Adult Congenital Heart Disease – “Anatomy” of a “Growing Problem”

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Overview

• Demographics of congenital heart disease in the adult
• General approach to evaluation of the adult with congenital heart disease
• Anatomy, physiology and management of selected lesions
• Focus on the major problems that plague these patients as adults
• References to recent clinical guidelines
Current Landscape of CHD

0.8% of live births in the U.S. excluding bicuspid AV, MVP
- Improved diagnostic techniques
- Improved medical, catheter-based and surgical techniques

Now >1,000,000 adults with congenital heart disease (adults>children)

Minimal exposure during training
Improving Natural History of Children with CHD = More Adults with ACHD

1960s

- Surviving to Adulthood: 50%
- Died in First Year: 15%
- Died within 18 Years: 35%

2010

- Surviving to Adulthood: 85%
- Died in First Year: 10%
- Died within 18 Years: 5%
The Trend to Geriatric CHD
Long-term Issues in the CHD Patient

• Patients believe they are cured – reality is they were usually only palliated
  – Lesions can recur
  – Palliative methods can cause problems
    – The right ventricle and tricuspid valve are ill-equipped to handle systemic pressure

• Shunt lesions can result in pulmonary hypertension, arrhythmias and heart failure, even after repair

Current Landscape of CHD

• Can separate into 2 general groups
  – Patients recognized and rx as children
  – Adults diagnosed *de novo*

• Due to complexity adults with complex CHD best followed by subspecialists

• Nearly all ACHD patients followed by internists +/- general cardiologists
### ACC/AHA Guidelines for Monitoring ACHD

#### Being seen at an adult congenital heart disease center:

<table>
<thead>
<tr>
<th>Complexity</th>
<th>Frequency</th>
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<tbody>
<tr>
<td>Simple</td>
<td>At least once to determine needs for future follow-up</td>
</tr>
<tr>
<td>Moderate complexity</td>
<td>Every 12 to 24 months</td>
</tr>
<tr>
<td>Great complexity</td>
<td>Every 6 to 12 months</td>
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Adult Congenital Heart Disease: Trends in Hospitalization

Annual Pediatric and Adult CHD Admissions

Number of admissions

Year


Pediatric
- Unclassified, <18
- Simple, <18
- Complex, <18

Adult
- Unclassified, ≥18
- Simple, ≥18
- Complex, ≥18

O’Leary J et al. JAMA 2013
General Approach to Adults with CHD

• Understand the anatomy and surgeries
  – Review the pediatric records

• Be aware of warning signs and sxs
  – Syncope
  – Progressive exertional dyspnea
  – Increasing palpitations
General Approach to Adults with CHD

• Careful clinical exam with auscultation
  – Look at JVP, feel pulses in all extremities
  – Listen for “hard to hear” murmurs

• Utilize noninvasive diagnostic modalities
  – ECG
  – Echocardiogram
  – CT and MRI
Cardiac MRI in the CHD Patient

• Useful when echo data is inadequate or inconsistent

• May avoid need for cardiac cath
  – Ionizing radiation exposure
  – Contrast
  – Vascular access issues

• Advantages of CMR
  – Evaluate extra-cardiac vasculature
  – Quantify ventricular volumes/mass/function and blood flow
  – Tissue characterization

Role of Diagnostic Cardiac Catheterization in ACHD Work-up

• Generally performed later in w/u than in the past
• MRI and CT provide similar images less invasively
• Remains “Gold Standard” for assessment of:
  – intracardiac pressures
  – oxygen saturations
  – cardiac output calculation
  – determining pulmonary vascular resistance and reactivity
• Careful data collection and proper documentation is critical
200 Consecutive Complex Congenital Cath Cases in the Cleveland Clinic Adult Lab
Shunt Lesions

- Most common form of ACHD
- Frequently diagnosed in the adult population
- Results in increased pulmonary blood flow
  - Right heart enlargement
  - Arrhythmias → atrial fibrillation
  - Pulmonary hypertension
Early Foramen Physiology

- In-utero passage of blood, bypassing pulmonary circulation

Maintains patency in ~25% of adults

Atrial Septal Defect vs Patent Foramen Ovale

**ASD**
- Incidence ~1/1000
- Usually L to R shunt
- Also has R to L shunt
- Association with stroke
- Can be concurrent with ASA
- Results in “flow” complications
  - Right heart enlargement
  - Pulmonary HTN
  - Atrial fibrillation

**PFO**
- Incidence ~1/4
- Usually only R to L shunt
- “Stretched PFO” can result in L to R shunt
- Association with stroke
- Can be concurrent with ASA
- No “flow” complications

Atrial Septal Defect

- Most common cardiac malformation in adults
- More common in females 2-3:1
- 75% are secundum defects
- Symptoms can be very subtle – Dyspnea and fatigue most common
- Commonly mistaken for other disorders

Krasuski RA  CCJM  2007;74(2):137-47.
Types of Atrial Septal Defects
Secundum ASD

- Rule of 10%
  - 10% with multiple defects
  - 10% with anomalous veins
  - 10% unrepaired can develop Eisenmenger’s

- Shunt determined by size of defect and compliance of ventricles

- Decompensation can occur in older patients
  - LV diastolic dysfunction (HTN, CAD)
  - Atrial fibrillation
  - Development of pulmonary HTN
What constitutes a significant ASD?

- Qp/Qs > 1.5
- RA+RV Enlargement
- ~ Normal PVR (<7-10 Wood units or PVR/SVR<0.3)
- Anatomy conducive to percutaneous repair
  - No anomalous pulmonary veins
  - Secundum defects only
  - Good septal rims
“Medical Therapy” vs. Surgical Correction

Atrial Fibrillation as Source of Morbidity Following Surgical Repair

Therapeutic Approach to ASD

• Medical
  – Antibiotic prophylaxis not necessary
  – Arrhythmia prophylaxis controversial

• Surgical repair
  – <1% mortality
  – Significant morbidity, discomfort, scar

• Percutaneous
  – Gold standard for simple, significant secundum defects
  – Role being challenged by erosion risks
ASD Occluders

- Amplatzer
- ASDOS
- Sideris Button
- Angel Wing
- CardioSeal
- Helex
- Guardian Angel
- StarFlex
Closure of an ASD either percutaneously or surgically is indicated for right atrial and RV enlargement with or without symptoms.

A sinus venosus, coronary sinus, or primum ASD should be repaired surgically rather than by percutaneous closure.

Surgeons with training and expertise in CHD should perform operations for various ASD closures.

Warnes CA, Williams RG, Bashore TM et al. J Am Coll Cardiol 2008;52(23): e1-121.
Primum ASD/AV Canal Defect

- Cleft mitral valve is often associated with a primum ASD or “Atrioventricular Canal Defect”
- Forms from failure of embryonic endocardial cushions to meet and normally partition the heart.
- “Complete” has 4 components:
  - Inlet VSD
  - Primum ASD
  - Cleft mitral valve
  - Widened antero-septal tricuspid commissure
- If the VSD is absent = “Partial” AV canal
Partial vs Complete AV Canal

Partial AV Canal

Complete AV Canal

ASD

VSD
Complete AV Canal Defect
Sinus Venosus ASD

- Defect is located near the junction of the SVC or IVC with the RA (posterior to the fossa ovalis)
- Often difficult to find—typically need TEE or MRI
- Suspect in unexplained right sided dilatation - perform agitated saline contrast for assessment
- Usually associated with anomalous connection of right pulmonary vein(s) to RA
Sinus Venosus ASD and Partial Anomalous Pulmonary Vein Return (PAPVR)
Surgical Repair of Sinus Venosus ASD and PAPVR
Ventricular Septal Defects (VSD)

- Most common congenital lesion seen in children (~25%)
- Less common in adults (2nd after ASD)
  - Smaller lesions often close spontaneously
  - Larger lesions present with heart failure and get repaired
- Many different types
  - Membranous
  - Muscular
  - Inlet
  - Outlet
Locations of VSDs

1. Muscular VSD - often multiple defects (10% of VSD)
2. Supracristal VSD - involving LVOT/RVOT - (5%)
3. AV canal defects - involving inflow portion of the septum
4. Membranous VSD – (80%)
General VSD Facts

• Membranous often have associated “aneurysmal” tissue

• Small (<0.5 cm) = restrictive
  – Loud murmurs
  – Asymptomatic

• Larger lesions
  – Softer Murmurs
  – LV volume overload and pulmonary hypertension
  – Most common cause of Eisenmenger syndrome
Patent Ductus Arteriosus (PDA)
PDA Epidemiology

- Incidence in isolation 1:2000 to 1:5000 births (10-15% of adult congenital heart lesions)
- Isolated defect in 75% (~all of those presenting as adults)
- 2:1 to 3:1 women (↑ ratio in older pts)
- Usually funnel-shaped with base on aortic side and neck at PA (~75% in largest series)
Pulmonary Hypertension Complicates a Variety of ACHD Lesions

- Complex
  - Mitral Atresia
  - d-TGA
  - CCTGA
  - DORV
  - Heterotaxy
  - Single ventricle
  - Conduits
  - Truncus
  - Cyanotic
  - Eisenmenger

- Moderate
  - TOF
  - SV defect
  - APV drainage
  - AVC
  - Primum ASD
  - Sub PS
  - AoCo
  - Ebstein
  - VPS
  - PR
  - Complex PDA or VSD

- Simple
  - Large defects (ASD > 2 cm, VSD > 1 cm, PDA > 0.6 cm)
  - Simple ASD
  - Simple Aortic Disease
  - Simple Mitral Disease
  - Simple PDA
  - Mild valvular PS

- 60% with prior operations
- 50% will have reoperation
- 3:1 interventions cath-based

Red font = associated with development of PAH

CHD-PH Accounts for 10% of PAH in the REVEAL Registry

N=2525

- IPAH 46.2%
- APAH 50.7%
- CVD/CTD 49.9%
- CHD 19.5%
- Drugs/toxins 10.5%
- Portal HT 10.6%
- Other** 5.5%
- HIV 4%

PH in CHD Greatly Impacts Long-Term Outcome

The Montreal CHD Database

PH Prevalence ~5.8%

Pulmonary Hypertension in the ACHD Patient

• Can be pulmonary venous or pulmonary arterial

• Depending on lesion, can have left ventricular dysfunction, right ventricular dysfunction or both

• Differentiation is essential and impacts management
Prognostic Indicators for Developing PAH in ACHD

- Type and size of defect
- Magnitude of shunt flow (Qp:Qs ratio)
- Surgical repair (correction, palliations, age at repair)
- Patient age

Duffels MGJ et al. *Int J Cardiol* 2007;120:198-204.
PAH Often Complicates Congenital Systemic-to-Pulmonary Shunts

- Patients with repaired and unrepaired defects can develop PAH (~2-10%)
- Increasing dyspnea, declining exercise capacity and progressive increase in PVR are clinical hallmarks
- 25% to 50% of CHD-PH patients progress to Eisenmenger Syndrome

Progression of PAH-CHD to Eisenmenger’s Syndrome:

1. Systemic-to-pulmonary (left-to-right) shunt
2. Increased pulmonary blood flow and pressure
3. Pulmonary vascular remodeling
4. Increase in pulmonary vascular resistance
5. Reversed (right-to-left) shunt: pulmonary-to-systemic
6. Cyanosis (Eisenmenger Syndrome)

References:
Progression of PAH-CHD to Eisenmenger’s

1. Systemic-to-pulmonary (left-to-right) shunt
2. Increased pulmonary blood flow and pressure
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References:
Insights into Pathophysiology

• Cardiac Biomarkers
  – Markers of Endothelial Dysfunction/Damage
    – ADMA
    – VEGF
  – Markers of Inflammation
    – hsCRP
    – IL-6
  – Markers of Right Ventricular Strain
    – BNP and NTpro-BNP
  – Potential Genetic Mediators

Eisenmenger Syndrome

• Classic description by Wood in 1958
• Pulmonary vascular disease progresses to systemic pressures and shunt reverses
• Multiple systemic complications (hypoxia)
  – Erythrocytosis
  – Proteinuria and ↓ GFR
  – Increased Uric Acid

Eisenmenger Syndrome

- Patients can rapidly deteriorate
  - ARF from contrast dye load
  - Arrhythmias
  - Anesthetic agents
- Phlebotomy should only be performed in patient is symptomatic
  - Check for iron deficiency and replete if necessary
  - Equal saline repletion if phlebotomy is performed
- Survival and functional capacity reduced significantly

Limited Previous Impact on Survival in Eisenmenger Syndrome

Impact of Vasodilator Response in CHD-PH

- Response to vasodilator challenge appears important in prognosis
- Challenge may also unmask pulmonary venous contribution to PH
- Reversal of PAH in some patients may make surgical correction eventually feasible

Therapeutic Options in ACHD-PH Patients

• Pharmacologic
• Percutaneous
• Surgical
• Hybrid Therapies
Impact of Shunt Repair and PAH on Survival

A. ASD

Follow-up (years)

Survival (%)

Patients remaining at risk

Eisenmenger Syndrome (75.4%)

Closed, no ‘PAH’ (99.1%)
Open, no ‘PAH’ (98.4%)
Open, ‘PAH’ (97.2%)
Closed, ‘PAH’ (94.8%)

B. VSD

Follow-up (years)

Survival (%)

Eisenmenger Syndrome (82.6%)

Patients remaining at risk

Closed, no ‘PAH’ (99.1%)
Open, no ‘PAH’ (98.7%)
Open, ‘PAH’ (96.7%)
Closed, ‘PAH’ (93.1%)
Lesion Repair in CHD: Severities of Disease State to Consider

<table>
<thead>
<tr>
<th>Condition</th>
<th>Pathology/Consequence</th>
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</thead>
<tbody>
<tr>
<td>Pulmonary Hypertension</td>
<td>Normal Vasculature Elevated Pulmonary Pressure</td>
</tr>
<tr>
<td>Pulmonary Hypertension and Pulmonary Vascular Disease (iPAH)</td>
<td>Permanently remodeled vascular bed Elevated Pulmonary Pressure and Elevated Pulmonary Vascular Resistance</td>
</tr>
</tbody>
</table>

Surgery or catheter intervention is possible

Surgery or catheter intervention may worsen prognosis

Beghetti et al. Congenit Heart Dis 2012;7:3–11
<table>
<thead>
<tr>
<th>Therapy</th>
<th>Role</th>
<th>Rationale</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oral Anticoagulants</td>
<td>Controversial</td>
<td>Clinical trial data lacking and potential for bleeding serious abnormalities</td>
</tr>
<tr>
<td>Nitric Oxide</td>
<td>Acute post-operative management to reduce PVR</td>
<td>Studies have shown inhaled NO to reduce PVR with minimal systemic effects in patients with ES</td>
</tr>
<tr>
<td>Long Term O₂</td>
<td>Controversial</td>
<td>Limited data and potential risk and side-effects</td>
</tr>
<tr>
<td>Calcium Channel Blockers</td>
<td>Not recommended in most patients with CHD/ES</td>
<td>In ES patients, potential for worsened right-to-left shunt, increasing cyanosis and inducing hypotension</td>
</tr>
<tr>
<td>Prostacyclins</td>
<td>Improved 6-MWD and hemodynamics</td>
<td>Several studies exist with epoprostenol, treprostinil and iloprost</td>
</tr>
<tr>
<td>Endothelin Receptor Antagonists</td>
<td>Improved 6-MWD and hemodynamics without worsening of SpO₂</td>
<td>Bosentan approved for use (BREATHE-5) Ambrisentan-no patients enrolled in RCT, but limited data available</td>
</tr>
<tr>
<td>Phosphodiesterase Inhibitors</td>
<td>Improved hemodynamics</td>
<td>A few observational and randomized studies exist with sildenafil and tadalafil</td>
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</table>
Suspected Diagnosis of PAH Associated with Congenital Heart Disease and/or Eisenmenger Syndrome

General measures
- Exercise education
- Prevention of infections and endocarditis prophylaxis
- Avoidance of smoking/recreational drug use
- Birth control
- Avoidance of anemia/iron deficiency
- Prevention of air embolism (air filters with IV lines)
- Avoidance of volume depletion
- Psychosocial support

Expert referral
Confirmation of PAH associated with CHD

Supportive treatment
- In the absence of hemoptysis, oral anticoagulant treatment may be considered when PA thrombosis is present
- Supplemental O2 therapy can be considered when it increases arterial oxygen saturation and reduces symptoms.
- Phlebotomy with isovolumic replacement should be considered only in the presence of symptomatic hyperviscosity.
- Heart failure therapy (diuretics, vasodilators, digoxin)

Acute vasoreactivity testing may be considered

No

Reparative surgery/intervention(s)

Heart/lung or lung transplantation with repair of CHD may be considered

WHO-FC I-II

Close follow-up in specialized clinic

WHO-FC III-IV

ERA or PDE-5 I or Prostanoids

Inadequate clinical response, no improvement or deterioration

Combination therapy may be considered in patients with Eisenmenger Syndrome

Prostanoids + ERA + PDE-5 I

Adapted from Kaemmerer. *Current Cardiology Reviews* 2010;6:343-355.
Long-term Effects of Advanced Therapy

Obstructive Lesions
Pulmonary Valve Stenosis

RA

RV

PA
Epidemiology of Pulmonic Stenosis

- Most common congenital valve lesion in adults (excluding BAV and MVP)
- PS considered “pediatric” by ACC Guidelines
  - Mean age 37 in largest valvuloplasty series
- Usually benign clinical course until symptoms develop
  - RVH and heart failure
  - Arrhythmias
- Valvuloplasty performed for
  - ≥ Moderate PS
  - Sx likely due to valve gradient
  - Provocable gradient during exercise

Intervention in Patients With Valvular Pulmonary Stenosis

**Balloon Valvotomy and Doppler Gradients**

Balloon valvotomy is recommended for asymptomatic patients with a domed pulmonary valve and a peak instantaneous Doppler gradient greater than 60 mm Hg or a mean Doppler gradient greater than 40 mm Hg (in association with less than moderate pulmonic valve regurgitation).

Balloon valvotomy is recommended for symptomatic patients with a domed pulmonary valve and a peak instantaneous Doppler gradient greater than 50 mm Hg or a mean Doppler gradient greater than 30 mm Hg (in association with less than moderate pulmonic valve regurgitation).

Coarctation of the Aorta
Coarctation of the Aorta

- Common lesion (8% of all CHD)
- Likely due to extraneous ductal tissue which contracts following birth
- 50-85% have associated BAV
- 10% with berry aneurysms
- Most common presentation in adult is during w/u for secondary HTN
- RAS activation – HTN seen after repair
CW Doppler of Aortic Coarctation
Pathognomonic CXR Finding

Rib notching
Coarctation Angioplasty/Stenting

• Native coarctation:
  – Reasonable success after 1 year of age
  – Long term concern re: aneurysm formation

• Post-op re-coarctation:
  – Good short term results
  – Persistent long term success
Interventional and Surgical Treatment of Coarctation of the Aorta in Adults

**Intervention and Peak-to-Peak Coarctation Gradient**

<table>
<thead>
<tr>
<th>I</th>
<th>IIa</th>
<th>IIb</th>
<th>III</th>
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<td>C</td>
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Intervention for coarctation is recommended in the following circumstances:

a) Peak-to-peak coarctation gradient greater than or equal to 20 mm Hg.

b) Peak-to-peak coarctation gradient less than 20 mm Hg in the presence of anatomic imaging evidence of significant coarctation with radiological evidence of significant collateral flow.

Choice of percutaneous catheter intervention versus surgical repair of native discrete coarctation should be determined by consultation with a team of ACHD cardiologists, interventionalists, and surgeons at an ACHD center.

Complex Lesions - Acyanotic
Transposition of the Great Arteries (D-TGA)

Atrio-ventricular Concordance

Ventriculo-arterial Discordance

Atrio-ventricular Concordance

RA, Ao, PA, LV, RV, LA

CCF ©2007
Demographics of D-TGA

- 5% of all congenital heart disease
- Male:Female = 4:1
- Cardiac associations
  - 40–50% VSD
  - 25% LVOT obstruction (subvalvePS/valvular PS)
  - 5% coarctation of the aorta
Demographics of D-TGA

• Natural (unoperated) history
  - TGA and intact interventricular septum: 10% survive >1y - need ASD or PDA
  - TGA with VSD and PS:
    - May survive if mixing results in a “balanced” cyanotic circulation - sufficient but not excessive pulmonary blood supply
Surgical Pioneers

Åke Senning
- Stockholm 1957
- Uses native tissue
- More demanding
- Better long term results
- Still used as part of complex repairs

Bill Mustard
- Toronto 1963
- Uses foreign materials
- Easier, so superseded Senning operation until poorer long term results apparent
Mustard/Senning Operations for D-TGA

- 1/3 to 1/2 of patients demonstrate ↓ systemic RV function 15-18 years after repair
- Systemic AV valve regurgitation also common
- Baffle issues not uncommon
- Sinus node dysfunction common, requiring pacing

Puley G et al. *Am J Cardiol* 1999; 83(7): 1080-4.;
Atrial Level Switch:
Definitive Palliation for TGA Until mid-1980s

Cumulative numbers of Mustard and Senning operations 1970 - 1998

Complications after Atrial Switches

- Impaired atrial function
- Arrhythmias
- Systemic AV valve regurgitation
- Systemic ventricular dysfunction
- Pathway obstruction
- Baffle leaks
- Pulmonary hypertension
Loss of Sinus Rhythm and SVT Common

Gelatt et al (J Am Coll Cardiol 1997;29:194–201)

Puley et al (Am J Cardiol 1999;83:1080–1084)

Cleveland Clinic
Congenitally-corrected Transposition of the Great Arteries (L-TGA)

Atrio-ventricular Discordance

Ventriculo-arterial Discordance

Atrio-ventricular Discordance
Transpositions at Risk for Heart Failure

SVC
IVC
LA
RA
RA
LV
PA
Ao
LV
RV
LV
RV
Systemic Ventricular Dysfunction

• Common and progressive
• Associations
  • Longstanding TR
  • Poor ventricular filling due to atrial surgery
  • Myocardial perfusion abnormalities
• Management
  • Standard heart failure therapies
    - HR control. ↑HR prevents ventricular filling through restrictive surgically-modified atria
    - ?ACEI benefit - may ↓ ventricular filling
• Conversion to arterial switch
• Consideration for transplantation
A Randomized Clinical Trial!!

- 88 repaired d-TGA or L-TGA patients with transposition and systemic ventricular failure identified through the CONCOR registry

- Valsartan 160 mg BID vs. matching placebo for 3 years

- Primary endpoint - $\Delta$ RV function by cardiac MR

A Randomized Clinical Trial!!

- Underpowered study but no significant Δ in RV function, exercise capacity, QOL or clinical events
- Lesser increase in RVEDV and RV mass seen with valsartan
- Valsartan well tolerated

Arterial Switch Operation for TGA
Admissions for Heart Failure in CHD

• ‘98–’05 ACHD HF hospitalizations ↑ by 82%
• 20% of ACHD hospitalizations in 2007 for HF
• Incidence for first admission ~10 fold higher than age-matched non-CHD population
• Following 1st admission
  – 24% mortality at 1 year
  – 35% mortality at 3 years

Type of Defect Determines HF Risk

Sudden Cardiac Death Important Contributor to Mortality (up to 50% in some series)
Complex Lesions - Cyanotic
Tetralogy of Fallot

- RA: Right Atrium
- RV: Right Ventricle
- LA: Left Atrium
- LV: Left Ventricle
- Ao: Aorta

Cleveland Clinic
Blalock-Taussig Shunt

Developed in 1945

Modified w/ Goretex

Delayed need for “complete repair”

“Something the Lord Made”
Classic Repair of Tetralogy of Fallot

AO, PA, LV, RV

TRANSANNULAR PATCH
Tetralogy of Fallot

• Surgically repaired adults usually do well for 2-3 decades, then have consequences due to PI
  – Right heart failure
  – Arrhythmias

• When to consider pulmonic valve replacement
  – Progressive decline in exercise tolerance
  – Progressive increase in indexed RVESV, RVEDV
  – Severe decrement in RV function
  – Severe widening in QRS (>180 msec)
Tetralogy Repair and It’s Residual
RV Outflow Tract Tachycardia After Tetralogy of Fallot Repair
Ebstein Anomaly

RA

ATRIALIZED RV
Ebstein Anomaly

- Characterized by apical displacement of the TV into the RV
- TV tissue is dysplastic with portions of the septal and inferior cusps adherant to RV away from the AV junction
- Large and redundant anterior leaflet of TV leads to “sail” sound
- 80% have ASD or PFO
  - Shunting results in cyanosis
- Surgical repair indications
  - Severe cyanosis
  - Severe TR and right heart enlargement
  - Development of right heart failure
Summary

• Adult CHD is more common than generally realized and a rapidly growing patient population

• Due to a shortage of CHD specialists, general cardiologists/primary MDs will continue as caretakers

• Thorough history and review of pediatric records is essential in initial evaluation

• Noninvasive imaging, particularly echo should be first-line in evaluation and is useful for serial follow-up

• Catheterization can help further clarify diagnosis and offers opportunity for novel therapeutic approaches, particularly if pulmonary arterial hypertension is present

• Outcomes data is growing and guidelines are stimulating research support