Arrhythmia Surgery for Adults with Congenital Heart Disease

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KEYWORDS

• Atrial fibrillation • Atrial flutter • Atrial septal defect • Ebstein anomaly • Univentricular physiology

KEY POINTS

- As survival following initial surgical repairs of congenital heart disease has improved, the late sequelae of heart failure and arrhythmias for patients with severe forms of congenital heart disease are increasingly recognized.
- Successful arrhythmia surgery requires a clear understanding of tachycardia mechanisms present in an individual patient, the specific operative techniques for each mechanism, and cooperation between the electrophysiologist and surgeon.
- Surgical repair of congenital heart disease can be viewed as both an anatomic and an electrical intervention, with the combined goals of improving hemodynamic status and minimizing morbidity from the development of later arrhythmias.

INTRODUCTION

The purpose of this article is to review arrhythmia surgical techniques that may be incorporated into concomitant repairs for congenital heart surgery patients. As survival to adulthood following initial surgical repairs of congenital heart disease has improved for most patients, the late sequelae of heart failure and arrhythmias for patients with severe forms of congenital heart disease are increasingly recognized.[1] The median age of adults with severe congenital heart disease was reported as 29 years, and one recent review described tachyarrhythmias as their "inevitable destiny."[2] Atrial arrhythmias negatively impact ventricular function, functional assessment, and long-term survival.[3–5] Among adults with congenital heart disease, atrial arrhythmia development results in a 50% increase in early mortality, a 2-fold increase in stroke and congestive heart failure, and a 3-fold increase in the need for cardiac interventions.[4]

Understanding the mechanisms of arrhythmias is essential to determine appropriate catheter or surgical intervention and have been summarized in recent publications.[2,5,6] For adult patients with arrhythmias undergoing cardiac surgery, the options are transcatheter ablation of arrhythmias preoperatively or postoperatively, or incorporation of arrhythmia procedure into the cardiac surgery. Patients with unsuccessful or difficult catheter ablations (Ebstein anomaly), complex anatomy including markedly thickened atrial walls with multiple reentrant circuits, difficult venous access, or those requiring atrial reduction or treatment of atrial fibrillation (AF) are most suitable for arrhythmia surgical procedures. As the pioneer arrhythmia surgeon James Cox stated in 1983, "The selection of patients for the surgical treatment of cardiac arrhythmias is based on several

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variables. These variables include the patient's age and general condition, the nature of the presenting arrhythmia, its response to medical treatment, and the presence of associated anomalies that may require the additional surgical correction."[7] These original guidelines are particularly important for adults undergoing repair or reoperations for congenital heart disease presently.

STRUCTURAL CONGENITAL HEART DISEASE AND ARRHYTHMIAS

Diagnostic substrates associated with the highest prevalence of supraventricular tachycardia (SVT) include Ebstein anomaly, atrial repairs of transposition of the great arteries, univentricular hearts, atrial septal defects (ASDs), and right heart obstructive lesions, such as tetralogy of Fallot (TOF) and double outlet right ventricle.[2,4–6] **Table 1** summarizes the prevalence of arrhythmias and incidence of reoperation for congenital heart disease.[8] ASD closure in childhood is associated with late atrial flutter (AFL) and AF in as many as 20% to 35% of patients,[9–11] whereas patients undergoing ASD repair as adults have a 30% to 50% incidence of atrial arrhythmias, particularly AF.[9–13] The development of arrhythmias following TOF repair was initially concentrated on the risk of ventricular tachycardia (VT)[14–19]; more commonly, atrial arrhythmias are recognized in as many as 12% to 43% of older TOF patients, contributing to morbidity and hospitalizations.[4,20] Ebstein anomaly of the tricuspid valve is associated with SVT in up to 42% of patients, which is related to accessory connections, AF, and AFL.[21–23] Older Fontan patients with atriopulmonary anastomoses have an increasing incidence of atrial tachycardia (AT) over time, exceeding 40% by 20 years postoperatively and steadily increasing to more than 70% by 25 years postoperatively.[24–30] Fontan modifications have decreased the incidence of AT to approximately 8% to 15% in extracardiac connections, and 13% to 60% in lateral tunnel connections,[24,30] yet is likely to increase with longer durations of follow-up.[25–27] The development of AT in Fontan patients is associated with right atrial thrombus formation, congestive heart failure, atrioventricular valve regurgitation, thromboembolic events, increased hospitalizations, and mortality.[4,28,29] Catheter ablation in the Fontan patient is associated with acute success of about 50% with at least 70% recurrence of tachycardia within 2 years.[12,31–33]

Mechanisms of Supraventricular Tachycardia

In adult patients with congenital heart disease, the most common mechanism of SVT is macroreentrant AT, which accounts for at least 75% of SVT and involves the cavotricuspid isthmus in more than 60% of circuits.[13,34–36] AT is a slower form of AFL, with isoelectric periods between successive P waves; AFL is characterized by sawtooth flutter waves at more rapid atrial rates, without intervening isoelectric periods. AT develops most commonly in patients with TOF or right heart conduit repairs, ASDs, Ebstein anomaly, atrial baffle repairs for transposition of the great arteries, and in patients with univentricular hearts following Fontan surgery.[2,4–6] In addition

Table 1
Reoperation rates and estimated prevalence of arrhythmias in adults with congenital heart disease

Congenital Heart Disease	Reoperation (%)	Atrial Arrhythmias (%)	Ventricular Tachycardia (%)
ASD	<2	16–50	<2
Ebstein anomaly	30–50	33–60	>2
Single ventricle	>25	>40–70	>5
TOF	26–50	12–43	10–15
Transposition of the great arteries, atrial switch	15–27	26–50	7–9
Transposition of the great arteries, arterial switch	12–20	<2	1–2
Congenitally corrected transposition of the great arteries	25–35	>30	>2
Truncus arteriosus	55–89	>25	>2
Atrioventricular septal defect	19–26	5–10	<2

Modified from Khairy P, Van Hare GF, Balaji S, et al. PACES/HRS Expert Consensus Statement on the recognition and management of arrhythmias in adult congenital heart disease. Heart Rhythm 2014;11:e35; with permission.