

Murmurs in the diagnosis of congenital heart disease

Diana Tsen, MD

Assistant Professor of Pediatrics, Division of Cardiology

University of Colorado School of Medicine

Children's Hospital Colorado Heart Institute



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Disclosure

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Objectives

- 1) Review physiology and practical aspects of the cardiac auscultation exam
- 2) Identify red flag symptoms for critical congenital heart disease (CHD) requiring more urgent referral
- 3) Discuss cases reviewing the presentation and natural history of common CHDs



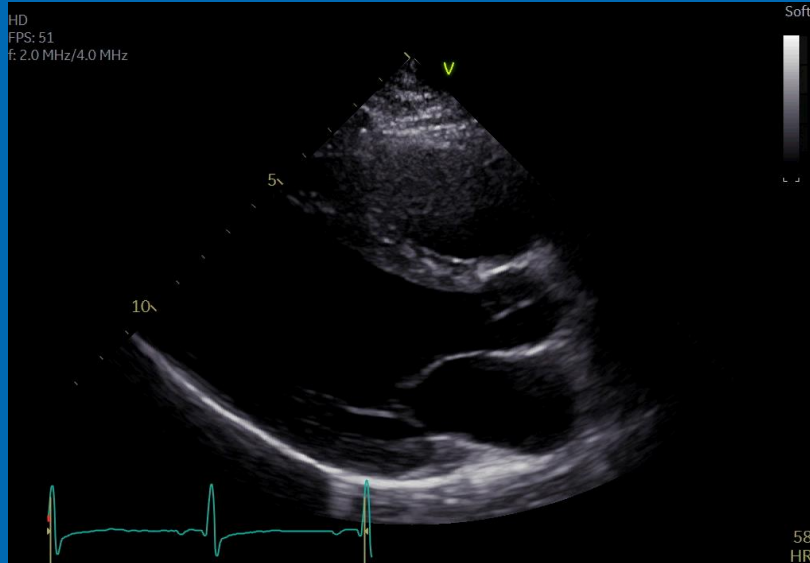
Outline

- Heart sounds physiology
- How to describe murmurs
- Qualities of innocent versus pathologic murmurs
- When to refer and referral urgency
- Cases



Physiology review: what produces heart sounds?

- S1: atrioventricular valves, T1/M1
- S2: semilunar valves, A2/P2

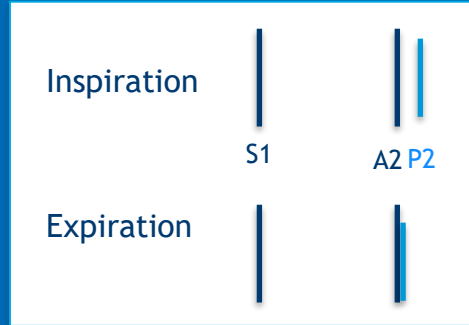


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Second heart sound (A2/P2)

Physiologic (normal) splitting

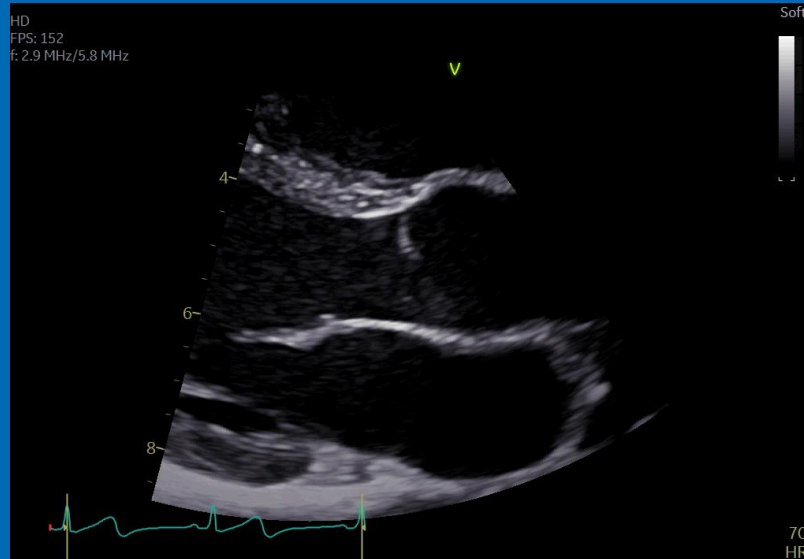


Fixed splitting (ASD, PAPVR)



Early systolic click

- Remember: heart sounds occur when valves close
- If a semilunar valve domes in systole, this results in an early systolic click as the valve “snags” in its partially-open position



Gallops

- Low-pitched, best heard using bell at the apex
- S3: often a normal finding in children and up to age 35-40, due to good LV compliance



- S4: considered abnormal in pediatrics, suggestive of poor ventricular compliance



What causes a murmur?

1. Increased flow across normal structures
2. Obstructed flow across abnormal structures
3. Regurgitant flow across incompetent valves
4. Turbulent flow of blood from one chamber/vessel into another (shunts)*



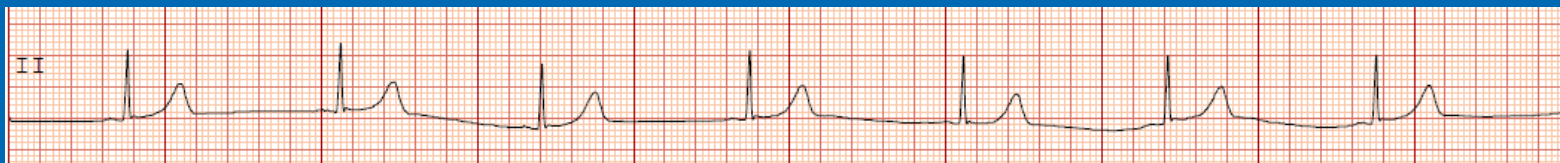
Describing murmurs

- Timing
- Grade
- Pitch
- Quality
- Location
- Radiation
- +/- *symptomatic?*

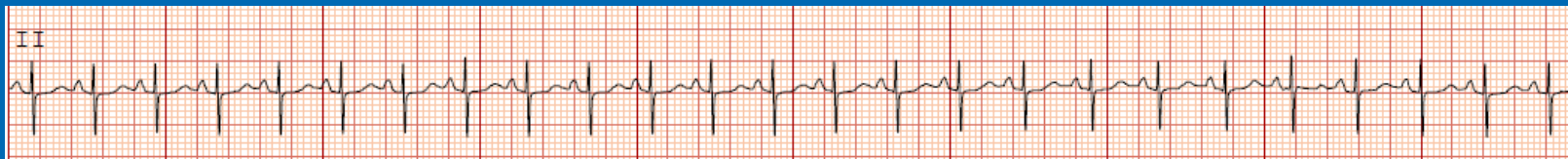


Timing - cardiac cycle

- When does it occur?
 - Systolic - Diastolic - Continuous
- How can I tell the difference?



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Clinical tip: palpate apical impulse while auscultating (practice on older kids with slower rates)



Grade

Systolic: I-VI

I: softer than S1/S2

II: same volume as S1/S2

III: louder than S1/S2

IV: heard with stethoscope on chest

V: heard with stethoscope partially off chest

VI: heard with stethoscope off chest



No thrill

+thrill



Grade

Diastolic: I-IV

I: softer than S1/S2

II: same volume as S1/S2

III: louder than S1/S2

IV: loud, +thrill



No thrill

+thrill



Pitch & Quality

- Low, medium, high-pitched
 - Lower frequencies better heard by bell
 - Higher frequencies better heard by diaphragm
- Quality
 - Musical
 - Vibratory
 - Harsh
 - Blowing
 - Crescendo-decrescendo
 - Holosystolic



What does “holosystolic” mean anyway?

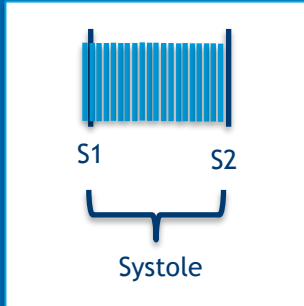


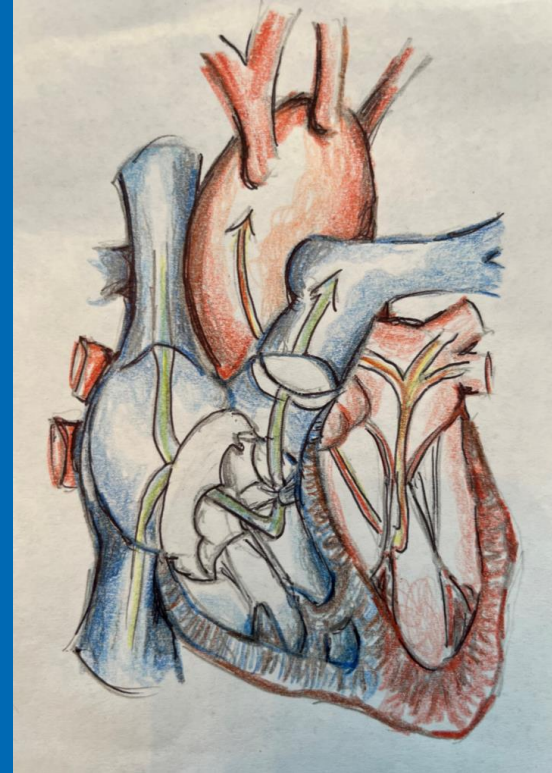
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Holosystolic
(S1 obscured)



Systolic ejection
(S1 audible)

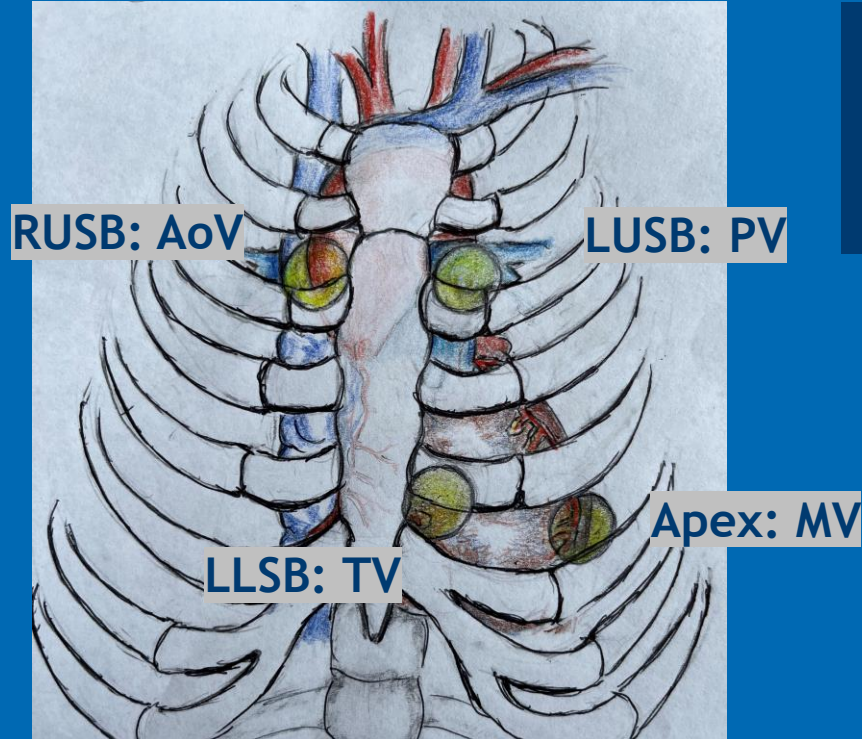


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Location: cardiac auscultation review

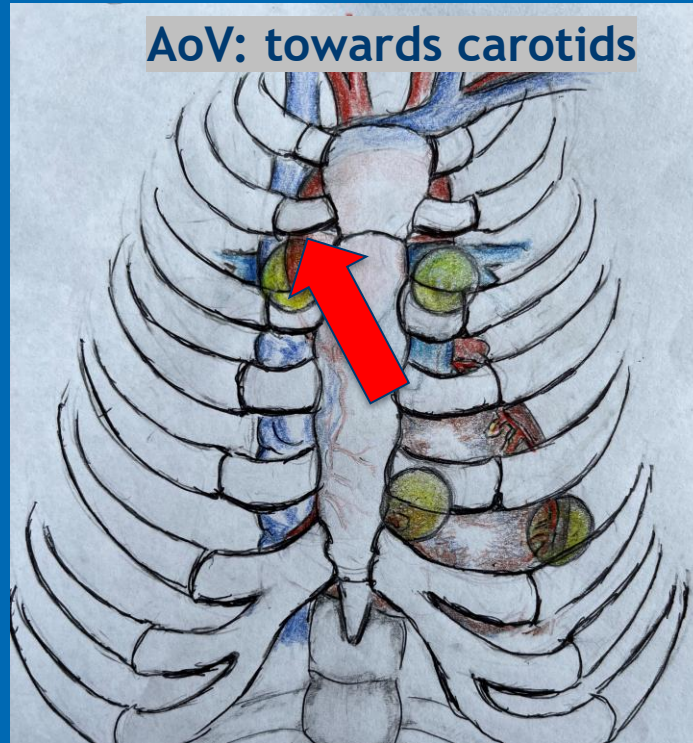


Don't forget to auscultate with your diaphragm AND the bell!

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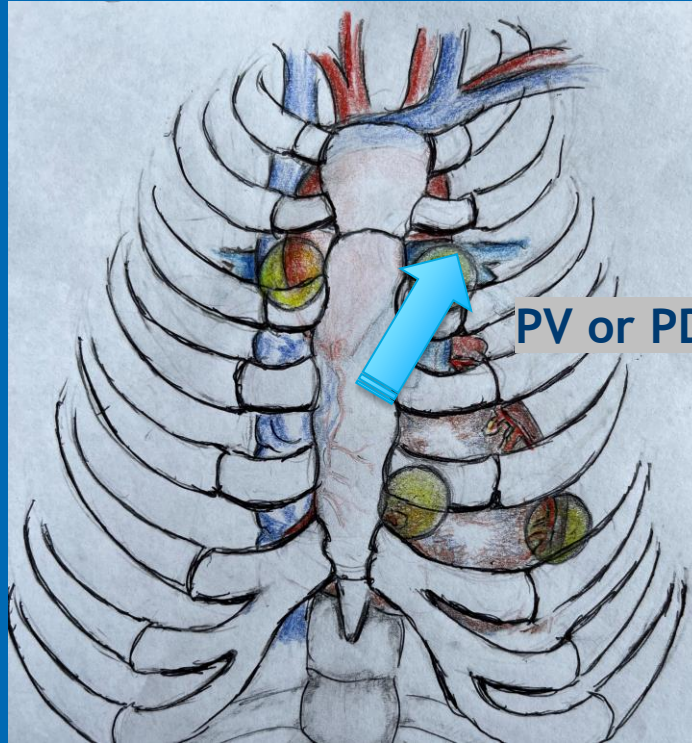
Radiation



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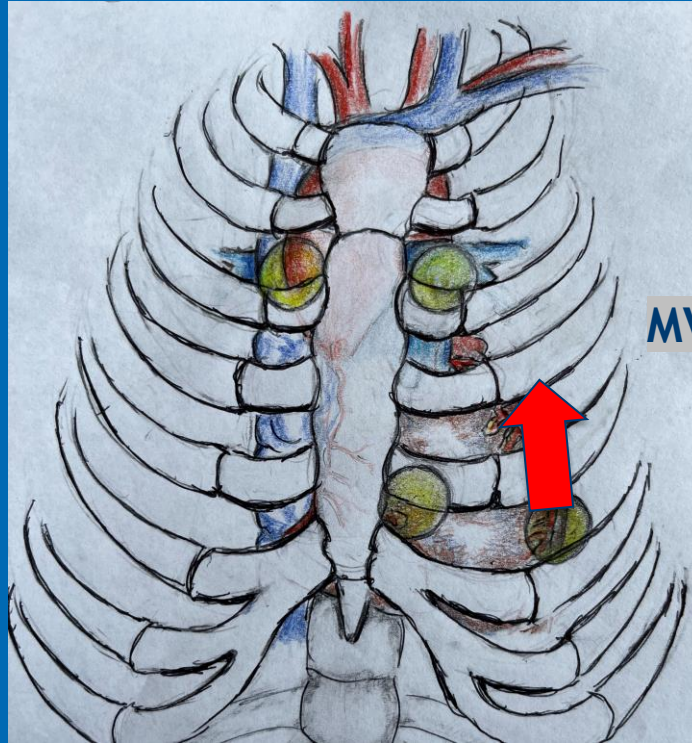
Radiation



PV or PDA: towards back



Radiation

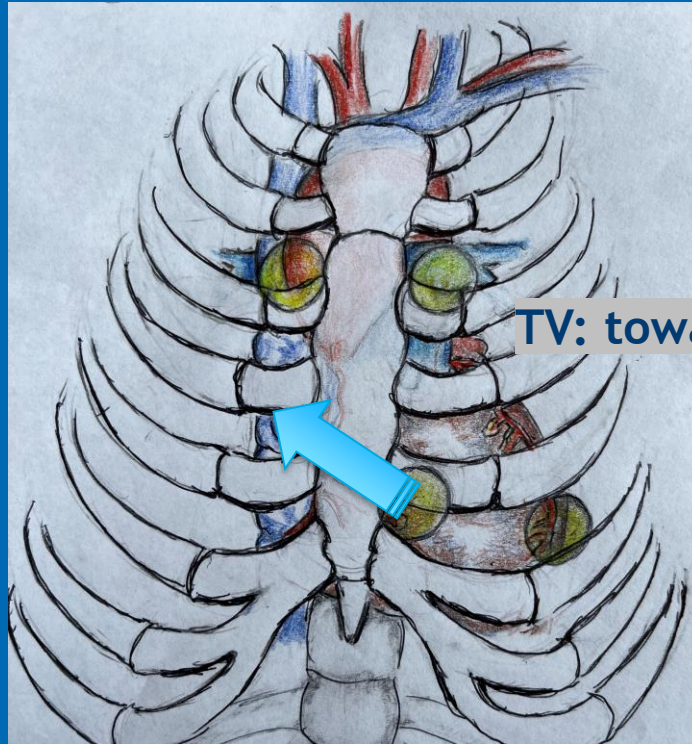


MV: towards left axilla

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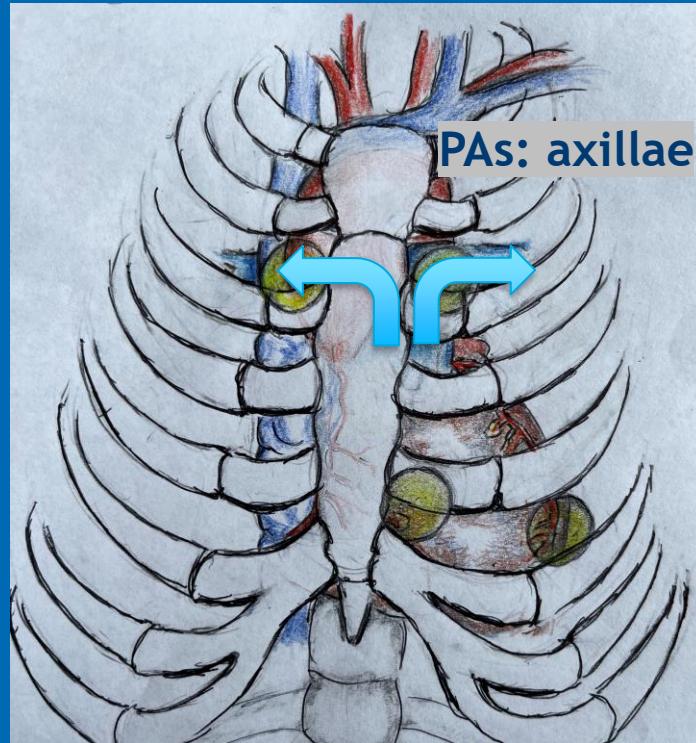
Radiation



TV: towards RLSB



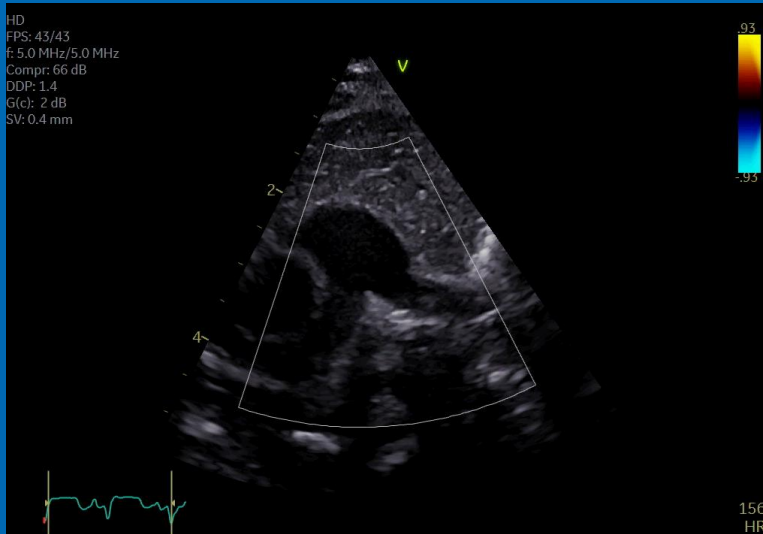
Radiation



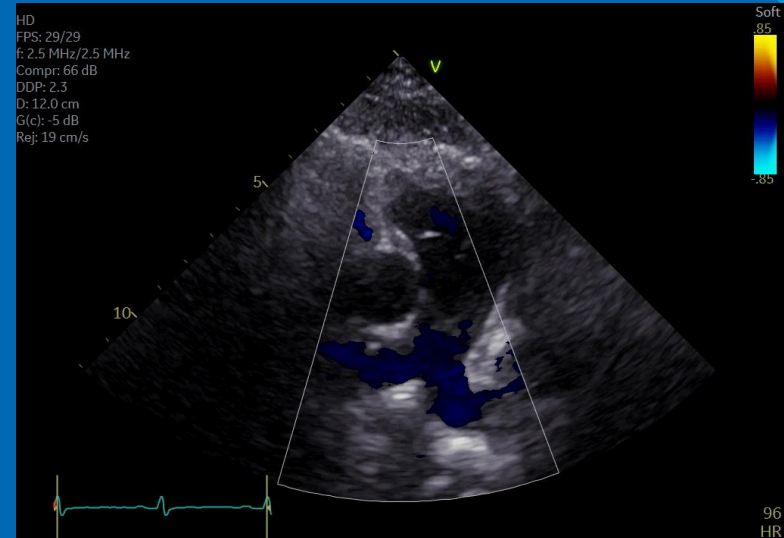
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Peripheral pulmonic stenosis



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- Flow acceleration in normal-sized pulmonary arteries
- Physiologic up to 6 months of age

- Less acute take-off angle of PAs as AP diameter of chest increases



Clinical tip: if you hear something, auscultate in all positions!

- Supine
- *(Sitting)*
- Standing
- Squat to stand



Qualities of innocent versus pathologic murmurs

Innocent	Pathologic
<ul style="list-style-type: none">• Soft• Vibratory/musical• Limited to systole• Loudest when supine, decreases with sitting/standing	<ul style="list-style-type: none">• Harsh or high-pitched<ul style="list-style-type: none">• Associated thrill• Associated click or gallop<ul style="list-style-type: none">• Diastolic*• Holosystolic



To refer or not to refer?

- Cardiology is always happy to lend a second set of ears
- How urgent of a referral / how worried should you be?
- Good news - it is an easier answer than you think!



If you listened to 100 babies with structurally normal hearts, how many would have murmurs?

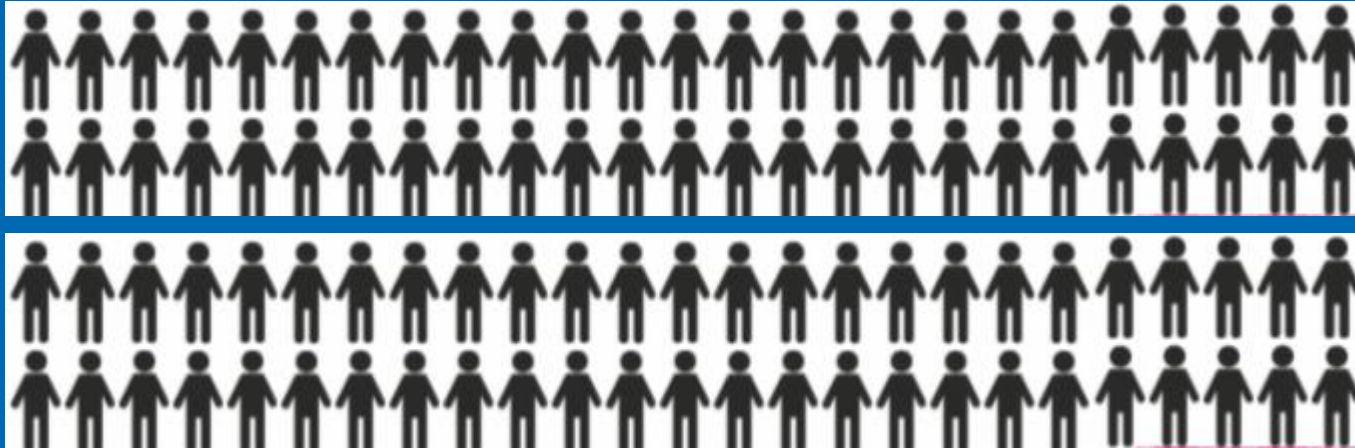


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Up to 30% may have innocent murmurs



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If you listened to 100 babies with critical CHD, how many would have murmurs?

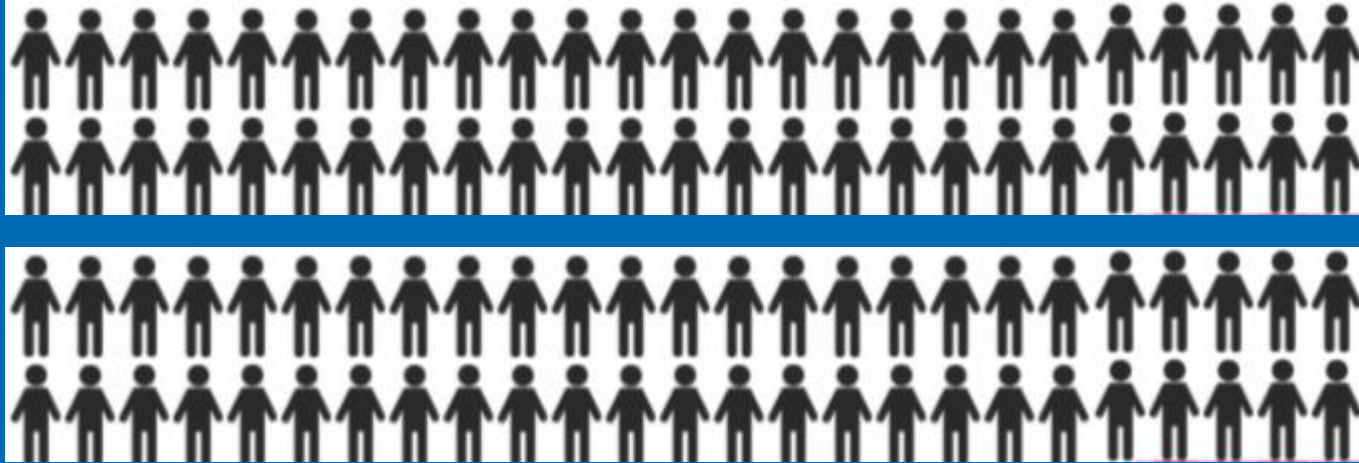


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Only half!



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Two important truths to consider:

- 1) The majority of newborn murmurs are innocent
 - * *1/3 to 3/4 of children have an innocent murmur auscultated at some point in their life*
- 2) About half of newborns with critical congenital heart disease DON'T have murmurs



Symptomatic vs asymptomatic



- 1) Cyanosis (mucous membrane color change, check SpO₂)
- 2) Hypoperfusion (brachial and femoral pulses, UOP, BPs)
- 3) FTT/increased WOB (weight curve, feeding tolerance, respiratory exam)

In school-aged children and teens who are outside the window of missed critical CHD, consider mid-exertional symptoms including chest pain, syncope, or palpitations



To refer or not to refer?

- How urgent of a referral / how worried should you be?
- Are they symptomatic of critical CHD? Cyanosis, hypoperfusion, FTT/increased WOB? Mid-exertional symptoms?
- 9-24 months can be a difficult time for physical exams or additional studies



Case 1

16yo M with newly auscultated murmur, needs sports clearance before starting cross country. Born on time, no prenatal or perinatal concerns, good growth, met developmental milestones on time. No exertional complaints, though relatively sedentary.

No FHx of congenital heart disease, arrhythmias, sudden cardiac or unexplained death, cardiomyopathy, pacemakers placed < 35 yoa, no early MI/stroke (< 55yo for males, < 65yo for females). Father with murmur as a child that he “grew out of”



Case 1

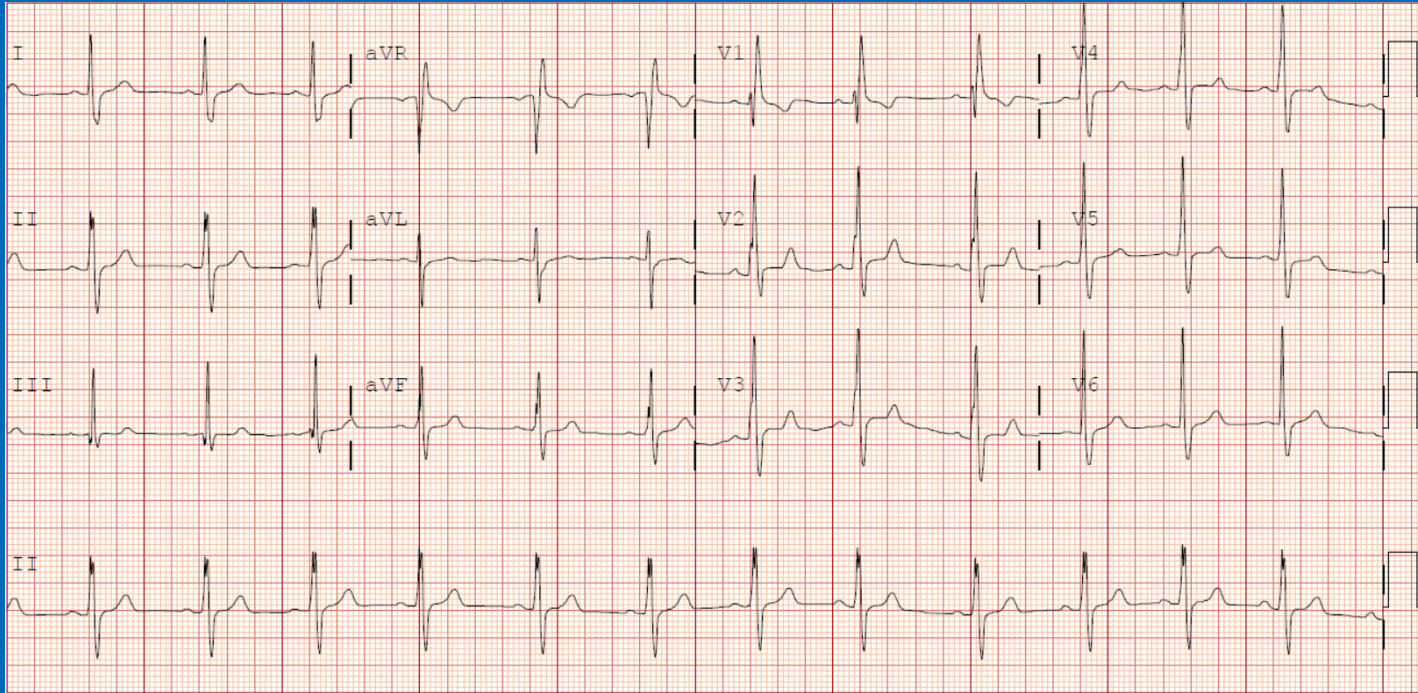
CV: RRR, quiet precordium. 2/6 medium intensity systolic ejection murmur best heard at LUSB. Diastolic murmur. Widened split S2. 2+ pulses without brachiofemoral delay. Otherwise unremarkable exam (lungs CTA, comfortable WOB)



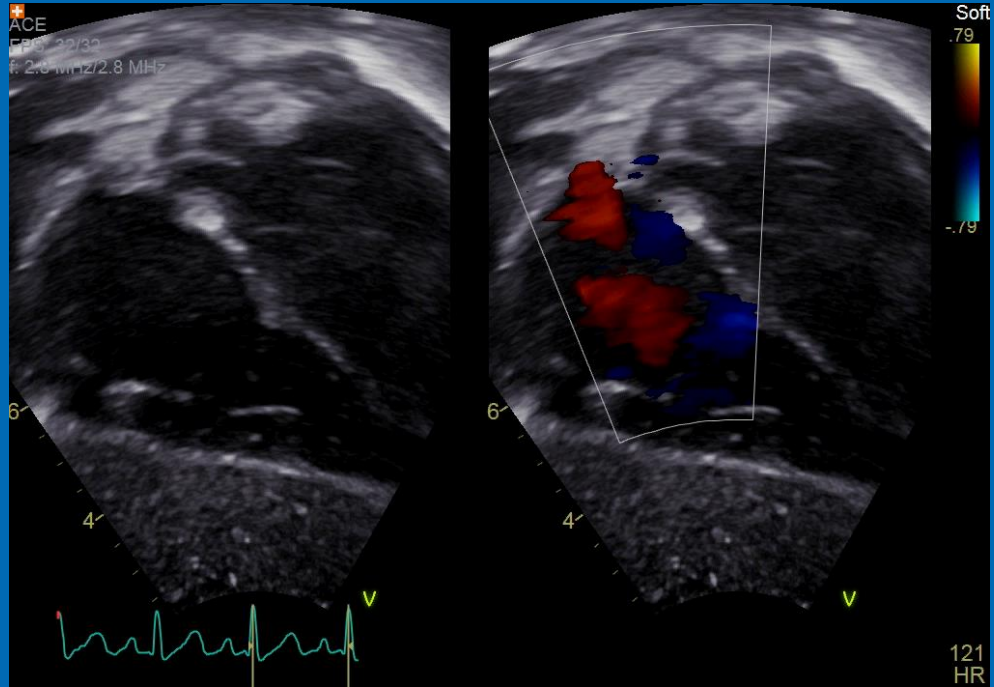
No clinical red flags (no cyanosis, hypoperfusion, FTT or exertional symptoms), but murmur doesn't sound benign. Non-urgent referral is reasonable



Case 1: OP cardiology referral



Case 1: echo



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Case 1: heart sounds

- What's causing the murmur?
“Relative pulmonic stenosis”
- Fixed split S2 strikes again!

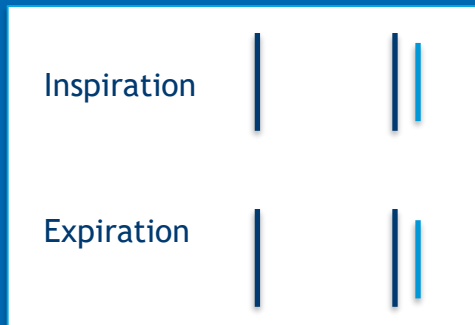
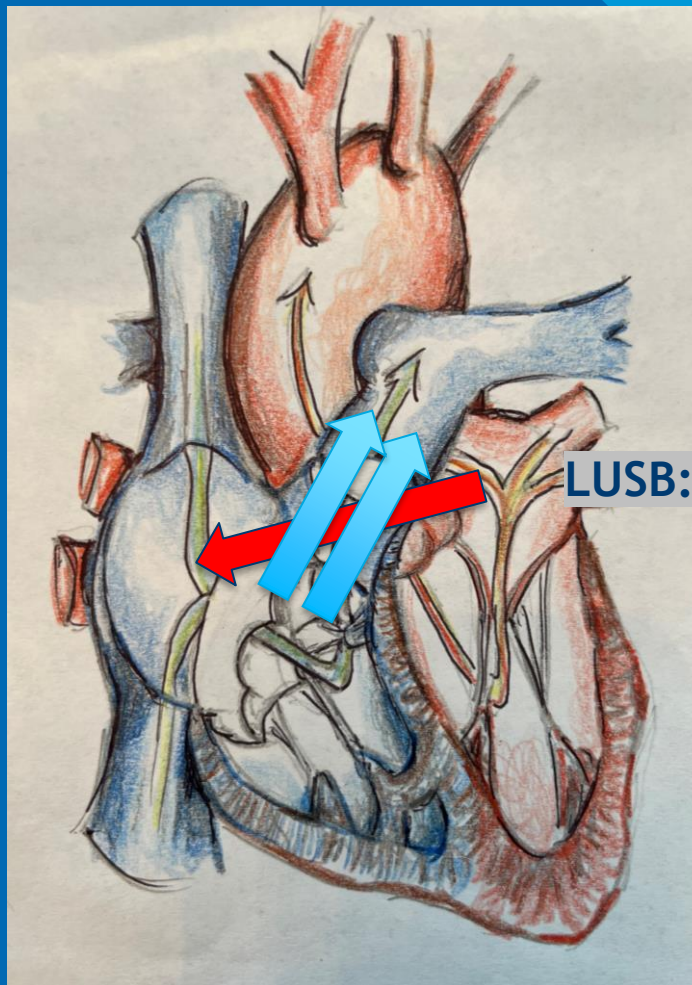


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- Clinical tip: in cooperative kids, have them exhale out and hold breath to see if S2 comes together



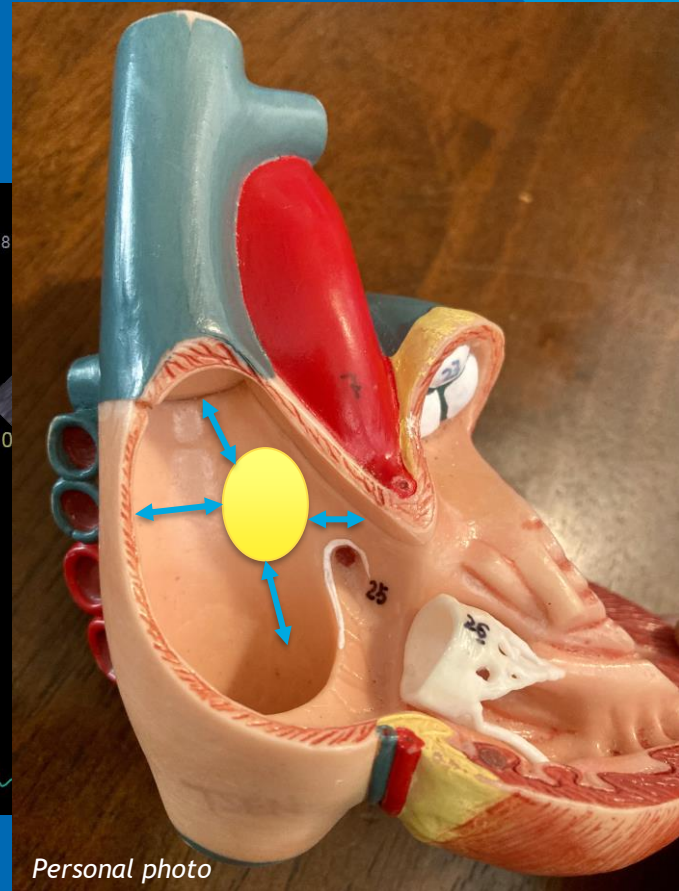
LUSB: PV

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Case 1: secundum ASD

- Left to right shunt leads to right heart volume-loading
- Right heart dilation generally well-tolerated (years to decades)
- Chronic right heart volume-loading may lead to pulmonary vascular disease (3rd, 4th, 5th decade of life)
- Usually closed ~4-5 years of age (percutaneously), or when discovered, if older/adequate weight and hemodynamically significant



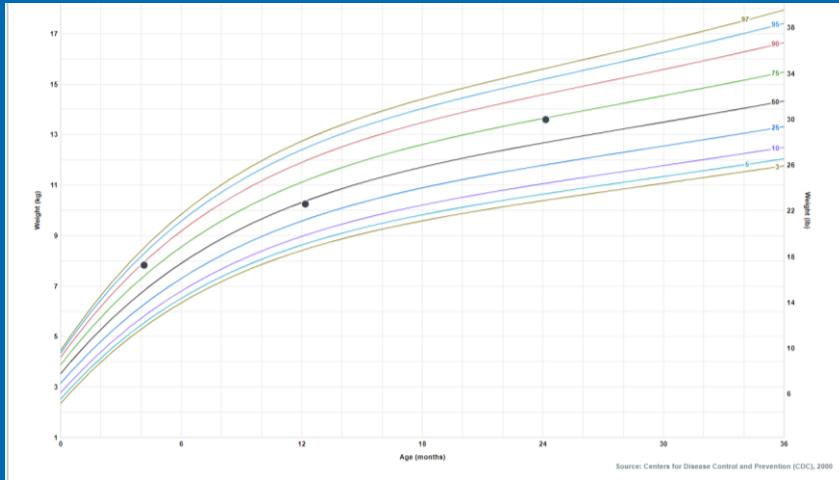
ACE
FPS: 106/
f: 2.9 MHz/5.8

10

120
HR



Case 1: what if this were discovered at 6 months of age?



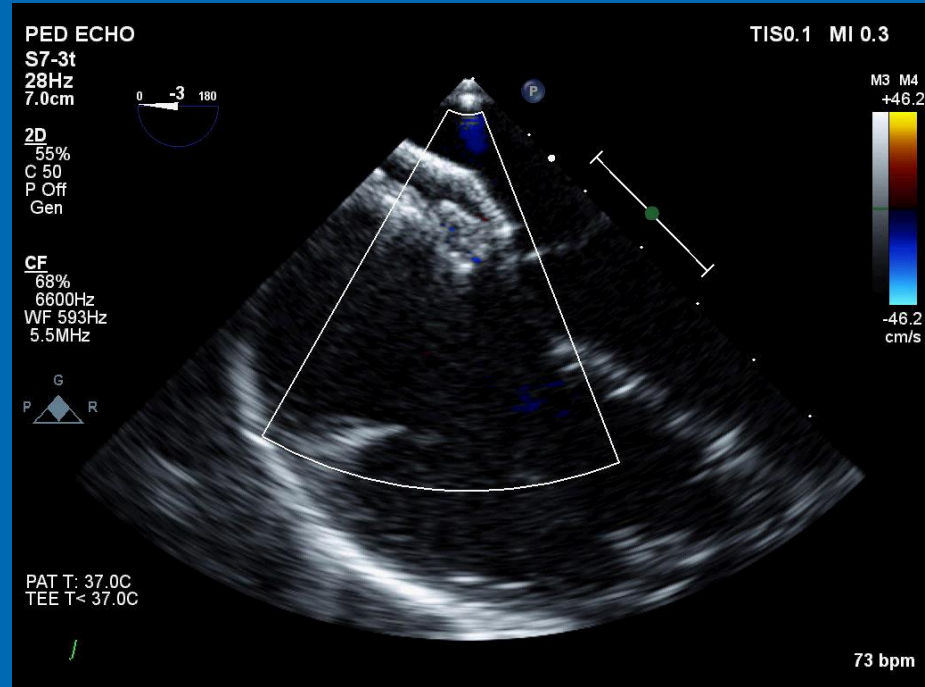
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- Expectant management, unless at risk for developing pulmonary arterial hypertension (e.g. T21, ex-premie/CLD)
- Small and moderate-sized defects have a ~60% chance of spontaneously resolving, or not requiring intervention with somatic growth and time (Hanslik et al)
- Murphy et al: if operated on before 25 years of age, survival is similar to age-matched controls



Case 1: ASD management

- Referred to cath lab for non-urgent ASD device closure
- Overnight stay, discharged on ASA and SBE prophylaxis x 6 months (minimum)
- Restricted from heavy lifting, contact sports, strenuous activity x 2 weeks



What about PFOs?

- Incidence in general population autopsies ~25%
- Rare potential complications from possible right to left (paradoxical) embolism
- Primary (preventative) closure not recommended in children
- Indications for closure (e.g. cryptogenic stroke, migraine) on a case-by-case basis with neurology and cardiology

Table 2

Summary of current evidence in relation to the management of patent foramen ovale in children

	Evidence
Incidental PFO diagnosis	
Counseling and follow up guidelines	None
Diagnostic modality of choice	
TEE versus TTE versus TCD	Weak
PFO closure indication	
PFO and cryptogenic stroke	Weak
PFO and deep diving	Weak
PFO and migraine	Weak
PFO and pregnancy	None
Risk stratification	
PFO and associated special situations	Weak/none

PFO: Patent foramen ovale, TCD: Transcranial Doppler, TTE: Transthoracic echocardiography

Saharan et al



Case 2

1 month old (CGA 33 weeks) ex-29 week gestation F in NICU with RDS, slow progression towards extubation. Poor somatic growth despite adequate caloric intake



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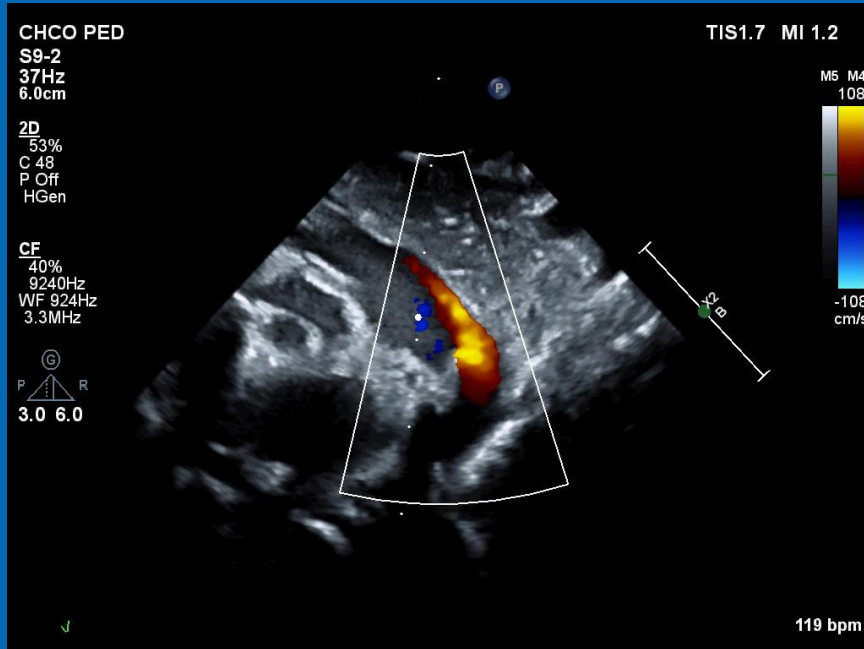
Case 2

Exam:

Resp: intubated, coarse lung sounds, hyperinflated chest, hyperresonance
CV: RRR, quiet precordium. 2/6 systolic murmur best heard at LUSB. Diastole inaudible. Femoral pulses
Abd: liver 1cm below the RCM
Ext: warm, well-perfused, no cyanosis



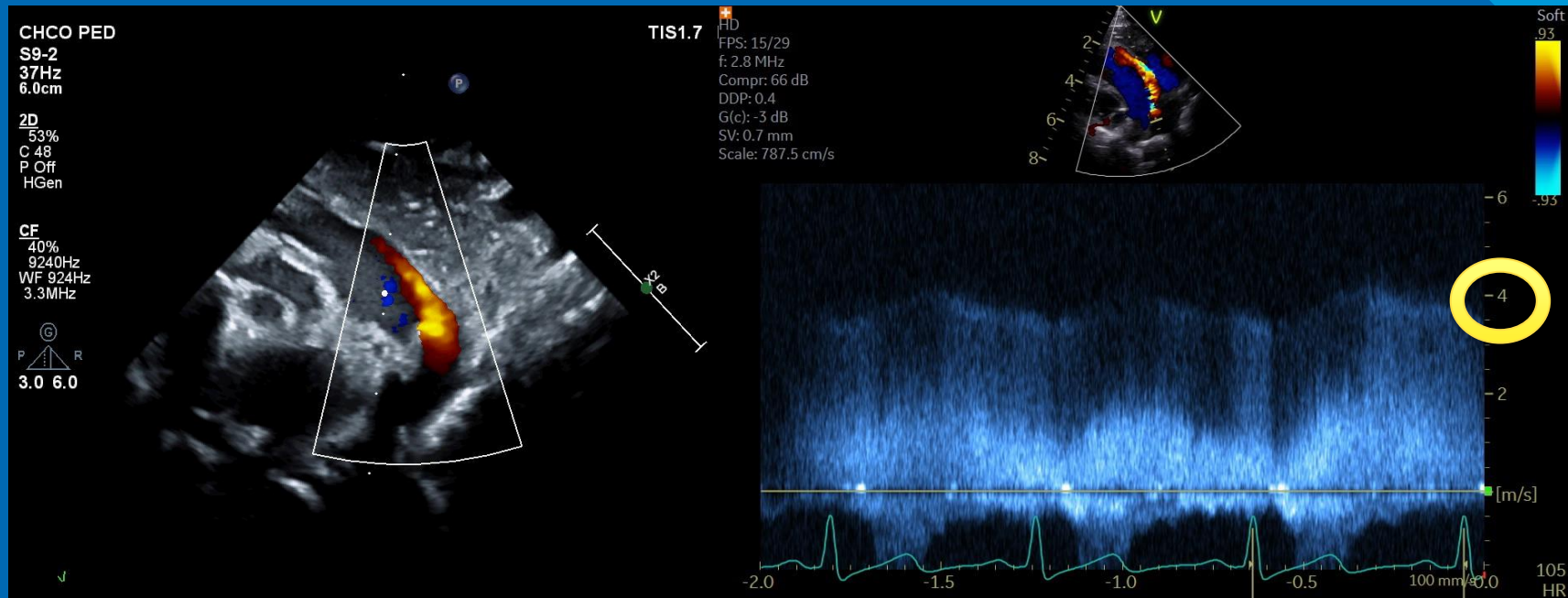
Case 2: echo ordered in NICU



- Large PDA with left to right shunt
- Excessive pulmonary blood flow → pulmonary edema → inability to wean respiratory support
- Wide pulse pressure - why?
- Pressure and volume-load to lungs
- What about a “continuous machine-like murmur”??

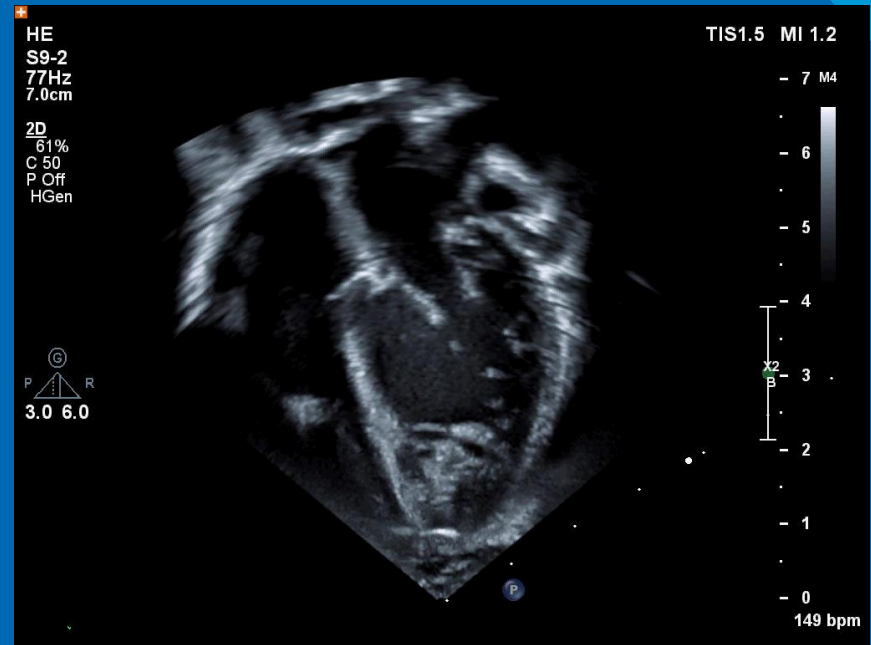


Case 2: why wasn't there a continuous murmur?



Case 2: PDA management

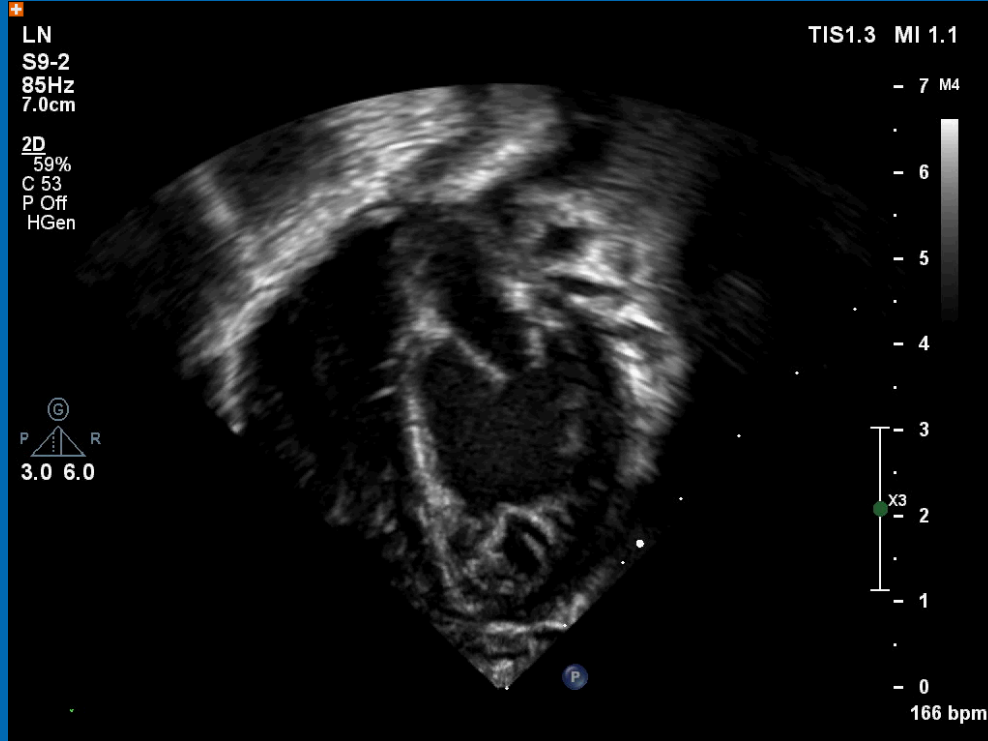
- “Hemodynamically significant shunt” definition varies: echocardiographic and clinical evidence
- Causation of associated premie morbidities (IVH, NEC) not proven
- Many studies leave management a gray area for preterm infants
- Medical therapy, then referral for transcatheter device if closure deemed necessary
- In term patients, no utility in medications. Watchful waiting as long as growing well with normal exercise tolerance



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Case 2: post-procedural LV “dysfunction”



Case 3

Otherwise healthy, ex-term 5yo M who just moved and is establishing care at your practice. Medical history is notable for “a murmur they’ve heard his whole life”. Pregnancy and birth history are unremarkable. He plays peewee soccer and keeps up with teammates, running around with excellent energy, no complaints of exertional chest pain, dizziness, or syncope. He has a good appetite and is growing well

FHx negative for congenital heart disease, arrhythmias, early MI/stroke, sudden cardiac death



Case 3

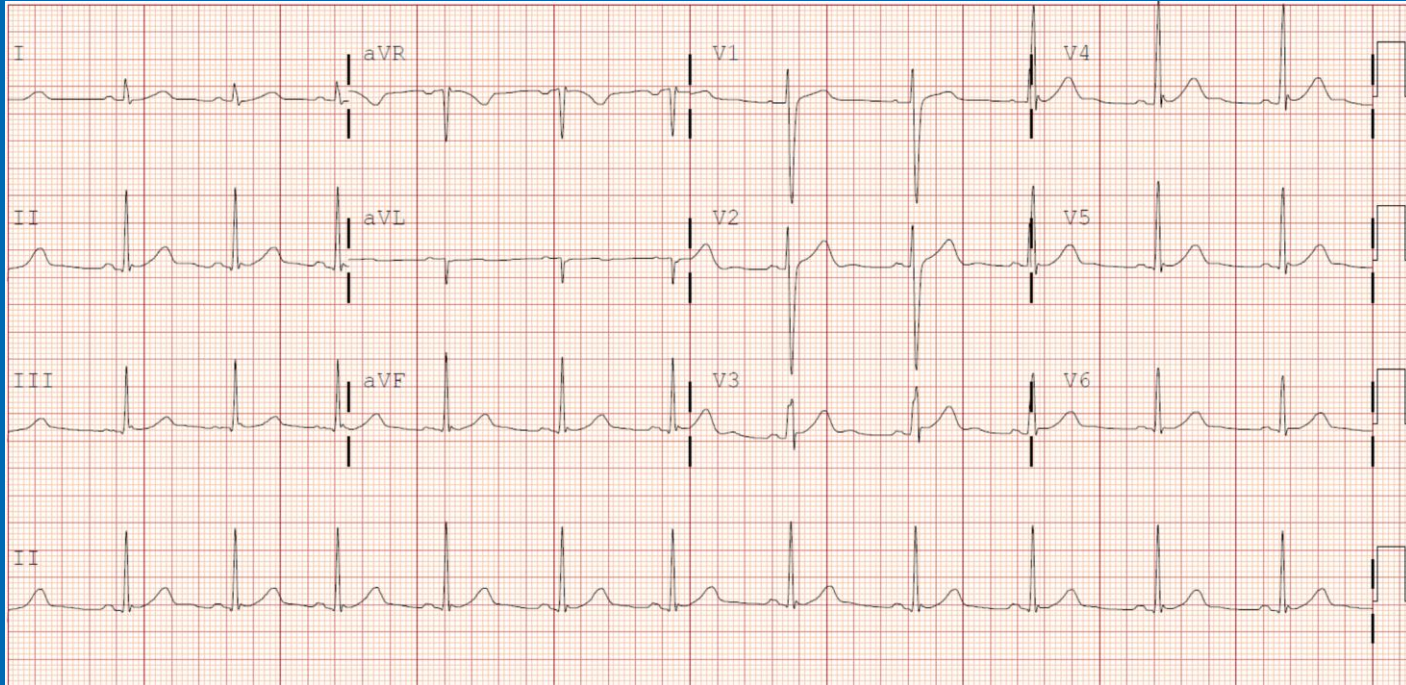
CV: RRR, quiet precordium, normal S1, low-pitched
(?vibratory?) systolic murmur louder while
standing? Diastole is silent. 2+ brachiofemoral delay
Otherwise unremarkable exam (lungs CTA,
comfortable WOB)



No clinical red flags (no cyanosis, hypoperfusion, FTT or exertional symptoms), but murmur may not be benign. Non-urgent referral is reasonable

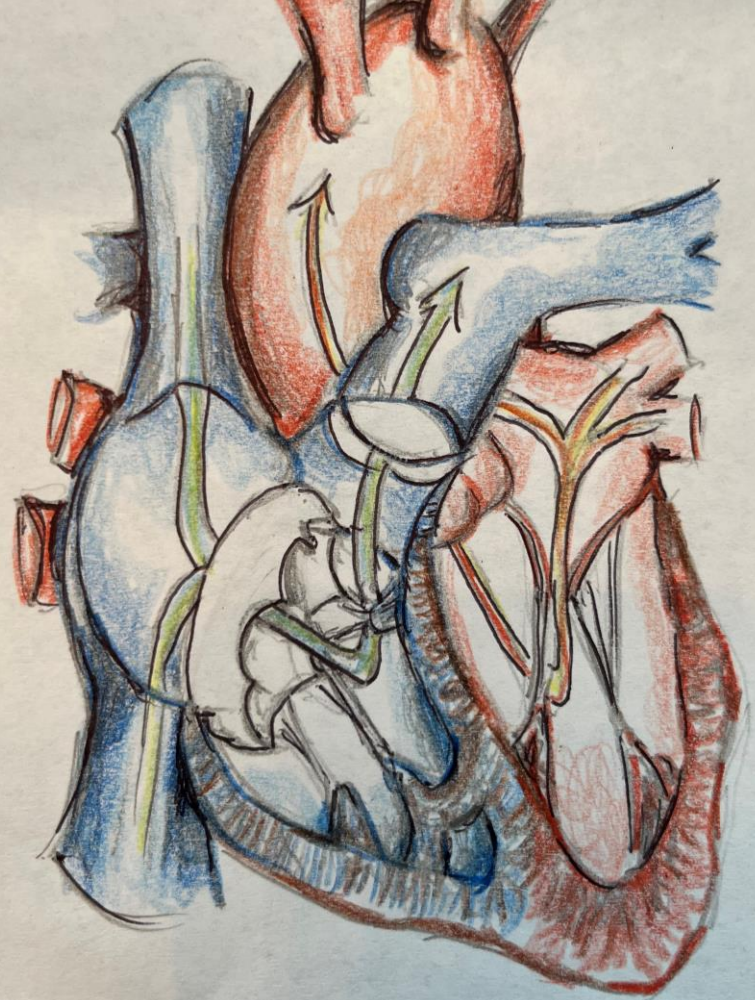


Case 3: OP cardiology referral



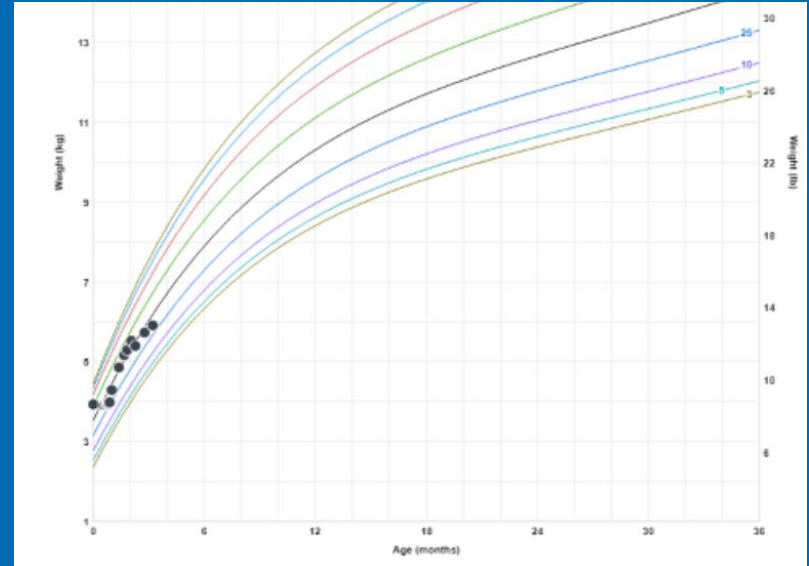
Case 3: murmur sounds classically Still's

- Loudest while supine,
softer/disappears when standing
- Sometimes we echo, sometimes
we don't
- How to counsel families?



Case 4

2mo ex-term baby presents to your office for tachypnea and increased work of breathing. No c/f infection. Previously referred to and seen by ENT for suprasternal retractions, flex scope normal. No pregnancy or peripartum complications, passed CCHD screen, previously was growing well. In the last month “hasn’t been eating”, taking over an hour to finish a 2-oz bottle due to taking breaks. Feels clammy and breathes hard while trying to eat.



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Case 4

GEN: thinner baby in mild respiratory distress

HEENT: no congestion or rhinorrhea

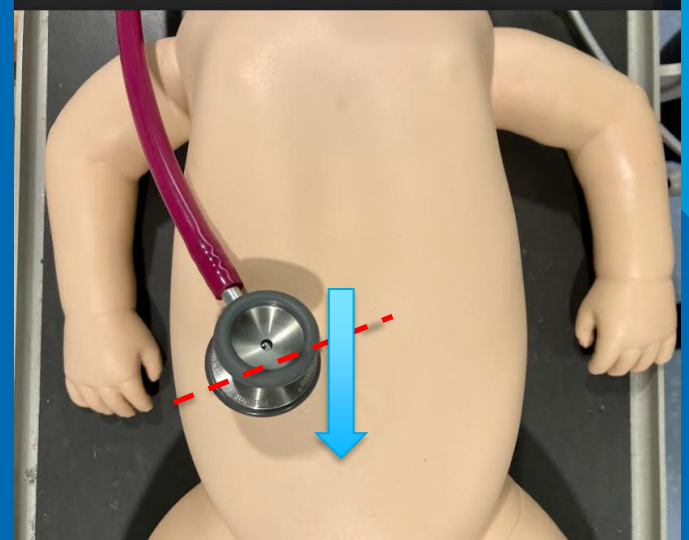
RESP: SpO2 88%, tachypnea, tachycardia, TA

CV: RRR, quiet precordial or diastolic
murmur heard. 2+ p femoral delay

ABD: liver edge 3cm

EXT: slightly cool, decreased cap refill 3 seconds, mild mottling

Feeding intolerance
WOB



Personal photo



Case 4: sent to ED for further work-up



- Placed on LFNC, O2 increased to 95%
- Pre/postductal SpO2 without split
- 4-extremity BPs without significant differential
- RVP negative
- CMP, VBG, lactate normal
- CBC: H/H 20/56
- ESR, CRP normal



Case 4: echo ordered

- Large perimembranous VSD with left to right shunting and significant left heart dilation
- No murmur heard because the defect is very large (no flow turbulence)
- If the defect began to get smaller, may start hearing a holosystolic murmur

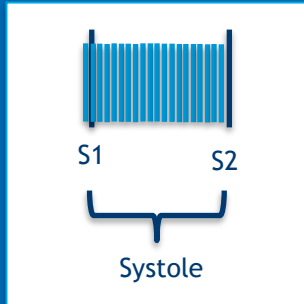
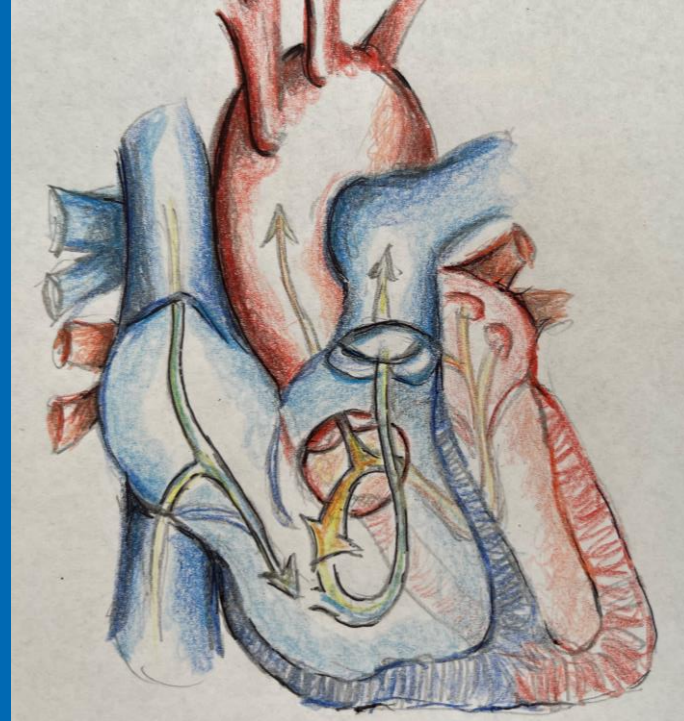


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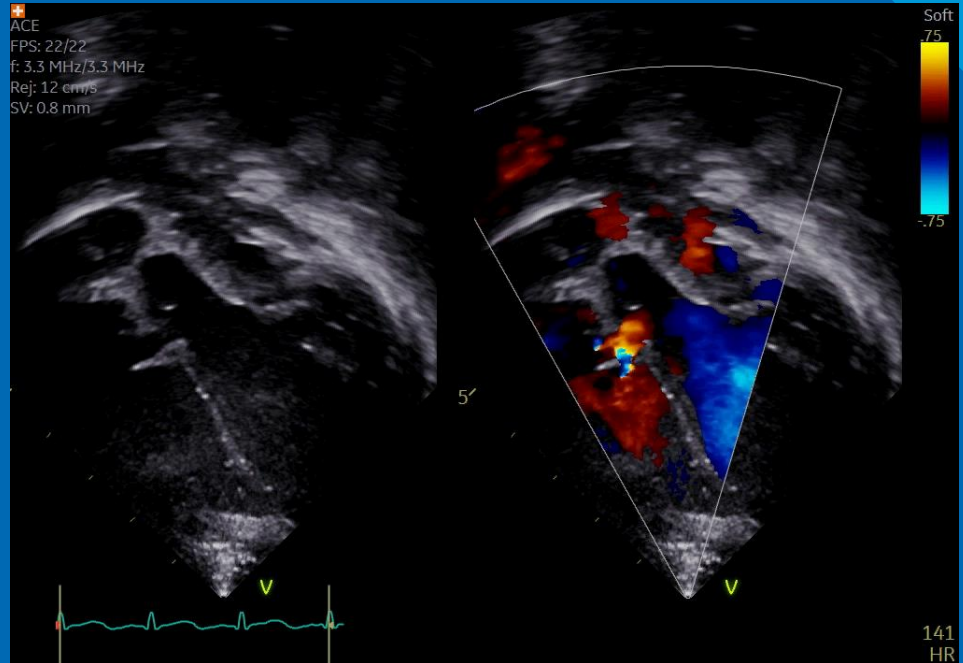


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Case 4: VSDs

- “Blood follows the path of least resistance”
- Excessive pulmonary blood flow leads to tachypnea
- Breathing > eating
- Natural history: dependent on muscular versus other locations; size
- Clinical tip: “Cold and sweaty” versus warm and sweaty

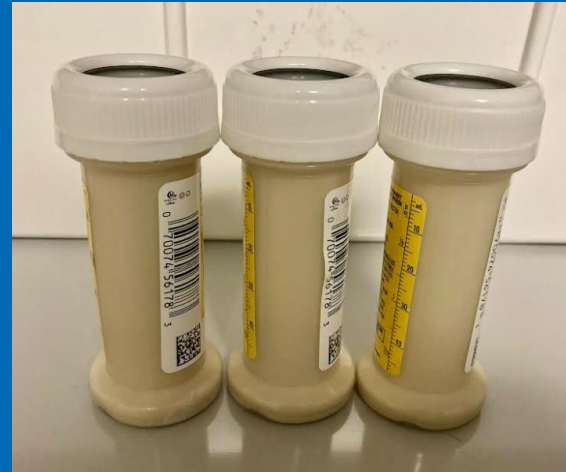


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Case 4: VSD management

- Diuretics, fortified feeds
- If hemodynamically significant defect persists, will repair around 4-6 months of age to prevent pulmonary vascular disease (pressure and volume load on lungs)
- Most are performed surgically
- May see post-operative LV “dysfunction” similar to PDA closure, 2/2 abrupt volume unloading of LV



Personal photo



Take-home points

- Do your best to describe the murmur, recalling characteristics of innocent versus pathologic murmurs
- Don't put too much pressure on your ears to absolutely identify innocent versus pathologic - how is the kid doing? (Sometimes there is heart disease but NO murmur!)
- Consider red flag symptoms of critical CHD when determining urgency of referral: cyanosis, hypoperfusion, FTT/increased WOB, and in older kids, mid-exertional symptoms

A small icon of a red flag on a black pole, positioned to the left of the third bullet point.

Cardiology is always available to answer questions
and evaluate patients with murmurs!





Thank you! Questions?

Personal photo

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