Utility of three-dimensional echocardiography for assessment of double-orifice mitral valve

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Double-orifice mitral valve is a rare congenital malformation of the mitral valve. A range of structural abnormalities of the subvalvular apparatus are found including a chordal ring, accessory papillary muscle, crossing/fused chordae tendineae, and a central fibrous subdivision creating two separate mitral valve orifices. More commonly, an iatrogenic double-orifice mitral valve is formed during percutaneous mitral valve repair (MitraClip). A congenital double-orifice mitral valve is associated with other congenital abnormalities including a bicuspid aortic valve, coarctation of the aorta, atrial and ventricular septal defects, and Ebstein’s anomaly. Approximately half of the patients have a functionally normal valve, although up to 25% of the patients have significant mitral regurgitation or stenosis. We present a 28-year-old male who previously underwent repair of aortic coarctation and replacement of stenotic bicuspid aortic valve.

Three-dimensional (3D) echocardiography was used to delineate the anatomical abnormalities of his mitral valve. (Panel A) Three-dimensional en face view of mitral valve allows identification of the two approximately equal size orifices (arrows) with a fibrous ridge (dashed arrow) dividing the two (see Supplementary data online, File S1). (Panel B) Three-dimensional sagittal plane. MV, mitral valve; LV, left ventricle. The posterior—medial papillary muscle with chordal attachments are visualized. An accessory papillary muscle and chordae is also identified. (Panel C) Three-dimensional transverse superior plane. Fusion of chordae tendineae and abnormal insertion of chordae tendineae are identified. Despite an abnormal subvalvular apparatus, there was only mild mitral regurgitation and no significant stenosis. This demonstrates the utility and incremental value of 3D echocardiography for characterization of anatomical abnormalities associated with this pathology.

Supplementary data are available at European Heart Journal — Cardiovascular Imaging online.

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