You’ve Come a Long Way, Baby:
The Adult With Congenital Heart Disease

Dallas, TX

December 12, 2008
3:30 PM – 4:45 PM
Session 7: You’ve Come a Long Way, Baby: The Adult With Congenital Heart Disease

Learning Objective

- Discuss the presentations and late complications of the most common untreated and palliated congenital heart conditions in today’s adult population.
- Explain the important absolute and relative contraindications to pregnancy in patients with palliated congenital heart conditions.

Faculty

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Dr Elyse Foster is professor of clinical medicine at the University of California, San Francisco and holds the Araxe Vilensky Endowed Chair in Cardiology. She is director of the Adult Echocardiography Laboratory and the Adult Congenital Heart Disease Service. She has published extensively in the field of echocardiography with specific interests in congenital heart disease, mitral regurgitation and cardiac resynchronization therapy. She serves on the editorial board of the Archives of Internal Medicine. Current research interests include the echocardiographic evaluation in congenital heart disease and percutaneous mitral valve repair, left ventricular remodeling following CRT therapy and animal models of stem cell repair of myocardial infarction.

Faculty Financial Disclosure Statements

The presenting faculty reported the following:

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Drug List

There is no drug list for this session.

Suggested Reading List


Craig B. Atrioventricular septal defect: from fetus to adult. *Heart*. 2006;92(12):1879-1885.


Growing Interest in Adult Congenital Heart Disease

- Most common birth defect
- 4 - 10/1000 live births
- Approximately 20,000 patients enter adulthood in U.S. each year
- Most patients treated for CHD as children are palliated, NOT cured

CHD = congenital heart disease

Congenital Heart Disease

You’ve come a long way, baby!

Growing Interest in Adult Congenital Heart Disease

- Most common birth defect
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CHD = congenital heart disease

Severe CHD

- Cyanotic or requiring intervention in childhood
- Requires ongoing care in specialized ACHD center due to residual disease
- Lesions
  - Atrioventricular septal defect (“Cushion defect”)
  - Tetralogy of Fallot
  - Truncus Arteriosus
  - Transposition complexes
  - Univentricular heart

Circulation 2007;115:163-172

Aims:
To determine the prevalence, age, proportions of adults relative to children with CHD in the population from 1985 - 2000 (in Canada)

Numbers and Proportion of Adults and Children with All CHD (A) and Severe CHD (B) in 1985, 1990, 1995, and 2000


Change in Prevalence of CHD from 1985 to 2000 Among Patients of Different Age Groups

CHD in the Adult

- Unrepaired
  - “Simple” lesions either mild or undiagnosed
    - eg. bicuspid aortic valve, small ASD or VSD
  - “Complex” inoperable lesions
    - Single ventricle
    - AVCD with Eisenmenger’s
    - Pulmonary atresia
- Surgically or non-surgically “cured”
  - eg. PDA, pulmonary stenosis, ASD or VSD
- Surgically or non-surgically palliated

CHD Classification

- Acyanotic
  - Obstructive lesions: Right vs. Left heart
  - Left to right shunt lesions
- Cyanotic
  - Tetralogy of Fallot
  - Transposition of the Great Vessels
  - Single ventricle (Tricuspid atresia, etc)
  - Truncus arteriosus
  - Eisenmenger’s syndrome
  - Ebstein’s with ASD

Incidence by Lesion

<table>
<thead>
<tr>
<th>Adult Born in 2010</th>
<th>Circulation. 2007;115:163-172</th>
</tr>
</thead>
<tbody>
<tr>
<td>Males</td>
<td>Females</td>
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<tr>
<td>Tetralogy of Fallot</td>
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<tr>
<td>Truncus Arteriosus</td>
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<tr>
<td>Atrioventricular Canal Defects</td>
<td>20</td>
</tr>
<tr>
<td>Transposition Complexes</td>
<td>15</td>
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</tbody>
</table>

3 Most Common Severe Lesions in Surviving Adults

- Tetralogy of Fallot + Truncus Arteriosus
- Atrioventricular Canal Defects
- Transposition Complexes

Other Lesions in Surviving Adults

- Most common: ASD, VSD, PDA
- Congenital AV disease
- Pulmonary valve disease
- Coarctation of the Aorta
- Rare: Ebstein’s, cor triatriatum, etc

Coarctation of the Aorta

- 6-8% of CHD
- M:F = 3:1
  - XO Turner’s syndrome
  - 50 - 70% have BAV
  - Aneurysms Circle of Willis
  - Asyx with UE HTN
  - Syx: Headache, CP, claudication
**Coarctation: Diagnosis**

- Physical Examination
  - SBP: UE > LE (Measure all 4 extremities!)
  - Radial to femoral pulse delay
  - ↑Femoral pulses
  - Murmur at ULSB radiates to back
- EKG: LVH
- CXR: “3 sign”, rib notching
- Echo - Doppler
- MRI/MRA

**CXR**

**Echo: Suprasternal Notch Views**

- Coarctation site
- Location
- Length
- Presence of collaterals
- Suitability for percutaneous intervention

**MRA of the Aorta**

**Approach: Unoperated Coarctation Patient**

- Blood pressures in both arms and leg
- CXR, EKG
- Echo
  - Evaluate and confirm gradient
  - Exclude other lesions esp bicuspid aortic valve, MV disease
- MRA
  - Anatomy of coarctation segment
  - Collateral flow to descending aorta
  - Catheterization ± balloon/stent

**Adult Native Coarctation: Stent Placement**
Surgical Approaches

- End-to-end anastomosis
- Other techniques:
  - Interposition grafts
  - Subclavian flap aortoplasty
  - Patch aortoplasty
  - Percutaneous balloon angioplasty - 1982
  - Expandable stents in the 1990's

Post - Coarctation Repair

- Bicuspid aortic valve > 50%
- Cerebral Aneurysms usually small
- Hypertension
  - 25 - 75% at surgery
- Aortopathy
  - Rupture or dissection
- Recurrent coarctation
- Premature CAD
- Aneurysms
- Endocarditis

Coarctation Repair: Sequelae

- Recurrent coarctation
  - 3 - 10%
  - Higher risk if repaired in infancy
- Aneurysms
  - 5 - 20%
  - Patch aortoplasty or balloon angioplasty
- Premature CAD:
  - 5 - 23%
  - Duration of pre-existing HTN
- Endocarditis of AV or coarct site
  - 3 - 5% at 25 yrs

Causes of Death

- Perioperative deaths: 20/274 (7%)
- Late deaths: 45/252 (17%)
- CAD in 10 pts (mean age = 53)
- Perioperative at 2nd surgery in 7 pts
- Sudden Death in 7 pts
- Ruptured Aneurysm in 6 pts
  - 5 dissection
  - 1 at coarctation site
- Risk for death:
  - Age > 10 at surgery
  - Post-op hypertension
Approach to the Operated Coarctation Patient

- Operating note/other records
- Blood pressures both arms and leg
- Left arm pressure may be lower due to subclavian flow not recoarctation
- Echo:
  - LVH, LV function
  - Aortic valve anatomy and function
  - Gradient in descending aorta (often overestimated by Doppler)
- MRA or CT angiography: recoarctation, aneurysm
- Indications for intervention in recoarctation:
  - UE hypertension with gradient > 20 mmHg

Atrioventricular Canal Defect
(AKA: AV Septal Defect, Endocardial Cushion Defect)

- Large Primum ASD
- VSD
- Common AV valve

Partial AV Canal

- Level of shunting depends on attachment of bridging leaflets
- Ostium primum defect
- VSD of the AV canal type
- Cleft mitral valve

41-Year-Old Woman with Down's

- Symptoms: SOB, fatigue
- PE:
  - Sustained RV impulse
  - Cyanosis and clubbing
  - Widely split S2 with P2 (fixed if ASD)
  - TR, PI murmurs
  - Murmur across VSD, or PDA no longer audible
- Labs:
  - Increase in Hgb, Hct
  - Low platelets
  - High uric acid

EKG

- RV enlargement
- Prominent main PA’s
- Peripheral pulmonary artery pruning

CXR
AV Septal Defect: Unoperated

- Cyanosis
  - Eisenmenger’s inoperable
  - RVOT obstruction operable
- Heart failure
  - Large left to right shunt
  - L AV valve regurgitation
- Arrhythmias
  - Atrial arrhythmias
  - Complete heart block
- Angina due to LV outflow tract obstruction

Cyanotic Heart Disease: Right to Left Shunt

- Eisenmenger’s syndrome:
  - Previous left to right shunt with PHTN and shunt reversal
  - Closure of defect is contraindicated
- PFO with pulmonary HTN
- Intracardiac communication with distal obstruction
  - ASD with tricuspid atresia
  - VSD with RVOT obstruction (TOF)

Cyanotic Heart Disease: A Multisystem Disease

- Secondary erythrocytosis
  - Phlebotomy only for hyperviscosity syx
    - HA, lethargy, CHF
    - Hct > 55
  - Exclude iron deficiency which can mimic hyperviscosity
  - Associated bleeding diathesis
- Endocarditis prophylaxis!
- Increased incidence of:
  - Gout
  - Cholelithiasis
  - Renal insufficiency

Eisenmenger’s Syndrome

- Pulmonary vascular disease
- Shunt reversal
- ASD, PDA, VSD, AV Septal defect
- RVH, Small LV, TR, PI
**Pulmonary Vasodilators in Eisenmenger Syndrome**

- Role still uncertain with increasingly positive data
- Options:
  - Prostacyclin
    - Requires indwelling catheter
  - Bosentan - liver toxicity
  - Sildenafil - off label use
  - Iloprost - inhaled prostaglandin recently approved
- Advisability of defect closure uncertain even if PVR falls

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**Breathe - 5**

Galie, N. et al. Circulation 2006;114:48-54

- Randomized, double-blind placebo controlled trial
- 54 Eisenmenger pts in 2:1 ratio to bosentan (37) vs placebo (17)
- 16 weeks
- ↓ PVR, ↓ mean PAP (-5.5 mmHg), Increased exercise capacity

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**AV Septal Defect: Operated**

- Usually asymptomatic
- Left AVV regurgitation most common sequelae
- Residual atrial septal defect
- LV to RA shunts
- Require careful follow-up

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**Tetralogy of Fallot**

- Symptoms
  - Childhood: cyanosis, “tet spells”, squatting
  - Adulthood: rare uncorrected TOF
- Physical exam:
  - Cyanosis and clubbing
  - Prominent RV
  - Soft or absent P2
  - Harsh RV outflow murmur
- EKG: RVH, RAE
- CXR: Prominent RV, boot-shaped cor, small PA

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**Echo:**

26 yo Patient with Down's and Cyanosis

Override Ao
Approach: Unoperated Adult with TOF

- Rarely encountered in the US
- Complications of cyanotic heart disease
  - 2° erythrocytosis
  - Clubbing
  - Gout
  - Cholelithiasis
- Usually remains operative candidate
  - Definition of pulmonary artery and coronary anatomy crucial

Palliative Procedures for TOF

- Waterston shunt: Asc Ao to RPA
- Pott’s shunt: Desc Ao to LPA
- Blalock-Taussig: Subclavian artery to ipsilateral PA
- Modified BT shunt: Gortex graft from Subclavian to PA
- Central shunt: Gortex graft from Asc Ao to main PA

Tetralogy of Fallot: Total Repair

Residua:
- Persistent outflow obstruction
- RVH
- Peripheral PA stenoses

Sequelea:
- Pulmonary valve insufficiency**
- Outflow tract aneurysms
- VSD patch leak
- Arrhythmias
- Aortic root dilatation with AI

34 yo Woman Post Tet Repair

Dilated RV due to volume overload

Dilatd RV due to volume overload

TR jet with low velocity c/w no residual RVOT obstruction

Diagnostic Procedures

- Holter: NSVT, atrial arrhythmias
- Echo:
  - LV/RV size and function
  - Pulmonary valve obstruction and insufficiency
  - TR
  - Residual VSD, interatrial shunts
  - Aortic root size and AI
- MRA/Helical CT: branch PS
- Exercise testing

Management of Post-operative TOF

- Timing of surgical intervention uncertain
- Symptoms
- QRS duration > 180 msec
- Ventricular tachycardia
- ICD indicated for sustained VT and/or resuscitated sudden death
- Percutaneous pulmonary valve replacement a reality

DiGeorge Syndrome: 22q11.2 Deletion

Transposition of the Great Vessels

- Atrio-ventricular concordance
- Ventriculo-arterial discordance
- Malposition of the great vessels

Post-operative TGV

- Atrial switch
- Senning
- Mustard
- Arterial switch

Atrial Switch = “Physiologic Correction”

- Interatrial baffle
- Diverts systemic venous flow to MV into sub-pulmonic LV
- Diverts PV flow to TV into systemic, sub-aortic RV

Atrial Switch Surgery for TGV

- Residual lesions:
  - Systemically functioning RV
  - Pulmonic stenosis
  - Ventricular septal defect
- Potential Sequelae
  - Systemic RV failure
  - Tricuspid regurgitation
  - Venous obstruction
  - Baffle leaks
  - Endocarditis
  - Atrial arrhythmias
  - Sinus node dysfunction
  - Sudden death

TGV: Post-arterial Switch

- Pulmonary stenosis
- Anastomosis site
- Branch
- Neo-aortic root dilatation
- Neo-AV regurgitation
- LV dysfunction
- Myocardial ischemia
- Arrhythmias

Summary

- Increasing numbers of adults with CHD
- With some exceptions, most have significant issues require close follow-up
  - Residual hemodynamic derangements
  - Arrhythmias
  - Ventricular dysfunction
  - Pulmonary vascular disease
- Additional issues:
  - Pregnancy
  - Ability to work, insurance, psychological sequelae