Congenital malformation with absence of posterior mitral leaflet is extremely rare. A case of a 27-year-old woman with congenital malformation of unicuspid mitral valve is reported. To our knowledge, this is the first case of unicuspid mitral valve reported in Croatia.

This report demonstrates that real-time three-dimensional (3D) echocardiography can be an optimal tool to evaluate congenital malformation of the mitral valve. Real-time 3D echocardiography is important in diagnosing heart valve diseases because it enhances the evaluation of morphologic abnormalities and improves understanding of complex relationships through more precise imaging and presentation of interrelations of different parts of the mitral apparatus. The downside of real-time 3D echocardiography is that it extends the time required for diagnostic procedure and it demands higher level of training of the examiner.

In our patient, real-time 3D echocardiography revealed a complete absence of the posterior mitral leaflet. In absence of the posterior leaflet, the image was dominated by an elongated and thickened anterior mitral valve leaflet, accompanied with trivial mitral regurgitation. The subvalvular apparatus was anatomically normal, with appropriate chordal attachments and papillary muscles. There was no evidence of chamber enlargement or valvular dysfunctions. The patient was asymptomatic with normal function of unicuspid mitral valve.

The prognosis of asymptomatic patients with unicuspid mitral valve is uncertain. As an example, annular dilatation can progressively impair mitral regurgitation. For this reason such patients should undergo annual examination with transthoracic echocardiography to monitor progression of mitral regurgitation.

**KEYWORDS:** three-dimensional echocardiography, congenital malformation of mitral valve, unicuspid mitral valve.

**Literature**