



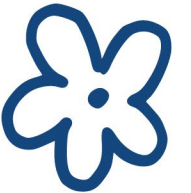
Pediatric Cardiology issues for the PCP in 2026

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No disclosures



Agenda

1

Review murmurs and when referral to a pediatric cardiologist is indicated

2

Review chest pain

3

Review Dizziness/syncope/POTS

4

Understand the better outcomes in children with complex congenital heart disease and important pearls to know when seeing these patients in your practice.



Murmurs

- The most frequent cause for referral to a pediatric cardiologist.
- Most children will have a murmur at some point in their life.
- Most murmurs are transient and of no clinical concern.
- Despite the above many PCPs lack the experience and confidence to distinguish innocent from pathologic.
- There is also the cultural anxiety of “missing” something that haunts us all.





Murmur

- Produced by perturbation in blood flow usually due to pressure gradients or velocity changes which is then transmitted through the chest wall.
- Most murmurs are innocent; however, 37% of murmurs in infants are diagnosed with congenital heart disease and 2.4% of infants with a murmur have a critical lesion.
- After infancy, most murmurs are innocent and usually disappear by adulthood.





Murmurs in Infants

- OB and Family history are very important
- Maternal exposure to teratogenic medications (lithium, cellcept, ACE inhibitors, retinoic acid or vitamin A derivatives)
- Maternal exposure to NSAIDS during the third trimester is associated with premature closure of the ductus arteriosus which can then result in fetal hydrops, PPHN, and RV failure.
- Maternal Diabetes Mellitus (especially preexisting)
- Maternal Obesity



Murmurs beyond infancy

- Family history is very important (CHD, genetic syndromes)
- Genetic syndromes (Trisomy 21, Williams syndrome, Turners syndrome, Marfans....)
- Signs and symptoms are very important and can separate out the innocent vs pathologic murmurs





Physical Exam in the infant

- VS and general exam are very important (other minor anomalies)
- 3 or more minor anomalies have a 90% chance of an underlying major anomaly, including structural heart disease
- CV exam focuses on any evidence of cyanosis, palpation of brachial and femoral pulses, precordial activity, abdominal exam for HSM and auscultation.

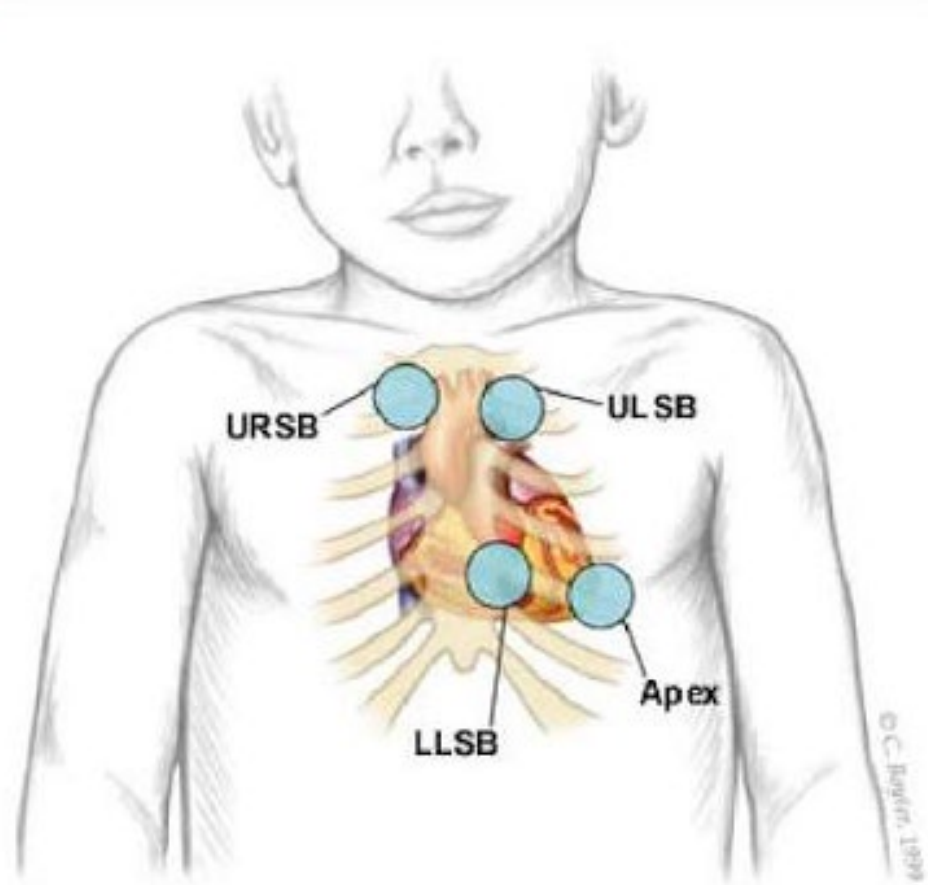


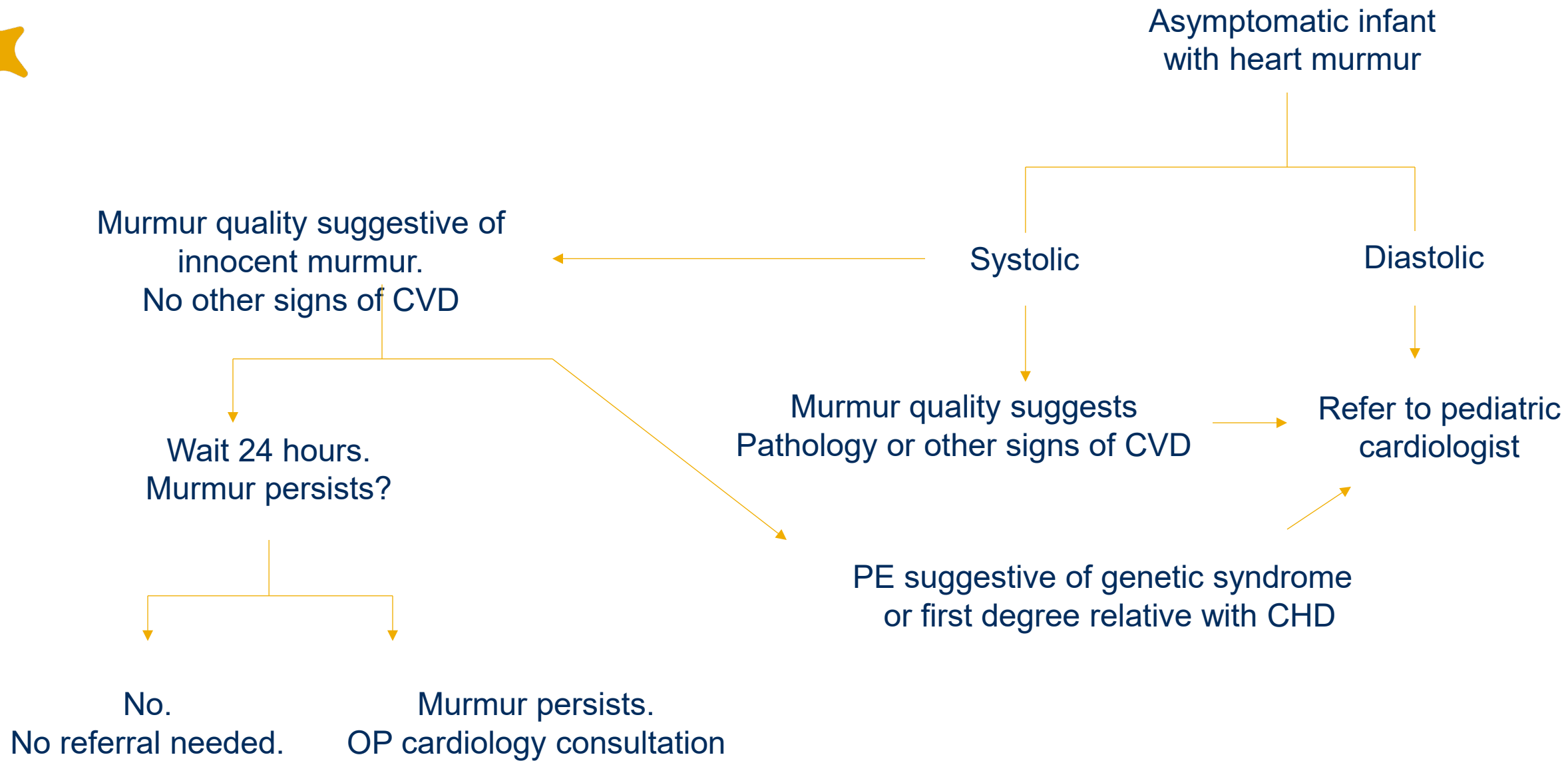


Critical CHD screen

- Positive - The oxygen saturation is below 90% in the right hand OR feet during 3 consecutive measurements separated by 1 hour.
- Positive - The oxygen saturation is below 95% and above 90% in the right hand AND feet OR more that a 3% difference between the right hand and feet saturation during 3 consecutive measurements separated by 1 hour.
- Negative - The oxygen saturation is above 95% in the right hand OR feet AND less than a 3% difference between the right hand and feet saturation at any point during 3 consecutive measurements separated by 1 hour.









Physical exam in the older child

- In older children the probability of a murmur being the first sign of congenital heart disease is low.
- In addition to all the steps taken in examining the infant the PE in older child should involve listening in multiple positions as well as dynamic testing.





Dynamic Testing of Heart Murmurs and Physiologic Correlates

Dynamic maneuvers	Physiologic effect	Murmurs expected to increase	Murmurs expected to decrease
Systolic murmurs			
Valsalva or Stand from squat or Forced heavy expiration	Decrease left ventricular preload	Hypertrophic obstructive cardiomyopathy (likelihood ratio = 6 to 14 for these maneuvers for diagnosis) Mitral valve prolapse	Aortic stenosis Mitral regurgitation Innocent murmurs
Squat or passive leg raises	Increase left ventricular preload	Most murmurs increase to include mitral regurgitation and aortic stenosis	Hypertrophic obstructive cardiomyopathy Mitral valve prolapse
Hand grip or Transient arterial occlusion (usually by blood pressure cuff)	Increase afterload	Mitral regurgitation Pulmonic stenosis Ventricular septal defects	Hypertrophic obstructive cardiomyopathy Aortic stenosis Mitral valve prolapse
Deep or exaggerated inspiration	Increased venous return; increase in left ventricular afterload	Tricuspid regurgitation Pulmonic stenosis	No effect on other murmurs
Diastolic murmurs			
Deep or exaggerated inspiration	Increased venous return; increase in left ventricular afterload	Tricuspid stenosis Pulmonary regurgitation (slight effect)	No effect on other murmurs
Hand grip or Transient arterial occlusion (usually by blood pressure cuff)	Increase afterload	Aortic regurgitation Mitral stenosis (slight increase or neutral) Ventricular septal defects	
Continuous murmurs			
Compression of the jugular veins or Movement from sitting to supine	Decrease in jugular venous return to the superior vena cava	Not applicable	Venous hum





Common innocent murmurs in children

- Still's murmur
- Pulmonary ejection/flow murmur
- Venous hum





Abnormal heart sounds

- Fixed split S2 – ASD
- S3 – conditions with ventricular dysfunction yet can also be a normal finding
- S4 – conditions with decreased ventricular compliance (cardiomyopathies, heart failure) AKA – gallop
- Thrills – VSDs (small), aortic and pulmonary valve stenosis
- Clicks – usually with aortic or pulmonic stenosis. Mid-systolic click is associated with MVP.





New murmur in an asymptomatic 2–18-year-old



Diastolic murmur?
Grade 3/6?
Abnormal heart sounds?
Click or Rub?



No.
F/u at next health maintenance visit.
Murmur still heard?

Yes.
Refer to Pediatric Cardiologist



No.
Reassurance.

Yes.
Continue to reassure vs referral.





Chest Pain

- One of the most common reasons for referral to a pediatric cardiologist.
- Often driven by parental anxiety due to the high morbidity and mortality associated with CP in the adult population.
- A review of a decade of pediatric cardiology chest pain referrals at Boston Children's Hospital (approximately 18,000 patient years) revealed that only 1% (n = 3700) of the patients had a cardiac etiology for their chest pain and that no patient discharged from the clinic with a diagnosis of noncardiac chest pain died because of a cardiac condition over an average follow-up period of 4.4 years (range: 0.5–10.4 years).





Reassuring S&S in the history

- Robust Exercise Tolerance
- Nonexertional chest pain
- Reproduction of CP with palpation
- Increased pain with inspiration
- Pain with food intake
- Pain that changes locations within or across the chest





Red Flags

- Family History of Sudden Death
- H/O CHD or heart transplant
- H/O Syncope with exertion
- H/O Palpitations
- Chest pain with exertion
- Abnormal ECG



Chest Pain in Children

Parents often worry when their child has any pain in their chest.

While chest pain in adults is frequently caused by serious cardiovascular problems, chest pain in children is often innocent. Research shows less than 1% of chest pain in children stems from the heart itself.

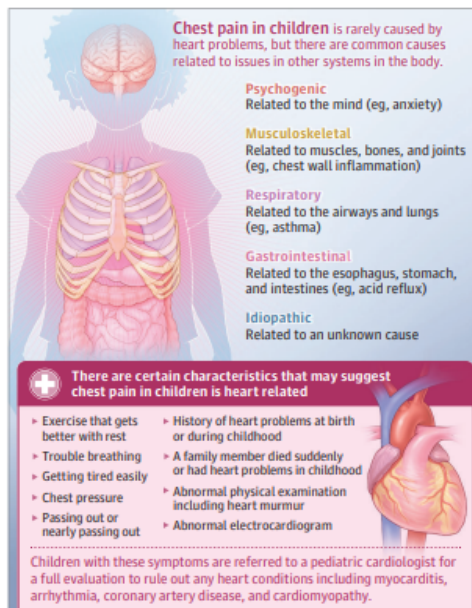
For children with chest pain, pediatric clinicians will take a full history, collect vital signs, and perform a physical examination. Your child may need testing, including a chest radiograph to see inside the chest and an electrocardiogram to look at the heart rhythm. Blood work may also be needed.

Common causes for a child's chest pain that does not stem from the heart can be split into several groups: (1) musculoskeletal, relating to the muscles, bones, and joints (eg, slipping rib syndrome and chest wall inflammation); (2) respiratory, relating to the airways and lungs (eg, asthma and pneumonia); (3) gastrointestinal, relating to the esophagus, stomach, and intestines (eg, acid reflux); (4) psychogenic, relating to the mind (eg, anxiety and panic attacks); and (5) idiopathic, relating to an unknown cause. While many of these causes of chest pain outside of the heart are not dangerous, they may still need evaluation and treatment by your child's health care professional or other specialists.

Features of chest pain in children that may suggest a heart-related cause include chest pain with (1) exercise that gets better with rest; (2) trouble breathing, getting tired easily, and/or chest pressure; (3) passing out or nearly passing out; (4) a history of heart problems at birth or during childhood; (5) a family member who died suddenly or had heart problems in childhood; (6) an abnormal physical examination result, such as a heart murmur; and (7) an abnormal electrocardiogram result.

If a heart-related cause is suspected, your child will need referral to a heart specialist called a pediatric cardiologist. A pediatric cardiologist will repeat a full history and physical examination. They may order an echocardiogram, or heart ultrasonography, to look at the structure and squeeze of the heart. Be reassured that most evaluations for chest pain at the pediatric cardiology office do not find any issues. However, it is important to have the full assessment to rule out any heart conditions. Possible heart-related causes of chest pain in children include (1) myocarditis (inflammation of heart muscle); (2) arrhythmia (unusual electrical patterns of the heart); (3) coronary artery disease (problems with the blood vessels to the heart); and (4) cardiomyopathy (problems with the heart's size and function).

Occasionally, children get sicker and may need evaluation sooner in the hospital, and they can see a pediatric cardiologist there. What-



ever is causing your child's chest pain, the medical team will work with you to understand your child's problem and to provide proper treatment when needed.

FOR MORE INFORMATION

<https://www.healthychildren.org/English/health-issues/conditions/heart/Pages/Chest-Pain-in-Children.aspx>

<https://www.healthline.com/health/childrens-health/chest-pain-in-children#outlook>

<https://www.choosingwisely.org/societies/american-academy-of-pediatrics-section-on-cardiology-and-cardiac-surgery/>

<https://www.ebmedicine.net/topics/cardiovascular/pediatric-chest-pain>
<https://www.aafp.org/pubs/afp/collections/choosing-wisely/453.html>

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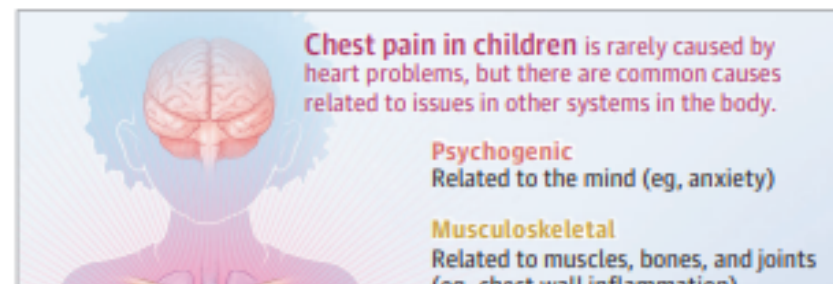


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For children with chest pain, pediatric clinicians will take a full history, collect vital signs, and perform a physical examination. Your child may need testing, including a chest radiograph to see inside





Appropriate Use Criteria

- Developed to promote effective patient care, better clinical outcomes, and improve resource utilization.
- Established multiple algorithms for several cardiac symptoms where the appropriate use of echocardiography was considered appropriate, may be appropriate or rarely appropriate.





Indication	Examples of appropriate and rarely appropriate indications
Palpitations/arrhythmias	Supraventricular tachycardia (A), ventricular tachycardia (A), palpitations with normal ECG (R)
Syncope/presyncope	Exertional syncope (A), vasovagal syncope (R)
Chest pain	Exertional chest pain (A), non-exertional chest pain (R)
Murmur	Pathologic murmur (A), presumed innocent murmur (R)
Symptoms/signs	Symptoms suggestive of congestive heart failure (A), central cyanosis (A), acrocyanosis (R)
Prior test results	Abnormal CXR or ECG (A), positive blood culture suggestive of endocarditis (A), previously normal echocardiogram without change in clinical status (R)
Systemic disorders	Systemic hypertension (A), concern for pulmonary hypertension (A), connective tissue disorder (A), cancer prior to or during chemotherapy (A), diabetes mellitus (R)
Family history of CVD ^a	Family history hypertrophic cardiomyopathy (A), family history of non-ischemic dilated cardiomyopathy (A), consanguinity (R)
Fetal concerns/complications	Suspected cardiovascular abnormality on fetal echocardiogram (A), isolated echogenic focus on fetal ultrasound (R)

A appropriate, CVD cardiovascular disease, CXR chest radiograph, ECG electrocardiogram, R rarely appropriate





Diagnostic yield of OP pediatric echocardiograms

- Non-exertional chest pain or syncope, distant family history of cardiovascular disease, and vague palpitations represent situations in which echocardiograms are highly unlikely to aid in medical management.
- Cost analysis using the model of cardiology consultation rather than direct primary care echocardiogram ordering estimated a 19.6% reduction in medical costs for the most common indication, murmur.





Syncope

- Frequent reason for referral to pediatric cardiology.
- Parents are typically concern about sudden cardiac death
- Common with peak incidence in 9-14 year old and then 15-19 year olds.
- Usually neurocardiogenic, aka vasovagal syncope.
- Typically includes prodromal symptoms like N/V, dizziness, lightheadedness and blurry vision before the syncopal event.
- Often have a history of poor hydration and exertion in hot weather.



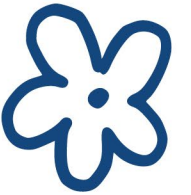


Red flags for syncope

- No prodromal symptoms
- Syncope during exertion
- Family history of sudden unexplained death
- Abnormal ECG – in particular QT abnormalities
- Systolic murmur that intensifies with Valsalva maneuver or stand from squat.



Orthostatic Intolerance and POTS





Orthostatic Intolerance

- *Dizziness*
- *Lightheadedness*
- Headaches
- Fatigue
- Heart Racing
- Difficulty concentrating
- Amplified Pain
- Nausea
- Syncope
- Difficulty sleeping



POTS

More severe
version

Daily Symptoms

Quality of life
impacted

Heart rate > 120
bpm with
standing or an
increase of 40
bpm with
standing or on
tilt test.



What causes POTS

- Increased tendency for venous pooling (bluish discoloration and swelling in lower extremities when upright)
- This causes less blood flow to the heart and then less to the brain resulting in dizziness and lightheadedness.
- The body responds by making adrenaline to get more blood to the brain.
- This over production of adrenaline leads to other symptoms: heart racing, fatigue, headaches, poor concentration, difficulty sleeping and GI symptoms







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POTS and syncope

With the increased adrenaline there is a mixed signaling that occurs which we all know as vasovagal syncope. The brain senses the increased adrenaline and responds with a drop in HR and BP resulting in syncope.



It's important to stress that the patient **WILL REGAIN CONSCIOUSNESS** once supine.



Seems to be more common in patients with hypermobility syndrome, adolescents, and in patients that have experienced some event associated with stress and a trigger to the immune system.

Treatment

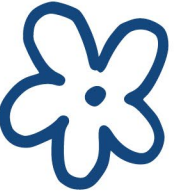
Patient education is essential

Patient must pay attention to dizziness which is specific for orthostatic intolerance and can be a way to measure improvement.

Patient must improve their blood volume by increasing salt and water intake (a minimum of 64 to 72 oz of water a day.)

Intake **MUST** be front loaded starting as soon as they wake up.

3-5000 mg of sodium daily



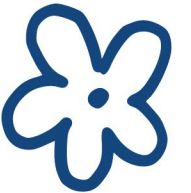
Increasing sodium intake

Doesn't have to be just chips and crackers

Fluids with added electrolytes

High in sodium foods that are low in carbs such as breakfast meats, pickles, and salted nuts.

Salt tablets can also be an option.



Treatment





Counterpressure Maneuvers

Dynamic Counter-Pressure Maneuvers

- Heel raises / toe extensions
- Marching in place
- Postural swaying



Positional Counter-Pressure Maneuvers

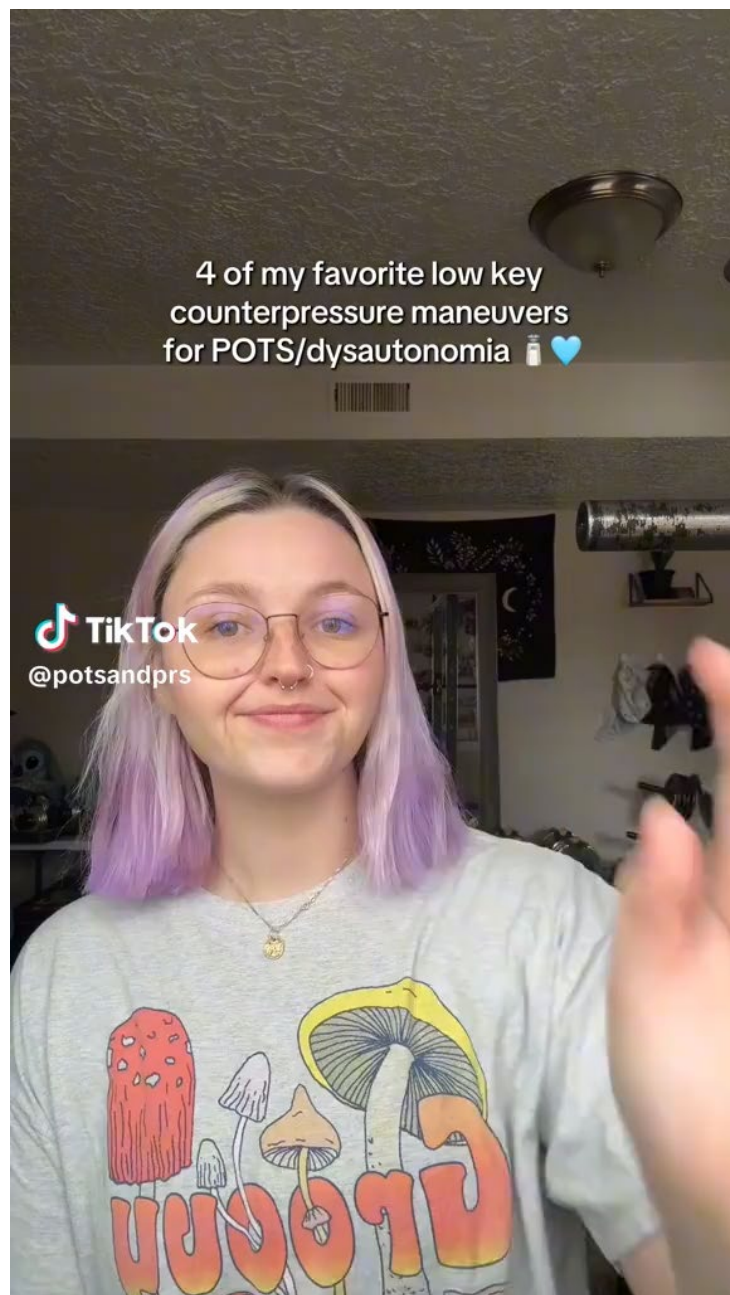
- Bending over as if to tie shoelaces
- Squatting

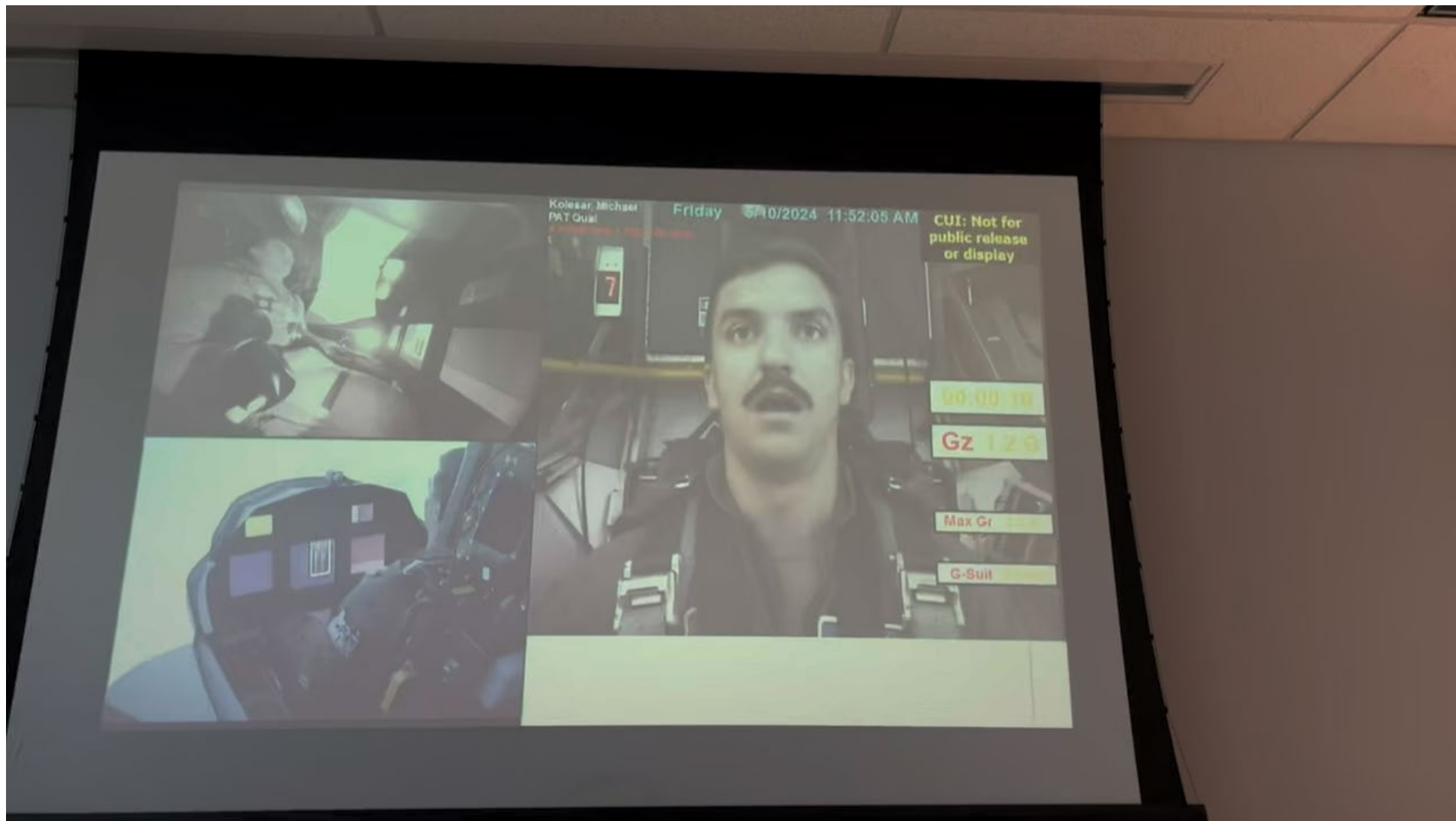


Isometric Counter-Pressure Maneuvers

- Leg crossing with muscle tensing
- Whole-body tensing
- Butt clenching







Medications that hurt

Medications that dry you out (diuretics, antihistamines)

Medications that lower your blood pressure.

Stimulants for ADHD can help with brain fog but can also be counterproductive because of the side effects of raising your heart rate and drying you out.

The pro and cons need to be weighed for each individual patient.



Medications to help with symptoms

- Fludrocortisone can help increase blood volume.
- Betablockers can help decrease the effects of adrenaline.
- Midodrine helps prevent blood pooling.
- All of these are only helpful if the nonpharmacologic measures are taken.





POTS and Anxiety

- Anxiety is not a cause of POTS but can certainly exacerbate the symptoms
- Treating the anxiety is important.
- Cognitive behavior therapy has been shown to be very beneficial in these patients.







The Care of Children with CHD in the Primary Medical Home

- CHD is the most common birth anomaly.
- More patients are surviving.
- Patients with CHD have complex health care needs that often need to be coordinated by the PCP.





Prenatal	Neonatal	Infancy	Childhood	Adolescence
a. Utility and limitations of prenatal diagnosis b. Parental counseling c. Prediction of neonatal needs d. Parental support	a. Recognition and diagnosis b. Pulse oximetry screening c. Genetic screening d. Coordination of subspecialist care e. Anticipation of major organ involvement f. Parental support	a. Nutrition b. Immunizations c. Infection prevention d. Antithrombosis e. Growth and development f. Parental support g. Coordination of care	a. Recognition of recurrent lesions b. Recognition of medication and pathologic complications c. Consequences of common childhood illness c. Academic performance d. Lifestyle initiatives e. Sports participation f. Parental support g. Coordination of care	a. Academic and future career support b. Sports participation b. Lifestyle initiatives c. High-risk behavior avoidance d. Teen pregnancy avoidance e. Fostering independence f. Transition to adult care providers g. Coordination of care h. Encourage medication compliance





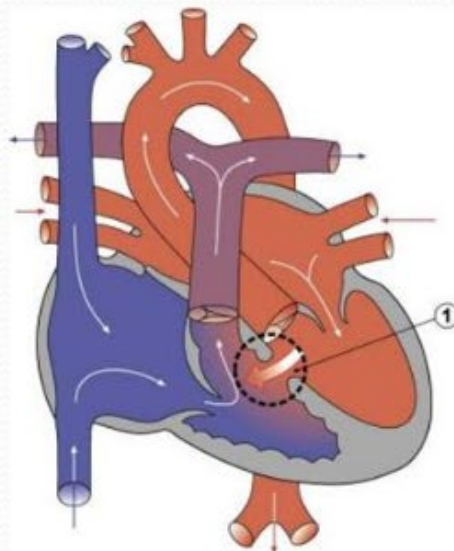
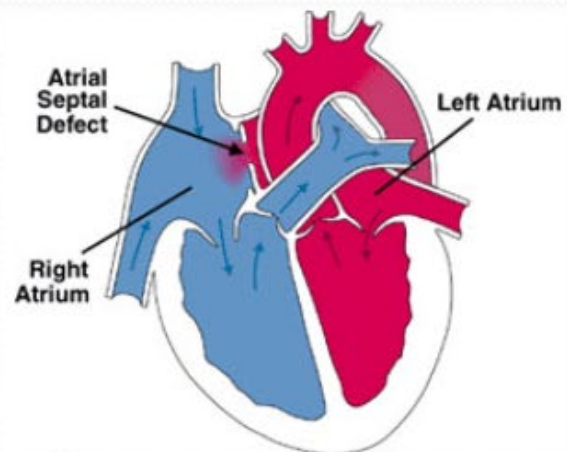
Palliation

To palliate a disease is to treat it partially and insofar as possible, but not cure it completely. Palliation cloaks a disease.



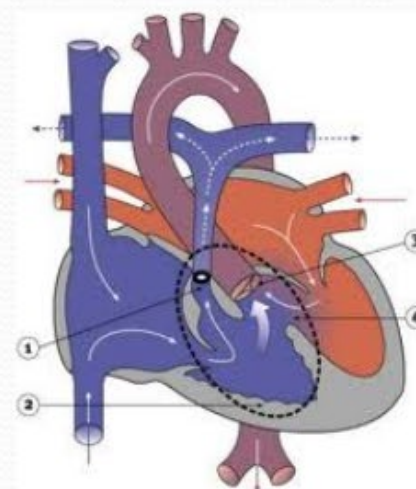


ASD



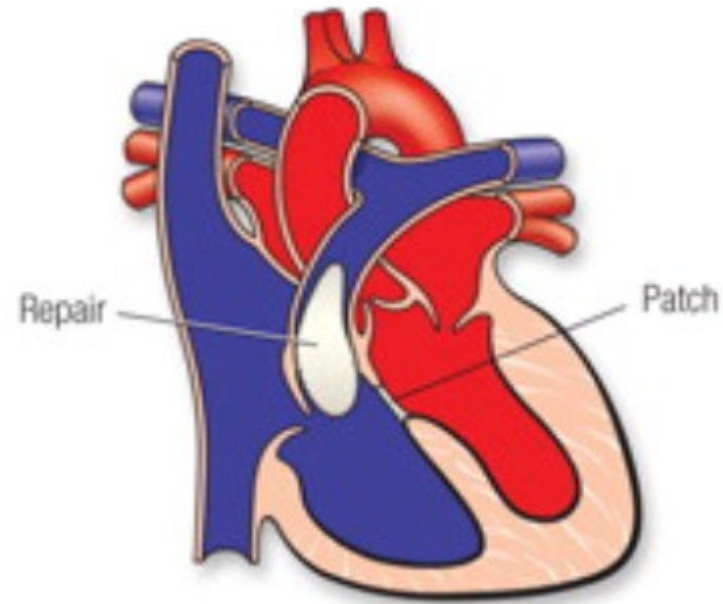
VSD

ToF





Tetralogy of Fallot





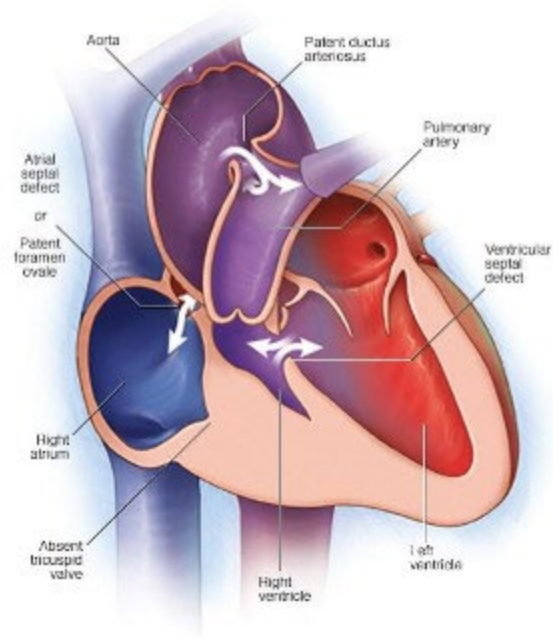
Single Ventricle

- In the US the mortality rate for congenital heart disease patients have substantially decreased, as much as 40% over the last 4 decades.
- Mortality in children has decreased by 31% and the age at death generally has moved steadily toward older age; now 76% of deaths occur in adulthood.
- Severe CHD patients, a group to which SV patients belong, have a prevalence of 1.76 per 1000 children with a live birth prevalence of 35 per 100,000.
- SV patients 10 year survival was 79% before 1990 but now with improved preoperative and postoperative management is estimated to be 95-98%.

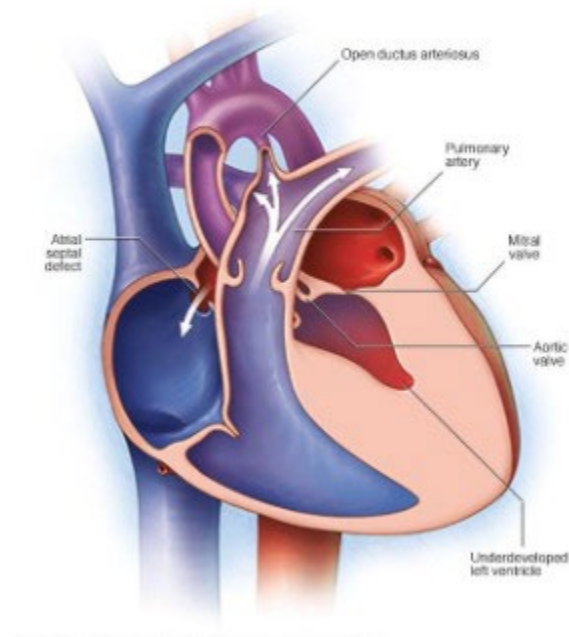




Single ventricle

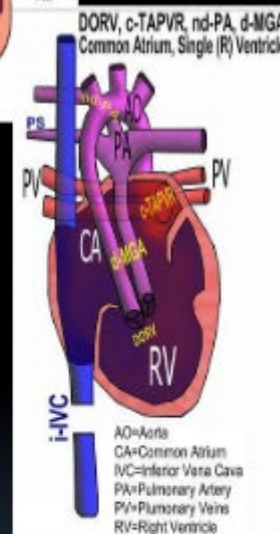
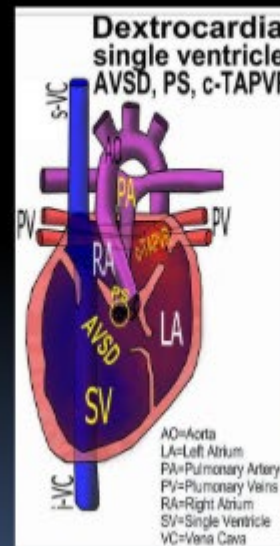
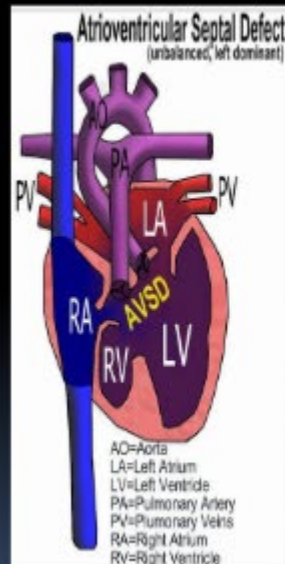
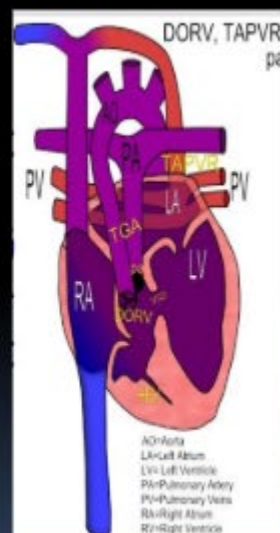
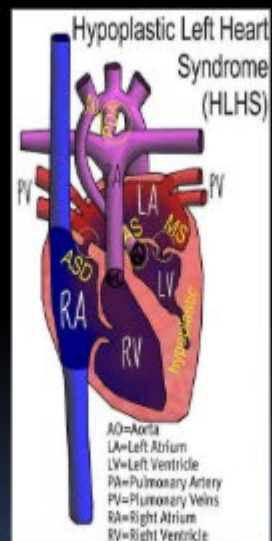
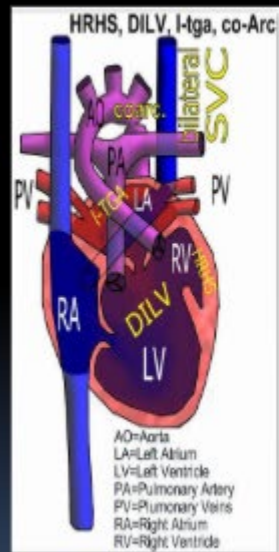
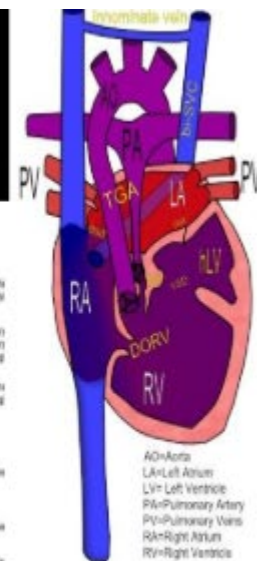
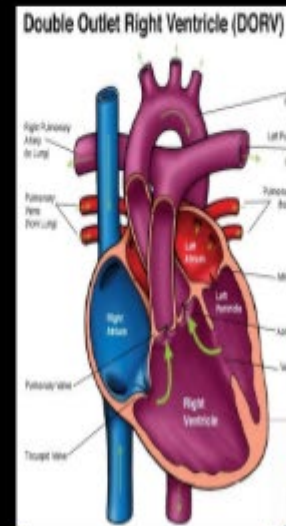
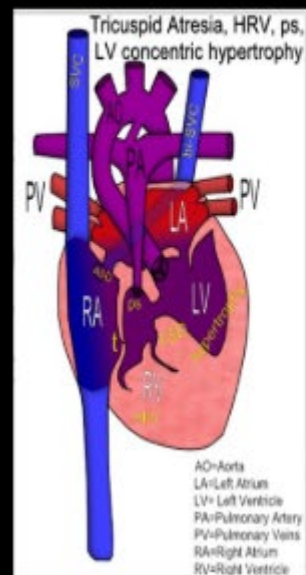
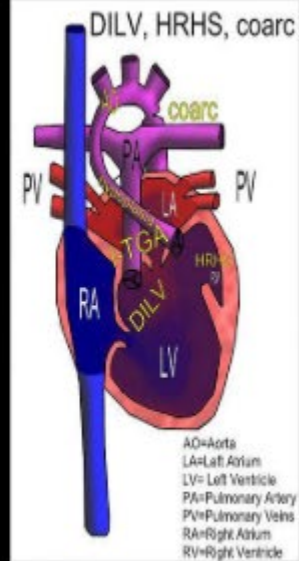
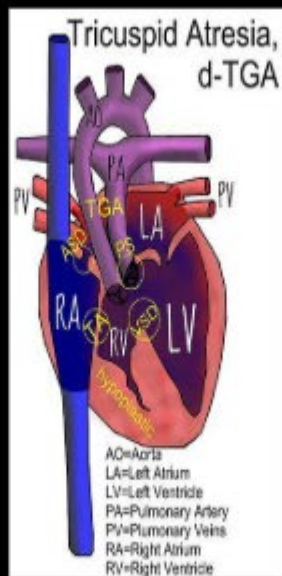
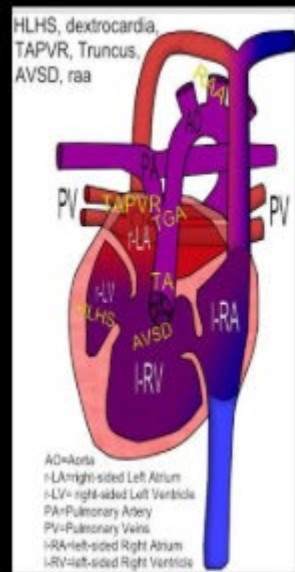


Tricuspid atresia



HLHS

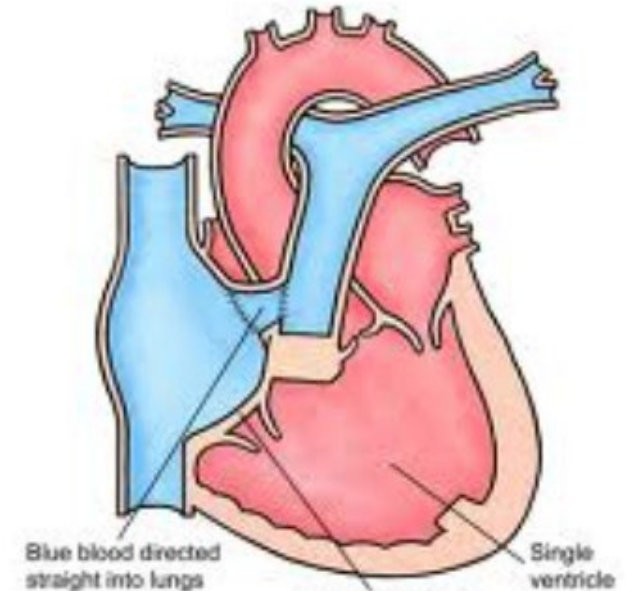






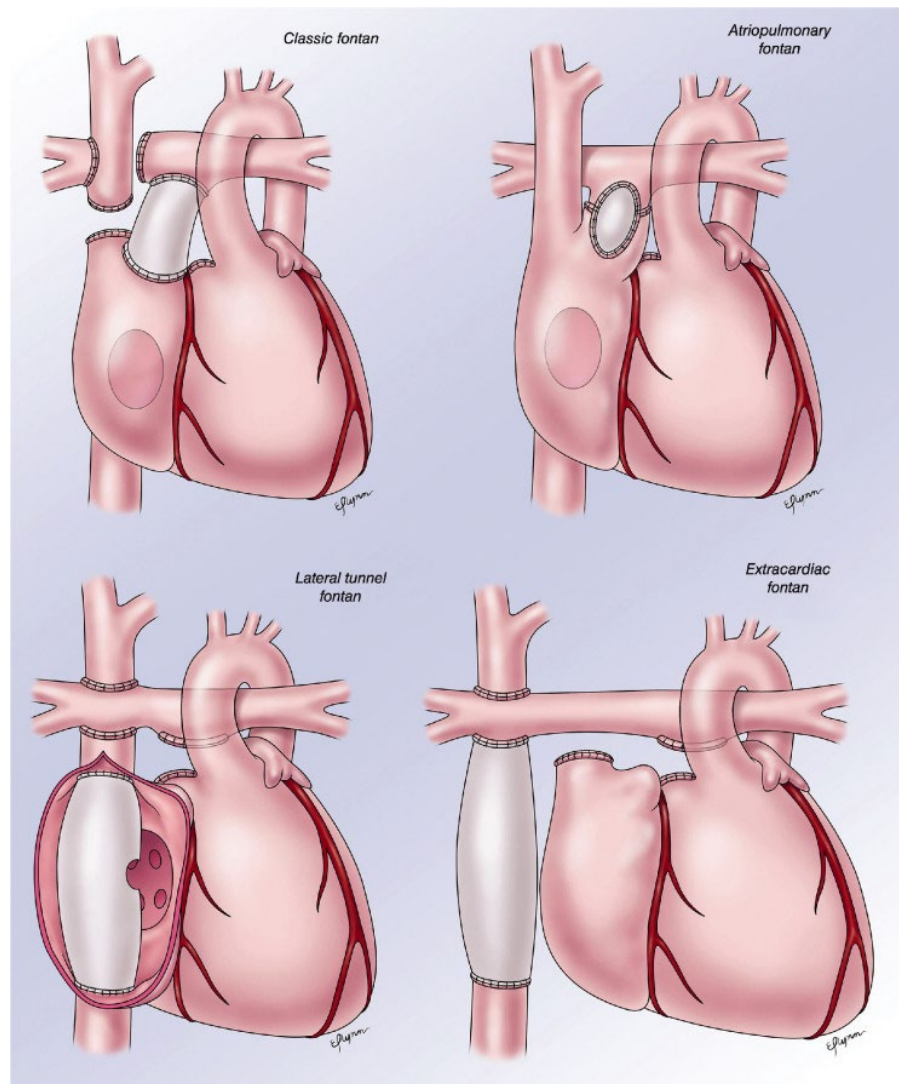
Fontan procedure

- First performed in 1968.
- Staged to improve saturations w/o increasing the volume load on the heart.





Palliation for Single ventricle - Fontan





Questions the PCP should ask the pediatric cardiology team?





Is this a systemic RV or LV?



Associated with decreased survival in Fontan patients

Single ventricle is morphologic right ventricle.

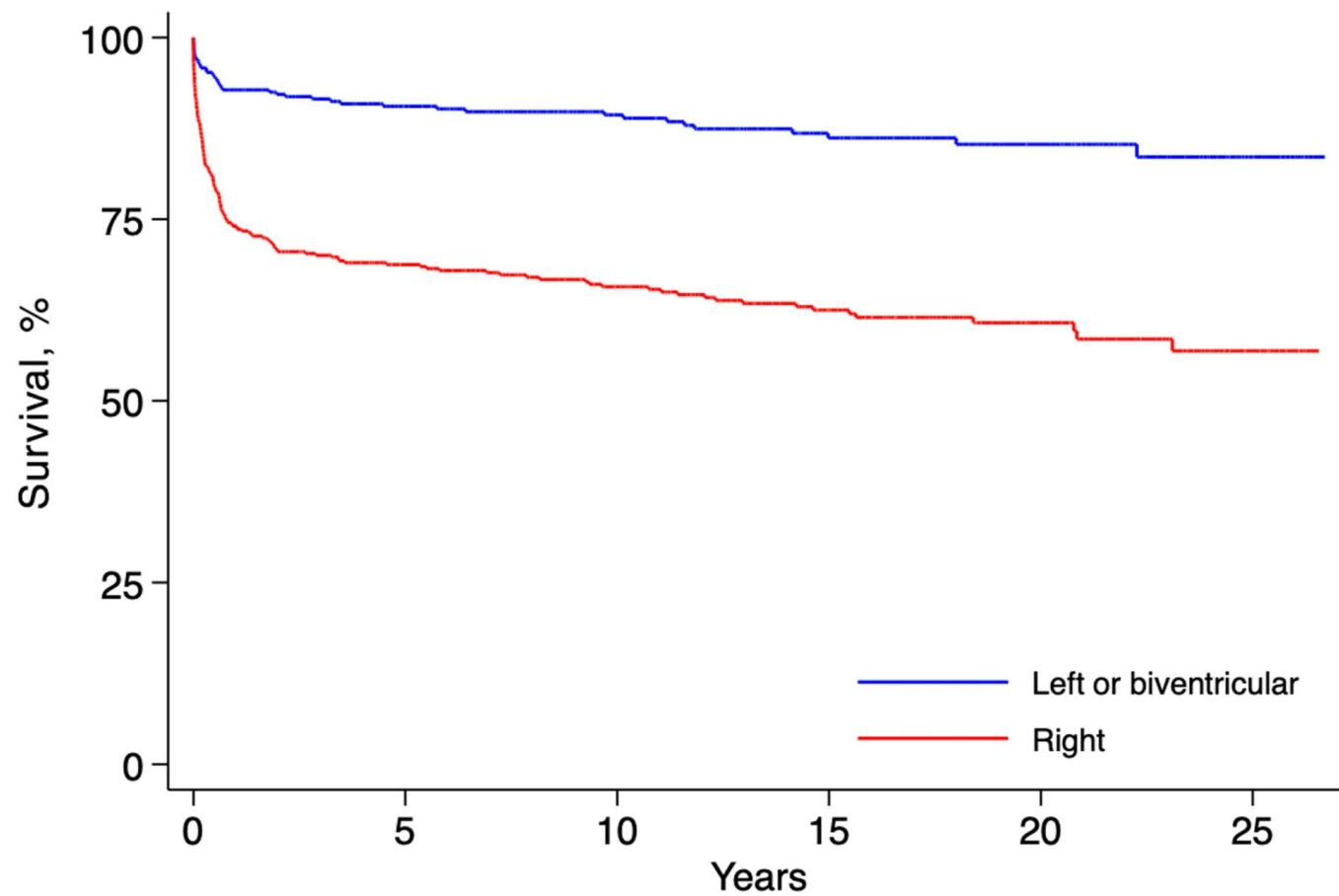
Heterotaxia syndromes

Increased pulmonary artery pressures.

AV valve dysfunction.

Decreased ventricular function.





No. at risk							
Left or biventricular	333	255	197	134	79	18	
Right	432	252	192	128	64	16	





What are acceptable sats in this patient?



Normal sats for a single ventricle patient

- 80s usually in the initial first stage after birth. These patients have a high level of care with home monitoring so everyone is watching them.
- 80s generally after the second stage.
- Most single ventricles after the Fontan (third stage) will have near normal sats. There are exceptions.





Are their vitals normal?





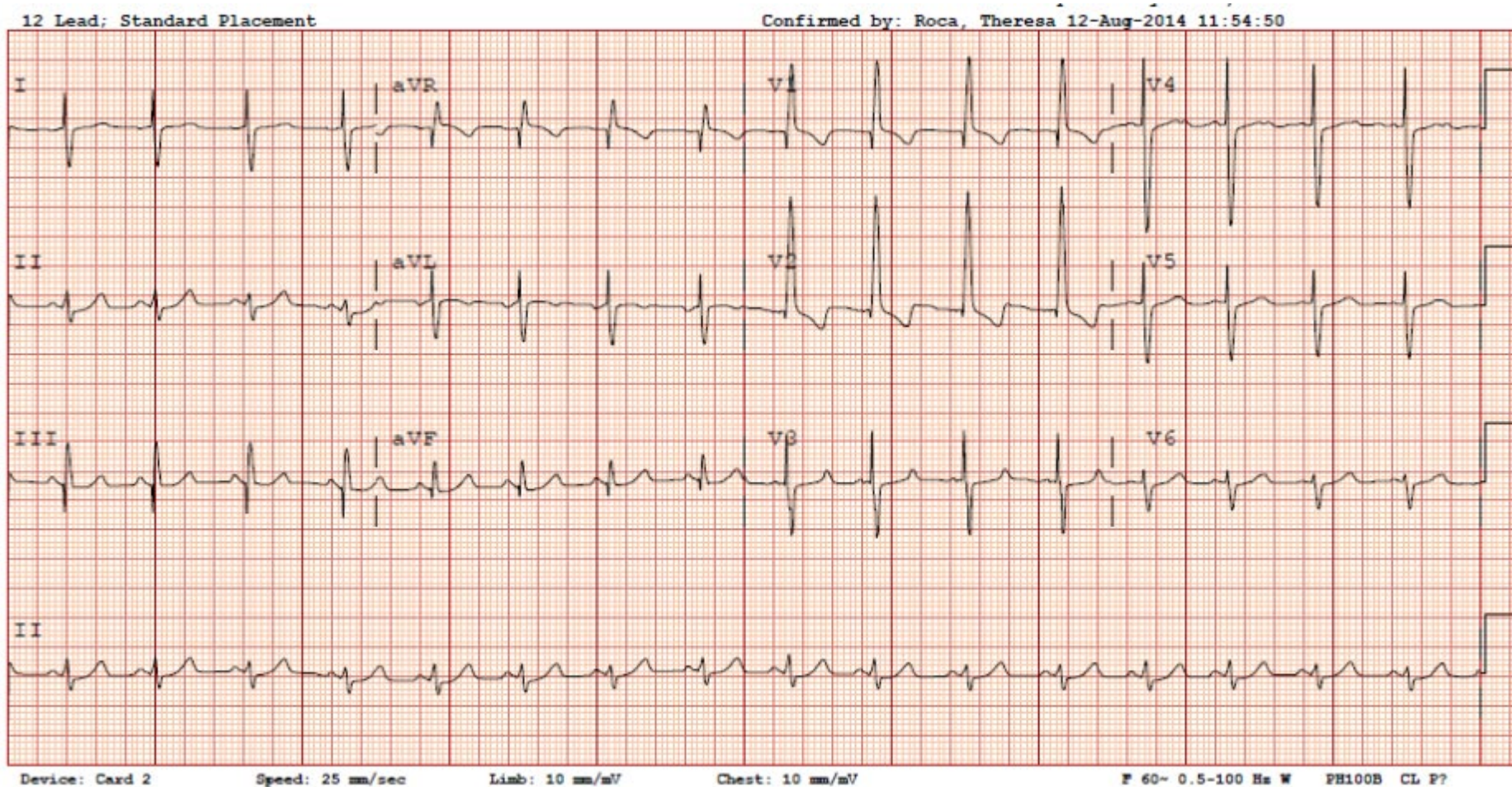
Do you have a copy of their baseline ECG?

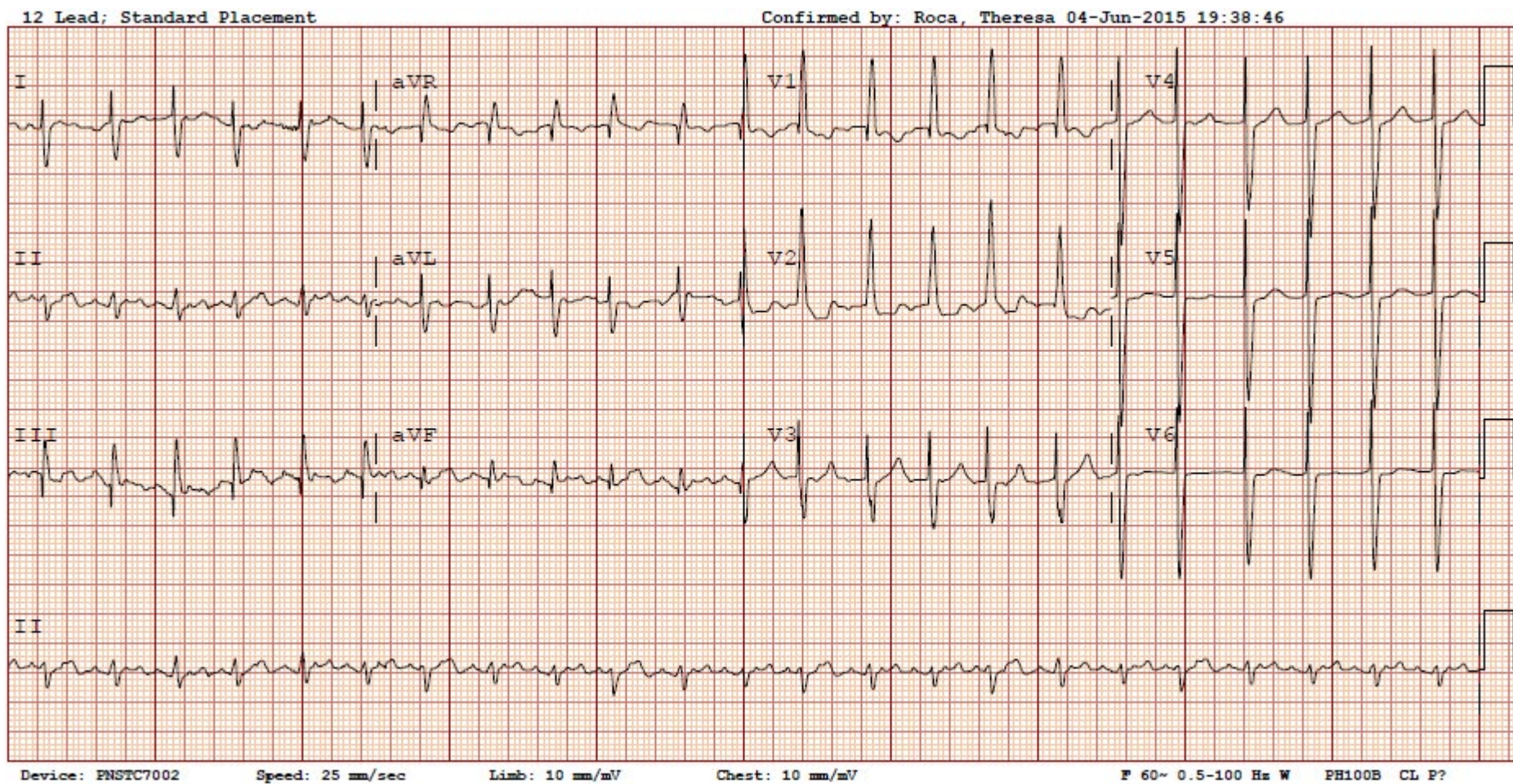


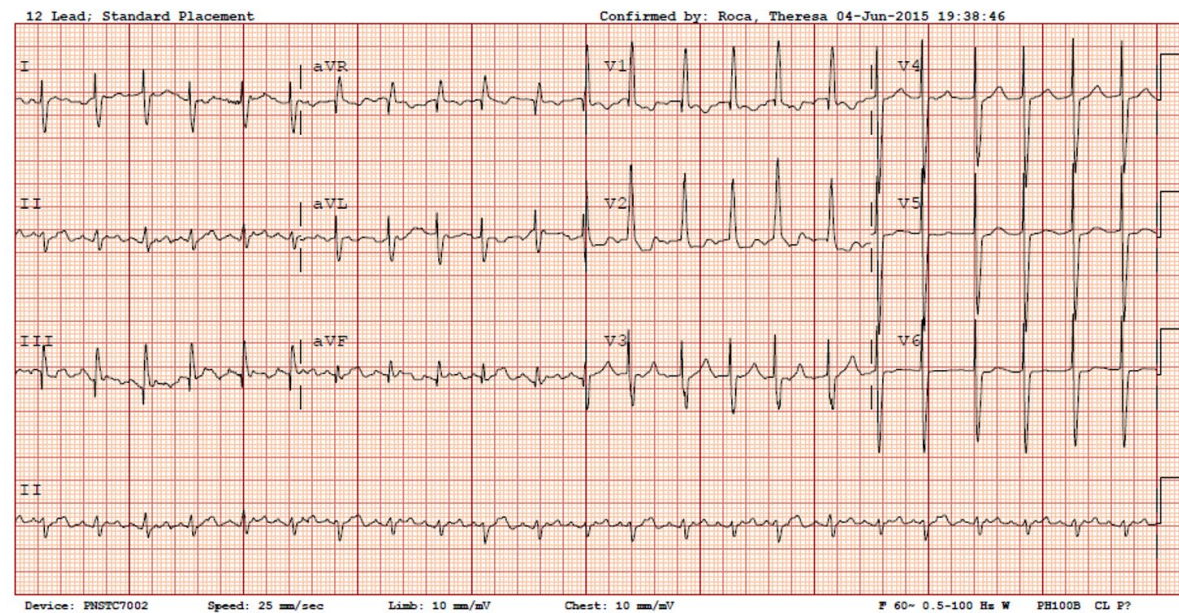
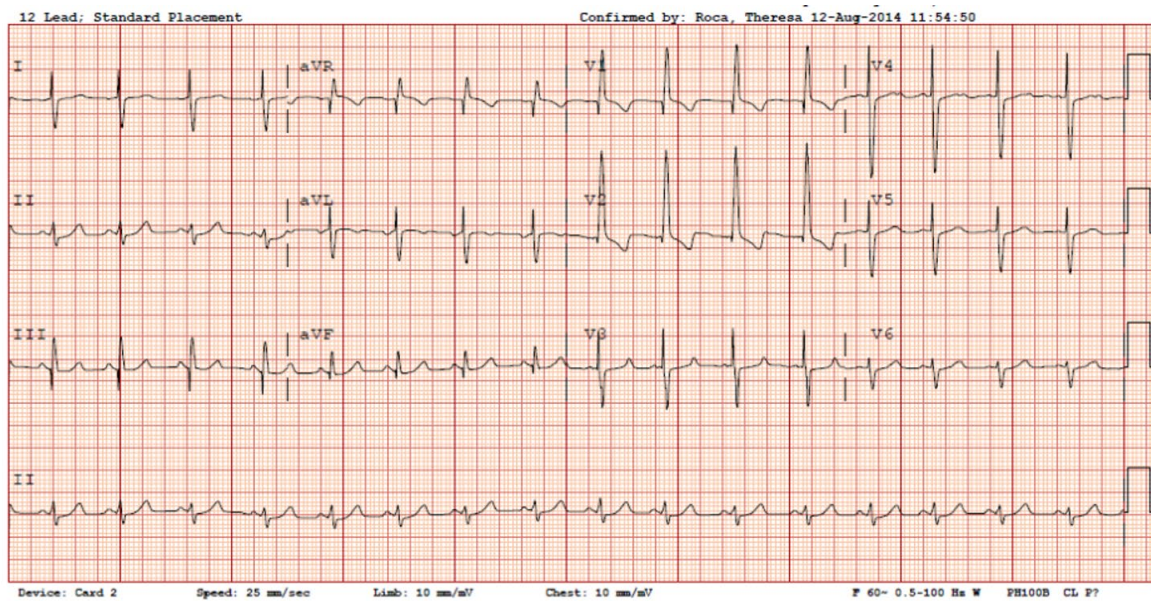


A 10 yo single ventricle presents with history of increased fatigue, poor color. Mother brings him to the local ER and his heart rate is 136 bpm.











Signs and symptoms of Fontan failure

- Increased fatigue
- Palpitations
- Shortness of breath
- Anorexia
- Diarrhea
- Weight loss or weight gain
- Increasing cyanosis
- New onset seizures
- fever of unknown origin
- Tia or stroke





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Thank you



