seimitar Syndrome

Seimitar syndrome is characterized by partial anomalous pulmonary venous drainage. The right-side pulmonary veins drain into the inferior vena cava and the left-side pulmonary veins usually drain into the left atrium. The right lung is almost always hypoplastic or completely absent, as well as the right bronchus. This syndrome is always associated with atrial septal defect and right cardiac chamber dilatation. Due to the hypoplastic right lung, the heart and the mediastinum tend to be shifted to the right.

2.6.1. Management

Management of Seimitar syndrome is by surgical repair of pulmonary venous drainage and the associated anomalies.

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