Ablation of Ventricular Tachycardia in Congenital and Infiltrative Heart Disease

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KEYWORDS

• Ventricular tachycardia • Catheter ablation • Congenital heart disease • Cardiac sarcoidosis

KEY POINTS

- Slow conducting anatomic isthmuses bordered by surgical scars, prosthetic material, and valve annuli are the dominant substrate for VT in repaired congenital heart disease.
- Identification and transsection of these anatomic isthmuses by catheter or surgical ablation leads to long-term VT-free survival in patients with repaired CHD and preserved biventricular function.
- Among infiltrative cardiac disease, cardiac sarcoidosis is the most important cause of ventricular arrhythmias.
- Catheter ablation in the advanced stage of cardiac sarcoidosis is challenging because of complex ventricular scars and an active inflammation.
- In most patients with cardiac sarcoidosis a significant decrease of the VT burden is achieved often requiring multiple procedures.

INTRODUCTION

Ventricular arrhythmias (VA) are frequently encountered in patients with structural heart disease and are an important cause of morbidity and sudden cardiac death (SCD). Most VAs in patients with ventricular scars are caused by reentry, which is facilitated by barriers of functional or fixed conduction block and zones of slow conduction. Radiofrequency catheter ablation (RFCA) targets the substrate and has the potential to prevent ventricular tachycardia (VT) recurrence. The nature and cause of ventricular scars and the prevalence and characteristics of VTs vary widely according to the underlying heart disease and surgical interventions. This article focuses on the substrate of VA and RFCA for VT in patients with surgically corrected congenital heart disease (CHD) and infiltrative heart disease.

VENTRICULAR ARRHYTHMIAS IN CONGENITAL HEART DISEASE

The incidence of moderate to severe CHD is 6 in 1000 live births.[1] Because of earlier surgical interventions and advances in medical care over the last decades more patients with repaired CHD (rCHD) survive to adulthood.[2] Accordingly, an increasing number of patients may be at risk for arrhythmias and SCD later in life.[3–5] SCD accounts for 19% of all deaths in adults with rCHD, often occurring in the fourth and fifth decades of life.[6] Sustained monomorphic VT and polymorphic VT/ventricular fibrillation (VF) are thought to be the dominant cause of SCD in this population.[6] Repaired cardiac defects associated with a high risk for VA and presumed arrhythmic deaths are tetralogy of Fallot (TOF) and transposition of the great arteries (TGA).[3,4,6,7] In patients with CHD

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ventricular dysfunction has been associated with the occurrence of VA and SCD as in other cardiac disease with increased wall stress and interstitial and replacement fibrosis.[5,6] However, two-thirds of patients with rCHD who died suddenly had a preserved ventricular function before the event.[6] This suggests that non-heart-failure-related VT mechanisms and substrates may play an important role.[8,9] Because VTs often recur despite antiarrhythmic drugs, and implantable cardioverter defibrillators (ICD) terminate but do not prevent arrhythmia recurrence, RFCA is an important treatment option.[10] A thorough understanding of the underlying mechanism and substrate of VT is crucial for risk stratification and treatment.

Tetralogy of Fallot

TOF is the most common form of cyanotic CHD.[1] It encompasses a subpulmonary stenosis, a subaortic ventricular septal defect (VSD), dextroposition of the aortic orifice, and as a consequence right ventricular (RV) hypertrophy. Since the 1960s a two-stage surgical procedure has been performed with initial palliative shunt operations and total repair later in childhood. Total repair includes patch closure of the VSD and relief of the infundibular or valvular RV outflow tract (RVOT) obstruction and was initially accomplished through a vertical or transverse right ventriculotomy often combined with the use of a large (transannular) patches. This often led to RV dysfunction and pulmonary regurgitation with chronic volume overload and subsequent RV dilatation and further functional impairment. Consequently, a transatrial-transpulmonary approach performed earlier in life, avoiding RV incision and the use of smaller transannular patches, has become the treatment of choice.[11] The type and timing of repair are decisive factors for the substrate of the late VTs.

Ventricular Arrhythmias and Anatomic Isthmus

The reported prevalence of monomorphic VT in the adult TOF population was 14.2% in a recent multicenter study accounting for 97.5% of all documented VAs.[12] Similarly, more than 80% of appropriate ICD therapy in patients with TOF with ICDs implanted for primary and secondary prevention is triggered by monomorphic VT.[13] These VTs are generally fast with heart rates greater than 200 bpm and may therefore cause hemodynamic compromise and SCD.[9,10,13,14]

Areas of dense fibrosis after surgical incisions, patch material, and valve annuli are the unexcitable borders of anatomic isthmuses (AI) that have been shown to contain critical isthmuses of VT reentry circuits. Four AI have been identified. Isthmus 1 is bordered by the tricuspid annulus and scar or patch material in the anterior RVOT/RV, isthmus 2 by the pulmonary annulus and the RVOT/RV incision or patch sparing the pulmonary annulus, isthmus 3 by the pulmonary annulus and the VSD patch, and isthmus 4 by the VSD patch and the tricuspid annulus in patient with a muscular VSD (**Fig. 1**).[14]

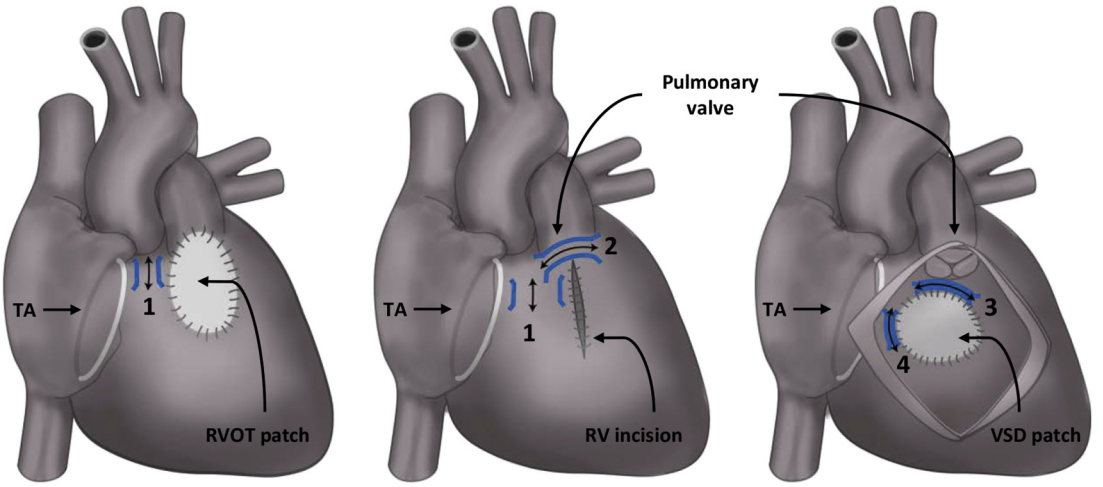

Fig. 1. Schematic overview of the four potential anatomic isthmuses (*blue brackets*): isthmus 1 bordered by tricuspid annulus and right ventricular outflow tract patch/right ventricular incision, isthmus 2 by right ventricular incision and pulmonary valve, isthmus 3 by pulmonary valve and ventricular septal defect patch, and isthmus 4 by ventricular septal defect patch and tricuspid annulus. TA, tricuspid annulus. (*Adapted from* Kapel GF, Sacher F, Dekkers OM, et al. Arrhythmogenic anatomic isthmuses identified by electroanatomical mapping are the substrate for ventricular tachycardia in repaired tetralogy of Fallot. Euro Heart J 2016. [Epub ahead of print]; with permission.)