

Arrhythmia in Congenital Heart Disease – A Current Perspective

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Abstract

Cardiac arrhythmia is a common presenting complaint in adult patients with treated congenital heart disease. (CHD) Advances in surgical and electrophysiological intervention, combined with improved mechanistic understanding, mean that effective arrhythmia treatment is a realistic expectation for this challenging patient group. This short commentary outlines the current paradigm for arrhythmia management across the spectrum of adult congenital heart disease.

Keywords

Arrhythmia, ablation, congenital heart disease

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Advances in surgical technique over the last two decades have heralded a dramatic improvement in survival rates in those born with congenital heart disease (CHD). With current methods, more than 85% of paediatric patients undergoing surgical repair survive into adulthood,¹ and there are now at least 1.2 million people living in Europe with CHD,² of whom 50% are adults. These patients pose unique challenges to the cardiologists who treat them, and arrhythmias represent one of the leading causes of morbidity in this group. It is estimated that more than half of adults with severe CHD will experience atrial arrhythmia (AA) by the age of 65.³

Despite the headline figures, there has been a general fall in prevalence of arrhythmia within lesion groups. This is likely to be related to the evolution of surgical procedures and the advent of catheter-based interventions, reducing surgical scar burden. However, 20% of hospital admissions in adults with repaired CHD continue to be arrhythmia-related⁴ and they represent a diverse group of patients who differ from the general population both in terms of the mechanism of their arrhythmia and its treatment with respect to catheter ablation. The ongoing development of catheter-based technologies has made the treatment of arrhythmia in the congenital population an exciting, evolving and rewarding field.

The type and prevalence of arrhythmia in CHD varies according to congenital cardiac diagnosis and management course. In general though, the development of arrhythmias can be broadly attributed to:

1. conduction abnormalities inherent to the lesion;
2. the long-term effects of adverse haemodynamic loading resulting in chamber remodelling, hypertrophy and fibrosis; or

3. the arrhythmogenic effects of prior surgical incisions and consequent scar formation.

AAs are more common than ventricular arrhythmias across the spectrum of CHD, particularly in those who have undergone extensive atrial surgery such as atrial switch procedures (Mustard/Senning operation). All AAs, including focal and reentrant atrial tachycardia and atrial fibrillation (AF), tend to occur more frequently in CHD than in the general population, but intra-atrial re-entry tachycardia (IART) is of particular concern, often presenting many years after surgical correction. The majority of IARTs are right sided circuits utilising the cavotricuspid isthmus, as in the non CHD population, but up to 40% have right or left sided circuits within scar or incisional regions.⁵ Ventricular arrhythmias occur mainly in the setting of more complex defects, including those with univentricular circulation, or in those with abnormal surgical and loading conditions of the ventricle, such as Tetralogy of Fallot or left ventricular outflow obstruction.

Many patients may initially be asymptomatic from their arrhythmia, potentially eluding timely diagnosis. However for some, particularly those with univentricular circulation, the onset of AA can precipitate rapid haemodynamic deterioration making prompt recognition and cardioversion of paramount importance.

Long-term management is often difficult, with pharmacological treatment rarely succeeding in achieving adequate arrhythmia control without substantial drug-related side-effects. Early catheter intervention should therefore be considered in all cases following a patient-specific evaluation of risk/benefit, and particularly for those in whom arrhythmia is associated with haemodynamic compromise.

Once a decision is made to proceed to cardiac ablation, a number of items must be considered prior to intervention. First and foremost, familiarity with the patient's anatomy and prior electrophysiological, interventional or surgical procedures is crucial, including detailed review of patient notes and imaging. Recent computed tomography (CT) or magnetic resonance imaging (MRI) for delineation of contemporary anatomy is often useful, and may also be integrated into electro-anatomical mapping (EAM) systems or provide an insight into potential arrhythmia substrate. Anaesthetic support is recommended in all but the simplest defects as the haemodynamic effects of inducing tachycardia can be unpredictable in this cohort. Special consideration should also be given to procedural access as this is often complicated by complex or unusual anatomy and prior interventional or surgical procedures. Venous access may be limited by vessel thrombosis, but intracardiac access to individual chambers may also pose challenges. For example, patients with atrial septal defect (ASD) closure devices or prosthetic patch material may necessitate novel atrial septal puncture techniques, often under intracardiac or transoesophageal echocardiography guidance.^{6,7} Similar approaches may also be used in those with intra- or extra-cardiac baffles or thickened native septum. Use of fluoroscopic landmarks alone is generally not as reliable in this cohort as in the general population.

Once the arrhythmia mechanism has been identified, and the critical region accessed, radiofrequency (RF) energy remains the cornerstone of ablation. However, in specific circumstances cryoablation techniques offer theoretical advantages, due to enhanced catheter stability (cryoadhesion) and a potential period of reversible tissue injury before permanent destruction. This is particularly relevant for those with

accessory pathways in difficult locations, such as Ebstein's anomaly, and those undergoing ablation of atrioventricular (AV) nodal re-entry tachycardia in whom the location of the AV node is displaced or unknown. There is also likely to be a role for remote navigation systems including non-magnetic and magnetic navigation systems, the latter utilising highly flexible atraumatic catheters. Advantages of these systems include a potential reduction in total procedure and fluoroscopy time, whilst enabling stable navigation of the ablation catheter to locations that may be relatively inaccessible using conventional manipulation techniques.^{8,9}

In terms of procedural success rates, analysis of existing paediatric registries relating to catheter ablation would suggest that the presence of CHD does not impact significantly on acute outcome rates^{10,11} with an overall procedural failure rate of only 7.5%. Nevertheless, data are scarce, particularly relating to long term outcomes. To this end The Multicentre Pediatric and Adult Congenital EP Quality Initiative¹² registry was established in 2010 with the expectation of providing long term data on the status of catheter ablation in the CHD population. The results of large scale initiatives such as these are likely to provide a vital insight into optimal patient management.

In summary, every patient with CHD-associated arrhythmia presents a new challenge and the potential benefits of successful treatment may be life-changing or even life-saving. We need to strive to integrate detailed understanding of the patient anatomy and arrhythmia substrate with the most appropriate technologies to give every chance of treatment success. Arrhythmia in CHD will continue to grow over the next 30 years, and the continuing evolution of reliable treatment modalities will have a profound impact on long-term outcome for this patient group. ■

1. Chubb H, Williams SE, Wright M, et al., Tachyarrhythmias and catheter ablation in adult congenital heart disease, *Expert Rev Cardiovasc Ther*, 2014;12:751–70.
2. Moons P, Engelfriet, Kaemmerer H, et al., Delivery of care for adult patients with congenital heart disease in Europe; results from the Euro Heart Survey, *Eur Heart J*, 2006;27:1324–30.
3. Bouchardy J, Therrien J, Pilote L, et al., Atrial arrhythmias in adults with congenital heart disease, *Circulation*, 2009;120:1679–86.
4. Vergheugt CL, Uiterwaal CS, van der Velde ET, et al., The emerging burden of hospital admissions of adults with congenital heart disease, *Heart*, 2010;96:872–8.
5. Lukac P, Pedersen AK, Mortensen PT, et al., Ablation of atrial tachycardia after surgery for congenital and acquired heart disease using an electroanatomic mapping system: which circuits to expect in which substrate? *Heart Rhythm*, 2005;2:64–72.
6. Santangeli P, Di Biase L, Buckhardt JD, et al., Transseptal access and atrial fibrillation ablation guided by intracardiac echocardiography in patients with atrial septal closure devices, *Heart Rhythm*, 2011;8:1669–75.
7. Meas S, Del-Carpio Munoz F, Cetta F, Friedman P, Transseptal access for pulmonary vein isolation in a patient with atrial septal closure device: use of electrocautery and atrial septostomy, *Heart Rhythm*, 2011;8:901–4.
8. Roudijk RW, Gujic M, Suman-Horduna I, et al., Catheter ablation in children and young adults: is there an additional benefit from remote magnetic navigation?, *Neth Heart J*, 2013;21:296–303.
9. Bradfield J, Tung R, Mandapati R, et al., Catheter ablation utilizing remote magnetic navigation: a review of applications and outcomes, *Pacing Clin Electrophysiol*, 2012;35:1021–34.
10. Kugler JD, Danford DA, Houston KA, et al., Pediatric radiofrequency catheter ablation registry success, fluoroscopy time, and complication rate for supraventricular tachycardia: comparison of early and recent eras, *J Cardiovasc Electrophysiol*, 2002;13:336–41.
11. Van Hare GF, Javitz H, Carmelli D, et al., Prospective assessment after pediatric cardiac ablation; demographics, medical profiles, and initial outcomes, *J Cardiovasc Electrophysiol*, 2004;15:759–70.
12. Seslar SP, Kugler J, Batra AS, et al., The multicentre pediatric and adult congenital EP quality (MAP-IT) initiative-rationale and design: report from the pediatric and congenital electrophysiology society's MAP-IT taskforce, *Congenit Heart Dis*, 2013;8:381–92.