

Case 1: 31 y/o with d-TGA



Case 1: 29 y/o with d-TGA

- **Mustard procedure and VSD closure at age 5**
- **Routine follow up; no cath; no further surgery**
- **Occasional palpitations**
- **16 y/o Holter: Sinus HR 44 – 155 bpm, few PAC's**
- **21 y/o Holter: Sinus HR 37 – 183 bpm, few PAC's**
- **22 y/o Holter: Sinus HR 39 – 163 bpm, few PAC's**
- **26 y/o Holter: Sinus HR 40 – 170 bpm, few PAC's**

Case 1: 29 y/o with d-TGA

- Presents for routine follow-up
- Two episodes of subjective tachycardia: 3 – 10 min
- No other symptoms
- Holter: sinus HR 44 – 149, frequent PACs, no pauses

- 9 months later: palpitations with lightheadedness
- In the ER, HR 140 – atrial flutter
- Cardioversion (Pt refused EPS/ablation)
- No meds (Pt refused)

Case 1: 29 y/o with d-TGA

- Continuing subjective palpitations
- Event recorder: PACs, couplets, short runs of atrial tachycardia with variable conduction (70/140 bpm)

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- 6 months later: pt agrees to EP evaluation
- EP recommendations:
 - No need for ICD
 - AV node blocking medication
 - or
 - Electrophysiologic study with ablation

Case 1: 29 y/o with d-TGA

- 1 months later: pt seeks 2nd opinion
- EP recommendations:
 - Low dose beta blocker or calcium channel blocker
 - Ablation for recurrent arrhythmias

Case 1: Questions

- **Surveillance**
- **Pacer indications**
- **Pre-pacer evaluation of venous pathways**
- **Response to PACs**
- **Response to IART**
- **Chemical vs. Electrical cardioversion**
- **Drugs for IART, when & and which ones**
- **Challenge of ablation in atrial switch**

Case 1: Guidelines

11.9. Recommendations for Electrophysiology Testing/Pacing Issues in Dextro-Transposition of the Great Arteries

Class I

1. Clinicians should be mindful of the risk of sudden arrhythmic death among adults after atrial baffle repair of d-TGA. These events usually relate to VT but may be caused in some cases by rapidly conducted IART or progressive AV block. (*Level of Evidence: B*)

2. Consultation with an electrophysiologist who is experienced with CHD is recommended to assist with treatment decisions.

(*Level of Evidence: B*)

3. Pacemaker implantation is recommended for patients with d-TGA with either symptomatic sinus bradycardia or sick sinus syndrome.

(*Level of Evidence: B*)

Case 1: Guidelines

11.9. Recommendations for Electrophysiology Testing/Pacing Issues in Dextro-Transposition of the Great Arteries

Class IIa

1. Routine surveillance with history, ECG, assessment of RV function, and periodic Holter monitoring can be beneficial as part of routine follow-up.

(Level of Evidence: B)



Case 2: 40 y/o Fontan

- Tricuspid atresia with normally related great arteries
- Neonate: TE fistula repair
- 4 y/o BT shunt
- 16 y/o Atrio-pulmonary Fontan procedure
- Followed for 20 years by adult cardiologist/internist
- Frequent arrhythmias
- Cardioversion at 36 y/o and 37 y/o
- Treated with sotalol, propafenone, flecainide...
- Coumadin for a short time, d/c'd for epistaxis
- Currently on amiodarone 200 mg daily

Case 2: 42 y/o Fontan

- 2 months of symptoms: decreased energy level, fullness
- Heart rate 95 bpm
- Past week: GI symptoms, LE edema
- Sudden increase in HR > 150 bpm, referred to ER
- IV diltiazem, heparin (HR now 95 bpm)
- Diltiazem d/c'd; metoprolol started
- Reversion to sinus rhythm 24 hours later

Case 2: 42 y/o Fontan

- No TEE
- MRI: no thrombi; LVEF 36%; massive right atrium
- Pleural effusion, LE edema, creatinine 1.3 → 1.6
- Albumin 3.6 g/dl

- EP study: IART induced, ablation unsuccessful
- DC cardioversion → SR 50 bpm
- 25 # diuresis, creatinine 1.1
- Discharged home:
 - amiodarone 100 mg, carvedilol 6.25 bid
 - lasix, lisinopril
 - coumadin


Case 2: 42 y/o Fontan

- All symptoms resolve; back to work
- Exercise study:
 $V_{O_2} = 15.2 \text{ ml/kg/min (41\%)}$, HR 44 ► 82 bpm
- Cardiac Cath:
 CI = 1.7 L/min/m² , Fontan pressure = 12; LVEDP = 11 mm Hg
- Asymptomatic for 17 months
- Recurrent IART – DC cardioversion

Case 2: 42 y/o Fontan

- 6 months later: bradycardia on routine exam
- Admitted to outside hospital; coreg and amiodarone d/c'd
- 2 weeks later: shows up in ACHD clinic in IART, HR 120

Case 2: Questions

- **Surveillance**
 - **Fib vs. flutter?**
 - **Recognition of IART**
 - **Rx of sinus node dysfunction**
 - **Chemical vs. Electrical cardioversion**
 - **Drugs for IART, when & and which ones**
 - **IART ablation**
 - **Fontan conversion**
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Case 2: Guidelines

14.10.1. Recommendations for Electrophysiology Testing/Pacing Issues in Single-Ventricle Physiology and After Fontan Procedure

Class I

1. Arrhythmia management is frequently an issue in patients after the Fontan procedure, and consultation with an electrophysiologist with expertise in CHD is recommended as a vital part of care.

(Level of Evidence: C)

2. New-onset atrial tachyarrhythmias should prompt a comprehensive noninvasive imaging evaluation to identify associated atrial/baffle thrombus, anatomic abnormalities of the Fontan pathway, or ventricular dysfunction.

(Level of Evidence: C)

Case 2: Guidelines

14.10.1. Recommendations for Electrophysiology Testing/Pacing Issues in Single-Ventricle Physiology and After Fontan Procedure Class I

3. Electrophysiological studies in adults with Fontan physiology should be performed at centers with expertise in the management of such patients.

(Level of Evidence: C)

4. Clinicians must be mindful of the high risk for symptomatic IART in adult patients who have undergone the Fontan operation. This arrhythmia can cause serious hemodynamic compromise and contribute to atrial thrombus formation. Treatment is often difficult, and consultation with an electrophysiologist who is experienced with CHD is recommended whenever recurrent IART is detected.

(Level of Evidence: C)

Case 2: Guidelines

“If IART episodes are infrequent (less than 1 per year or so), well tolerated, and recognized promptly, it may be sufficient to rely on periodic cardioversion rather than embark on therapy with potent antiarrhythmic drugs or invasive procedures. In such cases, a chronic AV node–blocking agent (digoxin, beta blocker, or calcium channel blocker) may be prescribed to reduce the risk of a rapid ventricular response during subsequent episodes, and chronic anticoagulation is usually prescribed as well.”

ACC/AHA 2008 Guidelines for Management of Adults with Congenital Heart Disease.
Circulation 2008, December 2, 2008

Case 2: Guidelines

“If the episodes are frequent, cause significant symptoms, go unrecognized by the patient for a long time, or are associated with atrial thrombus formation, 1 or more of the aggressive treatment options **must** be considered. This is particularly true for patients with extremely dilated right atria and patients with hemodynamic concerns such as reduced function of their single ventricle, AV valve regurgitation, or pulmonary vein compression.”


“As discussed in Section 1.9, Recommendations for Arrhythmia Diagnosis and Management, options for aggressive therapy include pacemaker implantation to reverse bradycardia or provide automatic atrial antitachycardia therapy, drug therapy, catheter ablation, and surgical revision of the Fontan connection combined with an atrial Maze operation. The choice must be tailored to the hemodynamic and electrophysiological status of the individual patient.”

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Circulation 2008, December 2, 2008

Case 3: Transvenous Pacing with an Intracardiac Shunt

- #1 S/p VSD closure with small patch leak
- #2 TOF How do you exclude PFO?
- #3

Case 4: The perioperative period

- ACHD patient undergoing cardiac surgery
 - Peri-op antiarrhythmics?
 - Indications for pre-op EP study
 - When to include a Maze?
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Case 5: Atrial fib in the ACHD pt.

- **E.g. Cleft mitral valve, s/p primum ASD repair**
- **How does management differ from the non-CHD population?**
- **Is A Fib an indication for MV repair?**
- **When to include a Maze?**
- **When is catheter ablation indicated?**
- **When should we “tolerate” and rate control Afib?**
- **Indications and threshold for antibradycardia pacing**

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