**Abstract.** The spectrum of mitral valve (MV) pathology in patients with connective tissue dysplasia (CTD) include conditions from myxomatous degeneration with excess tissue of the valve leaflets and subvalvular apparatus, which is more common among young patients, to fibroelastic deficiency of the MV leaflets that is usually diagnosed in older age groups. Mitral regurgitation (MR) in patients with dysplasia belongs to the category of primary MR that can progress and lead to surgical treatment.

It is known that surgical intervention on MV in patients with CTD is recommended in cases of symptomatic severe MR. In asymptomatic severe MR with such pathophysiological consequences as left ventricular systolic dysfunction, pulmonary hypertension, atrial fibrillation, surgical treatment is also indicated. The question of surgical treatment of asymptomatic severe MI without the mentioned above criteria remains controversial.

Using the clinical example of long-term observation of the asymptomatic severe MR we will present step-bystep algorithm for patients with severe primary MI considering the latest clinical guidelines on valvular heart disease of 2021.