Chapter 1

TETRALOGY OF FALLOT SURGERY: CURRENT PERSPECTIVE

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Introduction

Tetralogy of Fallot (ToF) is the most common cyanotic congenital heart defect, occurring in 1 in every 3600 live births (1) and represents approximately 10% of congenital cardiac malformations. (2) Neonates are often asymptomatic. Right ventricular outflow tract (RVOT) stenosis increases with time, and infants typically become symptomatic after a few months of age. (3-4) Without repair, a quarter of patients with severe obstruction will die within the first year of life. Surgical intervention is necessary for long-term survival. (2)

The conventional technique called transannular patch repair is still widely used in many centers. (1) As results of this technique, pulmonary stenosis is easily eliminated, but severe pulmonary insufficiency is inevitable. Right ventricular (RV) dilatation, caused by pulmonary insufficiency (PI), is one of the major causes of malignant arrhythmia, heart failure, and sudden cardiac death in patients with repaired tetralogy of Fallot. (5-6)

Based on long-term outcomes of conventional TOF repair, many centers have developed techniques designed to preserve pulmonary valve (PV) function and thus limit or eliminate the long-term consequences of chronic PV regurgitation on ventricular function. (7)

In this section, we aimed to discuss TOF repair from a recent surgical perspective instead of the conventional techniques known by everyone.

The Scope of the Problem

Tetralogy of Fallot is one of the earliest repaired congenital heart defects. The original repairs involved closure of the ventricular septal defect through a large right ventriculotomy and correction of the right ventricular outflow tract (RVOT) obstruction with a transannular patch (TAP) (8)In the first years, congenital heart surgeons have been focused on the complete elimination of RVOT stenosis in Fallot surgery. PV regurgitation was also considered unimportant. But now, chronic right ventricular volume loading caused by pulmonary regurgitation (PR) is recognized as injurious to the RV.(9) RV dilation, RV diastolic dysfunction, RV fibrosis, risk of ventricular

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Later surgical repair has been associated with fewer postoperative complications and a higher possibility of 1-stage repair, given more stable physiologic characteristics after the first month of life (4). The downside of later surgical intervention, however, is the potential for worsening of right ventricular hypertrophy as well as persistence of hypoxemia, both of which can lead to ventricular dysfunction and arrhythmias (4).

Cunningham et al. have demonstrated that early primary repair can be safely performed without increased hospital resource utilization(33) and without any compromise in the surgical technical performance scores (TPS) (34)

Both, age cutoff of 55 days at repair and the residual peak RVOT gradient are significant independent predictors of intermediate term reintervention. And also, patients 55 days of age and younger with optimal repair by TPS (RVOT gradient < 20 mm Hg) had a higher reintervention rate than those repaired at an older age (35)

Despite its multiple advantages (36), early primary repair of tetralogy of Fallot with pulmonary stenosis (TOF) has not been universally adopted because of concerns for increased morbidity and mortality in younger patients and higher risk for postoperative reintervention (35). The optimal timing of elective early primary repair of TOF remains debated, and wide variations in practice exist among various centers.(35)

Conclusion

In the current era, many congenital heart surgeons have studied in the TOF repair in order to obtain far more efficient surgical technique. As a result, we believe that the surgical techniques including pulmonary valve and right ventricle sparing procedures can be applied in the most of TOF patient to achieve more successful outcomes.

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