Arrhythmias in Complex Congenital Heart Disease

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KEYWORDS

• Atrial fibrillation • Atrial flutter • Catheter ablation • Congenital heart disease
• Ventricular tachycardia

KEY POINTS

- Atrial and ventricular arrhythmias are a common cause of morbidity and mortality in the growing population of adults with congenital heart disease.
- Patients with high-risk congenital heart disease lesions such as dextro-transposition of the great arteries, levo-transposition of the great arteries, or tetralogy of Fallot should be monitored routinely for arrhythmias and associated symptoms.
- With the aid of electroanatomic mapping and newer irrigated radiofrequency energy delivery, catheter ablation is an excellent therapeutic option for a variety of arrhythmias observed in these patients when performed in experienced centers.
- Implantation of an implantable cardioverter-defibrillator is recommended for cardiac arrest survivors and congenital heart disease patients with sustained ventricular tachycardia discovered on electrophysiology study.
- In planning catheter ablation and device implantation procedures, clinicians should review specific anatomy and surgical records, obtain imaging to define possible obstructions or stenosis in vascular pathways, and be aware of associated congenital abnormalities.

INTRODUCTION

More than one million adults are living with congenital heart disease (CHD) in the United States, and this group now outnumbers children with CHD.[1,2] Late after surgical repair of complex congenital lesions, atrial arrhythmias are a major cause of morbidity, and ventricular arrhythmias and sudden cardiac death (SCD) are a major cause of mortality.[3–7] Arrhythmia mechanisms include reentry caused by substrate from previous surgeries, the long-term consequences of hemodynamic abnormalities, such as chamber enlargement and hypertrophy, and direct results of congenital abnormalities, such as the presence of accessory pathways. It has been reported that the prevalence of atrial arrhythmias is 15% in adults with CHD; for patients with complex CHD, the lifetime risk of atrial arrhythmias is more than 50%.[8] Atrial arrhythmias in these patients are associated with increased risk of stroke, heart failure, and mortality.[8] Ventricular arrhythmias are also common in CHD, especially in patients with tetralogy of Fallot (TOF), ventricular septal defect, Ebstein's anomaly, and systemic right ventricles. Drug therapy is often inadequate for these patients. Amiodarone is often avoided in younger patients because of concerns over long-term toxicity; class IC agents may have

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lower efficacy than in other patient groups[9] and may be contraindicated because of underlying structural heart disease. In experienced centers, catheter ablation has emerged as the preferred therapeutic option for atrial and ventricular arrhythmias in the CHD population. As increasing numbers of patients reach adulthood, the burden of arrhythmias and SCD are expected to increase even further, and the need for device implantations[10] and catheter ablation procedures will continue to grow. This review focuses on 6 cases that highlight common and important electrophysiology problems in the adult CHD population.

Case I

A 45-year-old woman with history of perimembranous ventricular septal defect after patch repair, moderate residual right ventricle (RV) enlargement, and supraventricular tachycardia (SVT) after ablation at another hospital 8 years prior was admitted with palpitations and SVT (Fig. 1). Electrophysiology (EP) study found 2 intra-atrial reentrant tachycardias (IARTs) involving a posterolateral right atrial scar (Fig. 2). Radiofrequency catheter ablation of the isthmus within the scar terminated the arrhythmias (Fig. 3).

The most common arrhythmia in older adults with CHD is IART. This is a macroreentrant circuit involving abnormal atrial tissue resulting from atriotomy incisions, fibrosis, or patches[11–13] and characterized by large areas of low voltage with multiple heterogeneous channels.[14] IART may be seen in any patient who has undergone atriotomy, such as this patient, but the incidence is particularly high for patients with dextrotransposition of the great arteries (D-TGA) after Mustard[15] or Senning repair and patients with a single ventricle after Fontan. Fontan patients treated with older intra-atrial lateral tunnel operations are at higher risk than those treated with extracardiac Fontan operations.[16–20] Atrial rates in IART are typically 150 to 250 beats per minute (bpm), and 1:1 atrioventricular (AV) conduction can result in presyncope, syncope, or SCD.[21] As in this patient, multiple circuits are common.

Catheter ablation has been used with success in experienced centers. Complete procedural success is reported to be as high as 80% with the use of irrigated ablation catheters and electroanatomic mapping, but recurrence is reported in about 40% of patients. Arrhythmia recurrence is more common for those with multiple circuits, atrial fibrillation, and Fontan physiology.[22] Because IART has been associated with thromboembolism,[23] adequate anticoagulation with periprocedural transesophageal echocardiogram guidance according to standard guidelines is recommended.

Case II

A 50-year-old man with D-TGA after Mustard procedure presented with dyspnea on exertion and was found to have pulmonary venous baffle stenosis and right-to-left shunting, suggesting a systemic venous baffle leak. At the time of stent placement for the pulmonary venous baffle stenosis, he was in atrial flutter at cycle length of 280 milliseconds. Flutter waves were negative in the

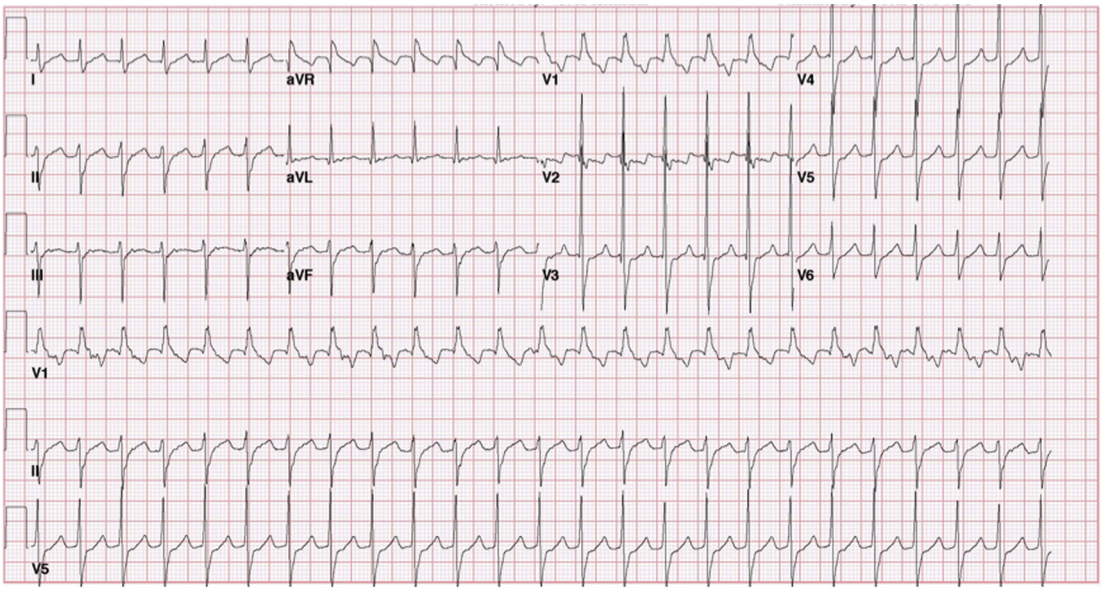

Fig. 1. Surface electrocardiogram of IART with 1:1 AV conduction.