Repair of Tetralogy of Fallot – Progress or Just a Moving Target?

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Classic repair for tetralogy of Fallot consists of a large transannular incision and has been well studied, and new techniques have been developed to improve upon its shortcomings. Mouws et al describe a single-center experience with repair of tetralogy of Fallot utilizing a transatrial and transpulmonary approach for complete repair.[1] Valve-sparing repair was performed in approximately 32% of the patients, whereas the remaining underwent transannular patch. As with many studies, the authors demonstrate excellent survival at short- and mid-term follow-up in this patient population. Several important observations were made regarding incidence of recurrent significant pulmonary stenosis (>moderate) in 44% of patients and development of pulmonary regurgitation in 76% at a median of 3.2 years. Pulmonary regurgitation developed even in patients who underwent valve-sparing repair, although onset of pulmonary regurgitation was delayed in these patients compared to trans annular patch patients.

This study provides several important teaching points. First, the techniques surrounding repair of tetralogy of Fallot have improved significantly over time, but there remains significant variability across centers regarding management of the pulmonary valve. As we interrogate colleagues around the world, we discover that each surgeon does something slightly differently than the other. The options for right ventricular outflow tract reconstruction can be broadly divided into transannular patch and valve-sparing repair, but upon further inspection there are many subtle differences that can have significant consequences. Let's examine the first controversy – limited vs extensive transannular incision. Proponents of the latter approach (the classic operation for TOF) report very low reintervention rate (less than 10%) and boast long-term freedom from reoperation for right ventricular dilation. On the other hand, proponents of the limited incision rightfully point to the incidence of ventricular tachyarrhythmias and right ventricular dysfunction in the area of extensive patching that can occur with the classic approach. One can speculate that a limited transannular incision approach would necessarily reduce the incidence of right ventricular injury and thereby reduce the rates of ventricular dysfunction and tachycardia. DeGroot et al illustrate an important (and possibly unforeseen) consequence of this strategy – early development of pulmonary stenosis and need for reintervention. The right ventricular outflow tract is a crowded area, with hypertrophied parietal and septal muscle bands converging upon a hypoplastic annulus and deviated conal septum. The extensive transannular patch raises the ceiling over this crowded area, whereas a limited incision necessitates extensive muscle bundle resection in order to avoid stenosis.

To spare or not to spare the valve – that is the next big question. Only one-third of patients in the study underwent valve-sparing repair. An important finding of this study is the high incidence of pulmonary regurgitation, even with valve-sparing approaches. Extensive manipulation of the pulmonary valve was not described as part of the valve-sparing repair in this study. Yet many centers have devised elaborate repair techniques ranging from simple commissurotomy to monocusp reconstruction, leaflet augmentation or leaflet delamination techniques.[2–4] At this point, it is unclear if 1 particular technique is superior to another in the prevention of long-term pulmonary regurgitation. Whereas elaborate repair techniques may yield a beautiful-looking valve early after surgery, it may be the biology of the valve leaflets that determine the fate of the valve in the mid to long term. Perhaps the onset of pulmonary regurgitation is delayed a few years by current valve-

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Central Message

New techniques have emerged to repair tetralogy of Fallot. We have made progress, yet have a lot to learn.

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sparing repair techniques, but whether this results in a healthier right ventricle in the long run remains to be seen. As with the limited transannular incision, reintervention for pulmonary stenosis can be an unintended consequence of the valve-sparing approach.

Scholars have argued that some degree of pulmonary stenosis may maintain just enough right ventricular hypertrophy to prevent right ventricular dilation associated with long-term pulmonary regurgitation. The speculation remains unproven. Is the theoretical long-term benefit of mild pulmonary stenosis worth the increased risk of reintervention that might become necessary in the short term? Unfortunately, we will not have the answers to such questions for many years since it takes decades for complications to develop.

Other considerations include role of initial aortopulmonary shunting or transcatheter intervention followed by complete repair for symptomatic neonates and infants, surgical approach (transatrial vs transinfundibular), method of annular enlargement (balloon vs surgical) and material used for RVOT reconstruction. The number of combinations of choices explains the wide variability in technique for TOF repair encountered.

In summary, we are starting to see results of iterations in repair techniques for tetralogy of Fallot that have evolved from the original large incision transannular patch technique. In our attempts to limit RV dysfunction, we have made smaller incisions on the right ventricle. To prevent PR, we have concocted a wide array of techniques to spare the pulmonary valve. In shooting for the moon, we have discovered that sparing the pulmonary valve is easier said than done, and that limited incisions sounds appealing but may create a new problem. We must keep sight of our goals to preserve long-term RV function while reducing rates of reintervention yet realize it will take decades to evaluate the performance of any new technique.

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