

## **Congenital mitral stenosis: old problems and new therapeutic options**

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**Introduction:** Congenital mitral stenosis (MS) is a rare but fatal anomaly and its course depends upon the entity of mitral damage and the severity of coexisting lesions. Valve repair could be difficult as the disease may involve the annulus, the leaflets and subvalvar apparatus. On the other hand, valve replacement is burdened by the size of the prosthetic valve. A light at the end of the tunnel could be represented by the use of the melody valve on mitral position. However, this approach is out of label and carries potentially fatal problems. Herein we describe our experience.

**Patients and methods:** In the last 30 years we operated on 279 patients with a congenital mitral disease. Among them, 40 presented with MS (age  $6,9 \pm 5,6$  years; body weight  $16,9 \pm 14,8$  Kg; 20 females). All patients were evaluated through an echocardiographic examination while a MRI or a cardiac catheterization were adopted to evaluate coexisting anomalies. According to Van Praagh classification, 19 patients had type I MS, none had type II, 12 had type III, finally 9 had type IV.

**Results:** The valve was repaired in 29 patients using different techniques whose purpose was to remove supramitral ring (14 pts), restore a proper leaflet motion (9 pts) and improve subvalvar apparatus opening (24 pts). Replacement was adopted in 11 patients: 7 received a mechanical valve and 4 had a Melody on mitral position. Overall mortality was 12,5 %, but it rose to 45% considering patients younger than 1 year of age. The melody valve was used in patients with the smallest annulus and the mortality rate among them was 50%.

**Conclusions:** Congenital MS is one of the most deadly congenital heart diseases, and the worst anomaly of the mitral valve. The conservative approach is difficult because of the anatomical disarrangement of the valvar and subvalvar apparatus. Nevertheless, replacement carries detrimental intraoperative and postoperative effects. Recently, surgeons looked kindly to the melody as the best candidate valve. In our experience, this valve suits also in small annulus, however, the results are quiet poor. There is still a way to go before achieving lasting and satisfactory results.