

ARRHYTHMIAS in Adult Congenital Heart Disease

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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. 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; congenitally corrected transposition of the great arteries (ccTGA)
Twin atrioventricular (AV) nodes	Heterotaxy syndrome
Intra-atrial reentrant tachycardia (atrial flutter)	Postoperative Mustard; post-op Senning, post-op Fontan; Atrial septal defect (ASD); Atrioventricular (AV) septal defect; Epstein anomaly; Eisenmenger physiology
Atrial fibrillation	Mitral valve disease; aortic stenosis; unrepaired single ventricle; ASD; Eisenmenger physiology
Ventricular tachycardia (VT)	Tetralogy of Fallot (TOF); congenital aortic stenosis; Transposition of great arteries (TGA); Epstein, single ventricle; Ventricular septal defect (VSD); pulmonary hypertension (PH)
<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; D-TGA
Congenital AV block	Endocardial cushion defects; ccTGA; L-TGA; AVSD
Acquired AV block	Ventricular septal defect (VSD) closure; subaortic stenosis relief; AV valve replacement
<i>Sudden Cardiac Death (SCD)</i>	TOF; TGA; ccTGA; AS; ventricular hypertrophy.

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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
- Surgical ablation with Atrial Maze procedure is beneficial for refractory IART or if surgery is planned for other revisions
- 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 and advanced imaging 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 recommended for atrial fibrillation
- 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
 - AAIR or DDDR recommended for symptomatic older ACHD pts
 - 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
 - Increases success to anticipate and react to hemodynamic change
 - Frequently have multiple pathway
 - Location of AV node
 - Specialists
 - Electrophysiologist (cardiologist specialized in electrophysiology)
 - Cardiovascular anesthesiologist
- Placement of pacing leads and pulse generator
 - Increased difficulty venous stenosis; anatomic lesions, fibrosis and scarring
 - Epicardial pacemaker implantation favored
 - Limited venous access
 - Need for lead and generator replacement
 - Epicardial sites
 - Multiple prior cardiac operations 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. 86 % of leads placed at operation function well when retrieved post-op.
 - Abdominal generator implantation may lessen external appearance decreasing psychological issues for adolescents
- Patients with Mustard or Senning procedures may require baffle stenting to alleviate any obstructions before attempting lead implants

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