Conduits, Shunts and Patches, Oh My!

The Adult with Congenital Heart Disease

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No disclosures
Congenital Heart Disease

- Recognized for centuries
- Most common of all major birth defects
- 5-9/1,000 live births
- Wide variety of lesions
- Natural and ‘unnatural’ histories

- Every 15 minutes a child is born with CHD… most will live to adulthood
ACHD

- >39,000 infants born annually with CHD
- 85-94% survive to adulthood
- 20-25,000 operations for CHD annually
- >1,000,000 adults with ACHD in US (Marelli, 2006)
- More adult than pediatric CHD patients in US
- 36,000 patients in 60 centers in 2005
- >50,000 patients in 82 centers in 2007
- Little formal training available for ACHD physicians
- ABIM approved ACHD subspecialty boards 2010
Birth Prevalence of ACHD Worldwide
Incidence/ Million Live Births

- Acyanotic CHD: 8205, 34%
- Cyanotic CHD: 1391, 6%
- BAV: 13556, 56%
- Syndromes: 900, 4%
### Relative Prevalence of ACHD

<table>
<thead>
<tr>
<th>Condition</th>
<th>Prevalence</th>
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<tbody>
<tr>
<td>Long QT syndrome</td>
<td>0.142/1,000</td>
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<tr>
<td>Hypertrophic cardiomyopathy</td>
<td>2/1000</td>
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<tr>
<td>HIV (living)</td>
<td>3/1,000</td>
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<tr>
<td>ACHD</td>
<td>4/1,000</td>
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<tr>
<td>Angina and/or MI</td>
<td>35-105/1,000</td>
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<tr>
<td>All invasive cancers</td>
<td>47/1,000</td>
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<tr>
<td>Hypertension</td>
<td>312/1,000</td>
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</tbody>
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Incidence CHD
Change in Prevalence

<table>
<thead>
<tr>
<th>Age in years</th>
<th>Children, year 1985</th>
<th>Children, year 2000</th>
<th>Adults, year 1985</th>
<th>Adults, year 2000</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age 1-12</td>
<td>1.00 (0.93-1.07)</td>
<td>1.31</td>
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<tr>
<td>Age 13-17</td>
<td>2.78 (2.44-3.17)</td>
<td>0.65</td>
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<tr>
<td>Age 18-25</td>
<td>2.35 (2.10-2.64)</td>
<td>0.45</td>
<td>1.07</td>
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<tr>
<td>Age 26-40</td>
<td>2.48 (2.19-2.80)</td>
<td>0.21</td>
<td>0.52</td>
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<tr>
<td>Age 41+</td>
<td>1.52 (1.30-1.79)</td>
<td>0.10</td>
<td>0.15</td>
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</table>

Prevalence Ratio 2000 vs. 1985 (95% CI)
Growth in ACHD

- Improved case finding – echocardiography
- Improved medical care - prostacyclin
- Improved diagnostic technology
- Surgery
  - Innovative surgical procedures
  - Reduced operative risk in infancy and childhood
  - Reducing long-term morbidity
- Interventional techniques
ACHD in the US

- Currently over 1,000,000 adults in US with CHD
- 88 self-identified ACHD centers
- Avg. 2 physicians per center… less than 20% of current need being met
Surgical Landmarks

- 1938 – Ligation of a patent ductus arteriosus (Gross & Hubbard)
- 1944 – First Blalock-Taussig shunt
- 1946 – Closed pulmonary valvotomy (Sellors, Brock)
- 1952 – PA band (Muller, UCLA)
- 1952 – Successful ASD closure (Lewis, U Minn)
- 1953 – Gibbon closes ASD successfully using heart-lung machine
Surgical Landmarks

- 1954 – Repair of Tetralogy of Fallot (Lillihei, U Minn)
- 1957 – Senning atrial switch for D-TGA (Stockholm)
- 1966 – Rashkind balloon septostomy (CHOP)
- 1968 – Fontan procedure (Bordeaux)
- 1981 – First stage Norwood procedure for HLHS (Boston)
- 1987 – Jatene arterial switch for D-TGA (Brazil)
TOF: Impact of Repair
ACHD Programs
Care Recommendations

✔ Every ACHD patient should be seen by an ACHD specialist at least once
✔ Every ACHD patient should have a ‘medical home’
✔ Every cardiologist/medical facility should have a referral relationship with an ACHD specialist/center
Clinical Challenges

- Relatively small numbers of patients/lesion
- High percentage of patients ‘lost’ to follow up
- Large burden of anatomically diverse and complex clinical care
  - History, lesion- and operation-specific
- High demands of imaging - anatomic, physiologic
- Imposition of acquired on congenital heart disease
- Consequences of aging
Societal Challenges

- Working relationship between Medicine and Pediatrics
- Transition of care
- Models of care
- Societal recognition (coding, insurance, disability)
- Reluctance to refer
- Insurance access and adequacy
- Neurocognitive issues
Lesion Categories

- Simple – 4 chambered heart, normal anatomic relationships, acyanotic, hemodynamically simple or trivial

- Moderately complex – 4 chambered heart, (near) normal anatomic relationships, mostly acyanotic; hemodynamically significant shunts, vascular or valve lesions

- Severely complex – <4 chambered heart, atretic valves, abnormal anatomic relationships, great vessel anomalies, often cyanotic; hemodynamically significant shunts, vascular or valve lesions; pulmonary arterial hypertension
Relative Prevalence of Acyanotic Lesions

After Hoffman, J Am Coll Cardiol, 2002
Cyanotic Lesions

- Single ventricle, tricuspid atresia
- RVOT obstruction/VSD - TOF, PS/VSD
- Pulmonary atresia
- TR or PS plus ASD, i.e. Ebstein’s
- Truncus arteriosus
- Eisenmenger’s reaction
Relative Prevalence of Cyanotic Lesions

After Hoffman, J Am Coll Cardiol, 2002
Aerobic Capacity in ACHD

Normal: 170 cm male, Stable weight 70 kg

Adapted from Weisman, 2001 and Diller, 2005
Residuae and Sequelae of CHD Surgery

- Electrophysiologic
  - Myocardial scars
  - Disorders of impulse formation and conduction

- Ventricular
  - Function!

- Valvular
  - Repairs and replacements
  - Prosthetic material

- Vascular

- Noncardiovascular

CHD Surgery is corrective, not curative!

Perloff, Circ, 1991
Causes of Death in ACHD

- **Non-cardiac**: 33 (17%)
- **Other cardiovascular**: 36 (18%)
- **CHF**: 41 (21%)
- **SD**: 51 (26%)
- **Perioperative**: 36 (18%)
Special Needs of ACHD Pts

- Limited exercise capacity
- Pregnancy, genetics, heredity
- Arrhythmia management
- Reoperation, intervention
- Hepatic, renal and pulmonary complications
- Employment, disability and insurance
- Neurocognitive issues
- Lack of patient knowledge concerning their illness
ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease

A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease)

Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons

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Summary

ACHD is increasing in prevalence

ACHD characterized by anatomic and clinical complexity

ACHD care and resources are available
Atrial Septal Defect
ASD Physiology

- Left to right shunt at the atrial level
- Volume load of right heart
- Often leads to tricuspid regurgitation and atrial dysrhythmias
- Can lead to right heart failure
- Generally recommend closure for shunt greater than 1.5:1
Amplatzer Device Closure
Adult Complications ASD

- Right heart volume overload
- Right heart failure
- Atrial dysrhythmias!
- Pulmonary HTN
- Paradoxical embolism and stroke

- Reasonable to close defect, even late in life
VSD
VSD Physiology

- Shunting is generally from LV to RV through VSD during systole, results in left heart dilation.
- Early in life, large VSDs will result in pulm. overcirculation leading to CHF, FTT, frequent pulm. infections and tachypnea.
- Over time, large VSDs will chronically flood the lungs, resulting in vascular changes and pulm. HTN
Adult Complications VSD

- If late repair, think pulmonary hypertension
- Residual VSD (5%)
- Aortic insufficiency
- Residual VSD/ patch leaks
- Complete heart block
- Tricuspid valve damage from repair
Tetralogy of Fallot
Physiology of TOF

- Pulmonary stenosis limits blood flow to the lungs
- Right ventricle hypertrophies
- Right to left shunt across VSD, resulting in desaturated blood being pumped to body
BT Shunt

Blalock-Taussig Shunt

Left subclavian artery

Left pulmonary artery (to the lungs)

The left subclavian artery is divided and connected to the left pulmonary artery. This allows blood to flow to the lungs to pick up oxygen.

Modified Blalock-Taussig

"BT Shunt"

Aortic Branch

Pulmonary artery (to lungs)

This procedure diverts blood from an aortic branch to the pulmonary artery, allowing blood to flow to the lungs to receive oxygen.

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Blalock Taussig Shunt
Transannular Patch Repair

- Patch closure of VSD
- Incision through pulmonary valve annulus and infundibulum with placement of large patch
Adult Complications TOF

- Pulmonary insufficiency
- RV dilation/ decreased function
- Ventricular dysrhythmias
- Atrial dysrhythmias
- Residual RVOT obstruction
- Aortic root dilation and insufficiency
- Sudden cardiac death
Physiology of D-TGA

- Systemic and pulmonary circuits run in parallel, not in series
- Must have mixing somewhere: ASD, VSD, PDA
Senning Repair
Arterial Switch
Adult Complications D-TGA

- Baffle leaks or obstruction
- Atrial dysrhythmias!!
- Right ventricular dysfunction
- Tricuspid regurgitation
- Pulmonary hypertension
- Sinus node dysfunction!
- Sudden cardiac death (15%)
Single Ventricle Lesions

- How to take a heart with only one pumping chamber and make it do the job of two ventricles
- Lots of types of “single ventricles”
Physiology

- How do we make one ventricle do the job of two?

- FONTAN PROCEDURE
General Anatomy

- Variations of single left ventricle
- Variations of single right ventricle
- Two ventricle hearts with potential single ventricle physiology
Variations of Single Left Ventricle

- Tricuspid valve atresia
- Double inlet left ventricle
- Unbalanced AV canals with hypoplastic right ventricle
- Pulmonary atresia with intact ventricular septum
Variations of Single Right Ventricle

- Mitral valve atresia
- Aortic valve atresia
- Unbalanced AV canal with hypoplastic left ventricle
Two Ventricle Hearts with Potential Single Ventricle Physiology

- Tetralogy of Fallot with pulmonary atresia
- Truncus arteriosus
- DORV with remote VSD
- AV canal with straddling AV valves
- Primitive common ventricle
Why Do We Fix This?

Goals of surgical procedures:

- Unobstructed systemic blood flow
- Limited PBF
  » minimize ventricular volume load and risk of pulmonary artery hypertension
- Unobstructed pulmonary venous return
  » minimize left atrial and secondary pulmonary artery hypertension
- Minimize likelihood of pulmonary artery distortion
- Avoid dysrhythmias
How Do We Fix This?

- **Systemic to PA Shunt**
  - Used in children with a single ventricle and restricted PBF (most kids)
  - Modified Blalock-Taussig shunt is the most common procedure
    - 3.5-4 mm Gore-Tex tube is placed from the brachiocephalic vessel to pulmonary artery

- **Atrial Septectomy**
  - Allows complete mixing of blood
  - Create pathway for blood to get from single ventricle to body ("Neo-aorta")
Norwood Procedure
Norwood Procedure

Fig. 30-8 The final appearance of the Norwood reconstruction is shown. AP, Aortopulmonary; RV, right ventricle.
Physiology

- Complete mixing of the systemic and pulmonary circulation at the atrial or ventricular level
- Pulmonary and aortic oxygen saturations are equal
Physiology

- Provide enough PBF for oxygen delivery
  - prevent acidosis
  - prevent excessive volume load to the single ventricle
- An arterial oxygen saturation of 75–80% represents a Qp/Qs ratio of approximately 1:0
Physiology

- Pulmonary flow is determined by:
  - Degree of pulmonary obstruction
  - Pulmonary arteriolar resistance (PVR)
  - Pulmonary venous and left atrial pressure

- Systemic flow is determined by
  - Presence of anatomic obstructive lesions (Valvar obstruction, arch hypoplasia, coarctation)
  - Systemic arteriolar resistance (blood pressure)
Separating the Circulation

- Goal: Separation of the systemic & pulmonary circuits will result in a near normal oxygen saturation

- Procedure: divert systemic venous return directly into the pulmonary vascular bed
  - (reduces the volume load on the single ventricle)
  - Done in 2 stages

- Postoperative: PBF derived by passive flow
Separating the Circulation

- Cavopulmonary connections
  - Bidirectional Glenn
  - Hemi-Fontan
  - Modified Fontan operation
Bidirectional Glenn/Hemi-Fontan

- Performed between 4-9 months of age
- Infants < 3 months of age have a higher incidence of cyanosis, pulmonary artery thrombosis, and vascular congestion

Procedure:
- SVC is divided
  - Cardiac end is over sewn
  - Cephalic end is anastomosed to the side of the ipsilateral pulmonary artery
Bidirectional Glenn Physiology & Advantages

- PBF is obligate
- All SVC return must pass through the lungs to reach the heart
- All IVC return passes to the systemic ventricle
- Advantages:
  - Reduces the volume work of the single ventricle
  - Removal of the aortopulmonary shunt
    » Improves myocardial perfusion due to increased coronary and diastolic pressure
  - Provides an opportunity to address distortion of the pulmonary arteries from previous bands or shunts
Hemi-Fontan
Fontan Operation

- <10 % operative mortality
- Long term outcomes are questionable
- Modifications of Fontan to improve late outcomes include:
  - Intra-atrial lateral tunnel
    » May decrease late tachyarrhythmias
  - Extra cardiac conduit
    » Eliminates atrial suture lines which may decrease atrial arrhythmias
  - Fixed fenestration in the lateral tunnel baffle
    » Decreases early effusions
    » Maintains ventricular preload in times of hemodynamic stress
Fontan Completion
Adult Complications

- Heart Failure
- Poor ventricular function
- Valvar regurgitation
- Protein losing enteropathy
- Endocarditis
- Thrombosis
- Atrial dysrhythmias
- Sudden death
- Hepatic dysfunction
Heart Failure

- The rule, not the exception
- Most patients NYHA class I or II, however- VO2 max averages less than 14 ml/kg/min (norm 42 ml/kg/min)
- Peak heart rate often limited
- Long term myocardial stress
- Elevated PA pressures
- Ventricular morphology plays a role
Atrial Dysrhythmias

- 50% with atrial tach
- Scar tissue
- Stretched out atrium
- Can be refractory to management
- Ablation, Cox-Maze, Fontan revision
- Some reasonably well tolerated (PAF)
Hepatic Fibrosis

- Commonly seen if looked for...
- Cirrhosis, thrombocytopenia, hyperbili
- Associated with ventricular dysfunction and absence of fenestration
- Increase with Fontan duration
- Frank hepatitis also common, but decreasing
- ALT, AST, GGT, liver US
Protein Losing Enteropathy

- Cause is still somewhat perplexing
- Loss of protein and immunoglobulins through gut
- May be related to increased Fontan pressures, but not always
- No good treatment
- Usually resolves after transplant
- Significant quality of life issue
Fontan Take Home Messages

- Most complicated group of ACHD patients
- Diuretics are almost always useful
- Put out “fires”, then call for help

- AVOID PREGNANCY
- Progesterone only OCP due to increased thrombosis risk
Pregnancy?!

 Loving You’ve got to be kidding me!

 Loving Many ACHD patients can carry a pregnancy successfully

 In general:
  – Good heart function
  – Minimal dysrhythmias
  – Normal saturations
Case- KH
Case- KH

- 25 year old woman with TOF, desires pregnancy
- Otherwise healthy
- Meds: Baby aspirin daily
- PE: vitals normal, 2/6 SEM, 2/4 decrescendo diastolic M, both at LUSB, otherwise normal
- CXR: sternal wires, otherwise unremarkable
Case- KH

- EKG: NSR, RBBB, QRS 180 ms
- Echo: Severe pulmonary insufficiency, RV severely dilated, mildly depressed RV function, mild TR, aortic root mildly dilated, normal left heart function

Stress: 12 minutes of Bruce protocol, Peak VO$_2$ 35 ml/kg/min, no desaturation, occ. Single PVCs
Case- KH

🔹 What To Do????
Take Home Message

- If you can’t draw it, you probably shouldn’t take care of it…
- Know if the patient should be pink or blue
- Get old records
- Ask for help!
New England Resources


♥ Boston Adult Congenital Heart (BACH) and Pulmonary Hypertension Program
(617) 355-6508

[ACCHA](http://www.achaheart.org)
Questions??
Thanks!
WE ARE!