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

Overview of Embryology
Understand Pediatric Echocardiography
Congenital Heart Disease
  • Common lesions
  • Complex lesions
## Congenital Heart Defects

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

<table>
<thead>
<tr>
<th>DIAGNOSIS (Balt-Wash)</th>
<th>PERCENT</th>
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<tbody>
<tr>
<td>Ventricular septal defect</td>
<td>26%</td>
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<tr>
<td>Tetralogy of Fallot</td>
<td>9%</td>
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<tr>
<td>Atrioventricular septal defect</td>
<td>9%</td>
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<tr>
<td>Atrial septal defect</td>
<td>8%</td>
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<tr>
<td>Pulmonary valve stenosis</td>
<td>7%</td>
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<tr>
<td>Coarctation of the Aorta</td>
<td>7%</td>
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<tr>
<td>Hypoplastic left heart syndrome</td>
<td>6%</td>
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<tr>
<td>D-Transposition</td>
<td>5%</td>
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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>
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<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>
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<tr>
<td>PDA</td>
<td>Repair</td>
<td>Repair</td>
<td>Repair</td>
<td>Repair</td>
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<tr>
<td>TOF</td>
<td>Palliate</td>
<td>Late Repair in adults</td>
<td>Repair after palliation</td>
<td>Repair 2-8 months or prn</td>
<td></td>
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<td>TGA</td>
<td>No survivors</td>
<td>Rare Survivors</td>
<td>Atrial Repair</td>
<td>Transitional Decade</td>
<td>Arterial Repair</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>
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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
From Dr. R. Anderson
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
Echo timeline

1793 Italian priest studied bats
1845 Austrian scientist Christian Doppler
WWII Sonar detected submarines
1954 Hertz & Edler
  • (A&B mode echocardiogram)
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
Echo in CHD

Doppler echo

• Pulsed wave Doppler
  • Quantitation of intracardiac hemodynamics
    (Modified Bernoulli Equation $\Delta P = 4 \times v^2$)
    – 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
Doppler Spectral Display

Pulsed Wave (PW)

Continuous Wave (CW)

Aortic Valve Velocity
RV pressure by TR estimation

The pressure in the right ventricle is 55 + 10 = 65 mmHg. The pressure in the LV is 83 mmHg; we know it from the blood pressure which was 83/61. So, it the RV systemic pressure is \( \frac{3}{4} \) of the systemic LV pressure. It means RV systemic pressure is elevated. It should normally be \( \frac{1}{3} \) systemic.
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
Questions....

1. How much time should you spend trying to obtain Doppler of TR when there is a HUGE ventricular septal defect?
2. What if your patient has a single ventricle, if you measure the TR what does that estimate?
3. Why is it important to Doppler a VSD?
4. If you see funny blood flow, should you invert your color scale?
5. The doctor wants to know if there is pulmonary hypertension in a NICU baby, but there is no TR, is there another way to answer that question?
Guidelines and Standards for Performance of a Pediatric Echocardiogram: A Report from the Task Force of the Pediatric Council of the American Society of Echocardiography

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Houston, Texas; and Philadelphia, Pennsylvania
<table>
<thead>
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<th>Table 4 Structures viewed from standard examination views</th>
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<td><strong>Subxiphoid (subcostal) views</strong></td>
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<tr>
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<td>Hepatic veins</td>
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<td>Abdominal aorta</td>
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<td>Diaphragm</td>
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<td>Superior vena cava</td>
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<td>Mitral valve</td>
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<td>Tricuspid valve</td>
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<td>Left ventricle</td>
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<td>Left ventricular papillary muscles</td>
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<td>Coronary arteries</td>
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<td>Main and branch pulmonary arteries</td>
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<td>Pericardium</td>
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<td>Apical views</td>
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<td>Inferior vena cava</td>
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<td>Ascending aorta</td>
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<td>Main and branch pulmonary arteries</td>
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Classification and Terminology of Cardiovascular Anomalies
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?
Cardiac base-apex axis and orientation in the chest

Levocardia  Mesocardia  Dextrocardia
Cardiac situs (sidedness)
Example: Cardiac sidedness

Situs solitus normal cardiac sidedness

Situs ambiguus, right isomerism
Differentiation between the atria

The morphologic RA has a smooth or sinusal portion, which is found between the interatrial septum and the crista terminalis. It receives the drainage of the superior and inferior venae cavae and the coronary sinus. The trabecular portion is characterized by the presence of pectinate muscles, which are directed from crista terminalis to the base of the right atrial appendage. The RA appendage is wide and its edge is blunt.

RA appendage is broad based and triangularly shaped (like Snoopy’s nose), with pectinate muscles that extend into the body of the right atrium.

The anatomic LA is totally smooth and lacks pectinate muscles. It receives the drainage of the pulmonary veins, and LA appendage has a narrow base and fingerlike appearance (like Snoopy’s ears) with pectinate muscles confined within the appendage.
RA and TV valve characteristics

**Right atrium:**
- Limbus of fossa ovalis (limb of oval fossa)
- Large pyramidal appendage (Snoopy’s nose)
- Crista terminalis (terminal crest)
- Pectinate muscles
- Receives venae cavae and coronary sinus*

**Tricuspid valve:**
- Low septal annular attachment
- Septal cordal attachments
- Triangular orifice (midleaflet level)
- Three leaflets and commissures
- Three papillary muscles
- Empties into right ventricle
LA and MV valve characteristics

**Left atrium:**
- Ostium secundum
- Small fingerlike appendage (Snoopy’s ear)
- No crista terminalis
- No pectinate muscles
- Receives pulmonary veins*

**Mitral valve:**
- High septal annular attachment
- No septal cordal attachments
- Elliptical orifice (midleaflet level)
- Two leaflets and commissures
- Two large papillary muscles
- Empties into left ventricle
Differentiation between the atria

The only structures that are constant and allow differentiation between the right and left atria are the appendages!

The drainage of the systemic and pulmonary veins does not permit the conclusive identification of the atria, as drainage sites are sometimes anomalous. The atrial septum cannot always be used either, because it can have defects or be absent.
Ventricles-characteristics

**Right ventricle:**
Tricuspid-pulmonary discontinuity
Muscular outflow tract
Septal and parietal bands
Large apical trabeculations
Coarse septal surface
Crescentic in cross sections* (* variable)
Thin free wall (3–5 mm)*
Receives tricuspid valve
Pulmonary valve empties into main pulmonary artery
Ventricles-characteristics

**Left ventricle:**
- Mitral-aortic continuity
- Muscular-valvular outflow tract
- No septal or parietal band
- Small apical trabeculations
- Smooth upper septal surface
- Circular in cross section* (* variable)
- Thick free wall (12–15 mm)*
- Receives mitral valve
- Aortic valve
- Empties into ascending aorta
Ventricular features (summary)

- Features of the morphologic RV:
  - Coarse trabeculae with prominent septal band, parietal band, and moderator band.
  - Septophillic attachments of the tricuspid valve (attachments to septum and free wall)
  - Well-developed infundibulum (= conus= cone of muscle beneath the semilunar valve) which results in fibrous dyscontinuity between the tricuspid and semilunar valves

- Features of the morphologic LV:
  - Smooth septal surface, fine trabeculae
  - Septophobic attachments of the mitral valve (attachments only to free wall)
  - No infundibulum which results in fibrous continuity of the mitral and semilunar valves
Atrioventricular connections

Concordance
Discordance
Ambiguous

Double inlet (univentricular)
Single inlet (univentricular)
Common inlet (univentricular)
Examples of atrioventricular connections:
A. Concordance
B. Discordance
C. Double-inlet LV
D. Ticuspid atresia: absent right A-V connection
Overriding and straddling
Arterial Segment

A- normal, Pa- anterior, left
Ao-posterior, right
Ventriculoarterial connection - possible
To summarize…...The Cardiac Segments

Viscera and atria
  • Abdominal situs
  • Systemic and pulmonary venous return
  • Atrial anatomy

Atrioventricular canal
  • AV valves and atrioventricular septum

Ventricles
  • Ventricular anatomy (D- or L-looping)
  • Ventricular size and proportion
  • Ventricular septum

Conus
  • Ventricular outflow tracts

Great arteries
  • Semilunar valves
  • Great arteries
Common Lesions
ASD

RV Dilation

Diastolic Septal Flattening
Atrial Septal Defects

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

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

Part of spectrum of AV canal defects

Defect is contiguous with AV valves

Associated with cleft mitral valve
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
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

Normal first sound

Fixed split second sound

Diastole | Systole | Diastole
Devices for ASD Closure

Cardio-SEAL

Amplatzer

Children's National
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
Newborn infant noted to be breathing heavy in Newborn nursery

Chest xray demonstrates increased lung markings.
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
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
“Very loud murmur” heard prior to hospital discharge
Baby is well, feeding, growing, pink, passed new pulse ox screening
The Ventricular Septum

AV canal septum (1)
Muscular septum including the trabecular portion (2) and the septal band (3)
Conal septum (4)
Conoventricular
Membranous
Inlet
Malalignment
The Ventricular Septum

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

Spontaneous resolution
Or not...
Pulmonary disease
  • Eisenmenger’s syndrome
Aortic regurgitation
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 x 4) the gradient is 81 mmHg. (Courtesy of Ann Kavanaugh-McHugh, MD.)
Restrictive Membranous VSD
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
Definitions
Spectrum of defects

- 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
Bonus points...

You are doing the echo on a baby and diagnosis her with an Unbalanced AVC.

You are having a hard time imaging the aortic arch.

Are you concerned, or do you think to yourself, the arch is always hard to image, these babies have no necks, they can’t stand when I put my transducer there....I am sure its fine, I just can’t see it right now.....
You are called to NICU to echo 28 week premature baby, weight is 600 gm, every time you try to image the baby’s HR falls and alarms go off...
Patent Ductus Arteriosus
Patent Ductus Arteriosus
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
Doppler of the PDA (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)

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
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
Doppler of the PDA (R-L shunt)

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

DIVISION AND LIGATION
Patent Ductus Arteriosus

[Diagram of heart with labeled sections and arrows indicating division and ligation]
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
- 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
Pulmonary Artery Branch Stenosis
This adorable baby was just adopted from Russia. She has a history of a heart condition....
Tetralogy of Fallot: “Maladie Bleu” 1888
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
Variations in TOF

- “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
- Tetrology with pulmonary atresia
TOF: Clinical Correlation

- Cyanosis due to right to left shunting at ventricular level
- Degree of cyanosis is proportional to amount of right ventricular outflow tract obstruction
- 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

Single second sound

Systolic murmur

Diastole | Systole | Diastole

Diagram showing heart in diastole and systole phases with noted sounds and murmurs.
Tetralogy of Fallot
Transcatheter Pulmonary Valve - 2010

• 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
Double Outlet Right Ventricle (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
You are called to the emergency room to perform an echo on a baby that is listless and pale.
He has not been eating well over the last 24 hours
The ER doctor wants to know if they need to call cardiology....
You decide to start with parasternal imaging, you notice the LV function is very very bad....where should you image next?
Coarctation of the Aorta
Coarctation of the Aorta

Aberrant ductal tissue within the wall of the aorta

All coarcts are “juxtaductal”

Must look for other left heart Disease (aortic & mitral valve)
The next day you are staffing a Children’s clinic and the nurse tells you the blood pressure in the legs of the next patient are the same as the arms.

The doctor is busy and asks that you perform an echo while she finishes the previous patient...
Descending AO Doppler
Doppler “drag”
Interrupted Aortic Arch

- Type A = After the subclavian artery, probably an extreme form of coarctation with obliteration of the lumen
- Type B = Between the LCC and LSCA, most common, defect of arch remodeling/neural crest
- Type C = Between the Carotid arteries, most rare
Complex Lesions
The nurse from the nursery calls you frantic, there is a baby that is blue. He was born earlier today, he seemed ok, his birth weight was 8 pounds, uncomplicated pregnancy and delivery...
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-Transposition
Balloon Septostomy
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

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

VAN PRAAGH
AP Window

Communication between aorta and PA
Hypoplastic Left Heart Syndrome
Hypoplastic Left Heart Syndrome

LA, RA, RV, LV, MPA, AAo, 1 mm
1924: Failing to obtain a surgical residency at Hopkins, Alfred Blalock goes to Vanderbilt and begins research on traumatic shock

1930: Vivien Thomas hired as Alfred Blalock’s lab assistant

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 to PAs in diastole
- Enhances coronary perfusion
Sano Shunt
Bidirectional Glenn: Anatomy

- 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
Fontan: Variations

Lateral tunnel runs within RA, using free wall plus conduit as baffle for IVC blood

- Fenestrations: R-to-L shunting through the fenestration → hypoxemia
- Improve cardiac output, minimize systemic venous hypertension, decrease post-op thoracostomy drainage
- Can later be closed by cath

Extracardiac is IVC to MPA
- Generally has lower rate of complications
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...

GAAAAH!

"POWERPOINT" POISONING.
QUESTION 1
A tachypneic 2 month old is not growing well and has a murmur. An echocardiogram is obtained:

SYSTOLE

DIASTOLE
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 may not need surgery
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 pulmonary artery gives rise to the coronary arteries.
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
QUESTION 3
A 40 year old with atrial fibrillation has the following echo:

SYSTOLE

DIASTOLE
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
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:
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
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