

Tri-leaflet mitral valves – when lightning strikes thrice

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Mitral valves are well known to be bi-leaflet structures with attachments from both leaflets (anterior and posterior) to both papillary muscles (anterolateral and posteromedial). Congenital abnormalities of the mitral valve, although well described, are quite rare. These abnormalities can involve either the leaflet (cleft mitral valve) or the subvalvular apparatus (parachute mitral valve) or even occur as accessory mitral valve tissue (accessory mitral valve leaflet). These can occur in isolation, or in association with other congenital abnormalities. A tri-leaflet mitral valve is a novel echocardiographic finding that has only been described in 6 patients in 4 different case reports.⁽¹⁻⁴⁾ We report on 3 patients recently found to have trileaflet mitral

valves in the setting of atrioventricular concordance and normal offset of the AV valves at our out-patient clinic.

The first patient is a 22-year-old male referred to our congenital heart disease clinic for follow-up of a restrictive VSD. Echocardiography revealed a quadricuspid aortic valve and an unusual appearance of the mitral valve. There were 3, evenly spaced commissures with central coaptation present (Figure 1a and b) as well as 3 papillary muscles – a medial, a small or rudimentary antero-lateral and a posteriorly positioned papillary muscle (Figure 1c). Despite these morphological abnormalities, only trace mitral regurgitation (MR) was detected. Three-dimensional (3D) echocardiography allowed for a more detailed morphological and functional assessment (Figure 1a).

The second patient is a 49-year-old male evaluated for chronic severe MR. Echocardiography confirmed a dilated, well-functioning left ventricle with severe MR due to a tri-leaflet mitral valve (Figure 2a). There were 3 (medial, lateral and posterior) papillary muscles present (Figure 2b) with chords to all 3 papillary muscles noted. There were no other structural abnormalities present.

The third patient is a 50-year-old male evaluated for chronic severe mitral regurgitation. Initially this was assessed as being due to prolapse, but more careful scrutiny confirmed a dilated, well functioning left ventricle with severe mitral regurgitation due to a tri-leaflet mitral valve (Figure 3a and b). Again there were 3 papillary muscles present (Figure 3c): a rudimentary basal medial PM, an anterolateral PM and a more apical posterior PM. Three-dimensional (3D) TOE echocardiography clearly showed the 3 commissures (Figure 3a).

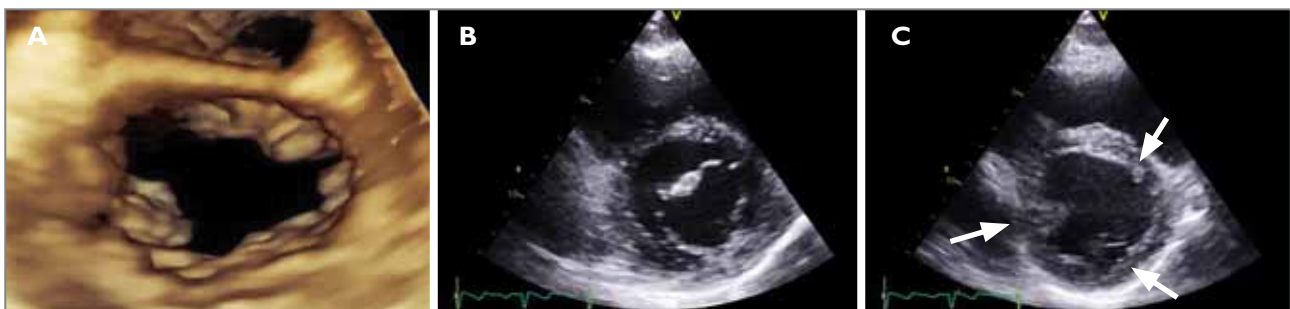


FIGURE 1: A 3D image from patient 1 obtained by TOE clearly shows the novel finding of a tri-leaflet mitral valve (A). Two-dimensional (2D) short axis images taken by TTE show 3 commissures (B) and 3 papillary muscles (C: white arrows).

The mitral valves of all 3 patients mimic the normal morphology of the tricuspid valve with 3 leaflets, 3 commissures, 3 papillary muscles (of which one is small/rudimentary) and chords from each papillary muscle (including from the rudimentary papillary muscle attaching to 2 separate leaflets). There appears to be a wide spectrum of functional impairment associated with these valves, ranging from only trace MR to chronic severe MR.

A literature search revealed only 4 case reports of this finding. The first case was a 13-year-old girl with a tri-leaflet mitral valve as well as severe subaortic stenosis. The presence of 3 papillary muscles associated with the mitral valve were also noted with mild regurgitation of the tri-leaflet mitral valve. The other 5 cases all occurred in the presence of hypertrophic obstructive cardiomyopathy with the MR ranging from mild to severe. The presence of 3 associated papillary muscles was confirmed in one case report, but its presence/absence was not reported on in the other 4 cases. These 5 cases were all identified incidentally on intra-operative trans-esophageal echo-cardiography (TOE)/3DTOE during surgical myectomy and it is therefore unclear if there is a true association.

The tri-leaflet mitral valve is a rare congenital abnormality that is increasingly being recognised with advances in imaging techniques. While 3 commissures and 3 papillary muscles are consistently present in our cases, there is a wide range in the functional impairment associated with these valves, ranging from essentially normal to severe regurgitation. It appears to be commonly associated with other congenital abnormalities, although it can occur in isolation.

Conflict of interest: none declared.

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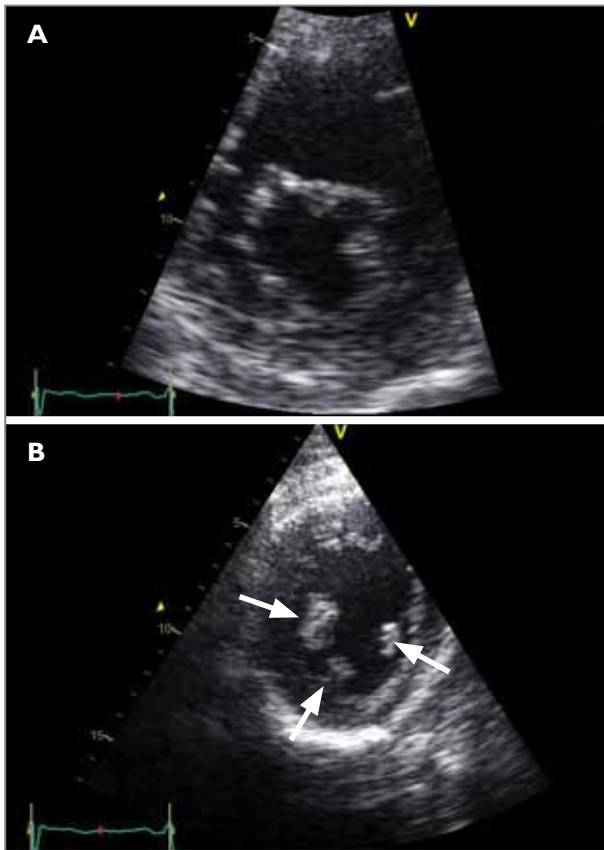


FIGURE 2: Short axis images obtained from patient 2 by TTE shows 3 commissures (A) and 3 papillary muscles (B: white arrows).



FIGURE 3: A 3D image obtained by TOE from patient 3 again shows 3 commissures (A: white arrows). This can also be seen on the short axis taken by TTE (B) with again 3 papillary muscles clearly visible (C: white arrows).