

SHONE COMPLEX

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A 10-year-old child was referred to our department for the evaluation of dyspnea on effort of New York Heart Association functional class III. General examination was unremarkable. Cardiac examination revealed a normal first heart sound, loud second heart sound, no opening snap, a mid-diastolic and a pansystolic murmur at the apex and a grade III/IV ejection systolic murmur at the right second parasternal area. Two dimensional transthoracic echocardiography with color Doppler showed the presence of a partial supra mitral ring and a parachute mitral valve (PMV) leading to severe sub valvar mitral stenosis along with severe eccentric mitral regurgitation (Fig. 1A, B, and C, Supplementary movie 1 and 2). There was also a subaortic membrane which resulted into severe subvalvar aortic stenosis (Fig. 1A). The aortic valve was quadricuspid (Fig. 1D) and there was no coarctation of aorta. Based upon these findings, a diagnosis of a partial form of Shone complex was made. Three dimensional (3D) transthoracic echocardiography clearly delineated the unifocal attachment of the PMV as well as the slit like opening in the inter chordal region that caused severe subvalvar mitral stenosis (Fig. 2A and B, Sup-

plementary movie 3). The near-circular morphology of the sub aortic membrane with a central opening was also evident on 3D echocardiography when viewed in the parasternal orientation (Fig. 2C, Supplementary movie 4) and from the left ventricular side (Fig. 2D). Patient is now planned for surgical intervention with possible repair of the mitral valve and resection of sub aortic membrane.

Shone et al.¹⁾ first described a complex congenital cardiac anomaly in 1963 which included PMV, supra mitral ring, sub aortic stenosis and coarctation of aorta. Partial forms of Shone complex are also described where only two or three out of four obstructive lesions are present. A good outcome is possible in patients with this rare anomaly if the surgical intervention is undertaken early before the onset of pulmonary hypertension.²⁾ Precise anatomical imaging of the PMV as well as the sub aortic membrane is necessary before the surgical intervention. Our case demonstrates how cropping the 3D data set can give rise to unique imaging views for sub valvar structures like chordal attachments of the PMV as well as the near-circular morphology of the sub aortic membrane which were not

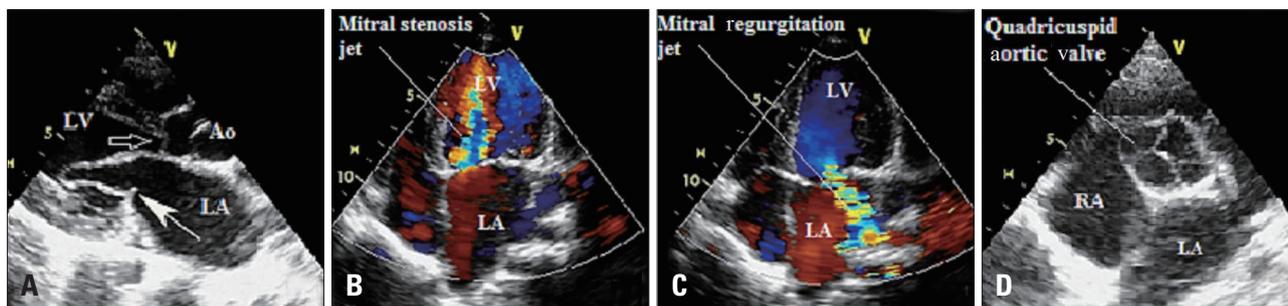


Fig. 1. Two dimensional echocardiography with color Doppler. Parasternal long axis view (A) showing the sub aortic membrane (open arrow), partial supra mitral ring attached near the posterior mitral annulus (white solid arrow) and abnormal anterior and posterior mitral leaflets. Diastolic frame in apical four chamber view showing severe degree of mitral stenosis with color flow turbulence starting at the supra mitral ring level (B). Systolic frame in apical four chamber view showing severe eccentric mitral regurgitation (C). The basal short axis view showing the quadricuspid nature of the aortic valve (D). Ao: aorta, LA: left atria, LV: left ventricle, RV: right ventricle.

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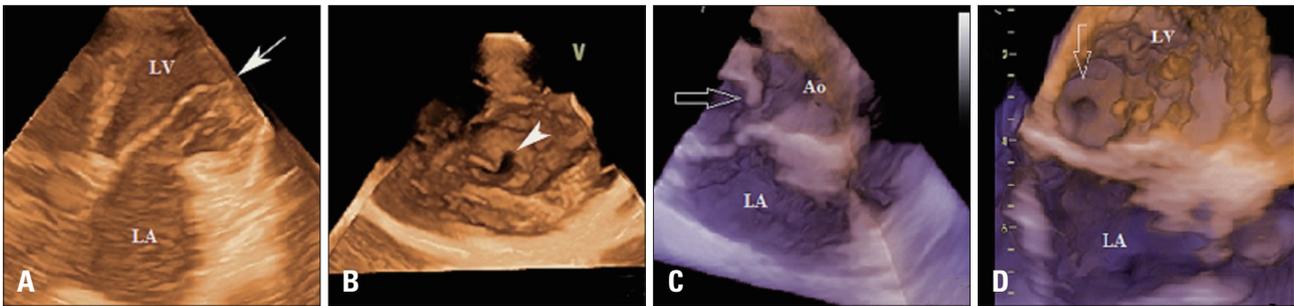


Fig. 2. Three dimensional transthoracic echocardiography with color Doppler. The unifocal attachment of the parachute mitral valve can be clearly delineated in apical two chamber orientation (arrow in A). The sub valvular mitral stenosis was in fact caused by a slit like opening (arrowhead in B) in the parachute mitral valve. Parasternal long axis image in a systolic frame (C) clearly showing the sub aortic membrane (arrow in C). Three dimensional echocardiography clearly showing the anatomy of the sub aortic membrane (arrow in D) with a near circular membrane morphology and a central opening as viewed from the LV side. Ao: aorta, LA: left atria, LV: left ventricle.

evident on two dimensional echocardiography.

SUPPLEMENTARY MOVIE LEGENDS

Movie 1. Two dimensional transthoracic echocardiography. Parasternal long axis view has been slightly modified showing the partial supra mitral ring near the posterior leaflet, parachute mitral valve with unifocal attachment and sub aortic membrane. The LA is dilated and there is concentric left ventricular hypertrophy. Ao: aorta, LA: left atria, LV: left ventricle.

Movie 2. Two dimensional transthoracic echocardiography. Apical 4 chamber view showing pear shaped mitral valve with turbulent flow across the sub mitral valve region indicative of mitral stenosis and eccentric jet of severe mitral regurgitation reaching posterior wall of LA. LA: left atria, LV: left ventricle.

Movie 3. Three dimensional transthoracic echocardiography. Full volume data set cropped in apical two chamber ori-

entation clearly showing the unifocal attachment of the parachute mitral valve.

Movie 4. Three dimensional transthoracic echocardiography. Real time image after cropping with depth colorization clearly showing the anatomical relationship and the morphology of the sub aortic membrane (green arrow). Ao: aorta, LA: left atria.

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