Management of Arrhythmias in Adults with Congenital Heart Disease

Introduction

• What makes the adult with congenital heart disease special:
  – Lifelong contact with medical professionals
  – Often multiple surgeries, multiple providers and centers
  – Arrhythmias may only be an issue as an adult

Introduction

• Bridging the gap
  – Pediatric Cardiologists
    • Not familiar with “adult” problems
    • Privileging issues
  – Adult Cardiologists
    • Not familiar with congenital heart disease
  – Adult Congenital Heart programs
    • Probably the best “fit”, but few centers exist
    • Difficult to organize
  • Numbers limited
Arrhythmias in Adults with CHD

Lesion types and arrhythmias

- Unoperated CHD
  - Acyanotic:
    - Atrial septal defect
    - Ventricular septal defect
    - AV septal defects
    - Patent ductus arteriosus
    - L-Transposition of Great Arteries ("Corrected" TGA)

- Atrial septal defect (ASD)
  - SAN dysfunction (prolonged SNRT)
    - Older patients
    - Large left to right shunt
    - Sinus venosus type ASD
  - Tachyarrhythmias
    - Older patients (over 50yrs = 50%, Paolillo)
    - Higher pulmonary vascular resistance
    - AFIB, AFL
Arrhythmias in Adults with CHD

Lesion types and arrhythmias

- Unoperated CHD (con't):
  - Cyanotic:
    - Tetralogy of Fallot
    - Pulmonary valve stenosis,
    - Ebstein’s malformation of tricuspid valve

- Ebstein’s
  - malformation of tricuspid valve
    - Valve remains adherent to RV wall
    - “Atrialized” RV
    - Obstruction to RVOT
    - ASD with right to left, chronic cyanosis

- First degree AVB, RV conduction delays
- Older age
- Atrial tachyarrhythmias
- WPW/AVRT; multiple pathways; annulus poorly defined
- Sudden death – VT/VF
Arrhythmias in Adults with CHD

Lesion types and arrhythmias

- “Other” pre-operative CHD
  - “Heterotaxy” syndrome
  - Single Ventricle
  - Coronary artery abnormalities
  - Eisenmenger physiology (ASD, VSD, etc)
  - Cardiac Tumors
Arrhythmias in Adults with CHD

Lesion types and arrhythmias

• “Heterotaxy” syndrome
  – Thoracic and abdominal situs abnormalities
  – Bilateral right or left “sidedness”
  – Mild to severe congenital heart disease
  – Congenital AV block, SAN dysfunction, others

5yr old “murmur”, left atrial rhythm

Arrhythmias in Adults with CHD

Lesion types and arrhythmias

• Perioperative arrhythmias
  – Junctional tachycardia
    • Narrow QRS with (usually) AV dissociation
    • Probably transient edema of His bundle
    • Fix electrolytes, induced hypothermia, amiodarone
  – Anything goes
    • Surgical AV block
    • Brady and tachyarrhythmias
Arrhythmias in Adults with CHD

Lesion types and arrhythmias

- Chronic post-operative arrhythmias
  - Atrial surgery
    - Mustard or Senning for d-TGA – the old “atrial switch” operation
      - SND with junctional rhythm (approx. 10% paced at mean 8 yrs post-op)
      - Flutter/atrial reentry, AFIB
  - Fontan operation for “single ventricle”
    - Quite variable anatomy (hypoplastic RV or LV)
    - Complex surgery, RA stretch, decreased ventricular function, AVV insufficiency
    - SND, AFL/FIB
    - “Old style” Fontan – up to 50% atrial arrhythmias at 10 yrs

Arrhythmias in Adults with CHD

Lesion types and arrhythmias

- Chronic post-operative arrhythmias; Atrial surgery (con’t)
  - Fontan operation for “single ventricle”
  - Medical management – type 3 drugs exacerbate bradycardia, potentially pro-arrhythmic
  - Ablation for atrial reentry
    - Access issues
    - Poor long term results
  - Surgical “re-do”

Arrhythmias in Adults with CHD

Lesion types and arrhythmias

- Treatment of chronic atrial arrhythmias in single ventricle and atrial switch
  - Prevent severe bradycardia – pacing alone
    - Symptomatic, Bradycardia w/ CHF, tachy-brady syndrome
  - Medical management – type 3 drugs exacerbate bradycardia, potentially pro-arrhythmic
  - Ablation for atrial reentry
    - Access issues
    - Poor long term results
  - Surgical “re-do”
Arrhythmias in Adults with CHD

Lesion types and arrhythmias

• Chronic arrhythmias after ventricular surgery (eg. Tetralogy of Fallot)
  – Conduction problems:
    • Nearly all have RBBB
    • Most concerning are symptomatic pts, slow AV Wenckebach rates, Mobitz II, CAVB, marked HV prolongation at EPS

• Ventricular arrhythmias in tetralogy of Fallot
  – Sudden death not insignificant
  – Predisposing factors
    • Older age at repair
    • Higher RV pressure (residual obstruction)
    • Long time since repair
  – Scar-mediated macro-reentry
    • Medical management
    • Catheter ablation – pace mapping, entrainment, slow zones of conduction
    • Surgical reconstruction, cryoablation, RF ablation

• Tetralogy of Fallot
  – 15yr old
  – Definitive repair at 8yrs
  – Syncope, “SVT” diagnosed in Puerto Rico, not responsive to adenosine; Rx w/ amio
  – EPS – 2 sustained VT morphologies with V stim testing
  – Reconstruction of RVOT, surgical cryoablation
  – Repeat EPS in 6 months - negative
**New Mapping Technologies**

- Electromagnetic field mapping
  - Magnetic field generated by device under pt
  - Sensor in catheter tip
  - Simultaneously collects beat-by-beat electrogram and position of catheter tip
  - Generates 3D map

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**PA view of RA in Ectopic Atrial Tachycardia**

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**New Mapping Technologies**

- 50 year old Holt Oram
  - ASD repair
  - S/p TVR, CAVB
  - IART (“flutter”)
  - S/p failed “standard” isthmus RF ablation
  - S/p failed right atrial surgical Maze
  - Biosense® showed RA as “bystander”
  - Left atrial flutter; zone of slow conduction betw ASD patch and MV
  - Transeptal via patch
Arrhythmias in Adults with CHD

Pacemaker issues

- Unique anatomic issues
  - Epicardial access only?
  - Left SVC
  - Individualized treatment
- Long term pacing
  - “Saving” venous access
  - Lead extraction WILL be needed
  - Accounting for growth

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Arrhythmias in Adults with CHD

Pacemaker issues

- 40 year old Ebstein’s, tetralogy of Fallot, left SVC and no bridging vein
  - Surgical AVB
  - Left SVC VVI; lead failure w/ partial extraction
  - Right sided implant
- Atrial Tachycardia
  - ? RF ablation

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Arrhythmias in Adults with CHD

Pacemaker issues

15 year old d-Transposition, s/p atrial switch operation, bradycardia
Arrhythmias in Adults with CHD

Pacemaker issues

• 50 year old Holt Oram
  - ASD repair
  - Severe RV dilation, s/p tricuspid annuloplasty
  - S/p TVR, CAVB
  - Epicardial VVI pacer
  - S/p failed surgical Maze
  - S/p Left sided RF ablation
  - Endocardial atrial lead, tunneled to abdomen
  - DDD-R device; marked improvement in QOL

Arrhythmias in Adults with CHD

Pacemaker issues

• 16 year old Down Syndrome
  - VSD repair, surgical AVB; epicardial VVI
  - Endocardial pacer from left
  - Leads failed and removed
  - Right implant VVI, lead failed and DDD implanted
  - High thresholds

Arrhythmias in Adults with CHD

Pacemaker issues

• 8 year old Heterotaxy
  - Single atrium
  - Abdominal situs inversus
  - Congenital complete AVB
  - Endocardial VVI system at 3 yrs
Arrhythmias in Adults with CHD

Pacemaker issues

- 14 year old
- LQT, borderline
- Unexplained syncope
- Reveal® Plus
- Torsades
- ICD implanted

14 year old Long QT Syndrome

Arrhythmias in Adults with CHD

Pacemaker issues

Implant 18 months
**Arrhythmias in Adults with CHD**

**Conclusions**

- Very special people
- Lots of “baggage” from a “prior life”
  - New arrhythmias
  - Growth
- Individualized care
- Appreciate the extra time and patience

**Questions?**