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## Keywords

Congenital Mitral Stenosis • Parachute deformity • Supra valvular ring • Mitral valve regurgitation • Balloon valvuloplasty

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## Morphology and Associated Lesions

Congenital mitral stenosis (MS) refers to the parachute deformity of the valve. In this condition, condensed chordae tendineae join and attach into a single large papillary muscle, usually with thickened leaflets of the mitral valve (MV), fusion of the chordae tendineae, accessory MV tissue, and supralvalvular ring of the connective tissue rising at the base of the atrial aspect of the MV leaflets. Associated cardiac lesions are common and consist of coarctation of the aorta, patent ductus arteriosus, left ventricular outflow tract obstruction, and persistent left superior vena cava [1].

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## Clinical Findings

Findings and diagnosis are incidental in many cases at the time of the study for other left-sided obstructions such as coarctation of the aorta and/or aortic valve stenosis. The typical auscultatory findings seen with rheumatic MS are frequently absent in the congenital type of this lesion. Classic findings consist of normal S1 and mid-diastolic murmur with or without some presystolic highlighting [1–3].

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## Electrocardiography

In mild forms, electrocardiography is generally normal. However, there may be left atrial overload and enlargement, with or without right ventricular hypertrophy due to related pulmonary hypertension [1, 4].

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**Electronic supplementary material** The online version of this chapter (doi: [10.1007/978-1-4471-6383-1\\_40](https://doi.org/10.1007/978-1-4471-6383-1_40)) contains supplementary material, which is available to authorized users.

## Chest Radiography

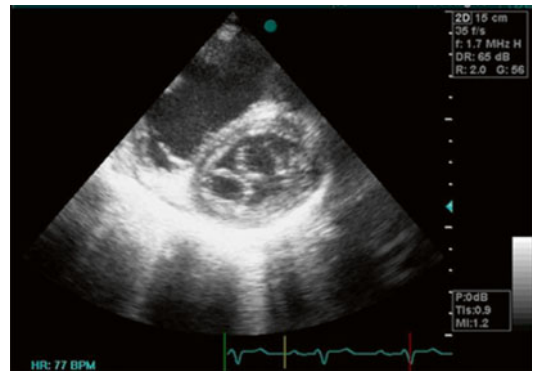
Chest X-ray is normal in mild types, with signs of pulmonary edema in patients with more severe MS [1].

## Echocardiography

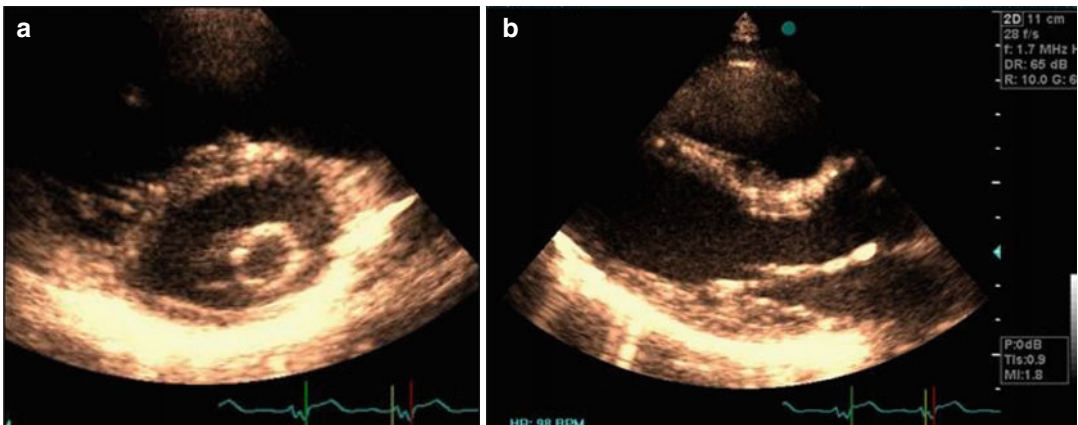
Two-dimensional and more recently three-dimensional echocardiography, concomitant with color and Doppler assessment, generally provides a comprehensive study of the anatomy and function in patients with congenital MS. The form, shape, and location of the papillary muscles are best seen in the parasternal short-axis view. In congenital MS, if both papillary muscles are present, usually they are closer together than is seen in the normal heart. The four-chamber view and also parasternal long-axis view permit the diagnosis of supra-valvular mitral rings. Color flow Doppler helps determine the level of stenosis and obstruction and also confirm the presence of MV regurgitation. Also, Doppler study provides a precise study of the mean gradient and pressure half time through the MV. 2D transthoracic echocardiography in parasternal short-axis view can determine the number of papillary muscles and mitral valve anatomy [1, 3–6] (Figs. 40.1a, b and 40.2).

## Management

Clinical and echocardiographic follow-up is all that is required in asymptomatic cases. When the patient begins to have increased pulmonary pressure or develop symptoms, surgical approach is generally indicated. MV balloon valvuloplasty is not as successful as it is in rheumatismal MS. Surgery removes the supra-mitral ring if it is present. Also, the splitting of the papillary muscles and fused chordae is indicated in cases with more common types of congenital MS. In general, surgery provides short-term relief, with many cases demanding MV replacement later [1, 5, 6].



**Fig. 40.2** Transthoracic echocardiography in short-axis view showing divided mitral orifice into two components *DOMV* double orifice mitral valve (Video 40.1)



**Fig. 40.1** (a, b) Parachute mitral valve with dominant anterior papillary muscle by 2D echocardiography in short- and long-axis views

## References

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