

Congenital Mitral Valve Regurgitation in Adults: Hammock or Arcade Mitral Valve

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The images correspond to a 28 year-old woman with congenital mitral valve disease. The patient is asymptomatic. She presents with severe mitral regurgitation (effective regurgitant orifice: 0.45 cm²; regurgitant volume: 78 ml), mild stenosis (area: 2.8 cm²), left ventricular diastolic diameter: 56.6 mm, systolic diameter: 39.7 mm, ejection fraction: 58%, systolic pulmonary pressure: 45 mm Hg, and sinus rhythm. The patient consults to evaluate her risk of becoming pregnant. Transthoracic and transesophageal echocardiography studies reveal that the posteromedial papillary muscle is larger and directly connected to the mitral leaflets without interposition of chordae tendineae, and the anterolateral muscle shows thick and short chords (Figure 1). A fibrous tissue bridge connecting the two leaflets is observed at the subvalvular level (Figures 1A and 2A). The posteromedial part of the mitral valve is completely fused and does not

open (Figure 3, Video 2). In systole, the leaflets are pulled by a thickened and shortened subvalvular apparatus and do not reach the closure level, causing severe mitral valve regurgitation (Figure 2 B, Video 1). Based on these findings, mitral arcade was diagnosed.

Congenital mitral valve disease is a rare entity, especially in the adult population, with a reported prevalence of 0.5%. (1) Mitral arcade, or hammock mitral valve, is a less prevalent congenital valve disease that involves the valve and the mitral tensor apparatus. This anomaly, described in 1967, (2) is characterized by elongated papillary muscles connected to one another and to the tip of the anterior mitral leaflet by a fibrous tissue bridge with no interposition of chordae tendineae or with very short and thick chords. This fibrous continuity restricts leaflet movement and interferes with the normal closure mechanism. (3, 4)

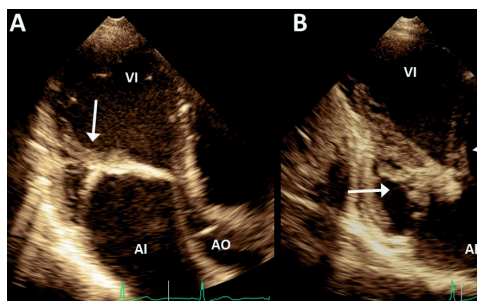


Fig. 1. A. Three-chamber apical view in diastole: restricted valve opening with the two leaflets connected by a fibrous tissue bridge (arrow). **B.** Two-chamber apical view in systole: posteromedial papillary muscle (long arrow) directly connected to mitral leaflets without interposition of chordae tendineae, and anterolateral papillary muscle (short arrow) with thick and short chords. Ao: Aorta. LA: Left atrium. LV: Left ventricle.

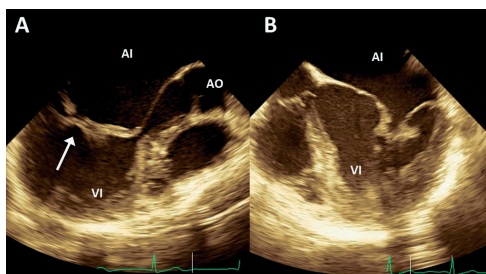


Fig. 2. Transesophageal echocardiography. **A.** Mid-esophageal view at 120° in diastole: anterior and posterior leaflets connected by a fibrous tissue bridge (arrow). **B.** Mid-esophageal view at 20° in systole: thickened and shortened tensor apparatus that prevents mitral leaflets from proper coaptation. Ao: Aorta. LA: Left atrium. LV: Left ventricle.

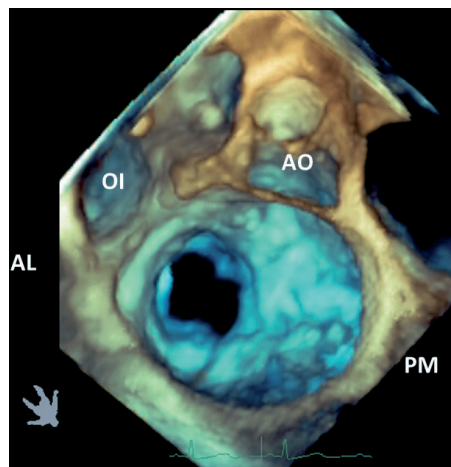


Fig. 3. Three-dimensional transesophageal echocardiography. View from the left atrium in diastole: total fusion and lack of opening of the mitral valve posteromedial part. AL: Anterolateral. AV: Aortic valve. LAA: Left atrial appendage. PM: Posteriomedial.

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