Learning Objectives

At the end of this presentation the participant will be able to:
1. Discuss adult congenital heart disease anatomy
2. Define common forms of adult congenital heart disease
3. Characterize sequelae of “successful” congenital heart disease treatment

“A longer view of congenital heart disease”

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Adult Heart Disease - simplicity

• Muscle
• Electrical
• Valve
• Blood Supply
• No shunts, no anatomic anomalies, different complexity

Congenital Heart Disease

• History
• Prevalence
• Types
• Terminology/classification
• Single Ventricle pathway
• Common adult scenarios
• Final thoughts

Historical

• Etienne-Louis Arthur Fallot
  “cyanosis, especially in the adult is the result of a small number of cardiac malformations well determined. One of these is much more frequent than others…”
• Maude Abbott
  autopsy series replete with ASD, “but where are the Maladie de Roger”
Demographics

- Over 1 million in US > 20
- Adult Congenital Heart Disease growing
- 200/100,000 persons
- 0.8% population has CHD
- Clear that we can expect that number to grow to 800/100,000 or close
- SA – serve roughly 10,000 folks

<table>
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<th>Year of Birth</th>
<th>Birth Rate/Year</th>
<th>Persistence</th>
<th>Survival Rate (First Year)</th>
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<td>Totals</td>
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32nd Bethesda Conference: Care of the Adult With Congenital Heart Disease

Med School – Basics Review

- Blue kids = cyanotic
- TOF
- TGA
- TAPVR
- Truncus
- Tricuspid Atresia
- Pink kids = failure
- VSD
- ASD
- PDA
- CAVC
- PS, Coarctation

Acyanotic Disease: VSD, ASD and PDA

- Good repair available for long time now
- Residua uncommon (except CAVC)
- Occasional associated defects
- 40% CHD
- Residual defects addressed directly

Cyanotic Disease (5 T’s)

- Tetralogy – many to adulthood
- TGA – many to adulthood now
- TAPVR – most to adulthood without sequelae
- Truncus – frequent reoperations
- Tricuspid Atresia/ Single Ventricle complexes – rapidly growing set of survivors
**Learning the language**

“Speak Congenital”
(And think it)

**Comprehensive consult components**
- Symptoms
- AVV anatomy/function
- Semilunar valve
- Muscle
- Coronaries
- Prior operation reports
- Prior cath reports
- Associated lesions/issues (genetics, etc.)

**Anatomic terminology (segments)**
- Atria – solitus, inversus, ambiguous
- Ventricles – D-loop, L-loop, indeterminate
- Great vessels – Transposed (D or L), normal
- Connections – AV Valves, Semilunar valves
- Septal defects – atrial or ventricle
- Venous connections – systemic, pulmonary
- Dextrocardia, levocardia, mesocardia

**Chambers and Vessels**

**Other terms**
- Concordant
- Discordant
- Straddling
- Overriding
- Hypoplastic
- Atretic
- Conal
- Infundibular
- Apex forming
Semilunar valve description

ACHD Composition

- PDA, ASD, Coarctation repaired early – 25% of all CHD
- VSD repaired early – 20% all CHD
- Less residua of these defects repaired well
- Other defects – near half will have problems for life

21st Century Patient Population

- Residua of previous surgical efforts
  - Medical concerns
  - Electrophysiology
  - Congestive failure, cyanosis, folan
  - Surgical concerns
  - Conduit changes, valvular defects, persistent septal defects, conduction issues

Tetralogy of Fallot

- 7% of all CHD
- Occasional survival to adulthood unrepaired
- Repair – highly successful, but usually inadequate pulmonary valve tissue
- Palliation was common, more rare today
- Residua – AI, PS/PI, VSD, pulmonary artery stenoses, ventricular irritability, PFO

Transposition of Great Vessels

- Near 10% of CHD
- Two repairs
- Never survive without surgical intervention (VSD, maybe)
  - Residua depend on repair done

Senning/Mustard

- RV failure
- Tricuspid insufficiency
- Venous pathway obstruction
- Atrial dysrhythmias
- Residual VSD
Arterial Switch

- Pulmonary Stenosis
- Coronary insufficiency
- Residual VSD
- AI
- PI
- Data emerging

PAPVR and TAPVR

- TAPVR - ~1% of CHD
- PAPVR - ~10% of all ASD’s (10% of CHD)
- TAPVR rare to adulthood
- PAPVR commonly presents at adult ages
- One is always blue, the other slowly overcirculates
- Right > left, Scimitar – 20%

Embryology

- 3.5 weeks – lung buds develop
- Primitive venous plexus surrounds these
- No definitive connections, but shares routes with splanchnic, umbilicovitelline and cardinal systems
- Primitive common pulmonary vein sprouts from posterior left atrium near septum and coronary sinus – usually other then atrophies
L to R Shunt calculations

**Systemic sat. – mixed venous sat.**
**Pulmonary vein sat. – PA sat.**

Stewart, Backer, Mavroudis 2007

- 54 Sinus Venosus, 52 associated PAPVR
- Median 10 +/- 12 1990 – 2006 mean age 4.6
- 8 – RA, 17 RA/SVC, 27 to SVC
- Single patch - 22 , two patch - 21 , Warden – 5
  12/22 (55%) 5/21 (24%) Ofer (%)
- Conclude single ok for RA or RA/SVC jxn, but
  Warden preferred for those to SVC

References

- Warden HE, Gustafson RA, Tarnay TJ, et al. (1984) An alter-
  native method for repair of partial anomalous pulmonary
  venous connection to the superior vena cava. Ann Thorac Surg
  38, 601–605.
- Evolving surgical strategy for sinus venosus atrial septal
  defect: effect on sinus node function and late venous
  discussion 1655. Stewart, Backer, Mavroudis, et al

Tricuspid Atresia/ Single Ventricle complexes

- 5-10% all CHD
- Residua abundant
- Dysarrhythmias
- CHF (AVVR, poor systemic ventricular fxn)
- Obstructed conduits/ pulmonary stenoses
- Pulmonary arteriovenous malformations
- Protein losing enteropathy

Fontan Concept Evolution

- Principle outlined by Glenn in 50’ s
- Sade – early 70’ s experimental work on
  “disposable right ventricle”
- Fontan – 1968 did first case in highly selected
  4 yo, published series in 1971
Fontan

- Best mortality in some 200 pts from Mayer et al, BCH 17% - 1987
- Highly selected only tricuspid atresia patients did this well with most if not all of the 10 commandments met
- Significantly worse with HLHS, complex heterotaxy, SV, etc.

Late 80’s to present – focus on a pathway

- Early on- smaller shunts to limit systemic ventricle volume load and thus compliance changes
- Bidirectional Glenn became a second step – Hopkins 1985 DC Childrens
- Bridges in BCH described fenestration of the conduit to allow for “pop off” and less volume load to pulm. circuit

Fontan pathways – many forks in the road...

- Blaylock vs. Sano
- Hemifontan vs. Bidirectional Glenn
- Classic
- Lateral tunnel
- Extracardiac
- Onpump vs. off pump
- Cath lab or interventional
Sano Modification

Stage I

- Sano – less mortality initially, early stage 2 for proximal obstruction – is PA growth as good?? – late arrhythmias? RV function??
- Norwood – harder to get through, better PA growth?, no RV incision, shunt thrombosis more common?
Fontan

- Lateral tunnel – extracardiac
- Lateral tunnel – intracardiac
- Fenestrated
- On or off pump
- Depends on substrate, ie AVVR, function, PVR, laminar flow pattern

Stage II

- Bidirectional Glenn – lowers Fontan mortality – no question
- Hemifontan – better theoretical blood flow, less sinus rhythm??
- Bidirectional Glenn – excellent blood flow, less SA node dysfunction??
- What is best interstage palliation - ?
Dysrythmias

- BCH JCTVS 2001
- 220 pts lateral tunnel Fontans
- 10 years or more follow up
- Fenestrated and nonfenestrated
- 20% - bradycardic
- 9% - SVT
- Late failure risk – previous coarctation

Single ventricular diastolic properties

- 55 pts pre and post Fontan
- Measured diastolic pulmonary vein blood flow relative to normal healthy controls
- No changes after operation, but clearly abnormal ventricular relaxation present in the single V pts regards controls
**Functional outcomes**

- Worse
- JCTVS BCH Wernovsky, et al 1995
- 1st 500 Fontans – all types
- 363 long term survivors 91% - I or II
- Identified longer duration of follow up, prior atrial septectomy, and prior Ao-PA anastomosis as risk factors III or IV

**Functional outcomes**

- JACC Toronto 1995
- Of 47 adult fontan pts – 30 underwent exercise testing
- 93% I or II clinically but...
- Work load was half anaerobic threshold half, and Max O2 1/3 of controls
- EF 38 vs 58 rest & 40 vs 70 with exer.

**Heparin and PLE**

- Early cases from U. of M.
- Noted two patients with improvement of PLE sxs when on heparin for procedures
- No benefit from LMW heparin
- Does have benefit – unclear for how long, no cases of HIT so far

**Plastic Bronchitis**

- Images of bronchitis showing inflammation and narrowing of bronchial tubes.
21st Century Fontan Population

- Residua of previous surgical efforts
- Potential foci of dysrhythmia
- Diastolic dysfunction
- Brady-tachyarrhythmias
- PLE
- Valvular failures
- Poor fluid hemodynamics

Best Fontan

- Careful pathway
- Limit diastolic injury
- Avoid pulmonary vein HTN
- Preserve AVV function
- Best possible conduction preserved
- Optimal laminar flow pattern
- When it fails - transplant

Summary: successful consult

- Symptoms
- AVV anatomy/function
- Semilunar valve
- Muscle
- Coronaries
- Prior operation reports
- Prior cath reports
- Associated lesions/issues (genetics, etc.)
- Connections
- Conduction issues
- Venous and arterial acquired lesions
- Exercise tolerance
- Medications
- Family pathology
- Eligibility/coverage
- Team approach