

## Mitral Percutaneous Repair in Congenital Heart Disease

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Mitral valve disease is the most frequent valvular heart disease in the world and its prevalence has been on the rise due to the aging population [1]. The etiologies of mitral regurgitation (MR) include mitral valve prolapse, rheumatic heart disease, ruptured chordae tendineae, infective endocarditis, ischemic cardiomyopathy [2]. There are two types of MR: primary or degenerative and secondary or functional. MR is a progressive disease and results in left atrial and ventricular enlargement, atrial fibrillation, pulmonary hypertension and most importantly, heart failure if left untreated. For treatment of MR, there are three options: medical, surgical and percutaneous therapy. Medical therapy has a limited to no role in the treatment of primary MR, however, appropriate guideline directed medical therapy is recommended in patients with hypertension or HF with reduced ejection fraction. Surgical therapy is the treatment of choice in treatment of primary MR. For adults, according to the recent valvular guidelines, a weak recommendation [3] (Level of recommendation: Class IIb) suggest for surgical intervention in patients with severely symptomatic grade 3 to 4+.

Secondary MR despite optimum guideline-directed management, treatment of coronary disease and cardiac resynchronization therapy [3]. In general, either MV replacement or repair has been shown to improve survival in the treatment of severe functional MR, only symptoms. The 2017 focused update of the 2014 AHA/ACC valve guideline suggested use of MitraClip in chronic severe primary MR (3 to 4+) among those who were highly symptomatic (New York Heart Association class III to IV) despite optimal guideline directed medical therapy (stage D), had favorable anatomy, reasonable life expectancy and a prohibitive surgical risk due to comorbidities (Table 1). For patients with primary MR who meet all criteria, the next step involves referral to the Heart Team for feasibility and potential risk vs. benefit from procedure. Most of these recommendations were made mainly in light of data from the EVEREST II trial [4]. The MitraClip is the treatment of choice for the high-surgical risk patients with severe and symptomatic MR for symptom improvement. However, the majority of the studies, which investigate the safety, feasibility and efficacy of the MitraClip, involve elderly patients.

30-day Society of Thoracic Surgeons (STS) predicted operative mortality risk
Score of > 8%
Porcelain or highly calcified aorta
Patient frailty
Severe liver disease
Severe pulmonary hypertension
Right ventricular dysfunction with severe tricuspid regurgitation
Others- chemotherapy for malignancy, major bleeding diathesis, immobility, AIDS, severe dementia

**Table 1:** Surgical Prohibitive risk for mitral valve surgery.

Table 1 Surgical Prohibitive risk for mitral valve surgery 30-day Society of Thoracic Surgeons (STS) predicted operative mortality risk score of  $\geq 8\%$  Porcelain or highly calcified aorta Patient frailty Severe liver disease Severe pulmonary hypertension Right ventricular dysfunction with severe tricuspid regurgitation Others- chemotherapy for malignancy, major bleeding diathesis, immobility, AIDS, severe dementia.

Most of pediatric patients with congenital heart disease are now living well into adulthood due to advances in medical and surgical techniques. Unfortunately, many patients with Tetralogy of Fallot, truncus arteriosus, double outlet right ventricle, and those who require the Ross operation undergo one or more repeat open chest operations during their lifetime, mostly in the first 2 decades of their lives [5]. Common indications include repair of residual defects, mitral and tricuspid valves, outflow tract conduit failure, and the development of native or prosthetic valve failure. Reoperation in this patient population can be technically challenging and fraught with high morbidity and mortality due to comorbid conditions, multi-organ dysfunction, and hostile chest. Recently, this progress has been made in the area of trans-catheter therapies for valvular disorders in congenital heart disease. The minimally invasive nature of these techniques combined with rapidly growing experience makes them especially appealing for the treatment of high-risk adult patients with congenital heart disease (ACHD) and valve failure. We discuss about use of trans-catheter therapies applied in pediatric patients or adult congenital heart disease. Pre-procedural consist of evaluation with imaging: echocardiography to quantify the extent of valve, ventricular dysfunction and magnetic resonance imaging (MRI) or cardiac computed tomography (CT) for both diagnostic purposes and to aid in procedural planning. Intraprocedural transesophageal (TEE) or intracardiac (ICE) echocardiography along with fluoroscopy is used to follow valve percutaneous valve repair or positioning and deployment The percutaneous mitral valve repair with MitraClip in pediatric patients and young adults has only been reported in some cases reports. Percutaneous mitral valve repair may be used in cleft mitral valve [5] and congenital cardiomyopathy with mitral valve disease<sup>6</sup>; these clinical cases reports use of Mitraclip both congenital structural disease with clinical improvement and good procedural results. Some authors [6,7] discuss about technical aspects, that although the MitraClip device is not designed for pediatric use, it might be considered in patients, who are not surgical candidates [6]. Moreover, the authors stated that they used standard adult guide wires, catheters, MitraClip delivery systems and trans-atrial puncture needles; and they emphasized that the sizes of the patient's vessels and heart chambers should be evaluated carefully, due to the unavailability of pediatric-sized MitraClip equipment. Now, for adults the only percutaneous technique currently approved by the US FDA is the MitraClip edge-to-edge repair (Abbot Vascular, Santa Clara, California). The clip was first introduced in 2004 and is commercially available in 40 countries. More than 10,000 patients have been treated with the clip, and the predominant indication in the non-US commercial setting is the surgically prohibitive patient with FMR. The device is currently approved in the USA only for the treatment of DMR and FMR despite this large worldwide experience. Percutaneous therapies have been adopted in much more limited settings to treat relatively younger adult patients with congenital heart disease at prohibitive surgical risk. Evidence for such an approach is rather limited now; however, case reports demonstrate feasibility with reasonable rates of success. Additionally, success of some of the above procedures in adult patients with acquired native or surgical valve disease may pave the way for the application and trial of these strategies in patients with ACHD. As our ability to perform transcatheter valve techniques improves and newer generation valve repair and replacement technology becomes available, it will be increasingly important to study them in adults with congenital heart disease as they may serve to delay or potentially obviate the need for repeated open-heart surgeries.

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