

Residual Problems With Repaired Tetralogy of Fallot

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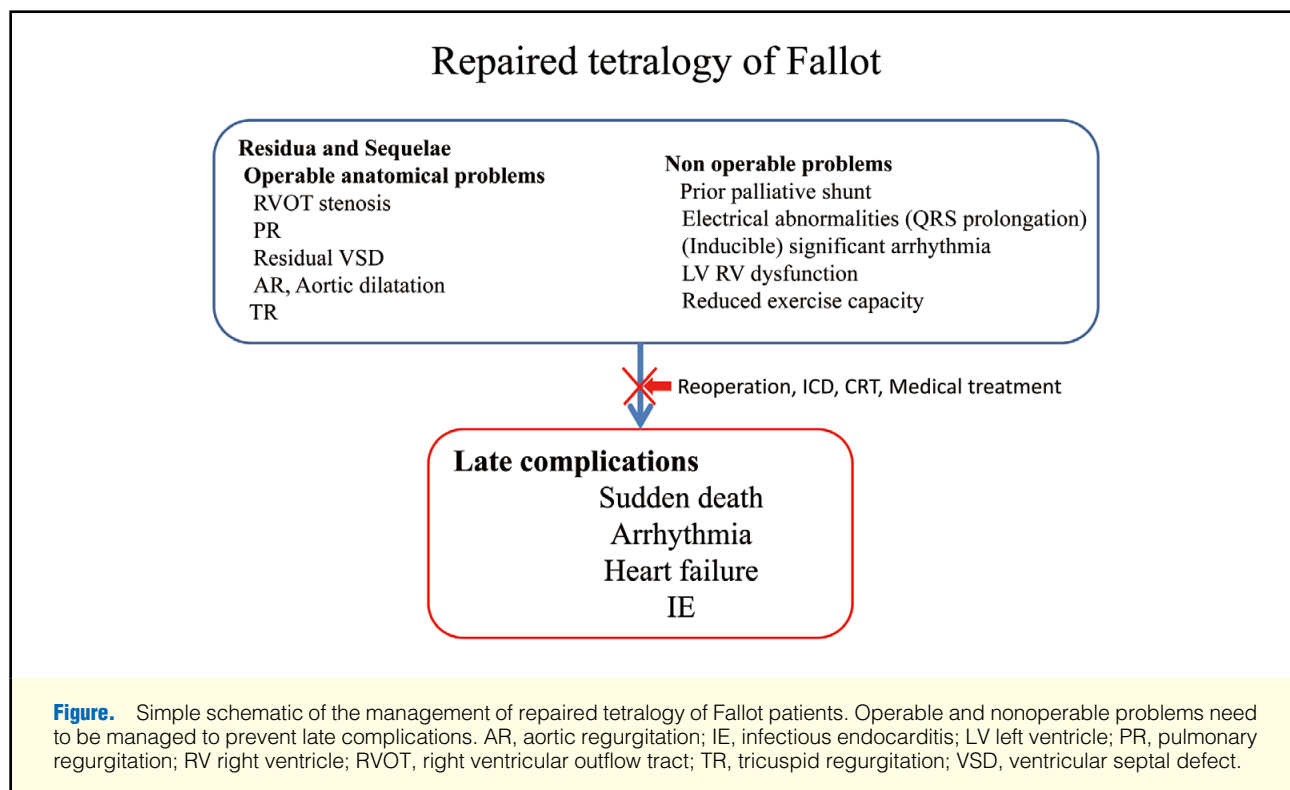
Tetralogy of Fallot (TOF) includes 4 major anatomical features: right ventricular outflow tract (RVOT) obstruction (hypoplasia); ventricular septal defect; deviation of the origin of the aorta to the right ventricle; and concentric right ventricular (RV) hypertrophy. TOF is the most common cyanotic congenital heart disease, accounting for 10% of congenital heart disease.¹ Despite improved mortality rates of TOF patients, thanks to recent progress in comprehensive management, including intracardiac repair, there are postoperative late complications.^{2,3} Prevention and management of these are of paramount importance. Sudden death in adulthood is commonly from either ventricular arrhythmia or heart failure. There are operable and non-operable residual and sequelae in adult TOF patients long-term after repair (Figure). It is recognized that patients with RV dilatation and dysfunction with or without severe tricuspid regurgitation have

a worse outcome.^{1,2} The predominant residual lesions responsible for poor prognosis are pulmonary regurgitation and RVOT stenosis.²

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Operable Problems

Pulmonary regurgitation and RVOT stenosis are anatomical complications that can be relieved only by anatomical repair including surgery and percutaneous catheter treatment.^{4,5} Therefore, appropriate timing of such intervention is important. However, definite criteria for reoperation are not yet determined. There were several advocated criteria for re-intervention using magnetic resonance imaging (MRI).^{6,7} We routinely decide the timing of reoperation according to the following parameters: RV and LV end-diastolic volume index;



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RV end-systolic volume index; RV and LV ejection fractions; RVOT aneurysm; exercise intolerance; and presence of heart failure symptoms, syncope, sustained ventricular tachycardia (VT).

Without reoperation, repaired TOF patients with severe anatomical residua and sequelae will develop irreversible RV myocardial damage and heart failure, but there is not conclusive evidence that reoperation for pulmonary regurgitation improves survival.

In Japan, there is also no cut-off value of RV volume or RV ejection fraction or other parameters about the timing of reoperation. MRI was used to assess the timing of re-operation in only 60% of repaired TOF patients in a Japanese multicenter survey.² Compared with Western countries, the most common reason for re-operation in Japan is RVOT stenosis. Conotruncal repair with the application of small RV outflow patching has been used for the primary repair of TOF.⁸ This technique may protect against the development of severe pulmonary regurgitation after repair, which results in a lower incidence of huge RV dilatation, RV dysfunction, sustained VT and sudden death than in previous reports from North America and Europe.^{6,9} In turn, our pre-re-intervention assessment might be reasonable for detecting RVOT stenosis rather than pulmonary regurgitation only by echocardiography. Because there is no consensus about the appropriate timing and also relative merits of preoperative imaging modalities in repaired TOF, we should discuss these residual clinical problems and late complications.

Nonoperable Problems

There are many prognostic variables to predict the long-term outcome of repaired TOF patients, such as RV and LV function, dilatation, inducible VT and cardiopulmonary exercise test results.^{10,11} These variables are used to predict sudden death from significant arrhythmias. One of the most popular variables used to predict long term outcome is QRS interval, which correlates with RV end-diastolic volume estimated by MRI.^{12,13} In order to prevent these fatal arrhythmias, the indication for implantable cardiac defibrillator implantation should be guided by several important variables: prior palliative shunt, inducible VT, QRS interval, ventriculotomy, nonsustained VT, and LV end-diastolic pressure.¹⁴ Longitudinal LV function also seems to be an important variable in repaired TOF patients to predict a composite endpoint of sudden cardiac death/life-threatening ventricular arrhythmias.¹⁵ In this issue of the Journal, Nakamura et al reveal that longitudinal LV dysfunction in repaired TOF is associated with RV dilatation,¹⁶ which is also a prognostic variable as mentioned. In Japan, MRI is less commonly used to assess ventricular function than echocardiography. Left longitudinal strain measurement could give additional prognostic information in real-world Japanese clinical practice.

Disclosures

Financial Disclosures and Conflicts of Interest: None.

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