Pediatric Echocardiography

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What is your career?

A. Adult Echocardiographic Sonographer
B. Pediatric Echocardiography Sonographer
C. Adult and Pediatric
D. Radiology
E. Other
Objectives

- Implement current best practice standards in pediatric echocardiography
- Describe the basic pediatric echocardiogram. (views, imaging techniques, etc.)
- Improve the ability of the sonographer to understand and perform high risk pediatric echocardiograms.
### Congenital Heart Defects

**7-10/1,000 Live Births**

<table>
<thead>
<tr>
<th>DIAGNOSIS (Balt-Wash)</th>
<th>PERCENT</th>
</tr>
</thead>
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<tr>
<td>Ventricular septal defect</td>
<td>26%</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>9%</td>
</tr>
<tr>
<td>Atrioventricular septal defect</td>
<td>9%</td>
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<tr>
<td>Atrial septal defect</td>
<td>8%</td>
</tr>
<tr>
<td>Pulmonary valve stenosis</td>
<td>7%</td>
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<tr>
<td>Coarctation of the Aorta</td>
<td>7%</td>
</tr>
<tr>
<td>Hypoplastic left heart syndrome</td>
<td>6%</td>
</tr>
<tr>
<td>D-Transposition</td>
<td>5%</td>
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</tbody>
</table>
CHD in Adults

30,000 babies born with CHD per year
20,000 surgeries for CHD per year
85% survive into adulthood
Over 1.2 million adults with CHD
Increasing at 5% per year
8,500 per year reach adulthood
Less than 10% disabled
<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>1950’s</th>
<th>1960’s</th>
<th>1970’s</th>
<th>1980’s</th>
<th>1990’s</th>
<th>2000’s</th>
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<tbody>
<tr>
<td>ASD</td>
<td>Rare Repair</td>
<td>Repair older child</td>
<td>Repair age 4</td>
<td>Repair age 2</td>
<td>Repair age 2-3</td>
<td>Device closure</td>
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<tr>
<td>VSD</td>
<td>Rare Repair</td>
<td>Repair &gt;10 kg or palliate</td>
<td>Repair &lt; 1 year or palliate</td>
<td>Repair 6 months or prn</td>
<td>Repair premature infants</td>
<td></td>
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<tr>
<td>PDA</td>
<td>Repair</td>
<td>Repair</td>
<td>Repair</td>
<td>Repair</td>
<td>Repair</td>
<td></td>
</tr>
<tr>
<td>TOF</td>
<td>Palliate</td>
<td>Late Repair in adults</td>
<td>Repair after palliation</td>
<td>修</td>
<td>Repair 2-8 months or prn</td>
<td></td>
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<tr>
<td>TGA</td>
<td>No survivors</td>
<td>Rare Survivors</td>
<td>Atrial Repair</td>
<td>Transitional Decade</td>
<td>Arterial Repair</td>
<td></td>
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<tr>
<td>Single Ventricle</td>
<td>Comfort care</td>
<td>Palliate</td>
<td>Rare Fontan</td>
<td>Fenestrated Fontan</td>
<td>Lateral Tunnel</td>
<td>Extracardiac Fontan</td>
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<td>HLHS</td>
<td>Comfort care</td>
<td>Comfort care</td>
<td>Surgery in Boston</td>
<td>Comfort vs. high risk surgery</td>
<td>Surgery &amp; Fetal Diagnosis</td>
<td></td>
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</table>
19 Days: Two endocardial tubes have formed – these tubes will fuse to form a common, single primitive heart tube
22 Days: Heart tube begins to beat
23 Days: Folding commences
30 Days: Primitive circulation
9 weeks (56 Days): All major structures identified

(In humans, several months of gestation remain for emergence of HLHS, PS, etc)
The Cardiac Crescent and the Tube Heart

From Heart Development, 1999
Looping and Septation

From *Heart Development*, 1999
How do Congenital Heart Defects form?

Complex interaction between environmental and genetic etiology

- Multifactorial
- 5-8% chance of recurrence

Environmental exposures may influence micro-uterine environment and either turn on or off needed protein development
Echocardiography

1793 Italian priest studied bats
1845 Austrian scientist Christian Doppler
WWII Sonar detected submarines
1954 Hertz & Edler
  • (A&B mode echocardiogram)
Reflection of US waves by target
Display based on
  • Intensity of returned signal
  • Time of “flight” or depth
Echo timeline

M-mode ultrasound early 1970’s
2D echo late 1970’s
Doppler Echo 1980’s
  • Pulsed wave Doppler
  • Continuous wave Doppler
  • Color Doppler
Pediatric Echo is Different

Anatomy and physiology over function
Segmental approach for complex patients
Improved resolution
  • Heart is closer to chest wall
  • Higher frequency transducers
  • TEE rarely necessary for diagnosis
Inversion of apical and subcostal images
Diagnostic accuracy depends on image quality

Improve signal/noise ratio
Improve image resolution
Appropriate transducer
Focus depth

OPTIMAL WINDOW SHOULD ALLOW US BEAM TO BE PERPENDICULAR TO AREA OF INTEREST FOR IMAGING AND PARALLEL TO FLOW JETS FOR DOPPLER AND COLOR
5 Standard Views

Subcostal
Left Parasternal
Apical
Suprasternal Notch
Right Parasternal
Subcostal structures

- IVC
- Hepatic veins
- Abdominal aorta
- Diaphragm
- SVC
- LA
- RA
- Atrial Septum
- Ascending aorta
- Branch PA
- Coronary sinus
- Pulmonary veins
- Mitral Valve
- Tricuspid Valve
- LV
- RV
- Ventricular Septum
- Aortic Valve
- Pulmonary Valve
- Pericardium
Left Parasternal

- IVC
- SVC
- LA
- RA
- Atrial septum
- Coronary sinus
- Pulmonary veins
- MV
- TV

- LV
- RV
- Ventricular septum
- Aortic Valve
- Pulmonary valve
- Ascending aorta
- Coronary arteries
- MPA/BPA
- Pericardium
Apical views

- IVC
- LA
- RA
- Atrial Septum
- Coronary sinus
- Aortic valve
- Pulmonary valve
- Ascending Aorta
- Pulmonary veins
- MV
- TV
- LV
- RV
- Ventricular Septum
- MPA/BPA
http://www.lai-echo.com/chapter4/video-4-4.asp
Suprasternal notch

- SVC
- LA
- Pulmonary veins
- Ascending aorta
- Thoracic Aorta
- MPA/BPA
- Aortic Arch
- Left Innominate vein
Right Parasternal

- IVC
- SVC
- RA
- Atrial septum
- Right pulmonary veins
- Ascending Aorta
- Right pulmonary artery
Hemodynamic Measurements

Doppler insonation angle
Pressure gradients
Bernoulli equation
  • Modified Bernoulli Equation $\Delta P = 4 \times v_2^2$
Flow
  • $Q_p = RVOT\ CSA \times RVOT\ VTI$
  • $Q_s = LVOT\ CSA \times LVOT\ VTI$
  • $Q_p/Q_s = 1/1$ normal, abnormal $\geq 1.5:1$
PI velocity for PAEDP
Echo in CHD

Doppler echo

• Pulsed wave Doppler

  • Quantitation of intracardiac hemodynamics
    – Valvar regurgitation
    – Intracardiac shunts
    – LVOT/RVOT obstruction

• Ventricular function

  – Systolic
  – Diastolic (mitral inflow, pulmonary venous inflow)
Echo in CHD

Continuous wave Doppler

• Non-invasive measurements of mean and peak transvalvar gradients
  • Valvar stenosis
• Prediction of Ventricular Pressure (modified Bernoulli equation)
  • VSD→ LV: RV pressure gradient
  • TR/PR→ RV, PA pressure
Echo in CHD

Color Doppler

- Direction of cardiac flow
  - TAPVR vs. LSVC
- Velocity and Turbulence of cardiac flow
  - Conduit obstruction
  - Identification of intracardiac shunts
    - VSD, PDA, ASD
- Assessment of Post-op CHD
  - Shunt patency, residual intracardiac shunt
Morphologic/Segmental approach

Define morphologic—not spatial—anatomy
- Which atrium is the Right? Left?
- Which ventricle is the Right? Left?
- Which great artery is which?

Define segmental anatomy
- Segments: Atrium, Ventricles, Great Arteries
- What is the position of each segment relative to each other?
  - Is the RA on the right? Is it connected to the RV? Is it connected to the PA?
  - Is the LA on the left? Is it connected to the LV? Is it connected to the Aorta?

Predict the physiology
- What is the physiology predicted by the segmental connections?
  - Normal? Transposition? Obstructed flow?
- What is the physiology predicted by flow in the ductus? Across the foramen?
The Cardiac Segments

- Atria
- Ventricles
- Great arteries
- AV canal
- Conus (infundibulum)
Abdominal and Atrial Situs

Cardiac position
- Levocardia, Dextrocardia, Mesocardia, Dextroposition

Situs abnormalities
- Inversus
  - Not often associated with CHD
- Ambiguous
  - Heterotaxy syndromes
    - Asplenia/polysplenia
    - Abdominal malrotation
    - Cardiac defect
      » AV canal defect
      » Conotruncal defects
      » Systemic and pulmonary venous anomalies
The Endocardial Cushion

Define the connections

• Concordant: RA to RV, LA to LV
• Discordant: RA to LV, LA to RV
• Common inlet: AV canal defect
• Atretic inlet: mitral, tricuspid valve atresia
• Double inlet

Assess AV valve anatomy and function

• Morphology
  • Ebstein’s tricuspid valve, parachute mitral valve
  • Hypoplastic

• Physiology
  • Stenosis
  • Atresia
  • Insufficiency
The Endocardial Cushion

Normal

Common AV inlet

Atretic AV inlet

Balanced

Unbalanced
The Ventricles

The Right Ventricle

- Coarsely trabeculated
- Moderator band
- “Septophilic” tricuspid valve chordal insertions

The Left Ventricle

- Finely trabeculated
- 2 prominent MV papillary muscles
- No septal attachments of valve
The Great Arteries

Identify the great arteries:

- **Aorta**
  - Coronary artery origins
  - Origin of brachiocephalic vessels from arch
  - “Candy cane?”
- **Pulmonary artery**
  - Proximal bifurcation into branch PAs
  - No brachiocephalic vessel from the ductal arch
  - “Hockey stick?”

How many outlets?

- One? = truncus arteriosus or semilunar valve atresia
- Two? Are they normal? In position? In size?

Do the great arteries arise from the correct ventricles?

- Aorta from LV, PA from RV = solitus (normal) GA
- Aorta from RV, PA from LV = transposition of the GA
- Both from RV = DORV
Types of conus

Subpulmonary

Subaortic

Bilateral

Bilaterally absent

Figure 3.13 Type of infundibulum. In general, there are four types of conus
Concordant
(D-loop TGA)

Discordant
(L-loop TGA)

Double-outlet right ventricle

Double-outlet left ventricle

Pulmonary atresia

Aortic atresia

Truncus arteriosus (common outlet)

the aorta and biventricular origin of the main pulmonary arteries
Common Lesions
Atrial Septal Defects

Secundum ASD
Primum ASD
Sinus Venosus defect
  • Not truly a deficiency of the same physiology as an ASD
Common atrium
Atrial Septal Development

http://www.med.unc.edu/embryo_images/unit-welcome/welcome_htms/contents.htm
ASD: Clinical Correlation

Usually diagnosed in childhood
Asymptomatic
F>M
Systolic ejection murmur and widely split fixed S2
EKG may show RBBB or RVH
Atrial Septal Defect

Increased flow across the tricuspid valve causes diastolic rumble

Excess flow through the pulmonic valve causes systolic murmur

Pulmonic valve closes late causing a fixed split second sound

Diastole | Systole | Diastole
Atrial Septum Normal Variants

Patent Foramen Ovale

Atrial Septal Aneurysm
Secundum ASD

RV Dilation

Diastolic Septal Flattening
Secundum ASD

Subcostal Coronal

Subcostal Sagittal
Devices for ASD Closure

Cardio-SEAL

Amplatzer
Amplatzer Occlusion of Atrial Septal Defect

Clockwise from above: Transcatheter delivery of Amplatzer device, which is positioned across the atrial septal defect

Left: Amplatzer device in place
ASD device
Primum ASD

Part of spectrum of AV canal defects

Defect is contiguous with AV valves

Associated with cleft mitral valve
Primum ASD
Sinus Venosus Defects

Deficiency in the wall between the right pulmonary veins and the RA

PAPV-DRAINAGE
- SVC type = RUPV
- Inferior type = RLPV
Sinus Venosus ASD
Sinus Venosus ASD
Partial Anomalous Pulmonary Venous Return (PAPVR)
Right veins (more common):
RA
SVC (RUPV to the RA or base of the SVC-sinus venosus ASD)
IVC
Left veins:
Innominate vein
Coronary sinus
Rarely: SVC, IVC, right atrium, or left subclavian vein
PAPVR to IVC
PAPVR to SVC
Total Anomalous Pulmonary Venous Return (TAPVR)

I: Supracardiac: common pulmonary vein drains into the right superior vena cava from the left superior vena cava (vertical vein) and the left innominate vein (50%)

II: Cardiac: coronary sinus, right atrium (20%)

III: Infracardiac: subdiaphragmatic (portal vein, inferior vena cava, ductus venosus) (20%)

IV: Mixed: any combination of types I, II, III, the least common
TAPVR
TAPVR to Vertical Vein
TAPVR to IVC
TAPVR to CS
The Ventricular Septum

AV canal septum (1)
Muscular septum including the trabecular portion and the septal band (3)
Conal septum (4)
The Ventricular Septum

Left ventricular view
AV canal septum (1)
Muscular septum including the trabecular portion (2) and the septal band (3)
Conal septum (4)
Muscular VSD

Within the muscular ventricular septum
Apical (black)
Mid (pink)
Anterior (green)
Posterior/inlet (white)
“Swiss cheese”
Tend to get smaller with time
Conoventricular VSD

In the area where the AV canal septum, conal septum and muscular septum meet “Membranous VSD”
- “Para-” or “Peri-” (red)

Malalignment
- “TOF-type”, “VSD in the Y of s...
AV Canal Type VSD

Deficiency of the AV cushion contribution to the ventricular septum (red)

"Inlet VSD"

Different from "Inlet muscular" V surrounded by muscle (blue)
VSD: Clinical Correlation

Size and pulmonary vascular resistance determines clinical presentation

- Fetal transition

Symptoms are determined by the size of the shunt

- Size of defect
- Presence of other anomalies
- Extracardiac abnormalities
VSD: Clinical Correlation

Audible after several days (not immediately after birth), typically picked up at 1st visit

Large defects = congestive heart failure

- Tachypnea (RR>60)
- Poor feeding/poor growth
- Reflux/vomiting
Small Ventricular Septal Defects

First heart sound is obscured

Second sound normal & splits with inspiration

Left to right high velocity shunt causes systolic murmur

Aortic & pulmonic valves close normally

<table>
<thead>
<tr>
<th>Diastole</th>
<th>Systole</th>
<th>Diastole</th>
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Children's National
VSD: Clinical Correlation

Spontaneous resolution
Or not...
Pulmonary disease
  • Eisenmenger’s syndrome
Aortic regurgitation
Muscular VSD
Membranous VSD
Membranous VSD
w/ TV aneurysm tissue
Membranous VSD w/ Aortic Valve Prolapse
Continuous wave Doppler in ventricular septal defect. The echocardiographic frame demonstrated the Doppler determination of pressure gradient across a membranous ventricular septal defect (VSD) (white arrow). The direction of the continuous wave Doppler beam used to obtain the velocity across the ventricular septal defect is illustrated by the red arrow. The velocity ($V$) is 4.5 m/sec and based upon the modified Bernoulli equation, (pressure = [velocity] $^2 \times 4$) the gradient is 81 mmHg. (Courtesy of Ann Kavanaugh-McHugh, MD.)
Restrictive Membranous VSD

Max V = 466 cm/sec
Max PG = 86.9 mmHg
Unrestrictive Membranous VSD
Atrioventricular Canal Defect - Complete
Common AV Canal (CAVC)

Endocardial Cushion Defect (ECD)
Atrioventricular Septal Defect (AVSD)

Failure of the AV canal to develop properly and form tricuspid, mitral valves and portions of atrial and ventricular septae
Spectrum of defects
Definitions

Incomplete CAVC = lack the VSD component or ASD component
Partial CAVC = synonym for incomplete CAVC OR = primum ASD with cleft mitral valve
Transitional CAVC = small VSD component
Balanced/Unbalanced
Atrioventricular Canal Defect – Partial
AV Septal Defect

Complete

ASD

VSD

Anterior Leaflet

Posterior Leaflet
Best View of CAVC
Unbalanced
Patent Ductus Arteriosus

Patent Ductus Arteriosus

Closed Ductus Arteriosus

Aorta

PA
PDA: Clinical Correlation

Closed in 90% of infants by 48 hours of life
  • Prematuring, altitude

Anatomy
  • Derived from the left 6th embryonic arch

Closure
  • Muscular constriction→endothelium→thrombosis→fibrous strand

Physiology↔ shunting
  • Symptoms proportional to shunting

Murmur

EKG
  • Ventricular hypertrophy
Patent Ductus Arteriosus

First sound obscured

Second sound obscured

Systolic & diastolic murmur from patent ductus arteriosus

Diastole  Systole  Diastole
Patent Ductus Arteriosus
Doppler of the PDA (L-R shunt)

CW Doppler tracing (right) seen above the baseline indicating flow toward the probe from the descending aorta through the PDA to the PA. The peak velocity is reached in late systole 4 m/s. L-R shunt

Color flow Doppler (left) showing a L-R shunt from the descending aorta through the PDA to the PA (red: towards the probe)
Doppler of the PDA (bidirectional shunt)

CW Doppler from an infant with pulmonary artery hypertension and PDA. The negative deflection in systole below the baseline arises from the R-L shunt through the PDA from the PA to the Dao (away from the TDX). The positive deflection (late systole-into late diastole) arises from L-R shunt through the PDA from the Dao to the PA.

Bidirectional blood flow through the PDA can be a normal finding in newborn infants due to high pulmonary resistance.
The Doppler spectral tracing shows evidence of severe pulmonary hypertension and no evidence of a L-R shunt through the PDA. The shunt is R-L from the ductus arteriosus to the Dao (blue: away from the TDX)
Patent Ductus Arteriosus – Ligation and Division
Occlusion of Intracardiac and Vascular Shunts
Coil embolization of PDA

Left, top: Catheter crosses the PDA from the aortic side and delivers a coil.
Left, bottom: Withdrawal of catheter, leaving coil in PDA
Amplatzer Ductal Occluders

Amplatzer ductal occluder
Illustration courtesy AGA Medical Group

Aorta angiogram with device occlusion of PDA, lateral view
Right Heart Obstructive Lesions
Pulmonary Valve Stenosis

Valve anatomy

- Doming, fused commissures
- Thickened, immobile leaflets
- Subvalvar obstruction
- Supravalvar obstruction

Post stenotic dilation

RVH
PS: Clinical Correlation

Asymptomatic
Murmur at birth
EKG
  • RAD, RVH proportional to obstruction
Management
  • Balloon dilation
Excellent outcome
Pulmonic Stenosis

First heart sound may be followed by an pulmonic ejection click that varies with respiration

Systolic murmur

Second heart sound has fixed split

Ejection click caused by pulmonary valve opening

Murmur caused by stenotic pulmonary valve

Fixed split due to delayed closure of the pulmonary valve

Diastole | Systole | Diastole

Children's National™
Pulmonary Artery Branch Stenosis
Tetralogy of Fallot
Tetralogy of Fallot

“Maladie Bleu” 1888
Stensen 1671
Sandifort 1777

Arthus Louis Etienne
FALLOT
Variations of Tetralogy

Tet, pulmonary atresia:
MAPCAS
“Mexican Tet”
- Hypoplastic or absent conal septum
Tetralogy with absent pulmonary valve
- Rudimentary pulmonary valve leaflets result in fetal pulmonary regurgitation, PA dilation
- Airway and lung development is compromised in severe cases
Tetralogy with CAVC
TOF: Clinical Correlation

Most common cyanotic defect
Defective neural crest migration resulting in abnormal conotruncal development
Clinical presentation depends on degree of subpulmonary narrowing
  • This may change over time
Presentation
  • Fetal dx
  • Murmur
TOF: Clinical Correlation

Cyanosis due to right to left shunting at ventricular level
Degree of cyanosis is proportional to amount of RVOTO
Dynamic factors may worsen cyanosis
  • Tet Spell → no murmur → deeply cyanotic
EKG
  • RVH, RAD, RAE
CXR
  • Boot shaped heart
Tetralogy of Fallot

First sound sounds split due to pulmonic valve opening click

Pulmonic valve click

Systolic murmur

Single second sound

Diastole | Systole | Diastole
Tetralogy of Fallot
Transcatheter Pulmonary Valve

• Catheter delivered prosthetic pulmonary valve
• Made from bovine jugular vein
• Sewn within a platinum-iridium balloon expandable stent
• For use in patients with a surgically placed conduit from the RV to the PA
• Used to treat significant conduit valve insufficiency and/or stenosis that would otherwise require surgical conduit replacement
• FDA approved 2010
DORV

- Describes a relationship where the PA and Aorta both arise from the anatomic RV
- “DORV” is normal during heart development
- Incidence 1 – 1.5% of patients with CHD
- 1 per 10,000 live births
- Possible association with trisomy 13 and trisomy 18
- Van Praagh – both great arteries arise from the morphologically RV
- NO mitral - aortic fibrous continuity
- Two functional ventricles in which a VSD provides the only outlet for one ventricle
- Anderson - 50% override rule – “if >50% of the aorta is over the RV, its DORV”
Left Heart Obstruction
Aortic Stenosis

Valve, sub-valvar or supravalvar

Clinical manifestations

- Mild-moderate assymptomatic
- Severe
  - Depends on age of patient
- Management
  - Cath vs. surgery
Aortic Stenosis

- First heart sound is followed by an aortic ejection click
- Systolic murmur
- Narrowly split second sound

Aortic ejection click caused by aortic valve opening
Murmur caused by stenotic aortic valve
Aortic valve closes late

Diastole | Systole | Diastole
Coarctation of the Aorta

Aberrant ductal tissue within the wall of the aorta
All coarcts are “juxtaductal”
Pseudocoarctation = kinking at t WITHOUT obstruction
Coarctation of the Aorta

AAo

Narrowed Isthmus
Normal or CoA?
Descending AO Doppler
Coarctation
Doppler “drag”
Interrupted Aortic Arch

Type A = After the subclavian artery, probably an extreme form of coarctation with obliteration of lumen

Type B = Between the LCC and LSCA, most common defect of arch remodeling/neural crest

Type C = Between the Carotid arteries, most...
Complex Lesions
D-Transposition of the Great Arteries
D-TGA

First described by Baillie 1797
Natural history: >90% mortality in infancy
Incidence: ~5% of congenital heart disease
Rare association with syndromes or other anomalies
Male:Female = 2:1
Possible association with infant of diabetic mother
D-TGA

Ventriculo-arterial discordance
Circulation in parallel
RA=>RV=>Ao
LA=>LV=>PA
Must have mixing at atrial or survive
D-TGA
D-Transposition
D-Transposition
Balloon Septostomy
Again...
Arterial Switch Procedure
Long Term Postoperative Concerns
Arterial Switch Operation

Neo-pulmonary stenosis
Coronary abnormalities
  • Obstruction and stenosis
  • Decreased flow reserve
Neo-aortic insufficiency
  • Almost always trivial/mild
LV function
Mustard Repair

Transposition of the Great Arteries
Mustard Repair
Atrial Baffle Repair
Long Term Sequelae

On going late mortality risk
  • 20% mortality at 20 years

Arrhythmia
SVC obstruction -- 14-17%
IVC obstruction -- 1%
Baffle Leak -- Significant 1-2%
Systemic AV valve regurgitation -- 30%
Systemic Ventricular Failure -- 15-20%
Transposition of the Great Arteries
L Type

MV – mitral valve
TV – tricuspid valve
L-TGA

“Congenitally Corrected Transposition”
Atrio-ventricular and ventriculo-arterial discordance (“double discordance”)
RA ⇒ LV ⇒ PA
LA ⇒ RV ⇒ Ao
May be an isolated, asymptomatic finding or may be associated with other heart malformations
Truncus Arteriosus

A single vessel arising from the heart and giving rise to the coronary, pulmonary and systemic circulations

The VSD is the same as TOF
Truncus Arteriosus

COLLETT & EDWARDS

I  II  III  IV

VAN PRAAGH

A1  A2  A3  A4
AP Window

Communication between aort
Hypoplastic Left Heart Syndrome
Hypoplastic Left Heart Syndrome

RV

LV

RA

LA

MPA

1 mm

AAo
1930: Vivien Thomas hired as Alfred Blalock's lab assistant

1924: Failing to obtain a surgical residency at Hopkins, Alfred Blalock goes to Vanderbilt and begins research on traumatic shock

1938: Rabbit models with subclavian to PA anastomosis fail to produce pulmonary HTN

1941: Blalock and Thomas move to Hopkins

1941: Coarctation relief with subclavian to descending aorta shunt

1943: Helen Taussig, a Hopkins pediatrics residency graduate, approaches Blalock about help for “blue babies”

1944: “Anna,” a dog with a surgically created mixing lesion, successfully undergoes end-to-side subclavian-to-PA anastomosis, lives 15 years

November 29, 1944: Eileen Saxon, a 15-month-old 4.5 kg undergoes successful systemic-to-pulmonary shunt by Blalock with Thomas directly over his shoulder
Norwood I: Anatomy

1. Atrial septectomy
2. Ligation of main pulmonary artery and construction of neo-aorta
3. Sano Modification/Modified BT Shunt
BT Shunt
Norwood I: Sano

Sano modification

- RV-to-PA conduit
- Eliminates competitive flow
- Enhances coronary perfusion
Sano Shunt
• End-to-side anastomosis of SVC to undivided right pulmonary artery

• Includes takedown of BT shunt

• Allows flow to both lungs from SVC via passive flow
Glenn Shunt
Glenn Doppler
Original Fontan
Fontan: Variations

Lateral tunnel runs within RA, using free wall plus conduit as baffle for IVC blood
  • Fenestrations: R-to-L shunting through fenestration → hypoxemia
  • Improve cardiac output, minimize systemic venous hypertension, decrease post-op thoracostomy drainage
  • Can later be closed by catheter

Extracardiac is IVC to MPA
  • Generally has lower rate of complications
  • Foreign material requires anticoagulation
Fenestrated Fontan
Hypoplastic Left Heart Syndrome
Palliative Reconstruction

Stage I -- Norwood Procedure
  • Birth
Stage II -- Bi-directional Cavopulmonary Shunt
  • 4-6 months
Stage III-- Fontan Procedure
  • 18-24 months for lateral tunnel procedure
  • > 15 kg for extracardiac procedure
AS YOU CAN CLEARLY SEE IN SLIDE 397...

GAAAHH!

“POWERPOINT” POISONING.
QUESTION 1
A tachypneic 2 month old is not growing well and has a murmur. An echocardiogram is obtained:
QUESTION 1 (CONT)
All of the following statements are likely to be true except:

A. The patient is at increased risk to have Down Syndrome
B. The patient’s pulmonary artery pressure is normal
C. The patient has an endocardial cushion defect
D. The patient has a normal oxygen saturation
E. The patient may have a small mitral valve cleft after surgical repair
QUESTION 2
A cyanotic newborn has the following echocardiogram:
QUESTION 2 (CONT)
All of the following statements are likely to be true except:

A. The aorta is malposed anterior and rightward
B. The right ventricle pumps blood to the body
C. Oxygenated blood is pumped to the lungs
D. The left ventricle pumps blood to the body
E. The right ventricular pressure is greater than or equal to the left ventricular pressure
A 40 year old with atrial fibrillation has the following echo:
QUESTION 3 (CONT)
Subsequent imaging is most likely to reveal the following:

A. Tetralogy of Fallot
B. Large membranous ventricular septal defect
C. Large patent ductus arteriosus
D. Large secundum atrial septal defect
E. No structural cardiac defect
QUESTION 4
A 3 month old with a loud murmur and intermittent perioral cyanosis has the following echo:
QUESTION 4 (CONT)
All of the following statements are likely to be true except:

A. The aorta is overriding the left and right ventricle
B. There is a large ventricular septal defect
C. There is pulmonary stenosis
D. The right ventricular pressure is increased
E. The pulmonary artery pressure is increased

E
QUESTION 5
An asymptomatic 9 month old with a loud murmur and a BP of 79/48 and has the following parasternal long axis 2D and CW Doppler findings:
QUESTION 5 (CONT)
The most likely diagnosis is:

A. Membranous VSD, normal RV pressure
B. Membranous VSD, elevated RV pressure
C. Muscular VSD, normal RV pressure
D. Muscular VSD, elevated RV pressure
E. Tricuspid regurgitation, elevated RV pressure
Acknowledgements

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