Congenital Heart Disease: A NICU Perspective

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Objectives

- To review the basics of cardiac anatomy and embryology
- To review the fetal and transitional circulation and pertinent physiologic changes that occur at birth
- To define basic murmur nomenclature and grading
- To practice a systematic approach to differential diagnosis of cardiac murmurs by location
- To develop a strategy for identifying innocent murmurs vs. those that need subspecialty evaluation
- To introduce neonatal screening guidelines and algorithms

You are here...
Cardiac Anatomy: 101

Superior vena cava
Right atrium
Right ventricle
Inferior vena cava
Aorta
Pulmonary trunk
Left atrium
Pulmonary veins
Left ventricle

Cardiac Anatomy: 101

Pulmonary trunk
Aortic arch
Superior vena cava
Right pulmonary artery
Right pulmonary veins
Right atrium
Tricuspid valve
Right ventricle
Inferior vena cava
Descending aorta
Interventricular septum
Left atrium
Left pulmonary veins
Left ventricle
Aortic valve
Mitral valve
But how the heck did we get here??

Sherman, set the Way-Back machine to the second week of gestation, and HANG ON!!!
Fetal Circulation

- Shunts – PFO, PDA, placenta
- High PVR, low pulmonary blood flow
- Low oxygen tension**
- Gas exchange occurs in the placenta

**Most prominent factor in the elevated fetal PVR

Transitional Circulation

- Separation from the placenta $\rightarrow$ $\uparrow$ SVR
- Expansion of the lungs $\rightarrow$ $\uparrow$ Oxygen tension $\rightarrow$ $\downarrow$ PVR $\rightarrow$ $\uparrow$ Pulmonary blood flow $\rightarrow$ $\uparrow$ LA volume and pressure $\rightarrow$ Closure of the PFO
- PDA closure – Shunt becomes L to R as the PVR drops $\rightarrow$ $\uparrow$ Oxygen tension causes ductal constriction and functional closure
What about that duct...

- Oxygen induces the initial constriction along with a normal fall in endogenous levels of prostaglandins after birth in the term baby
- Persistence of the ductus in preterm infants is likely secondary to a combination of persistently elevated prostaglandin levels and also relative hypoxemia

Transitional Circulation

- By 24h of life, the PVR and PA pressure will fall to about ½ systemic
- This continues through the first 6w of life
- Related to involution of smooth muscle in the walls of small pulmonary arteries
And we’re back!

What about murmurs???
Murmurs

• What is a murmur?
  – Sound created by blood moving through the heart & blood vessels

• Timing - Systolic vs. Diastolic

• Location

• Grade

• Quality

• Radiation

Systolic Murmurs

• Start with or after S1 & terminate before or with S2.

• Graded I-VI
  – I – Barely audible
  – II – As loud as heart sounds
  – III – Louder than heart sounds, no thrill
  – IV – Readily audible with palpable thrill
  – V – Loud enough to be heard with diaphragm barely on chest wall; + palpable thrill
  – VI – Loud enough to be heard with the diaphragm off the chest; + palpable thrill

• S1- Coincident (holosystolic) vs. Systolic Ejection
Holosystolic Murmurs

- Also called “S1-coincident” murmurs since they don’t always persist for duration of systole
- Start as soon as ventricular pressure exceeds atrial pressure
- Example regurgitant murmurs:
  - VSD
  - High pitch with smaller VSD
  - AV valve regurgitation
  - “blowing”

Systolic Ejection Murmurs

- Start after closure of the AV valves
- Lesions associated with SEMs
  - Stenoses of the semilunar valves
  - Pulmonary “flow” murmurs
  - Still’s murmur
  - PPS
Diastolic Murmurs

- Start after closure of the semilunar valves
- Lesions associated with diastolic murmurs
  - Insufficiency of the semilunar valves
  - Stenosis of the AV valves

Continuous murmurs

- During systole and diastole
- Example:
  - Venous hum
  - PDA
  - Coronary fistulae
  - AP shunt
  - AVMs
The Strategy - Systolic

AS
PS
TR
VSD
MR
Still's

The Strategy - Diastolic

AI
PI
TS
MS
(Pi)
(AI)
The Strategy - Continuous

Innocent murmur “pearls”

- Innocent
  - “musical”
  - LLSB-Apex
  - Early to mid-systolic
  - No diastolic component
  - Grade 2-3/6 or less
  - No radiation to neck or back
  - No coincident symptoms or other worrisome exam findings
  - ↓d or eliminated with Valsalva, squatting → standing, laying → sitting
Phone a friend..

- Presence of a thrill
- Radiation to the neck or interscapular area
- Clicks, gallops, or other concerning exam findings
- Diastolic murmurs
- Hyperdynamic precordium or subxiphoid impulse
- Symptoms

So why don’t I care about murmurs??

- Can you hear anything with this thing??

- Lots of big bad heart disease has no associated pathologic murmur...
So what do we do then??

Newborn Screening for Critical Congenital Heart Disease

What is CCHD??

• Primary Screening Targets:
  – Hypoplastic left heart syndrome
  – Pulmonary atresia with intact ventricular septum
  – Tetralogy of Fallot
  – Total anomalous pulmonary venous return
  – Transposition of the great arteries
  – Tricuspid atresia
  – Truncus arteriosus
HLHS

1. Mitral atresia (or stenosis)
2. Aortic atresia (or stenosis)
3. Hypoplastic LV chamber
4. Hypoplastic aortic arch
5. Coarctation
6. ASD

PA/IVS

1. Mitral atresia (or stenosis)
2. Aortic atresia (or stenosis)
3. Hypoplastic LV chamber
4. Hypoplastic aortic arch
5. Coarctation
6. ASD
1. VSD
2. RVOT Obstruction
   2a. Valve PS or Atresia
3. Overriding Aorta
4. Secondary RVH

**TOF**

**TAPVR**
d-TGA

1. ASD
2. Transposed Great Vessels
3. PDA

Tricuspid Atresia

1. ASD
2. Tricuspid atresia
3. Hypoplastic RV chamber
4. Pulmonary Stenosis
5. VSD
6. PDA
Truncus Arteriosus

1. Common Trunk
2. Abnormal Truncal Valve
3. VSD

What is CCHD??

- Secondary Screening Targets:
  - Coarctation of the aorta
  - Interrupted aortic arch
  - Ebstein anomaly
  - Double outlet right ventricle
  - Double inlet single left ventricle
Coarctation

Interrupted Aortic Arch
Ebstein Anomaly

Double Outlet Right Ventricle

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Double Inlet (Single) Left Ventricle

1. Hypoplastic RV
2. VSD
3. Transposed Great Arteries
4. Double Inlet to the LV
5. ASD
6. Ventricular Inversion (AV discordance)
7. Left Aortic Arch

What do we know?

- Nearly 7,000 newborns with CCHD are born in the US annually.
- Children with CCHD are more likely to develop impairments in motor functions, speech and language, visual-motor-perceptual functions, and executive functions.
- Children with CCHD are more likely to utilize social services.
- Pulse oximetry screening for CCHD is a noninvasive procedure and may take as little as 5 minutes to conduct.
- Recent estimates put the cost of screening for CCHD around $6 per newborn.
- Recent estimates have demonstrated that newborn screening for CCHD is cost effective, with early detection leading to around $40,000 per life year gained.
What if my baby fails??

- Call your friendly, local pediatric cardiologist to request a consultation including a complete congenital echocardiogram
- It’s NEVER the wrong answer to start PGE until a diagnosis can be confirmed or refuted

I’m friendly!

Is the test really that good?

- Sensitivity – 76.5%
- SPECIFICITY – 99.9%

- That means that if your baby passes the screening, there is a 99.9% chance that they DO NOT have CCHD
- If your baby fails screening, there is a 76.5% chance that they DO have CCHD
1% of births will be affected by CHD – 40,000 babies
25% of that CHD is “critical” – 10,000 babies
Pulse ox screening (in addition to thorough H&P) is estimated to identify 1,200 more babies per year that otherwise would have gone undiagnosed
50 deaths per year will be avoided by pulse ox screening before discharge

Here’s a story...

Emily was born at term via uncomplicated SVD to a 37yo G8P8 mom after an uncomplicated pregnancy
Nonsyndromic infant with NO pathologic findings on physical exam
Pulse ox screening at 24HOL revealed pre and post ductal sats of 91%
Echo was done – dx of TGA was made
Key Points

• Mild-moderate cyanosis is virtually impossible to detect visually
  – Hgb 20.0g/dl → not visible until SaO2 of 86%
  – Hgb 9.0g/dl → not visible until SaO2 of 67%
• Bad disease sometimes sounds completely normal
thanks.