Clinical Aspects and Ablation of Ventricular Arrhythmias in Tetralogy of Fallot

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KEYWORDS

• Tetralogy of Fallot • Ventricular tachycardia • Risk stratification • Electroanatomical mapping
• Anatomic isthmus • Radiofrequency catheter ablation

KEY POINTS

- Changes in type and timing of surgical repair in tetralogy of Fallot (ToF) are likely to influence the potential substrate for late ventricular arrhythmias (VAs).
- Monomorphic ventricular tachycardia (MVT) is the most common arrhythmia subtype in repaired ToF (rToF).
- MVTs are usually fast and require a substrate-based ablation approach.
- Complete procedural success defined as noninducibility of any VT and transection of the slow conducting anatomic isthmus can be considered curative in patients with preserved cardiac function and no competing VA mechanism.
- The advances made in the understanding of the substrate for VT in rToF may have important implications for risk stratification and preventive treatment in contemporary patients with ToF.

INTRODUCTION

The reported incidence of tetralogy of Fallot (ToF) is approximately 0.42 per 1000 live births and has remained stable over time.[1] Life expectancy, however, has significantly improved over the last decades. Seventy-eight percent of all individuals born with ToF between 1990 and 1992 followed at one European center survived into adulthood, which is significantly better compared with patients born in preceding years.[2] The change in mortality, with favorable survival in infancy and a trend toward death at older age, is likely the result of earlier and improved surgical interventions and medical care. However, despite early repair, these patients are not cured and the improved long-term outcome generates new challenges.[3]

VENTRICULAR ARRHYTHMIAS

An age-dependent increase in the prevalence of atrial and ventricular arrhythmias (VAs) has been reported[4,5] with a high arrhythmia burden for both atrial and VAs. In contrast to other repaired congenital heart diseases, ventricular tachycardia (VT) is the most common arrhythmia subtype with a prevalence of 14.2%.[6] The prevalence of ventricular fibrillation (VF) was only 0.5% in a large North American collaboration study.[6] Although VF may be underestimated in this cohort, including adult survivors, with implantable cardioverter defibrillators (ICDs) being implanted in only 10.4%, the higher burden of monomorphic VT compared with polymorphic VT/VF has also been observed in ICD recipients. More than 80% of all VAs that triggered

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ICD therapy in a cohort of 121 patients with TOF who had received an ICD for primary or secondary prevention were monomorphic VTs. Of note, treated VTs were fast with a median heart rate of 213 beats per minute (bpm) (interquartile range [IQR] 182–264 bpm).[7] The clinical importance of fast, monomorphic VTs has been confirmed by recent data from patients with repaired ToF (rToF) referred for risk stratification or VT ablation. The median VT cycle length of the 41 spontaneous and induced VTs recorded in 28 patients was only 252 ms (IQR 231–312).[8] These rapid VTs, if untreated, may be fatal even in the presence of a preserved biventricular function. Of concern, two-thirds of patients with rToF who died suddenly or experienced life-threatening VT, typically early to middle-aged adults, had a preserved or only moderately impaired cardiac function before the first event.[3,8,9] The average left ventricular (LV) ejection fraction of more than 50% and the only mildly impaired or normal right ventricular (RV) function in 75% of those who have experienced an arrhythmic event strongly suggest that non-heart failure–related substrates for VA play an important role in rToF.[3,6]

RISK STRATIFICATION AND VENTRICULAR ARRHYTHMIA SUBSTRATES

For risk stratification and treatment of VA in rToF, it is important to emphasize that different arrhythmia substrates and mechanisms may be encountered depending on the variation of the malformation, type and timing of repair, and residual lesions. ToF is characterized by a subpulmonary stenosis, a subaortic ventricular septal defect (VSD), dextroposition of the aortic orifice, and RV hypertrophy. The last characteristic is the consequence of the volume and pressure overload caused by the VSD and the subpulmonary stenosis.

The morphology of TOF, however, encompasses a broad spectrum, from a mild appearance with small VSDs and minimal pulmonary stenosis to severe forms of the disease with pulmonary atresia. The spectrum of the malformation, and the different types and timing of surgical interventions with variable hemodynamic outcomes, results in a heterogeneous population of adults with rToF. RV hypertrophy and interstitial fibrosis typically occur after long-standing cyanosis and high ventricular pressure overload due to repair at older age.[10]

Total repair usually includes (patch) closure of the perimembranous or muscular VSD, resection of the hypertrophic infundibulum, and insertion of an right ventricular outflow tract (RVOT) or a transannular patch to augment the restrictive RVOT or to relieve the stenosis of the pulmonary orifice. Surgery was initially performed through a vertical or transverse right ventriculotomy, often combined with the use of a large transannular patch with subsequent pulmonary regurgitation and RV dilatation and dysfunction.

Chronic volume and pressure overload can contribute to ventricular arrhythmogenesis due to nonreentrant mechanisms, such as triggered activity or abnormal automaticity typically observed in patients with nonischemic cardiomyopathies and end-stage heart failure.[11]

Accordingly, reported risk factors associated with any VA and/or sudden cardiac death in rToF are older age at repair, the presence of a transannular patch, the number of previous cardiac surgeries, moderate to severe pulmonary regurgitation, moderate to severe RV and LV systolic dysfunction, and a QRS duration of greater than 180 ms, which may be related to RV dilatation (**Box 1**). The number of prior cardiac surgeries, including prior palliative shunt operation, diastolic LV dysfunction, and, consistent with prior reports, QRS duration,[4] was independently associated with VT in a recent multicenter, cross-sectional study. Reasons why, in particular, diastolic LV dysfunction is associated with VA are unclear; the value of LV diastolic dysfunction in risk stratifying patients with rToF needs further evaluation.[6] Of note, most of the data on late morbidity, mortality, and risk stratification in rToF are based on patients who have undergone repair late in life, at a median age of 5 years (IQR 2.5, 7 years)[6] and 8 years (IQR 2, 9.4 years),[12] half of them after prior palliative shunt operation.

The detrimental effect of both late repair and right ventriculotomy on RV function and arrhythmogenecity has been recognized. As a consequence, the surgical strategy has evolved over time. Nowadays, patients often undergo repair early in life with a combined transatrial-transpulmonary approach. The RV incision is usually restricted to the pulmonary annulus; smaller patches are preferred, and effort is made to avoid or limit free pulmonary regurgitation. These changes are likely to positively influence the biventricular function and the occurrence of heart failure–related VA. In addition, they may also have significant impact on the substrate for monomorphic VT.

SUBSTRATE FOR MONOMORPHIC VENTRICULAR TACHYCARDIA IN REPAIRED TETRALOGY OF FALLOT

Intraoperative and catheter mapping studies have identified macro-reentry as the major underlying mechanism of spontaneous and induced

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