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. 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

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


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.


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