

ARRHYTHMIAS in Adult Congenital Heart Disease

Annette Haynes RN, MS, CCRN
Cardiology Clinical Nurse Specialist
Stanford Hospital and Clinics
Palo Alto, California

INTRODUCTION

Symptomatic arrhythmias occur with increasing frequency for adult congenital heart disease (ACHD) as they move through adolescence and into adulthood. Arrhythmias are associated with increased hospital admissions resulting in significant morbidity including exercise intolerance, heart failure, thromboembolic events and mortality. Arrhythmias may be intrinsic to the structural malformation of specific congenital defects or acquired related to the surgical scars of early treatments and changes over time related to hypoxemia plus volume /pressure changes. Sudden cardiac death (SCD) is the greatest concern in ACHD. The greatest risk of late SCD is seen in tetralogy of Fallot (ToF), Transposition of Great Arteries (TGA), congenitally corrected Transposition of the Great Arteries (ccTGA), aortic stenosis (AS) and ventricular hypertrophy (VH). Development of atrial arrhythmias is more common in ACHD and important because of atrial thrombus formation.

ARRHYTHMIAS AND ASSOCIATED DEFECTS IN ACHD

ARRHYTHMIAS	ASSOCIATED DEFECTS
<i>Tachycardias</i>	
Accessory pathways	Ebstein's anomaly; ccTGA
Twin atrioventricular (AV) nodes	Heterotaxy syndrome
Intra-atrial reentrant tachycardia (atrial flutter)	Postoperative Mustard; post-op Senning, post-op Fontan; others
Atrial fibrillation	Mitral valve disease; aortic stenosis; unrepaired single ventricle
Ventricular tachycardia (VT)	Tetralogy of Fallot; congenital aortic stenosis; others
<i>Bradycardias</i>	
Congenital sinus node dysfunction	Heterotaxy syndrome
Acquired sinus node dysfunction	Post-op Mustard; post-op Senning; post-op Fontan; post-op Glenn; others
Congenital AV block	Endocardial cushion defects; ccTGA
Acquired AV block	Ventricular septal defect (VSD) closure; subaortic stenosis relief; AV valve replacement

Modified from Walsh, Circulation 2007

CRITICAL THINKING POINTS TO USE WHEN ADDRESSING PROBLEM

- Arrhythmia onset may be signal of hemodynamic decompensation
- Risk associated with arrhythmias may be amplified by abnormal underlying circulation
- Catheter ablation should be considered in symptomatic tachyarrhythmia
- Antiarrhythmic drugs are often poorly tolerated due to negative inotropy and other side effects
- Sudden unexplained shortness of breath (SOB), severe palpitations, syncope are serious warning signs
- Spontaneous VT patients should undergo invasive hemodynamic and electrophysiology (EP) evaluation

DIAGNOSTIC EVALUATION OF PROBLEM

- Baseline evaluation essential to review all prior surgical reports for knowledge of congenital and surgical anatomy, structural nuances.
- Invasive hemodynamic evaluation- may indicate need for surgical solution or intra-operative ablation/ maze procedure.
- Angiography
- Echocardiogram (ECG); TEE; Intracardiography (ICE)
- Computerized tomography scan (CT)
- Magnetic resonance imaging (MRI) to monitor status of intracardiac structures
- Holter monitoring, yearly ECG, exercise testing
- Electrophysiology study; Electroanatomic mapping systems; 3D mapping
- Symptom presentation: Syncope, palpitations, dizziness or syncope

TREATMENT

- MEDICATIONS
 - Antiarrhythmics
 - Have limited successful treatments with no clear long-term benefit
 - Risk of pro arrhythmia effects,
 - Aggravate sinus node dysfunction
 - Compromise ventricular function through the negative inotropic effect
 - Beta blockade for atrial arrhythmias
 - Amiodarone for atrial fibrillation
 - Anticoagulation
- ABLATION
 - Catheter ablation
 - High-powered cooled tip catheters
 - 3-D mapping
 - Surgical revisions
 - Maze procedure with surgical revisions of defect
 - Combination incisions and cryoablations
- DEVICE THERAPY
 - Pacemakers
 - Dual chamber pacing for sinus node dysfunction
 - Bi Ventricular pacing for heart failure patients

- Reentry tachycardias interrupted with atrial tachycardia sensing and auto burst rate pacing
- A-V Synchrony for ventricular failure
- ICD (internal cardioversion defibrillator) for increased sudden cardiac death (SCD) risk

ASSOCIATED COMPLICATIONS

- Complete heart block (CHB)
- SCD
- Progressive Cardiac failure requiring transplantation

SPECIAL CONSIDERATIONS

- Electrophysiology procedures require specialists in ACHD
 - Increased success to anticipate and react to hemodynamic change
 - Specialists
 - Electrophysiologist (cardiologist specialized in electrophysiology)
 - Cardiovascular anesthesiologist
- Placement of pacing leads and pulse generator
 - Increased difficulty
 - Epicardial pacemaker implantation favored
 - Limited venous access
 - Need for lead and generator replacement
 - Epicardial sites
 - Multiple prior cardiac operation result in scarred mediastinum
 - Careful dissection required to expose with good sensing and pacing function.
 - May place epicardial leads when operating if likely need for future pacing. Eighty-six % of leads placed at operation function well when retrieved at a mean of 252 days post-op.
 - Abdominal generator implantation may lessen external appearance decreasing psychological issues for adolescents

REFERENCES

Biviano A: Atrial flutter catheter ablation in adult patients with repaired Tetralogy of Fallot: mechanisms and outcomes of percutaneous catheter ablation in a consecutive series, *Journal of Interventional Cardiology Electrophysiology* 28(2):125-135, 2010. Available at <http://www.springerlink.com/content/3900633ux5839127/fulltext.pdf>

de Groot N: Ablation of focal atrial arrhythmia in patients with congenital heart defects after surgery: role of circumscribed areas with heterogeneous conduction, *Heart Rhythm* 3:526-535, 2006.

de Groot N:) Long-term outcome after ablative therapy of postoperative atrial tachyarrhythmia in patients with congenital heart disease and characteristics of atrial tachyarrhythmia recurrences, *Circulation Arrhythmia Electrophysiology* 3:148-154, 2010.

Diller, G-P: Cardiac resynchronization therapy for adult congenital heart disease patients with a systemic right ventricle: analysis of feasibility and review of early experience. *Europace* 8, 267-272, 2006

ESC Guidelines 2010.

Kanter R: Pearls for ablation in congenital heart disease. *Journal cardiovascular electrophysiology* 21: 223-230, 2010.

Lim D: Illustrated field guide to adult congenital heart disease. Charlottesville, VA , 2009, Scientific Software Solutions, Inc.

Mavroudis C: Arrhythmia surgery in patients with and without congenital heart disease, *Annals of Thoracic Surgery* 86:857-68, 2008.

May L: *Pediatric heart surgery. A ready reference CE for professionals*, Milwaukee, WI, 2008, maxiSHARE.

Sable et al: Best practices transition to ACHD. *Circulation*, 2011.

Saul J: Role of catheter ablation in postoperative arrhythmias, *PACE* 31:S7-S12, 2008.

Snyder C: Can pediatric electrophysiologist safely perform electrophysiology studies on adults with congenital heart disease?, *The Ochsner Journal* 7:1: 16-19, 2007.

Walsh E: Arrhythmias in adult patients with congenital heart disease, *Circulation* 115: 534-45, 2007.

7/2011