AEMS: Recognition and Management of Patients with Congenital Cardiac Abnormalities

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Faculty Disclosure

- I have no financial relationship with any manufacturer of any commercial product and/or provider of commercial services discussed in the conference.

- I do not intend to discuss an unapproved/investigative use of a commercial product or device in my presentation

Objectives

- Define critical congenital heart disease and initial neonatal stabilization protocols for each specific type of CCHD
- Understand the anatomy and cardiovascular physiology of common surgeries performed for CHD
- Common emergencies in children with CHD
- Brief review of arrhythmias in pediatrics (Q&A)

Pediatric Cardiac Emergencies

- Structural heart disease
- Arrhythmias
  - Supraventricular tachycardia
  - Atrial flutter
  - Ventricular tachycardia/fibrillation
- Trauma
  - Cardiac contusion
  - Cardiac tamponade
- Ischemia
  - Kawasaki disease
  - Coronary artery anomalies
- Myocardial disease
  - Hypertrophic cardiomyopathy
  - Dilated cardiomyopathy
  - Restrictive cardiomyopathy

Which baby has central cyanosis?

acrocyanosis

central cyanosis

clubbing

Congenital Heart Disease

- Nearly one percent of all newborns have a congenital heart defect
  - CHD is the most common congenital disorder
- CHDs are the leading cause of birth defect-associated infant illness and death
- Up to 25 percent of infants with CHD have a "critical" defect
  - Critical CHD refers to lesions requiring surgery or catheter-based intervention in the first year of life
  - This translates to about 18 per 10,000 babies are born with critical congenital heart disease
- **All newborns are screened for CHD with pulsoximetry**

Pulse Oximetry Screening Algorithm

Child in well-infant nursery > 24hrs of age or right before discharge if < 24hrs

Place Pulse Oximeter on Right Hand (RH) & Any Foot (F)

< 90% in RH or F

< 90% in RH or F

< 90% in RH or F

Positive Screen

90-94% in RH & F **OR**

> 3% difference between RH & F

Repeat Screen in 1hr

90-94% in RH & F **OR**

> 3% difference between RH & F

Repeat Screen in 1hr

90-94% in RH & F **OR**

> 3% difference between RH & F

≥ 95% in RH or F **AND**

≤ 3% difference between RH & F

≥ 95% in RH or F **AND**

≤ 3% difference between RH & F

≥ 95% in RH or F **AND**

≤ 3% difference between RH & F

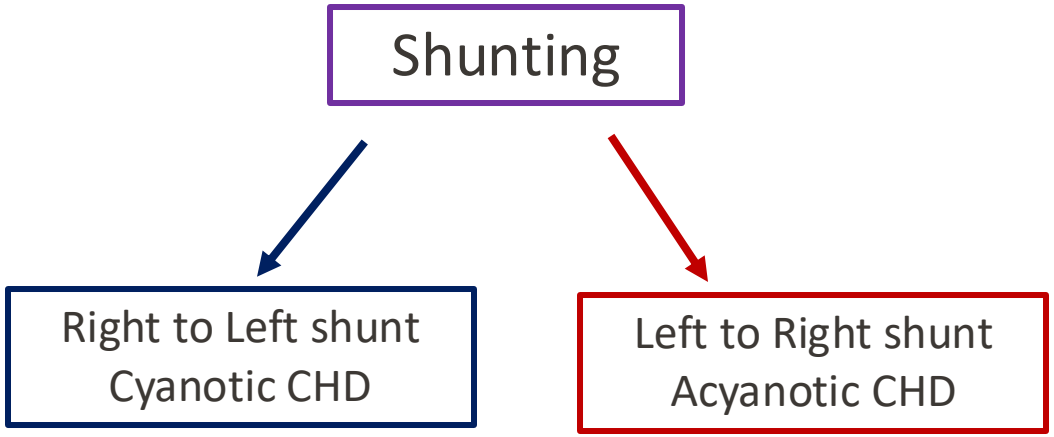
Negative Screen

Critical Congenital Heart Disease

- Significant CHD that requires neonatal stabilization and intervention within the first year after birth
- Structural
  - Transposition of the Great Arteries
  - Truncus Arteriosus
  - Total Anomalous Venous Return
  - Tricuspid Atresia
  - Tetralogy of Fallot
  - Hypoplastic left heart syndrome
  - Coarctation of the aorta
  - Aortic/pulmonary stenosis
  - Ebstein anomaly

"5 T's"

Categories of CHD



- Tetralogy of Fallot
- Transposition of Great Arteries
- Truncus arteriosus
- Tricuspid atresia
- TAPVR

- ASD
- VSD
- Atrioventricular septal defect
- PDA

Abnormal communications between cardiac chambers and blood vessels

Obstruction

- HLHS
- Aortic stenosis
- Pulmonary stenosis
- Coarctation of the aorta/interrupted aortic arch

Abnormal narrowing of valves or arteries that obstruct blood flow

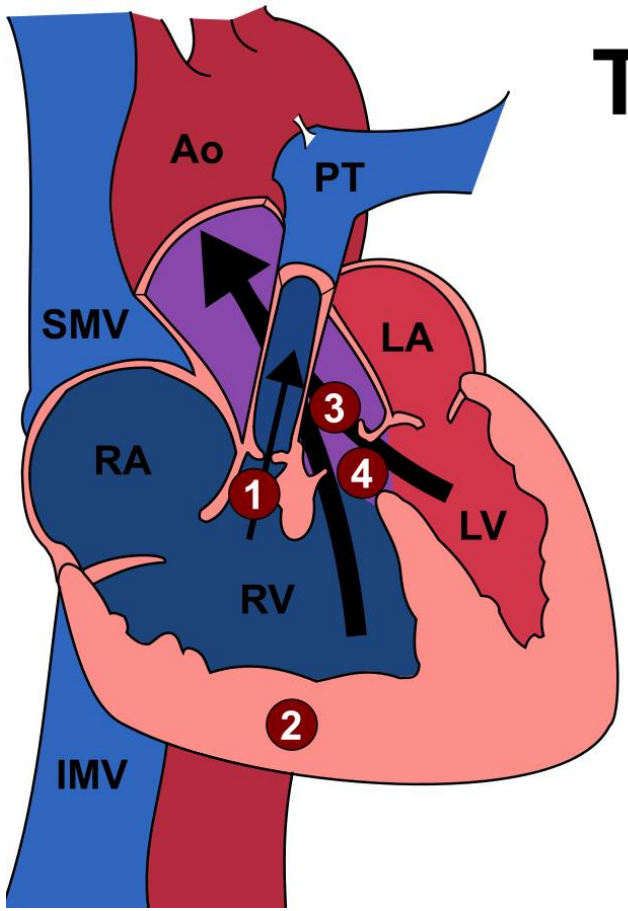
Cyanotic CHD – CXR Findings

- Decreased pulmonary vascular markings
  - Tetralogy of Fallot
  - Pulmonary atresia
  - Tricuspid atresia
  - Critical pulmonary stenosis associated with CHD
  - Ebstein anomaly

- Increased pulmonary vascular markings
  - Truncus arteriosus
  - Transposition of great arteries with VSD
  - Total anomalous pulmonary venous return

Tetralogy of Fallot

- Most common cyanotic CHD
- Four defects
  - Pulmonary stenosis
    - RV outflow obstruction
    - Subvalvar, valvar, and supravalvar
    - Determines severity of symptoms
  - VSD
  - Right ventricular hypertrophy
  - Overriding aorta
- Decreased pulmonary blood flow



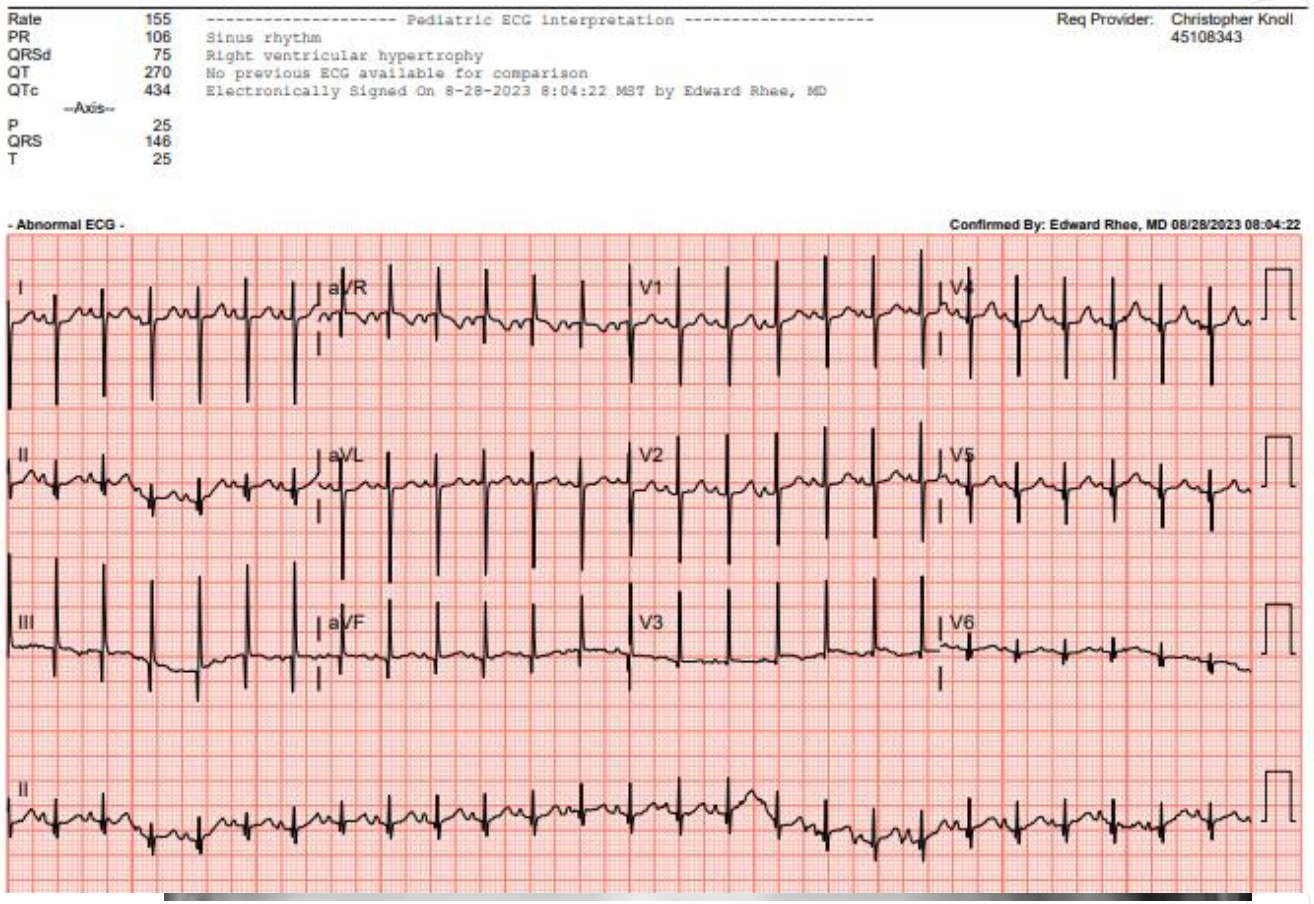
Tetralogy of Fallot

Major Defects

1. Pulmonary Stenosis
2. Right Ventricular Hypertrophy
3. Overriding Aorta
4. Ventricular Septal Defect

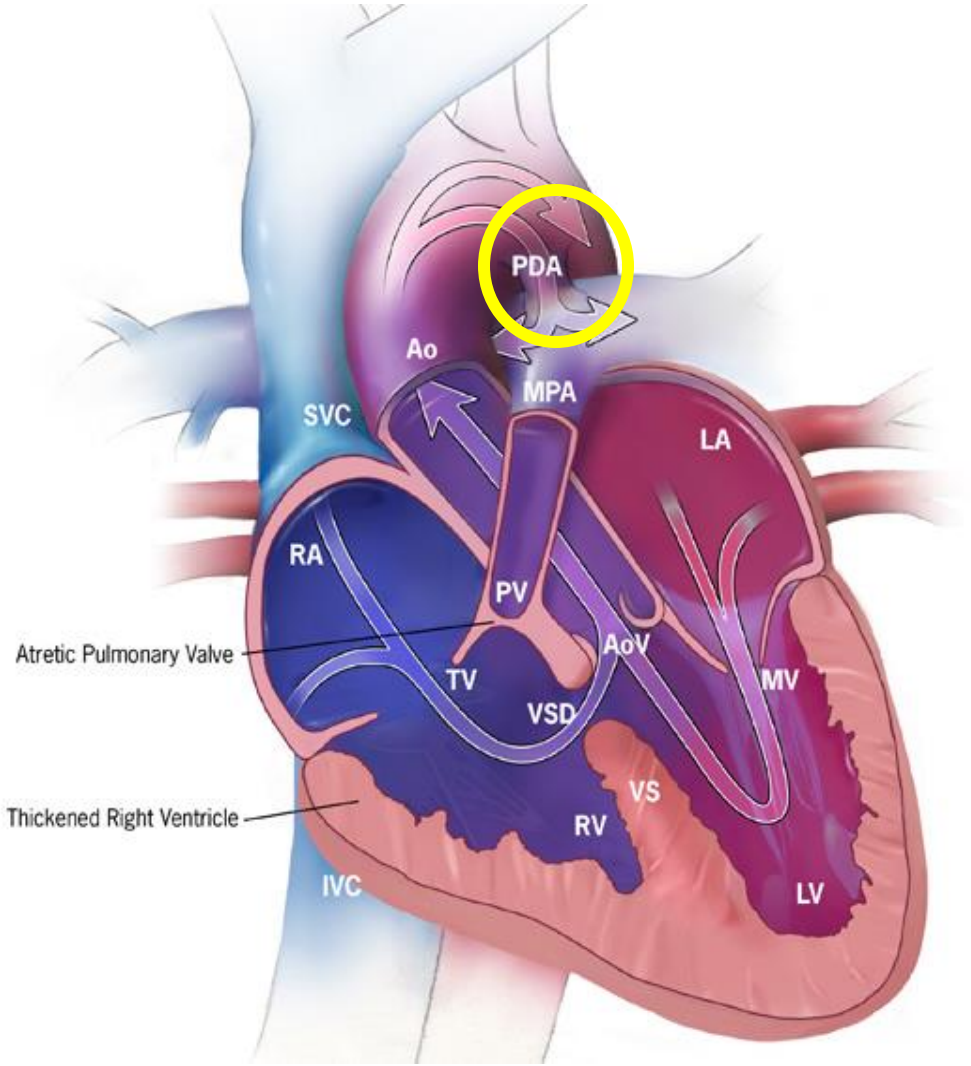
Tetralogy of Fallot

- CXR
  - Decreased pulmonary vascular markings
  - “Boot-shaped” heart
  - Concave RVOT/pulmonary artery
  - Narrow mediastinum (absent thymus)
- ECG: RVH, RAD
- Exam: Harsh systolic ejection murmur at LUSB radiating to axillae and back
- O2 sat goal: >80%



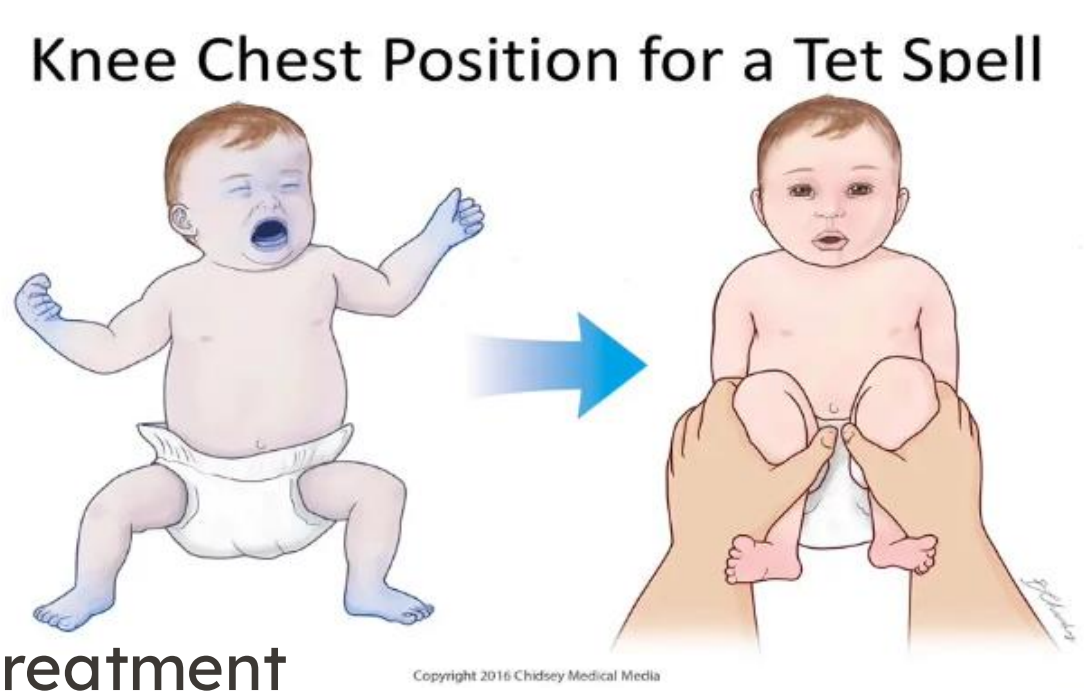
Variable severity of TOF

- “Pink Tet” – mild pulmonary stenosis
  - No neonatal intervention needed
- “Blue Tet” – moderate to severe pulmonary stenosis
  - Possible PGE infusion (0.01-0.02 mcg/kg/min)
  - Keep fluid positive
  - Avoid tachycardia, agitation
- TOF/Pulmonary atresia
  - Requires PGE infusion



Hypercyanotic (Tet) Spells

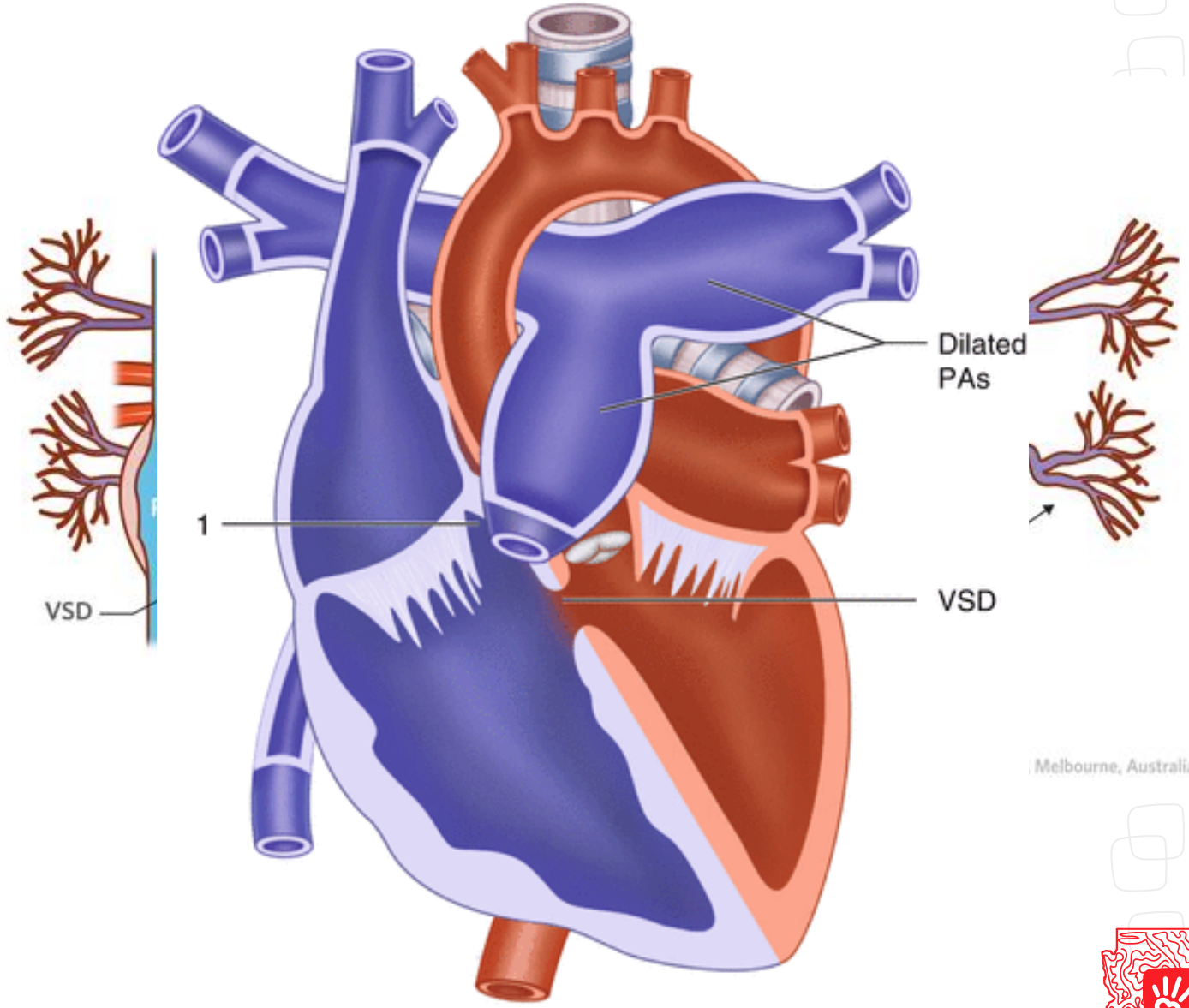
- Provoked by tachycardia (e.g. crying, fever, dehydration)
  - Increased RVOT obstruction
  - Increased right to left shunting across the VSD
- Four characteristics
  - Sudden onset of cyanosis or worsening cyanosis
  - Dyspnea
  - Altered consciousness
  - Decrease or disappearance of systolic murmur



- Treatment
  - Knees to chest (increasing SVR)
  - Oxygen
  - Phenylephrine
  - Volume repletion – PIV, IJ line, IO
  - Morphine
  - IM Ketamine
  - Consider PGE
  - Intubate & paralyze
  - Surgical repair vs BTT shunt

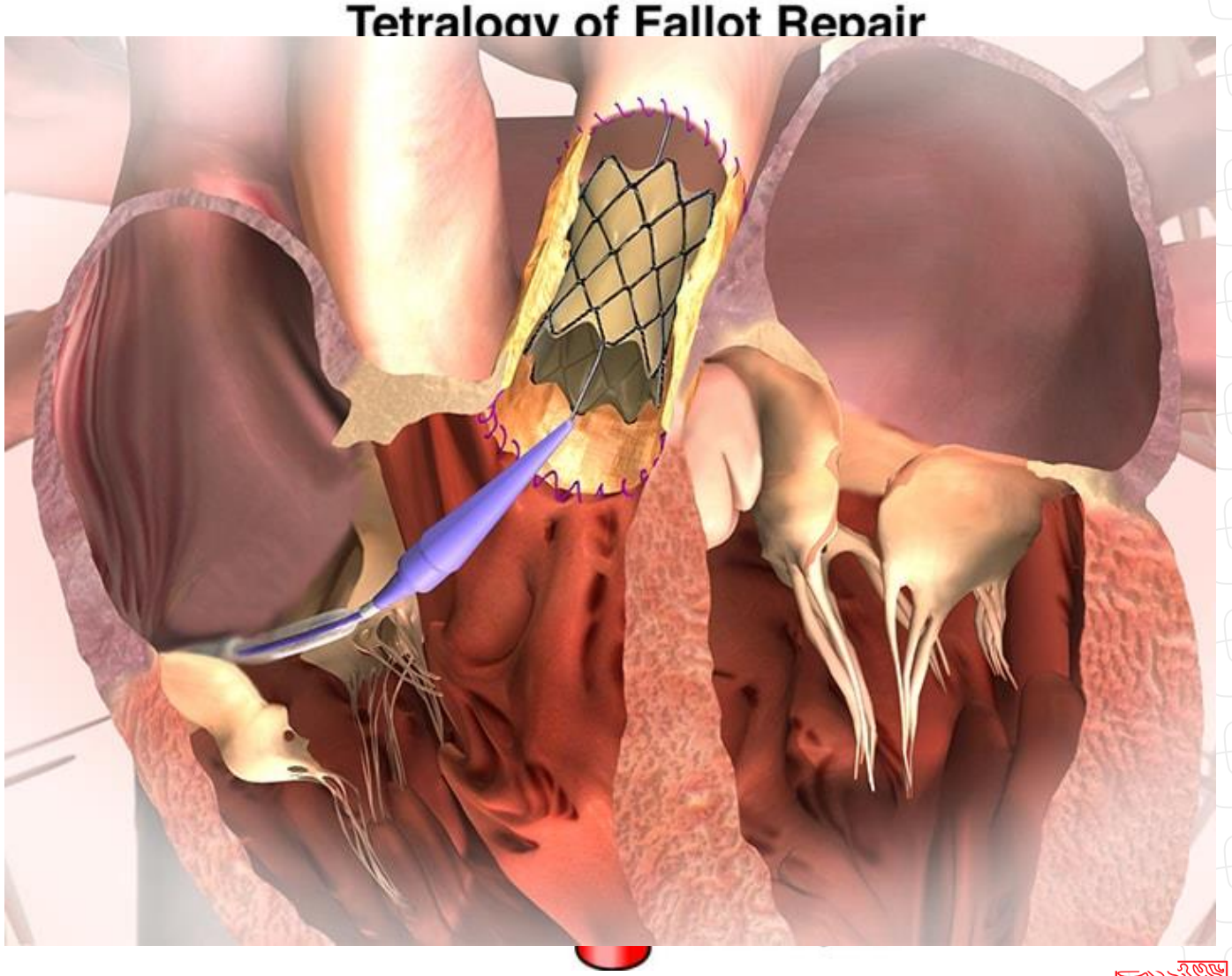
Important Variants of TOF

- TOF/PA with MAPCAs
  - Major Aorto-Pulmonary Collateral Arteries
  - May not need PGE
  - Complex repair involving unifocalization of the MAPCAs into the pulmonary arteries and placement of a RV-PA conduit
- TOF/Absent Pulmonary Valve syndrome
  - Bronchial obstruction
  - Prone position
  - May need positive pressure ventilation



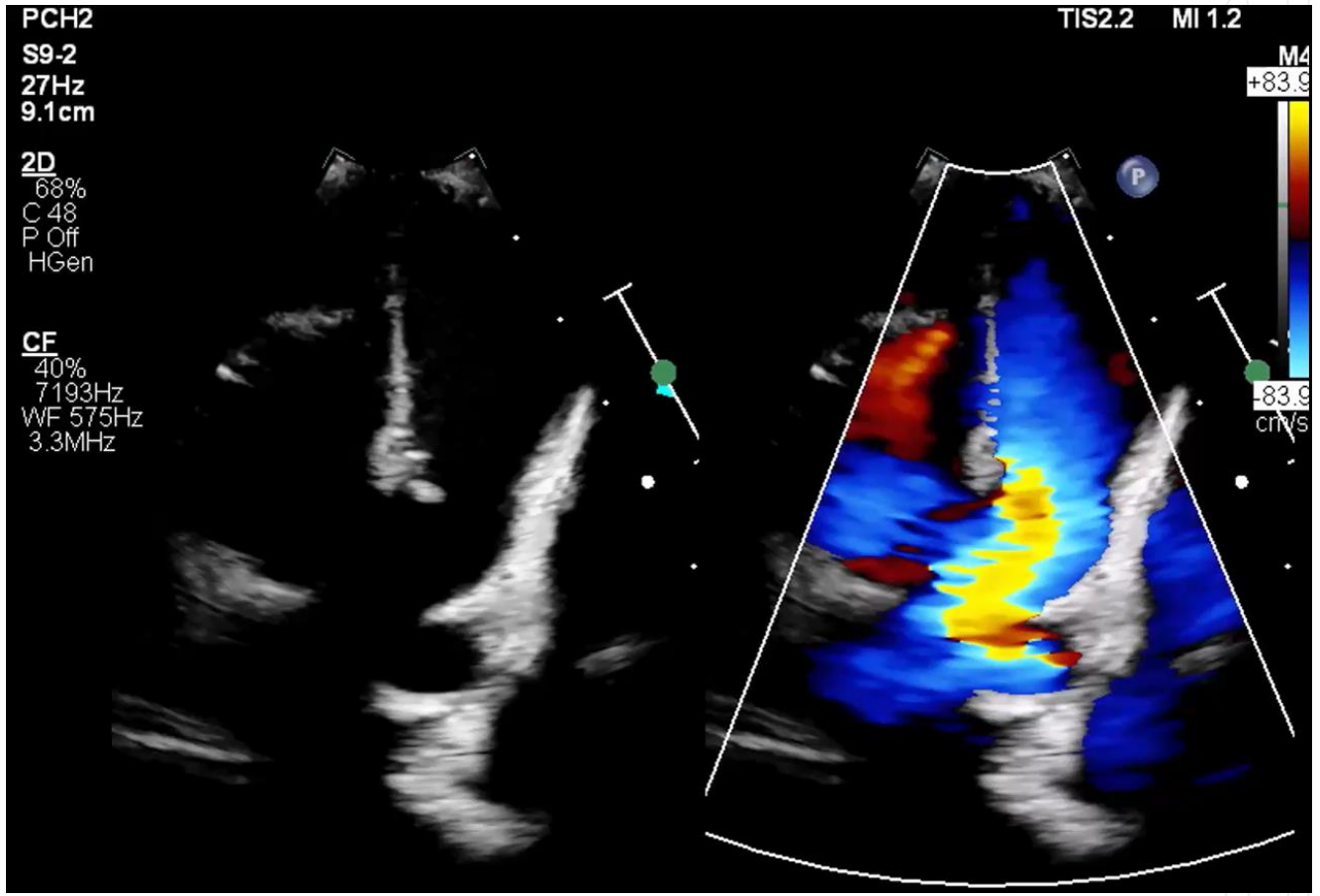
Surgical repair of TOF

- Initial surgery may be a BTT shunt or RF perforation and pulmonary balloon valvuloplasty
- Complete repair between 3-6 months of age
- May need a transannular patch resulting in free pulmonary regurgitation
- Future pulmonary valve replacement with catheter or surgical intervention



TOF: Post Surgical Emergencies

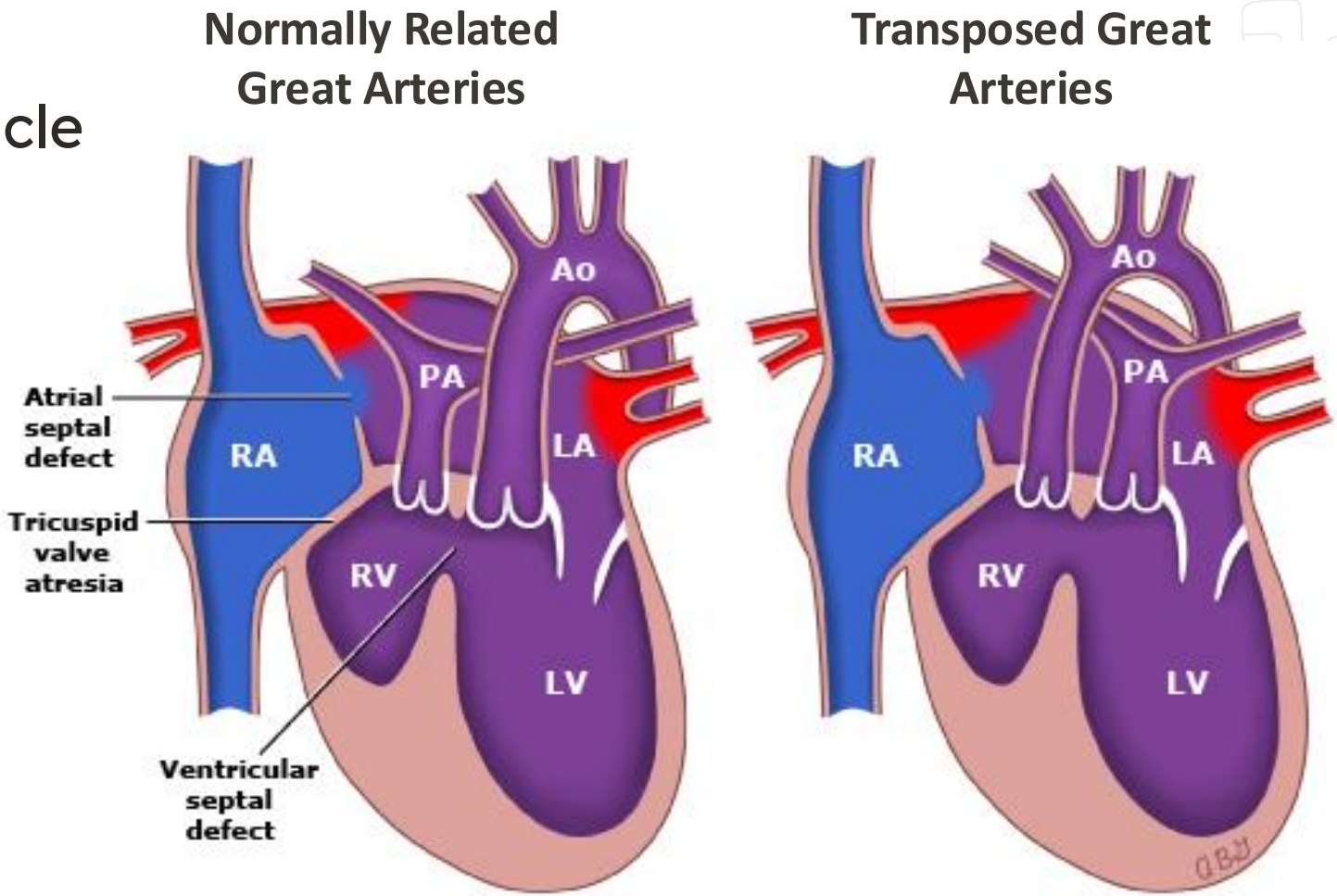
- Ventricular arrhythmias
- RVOT patch dehiscence
- Pseudoaneurysm rupture
- Stent fracture
- Endocarditis (higher risk with catheter PVR)
- Intervention
  - Lidocaine/amiodarone
  - Fluid resuscitation
  - Intubation and sedation
  - Antibiotics



2 year old with TOF post “valve sparing” repair

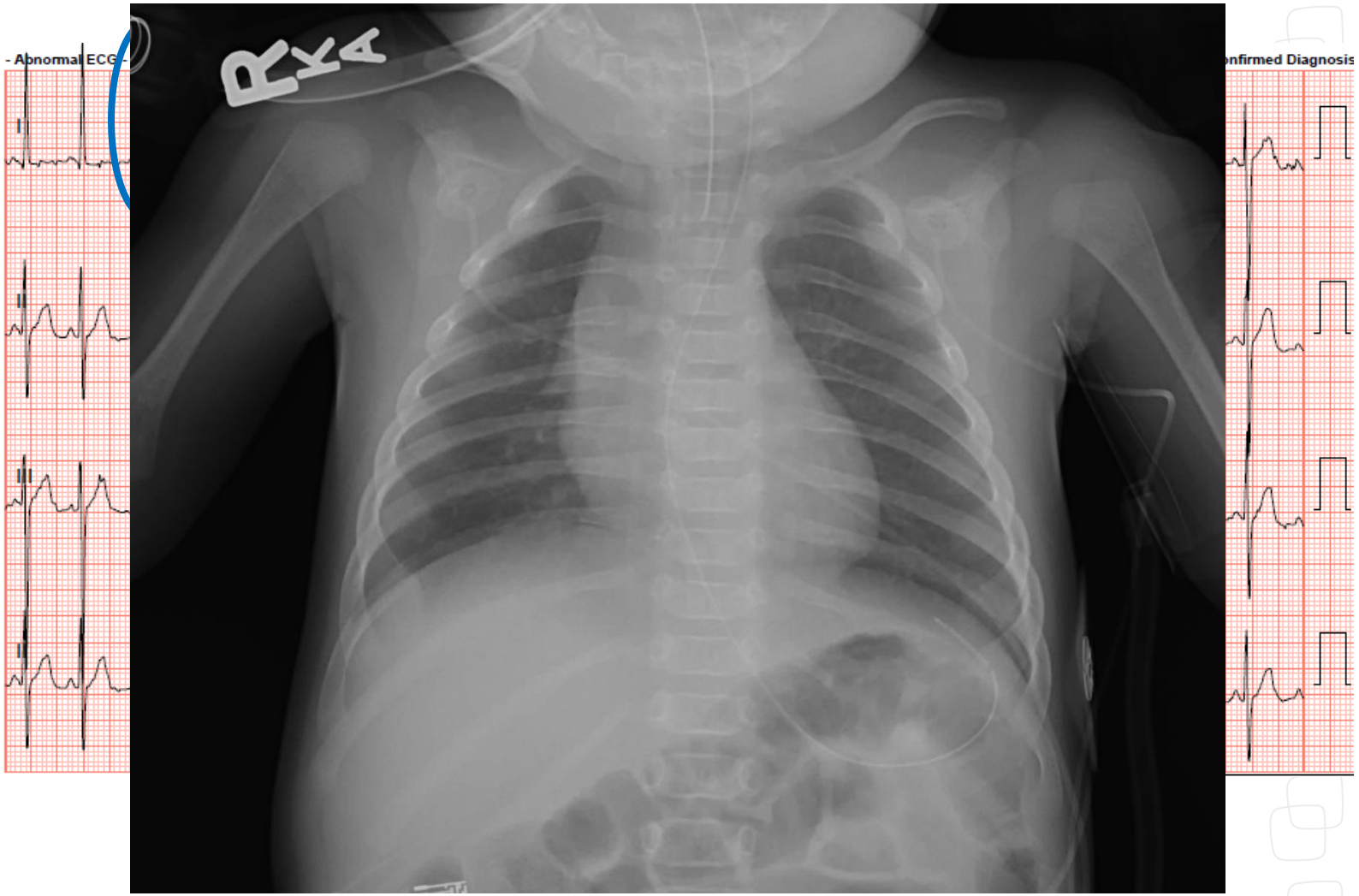
Tricuspid Atresia

- Absent connection between the right atrium and the right ventricle
- Right to left atrial level shunt
  - May need to be enlarged
- Normally related (cyanotic) vs transposed great arteries (acidotic)
  - TA/TGA associated with COA
- VSD
  - If VSD is small, cyanosis or decreased cardiac output will occur
  - May require emergent intervention

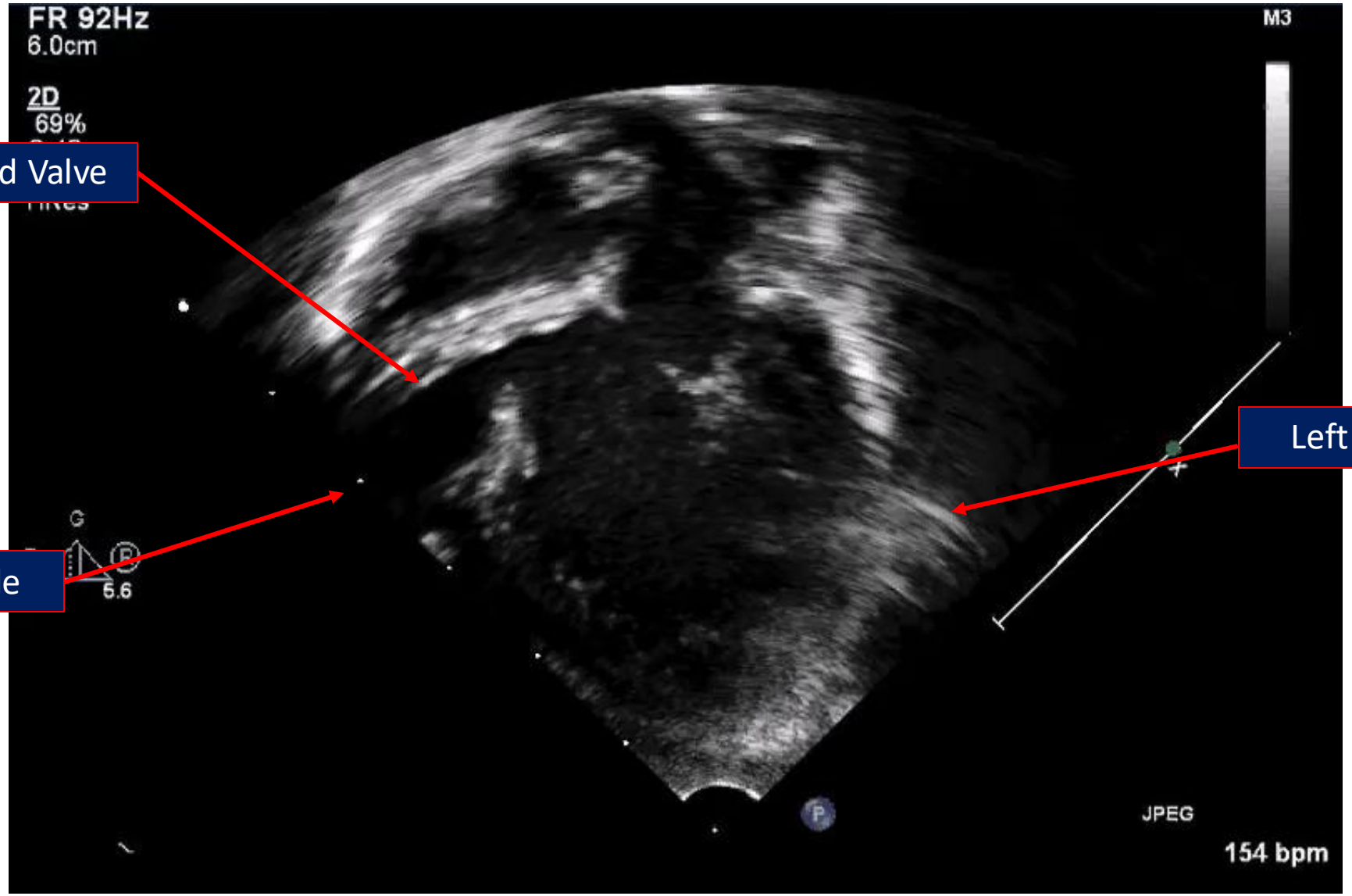


Tricuspid Atresia

- CXR: heart small-mildly enlarged, pulmonary vascular markings increased or decreased
- EKG: left axis deviation
- Exam: +/- systolic ejection murmur at LUSB
- O2 sat goal: 75-85%
- Likely need PGE
  - Depends on pulmonary stenosis, VSD size, and presence of coarctation of the aorta



Single Ventricle Physiology



Atretic Tricuspid Valve

Left Ventricle

Right Ventricle

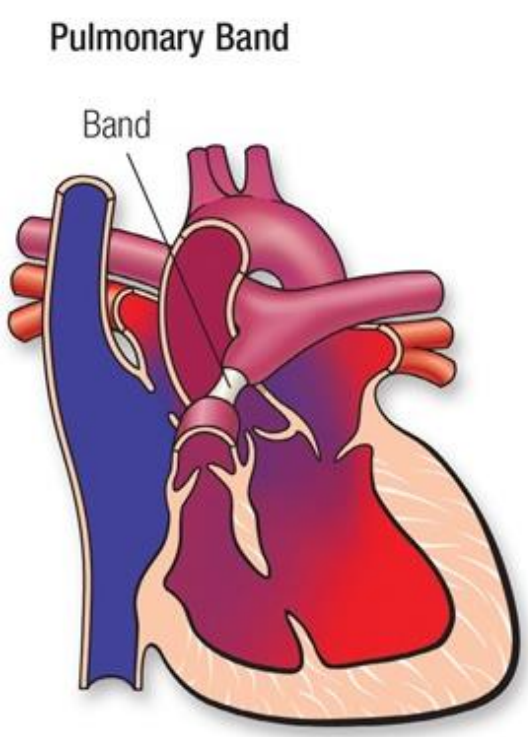
Phoenix Children's

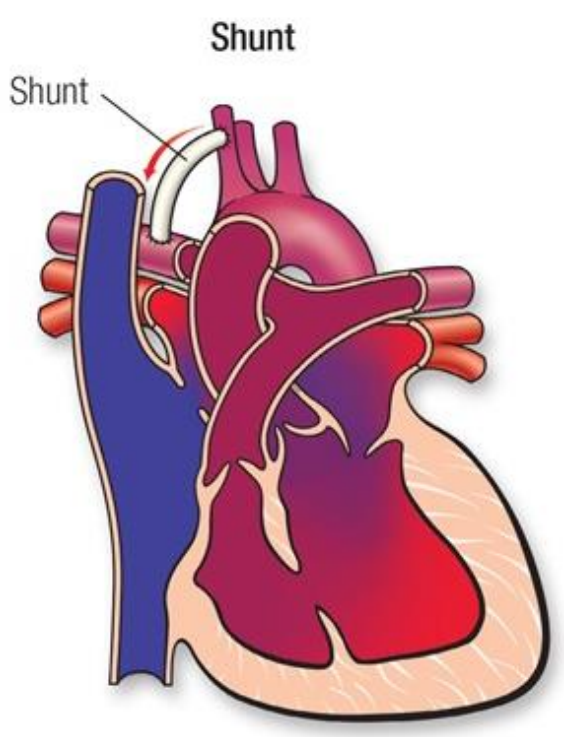
Tricuspid Atresia – Staged Surgical Palliation

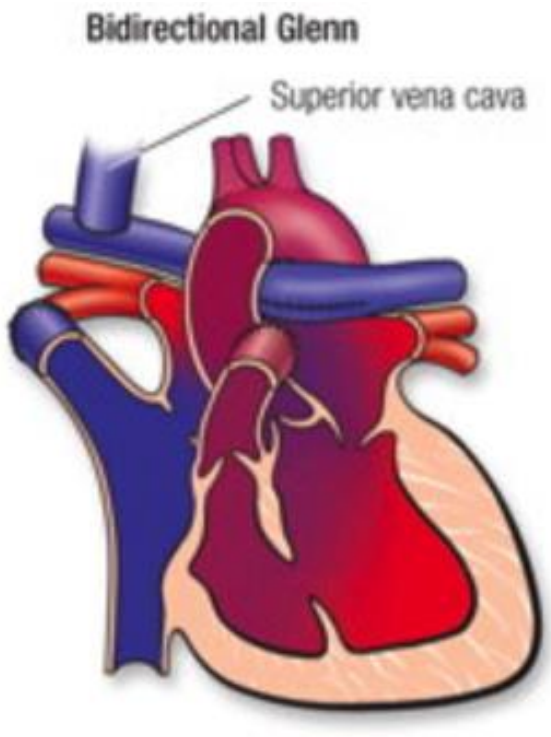
Stage 1

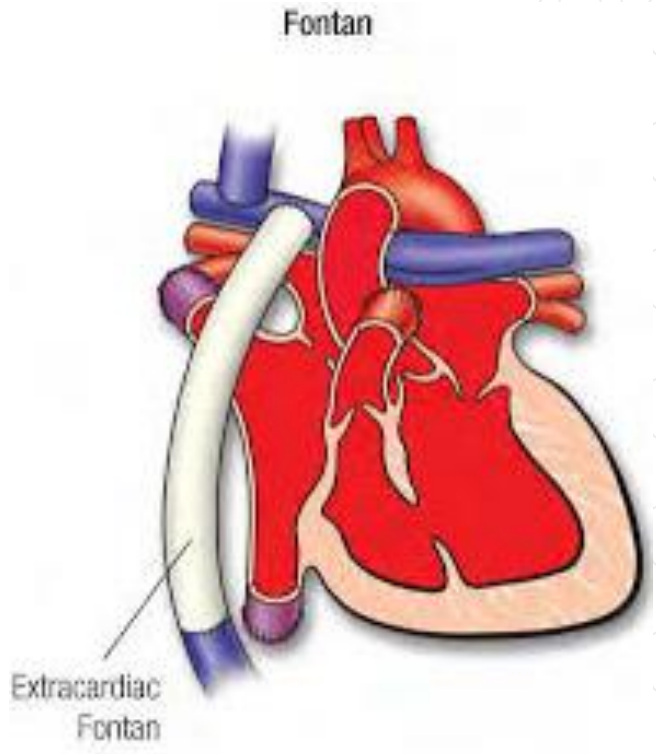
Stage 2

Stage 3









PA Band

Blalock-Taussig Thomas Shunt

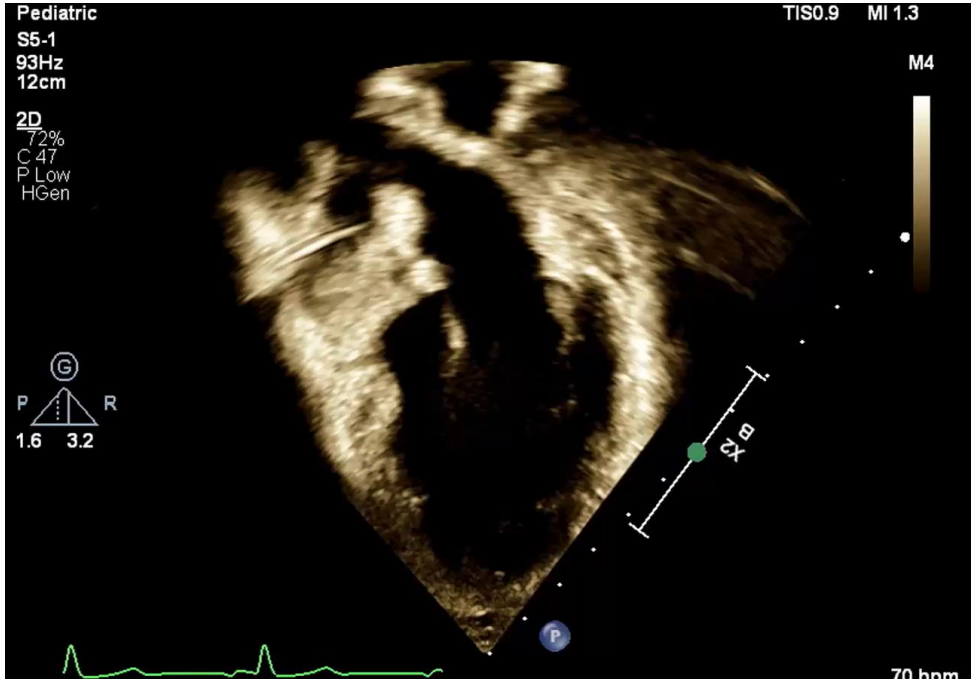
Glenn

Fontan

Tricuspid Atresia: Post Surgical Emergencies

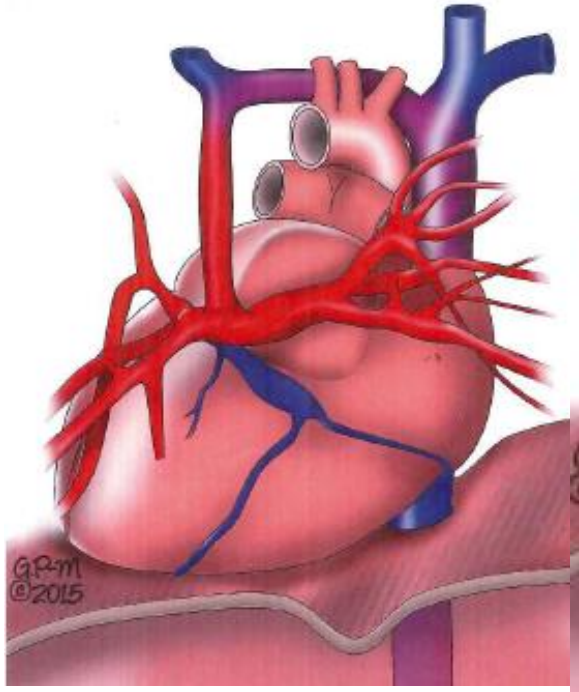
- After BTT shunt
  - Shunt occlusion
  - Respiratory failure
  - Cardiogenic shock
- After PA band
  - Severe hypoxia (band is too tight)
- After Glenn
  - Cardiac dysfunction
- After Fontan
  - Cardiac dysfunction
  - Respiratory failure
  - Liver failure

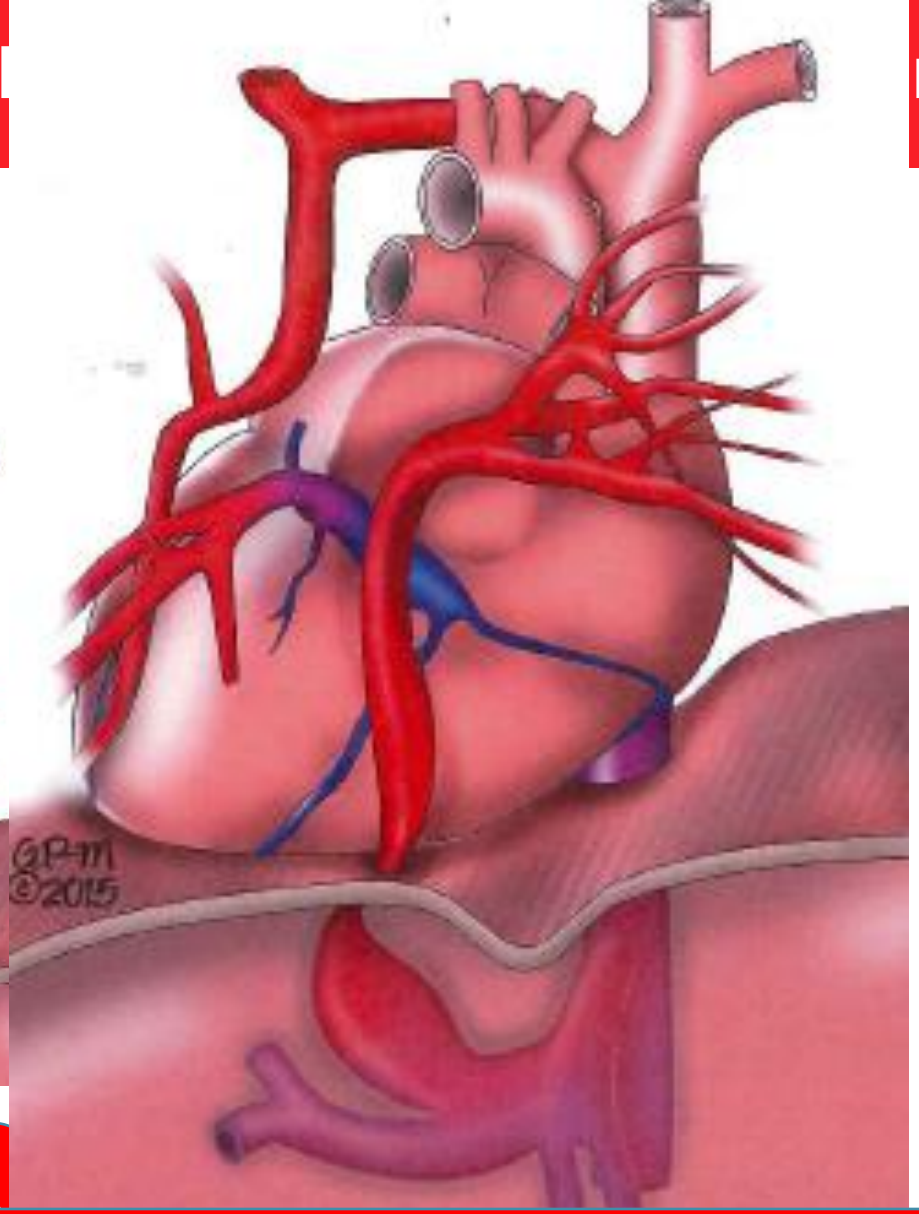
- Intervention
  - O2 if hypoxic (goal sats 75-85%)
  - Fluid resuscitation
  - +/- Mechanical ventilation
  - Inotropes

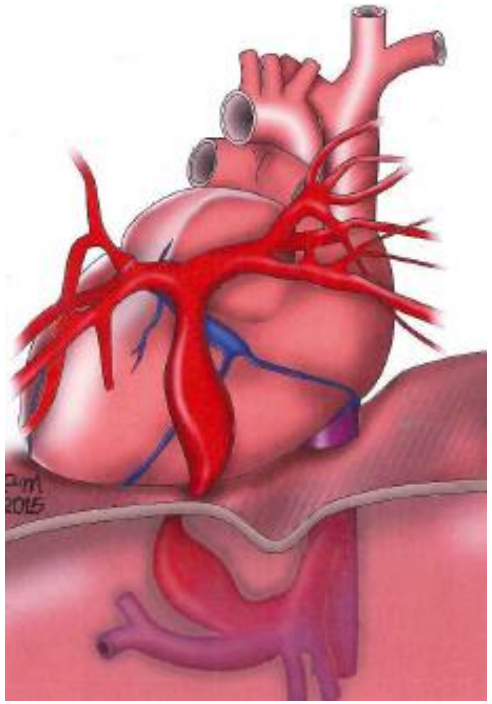


12 year old with tricuspid atresia post Fontan

Total Anomalous [illegible] rn







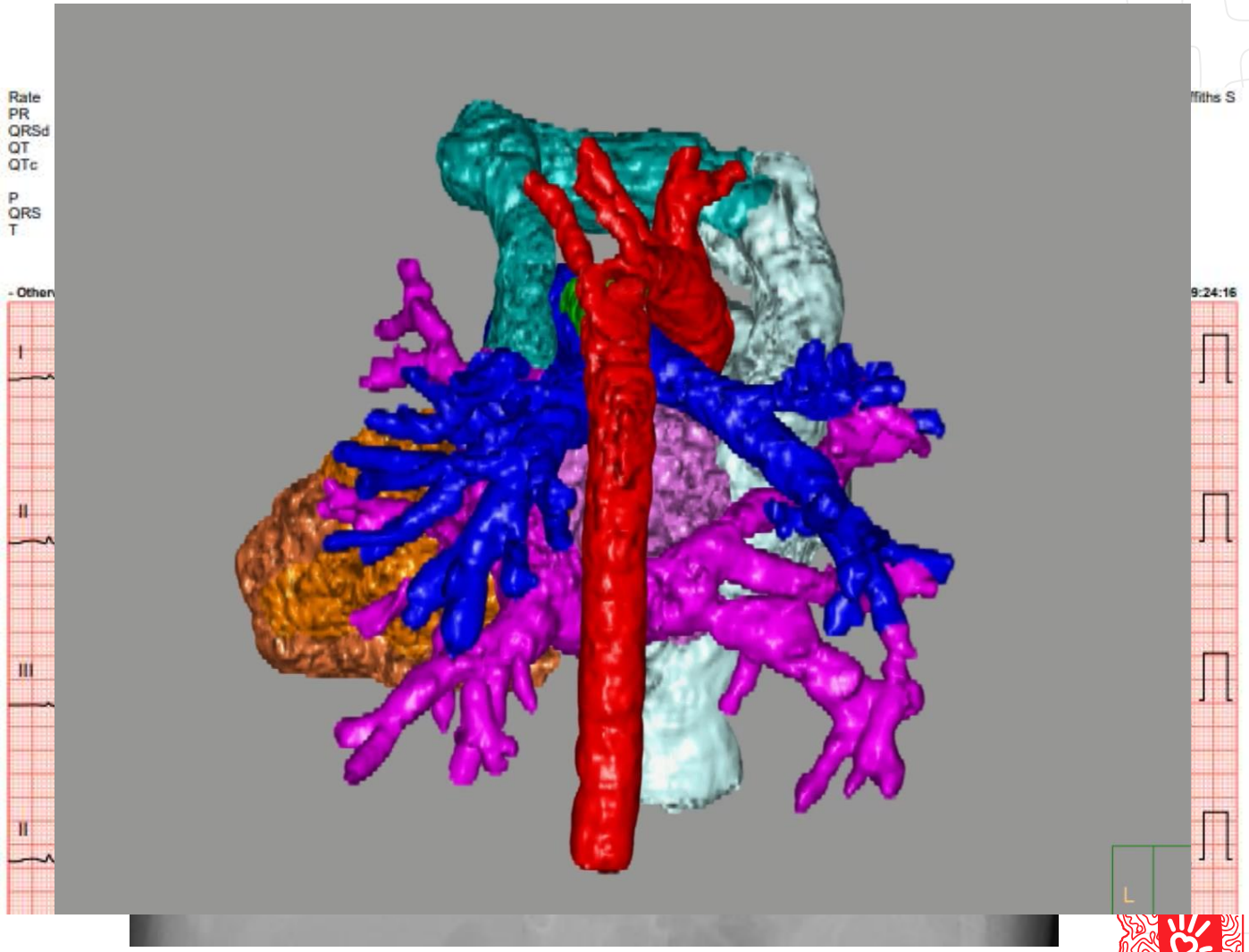
Type I Supra[illegible]
most con[illegible]

Type IV Mixed - least common
Combination of any 2 types

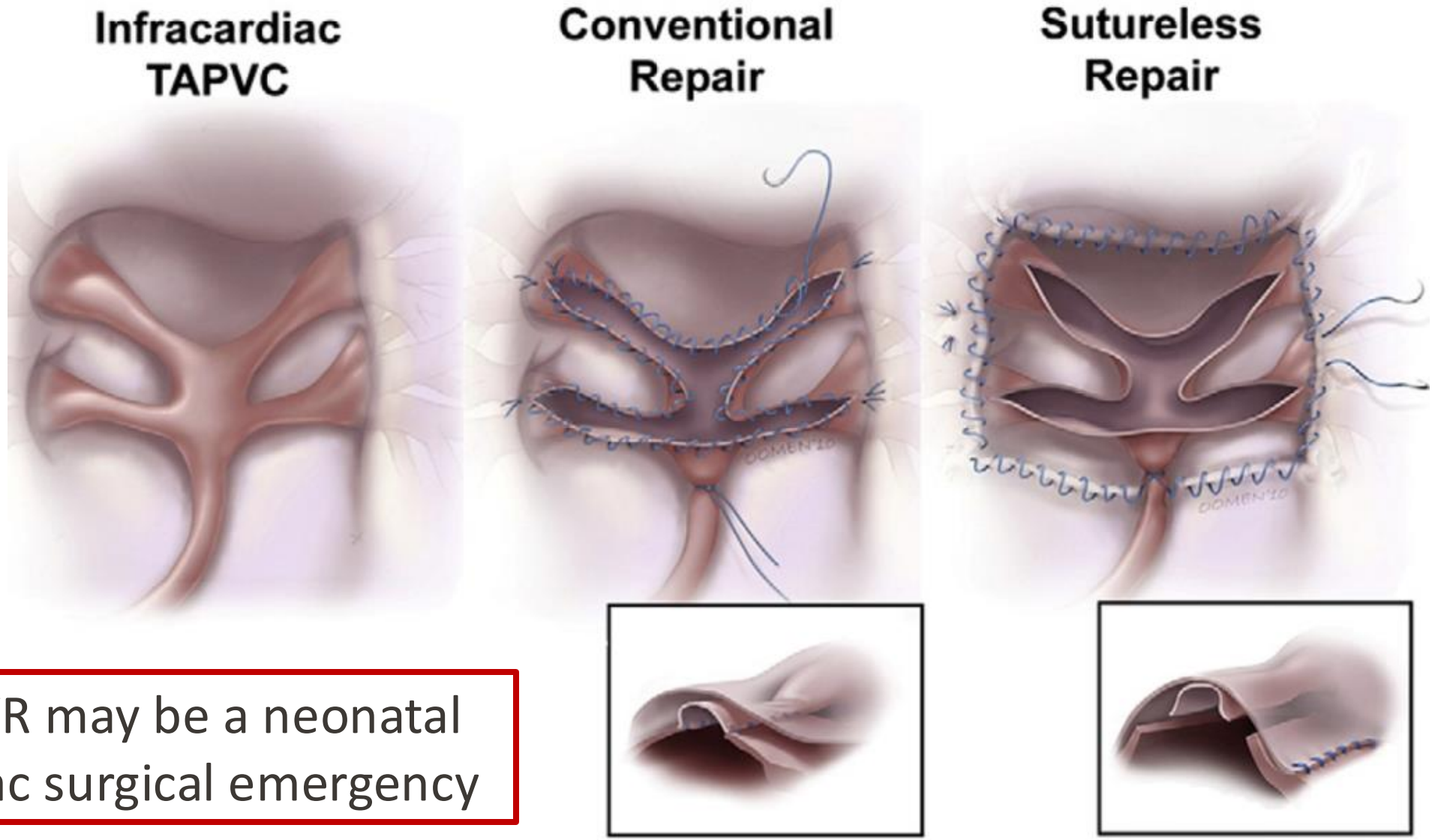
Type III Infracardiac
symptomatic

TAPVR: Presentation

- CXR
  - Supracardiac: “snowman heart”
  - RA/RV enlargement
  - Pulmonary edema with pulmonary venous obstruction
- EKG: RVH, RAE
- Exam (dependent on severity of pulmonary venous obstruction)
  - No murmur, loud S2
  - Infant with feeding difficulties and poor growth
  - Cyanosis, tachypnea, dyspnea, tachycardia
  - Metabolic acidosis, respiratory failure, cardiogenic shock
- O2 sat goal: >80%
- Do NOT give PGE!



TAPVR: Surgical Repair

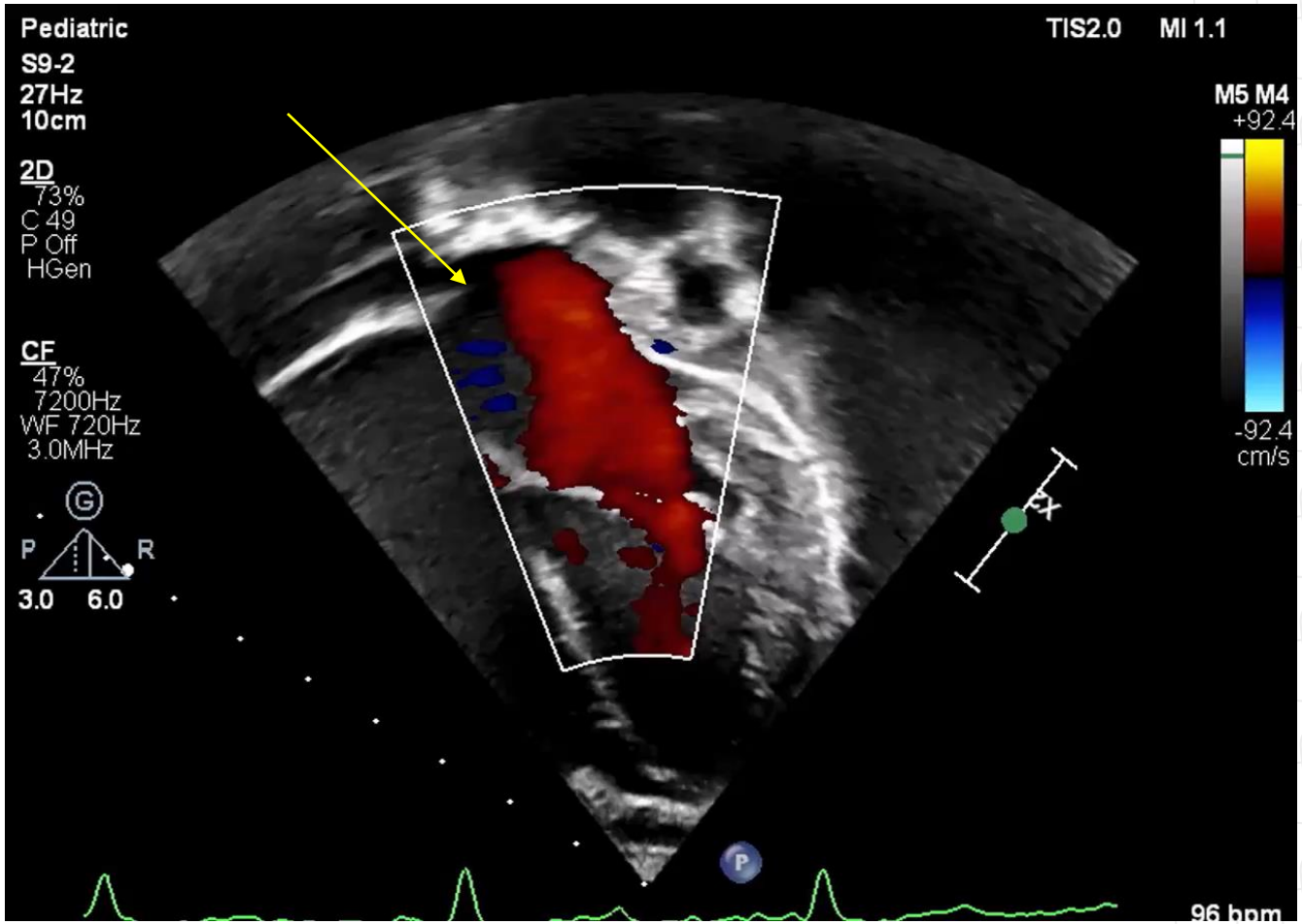


TAPVR may be a neonatal cardiac surgical emergency

Yanagawa et al, JTCVS2011;141:1346-54

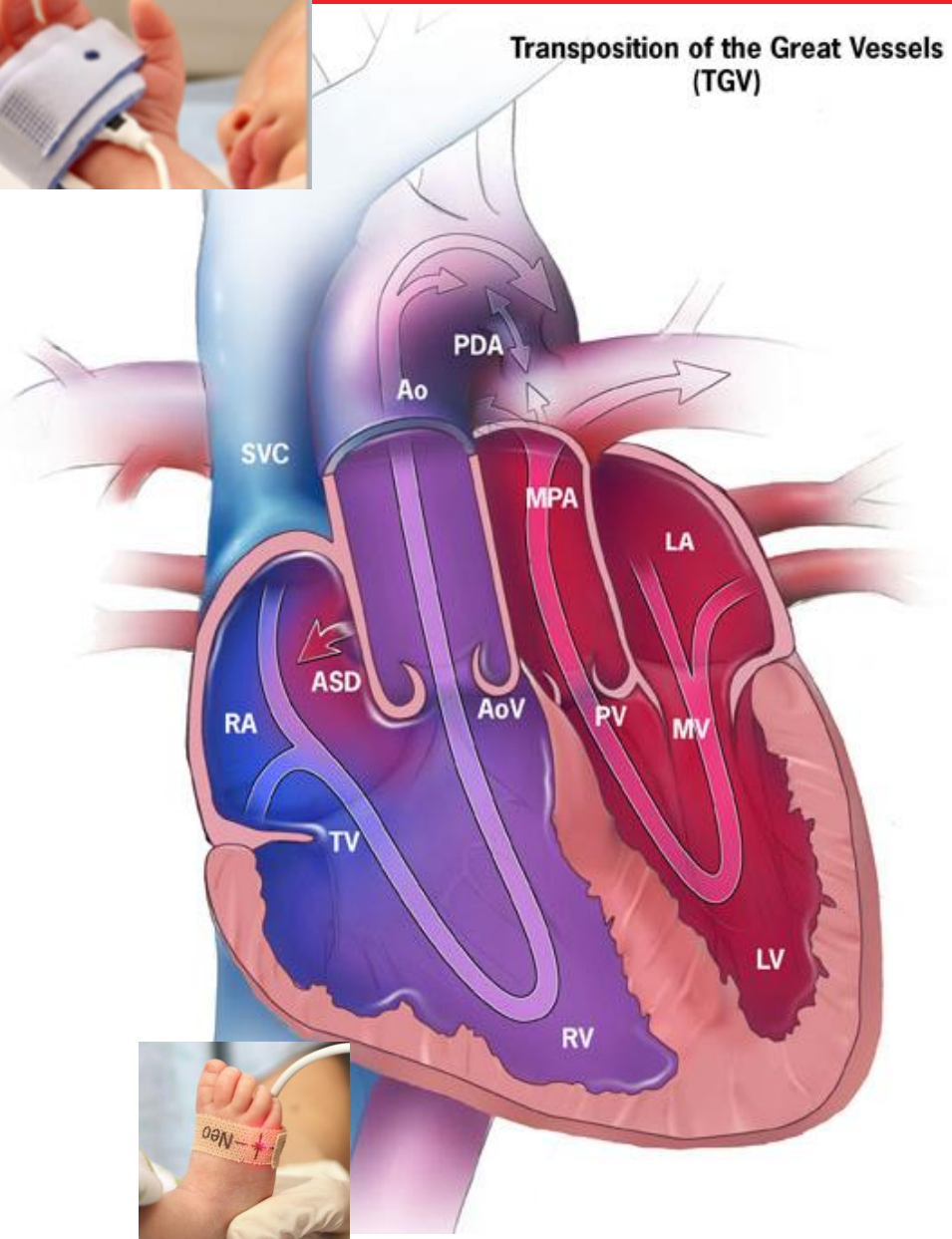
TAPVR: Post Surgical Emergencies

- Pulmonary vein anastomosis obstruction
  - Pulmonary edema
  - Hypoxia
  - Respiratory failure
  - Metabolic/respiratory acidosis
- Intervention
  - Oxygen
  - Intubation
  - Correct acid-base imbalance
  - Surgical or catheter intervention



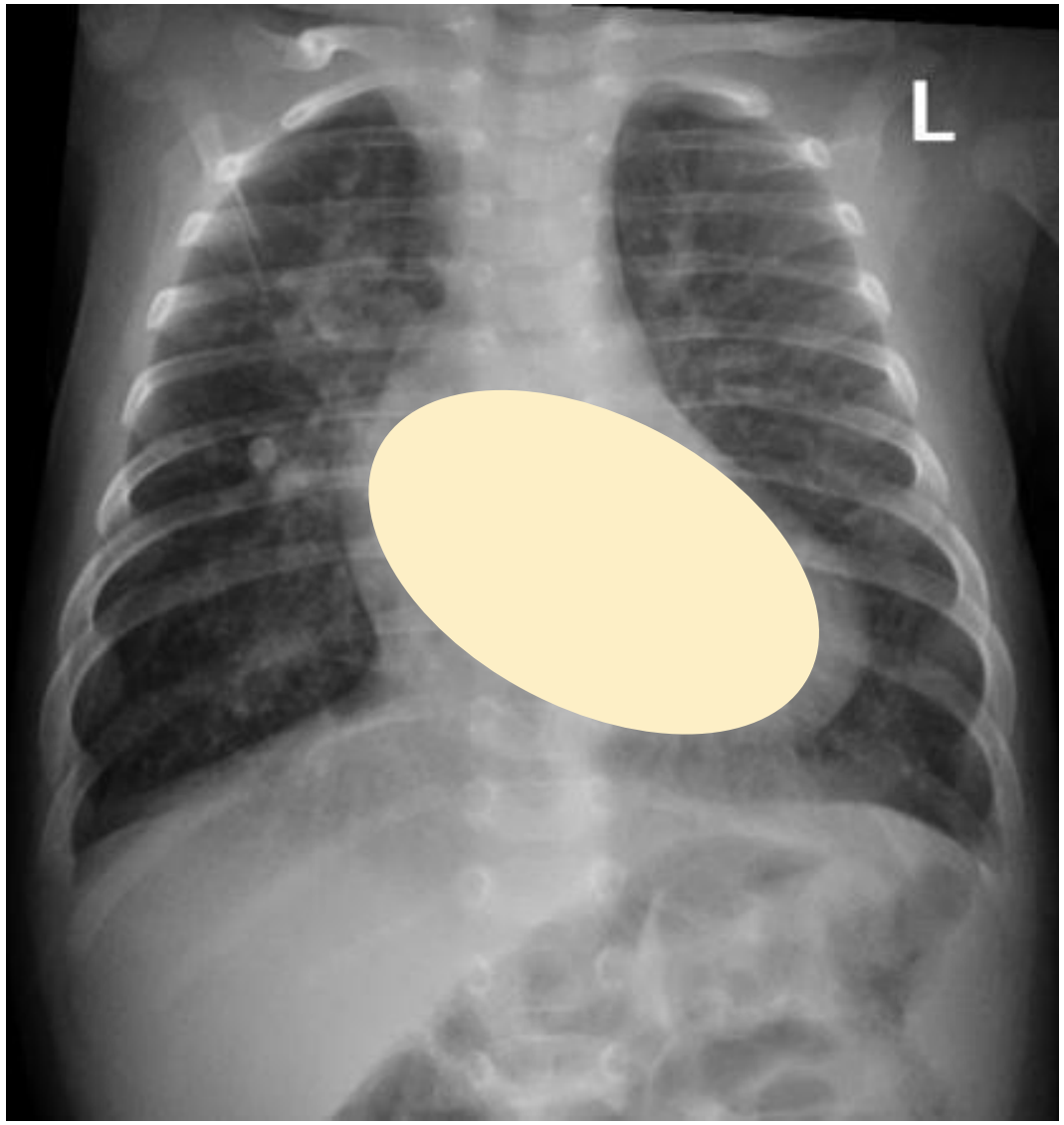
Transposition of the Great Arteries

- Most common cyanotic CHD presenting as a newborn
- Aorta arises from right ventricle and pulmonary artery arises from left ventricle
- Requires adequate intracardiac shunt to survive
  - ASD
  - PDA promotes atrial shunting
- “Reversed differential cyanosis”
  - O2 sat: Right UE < Lower extremity

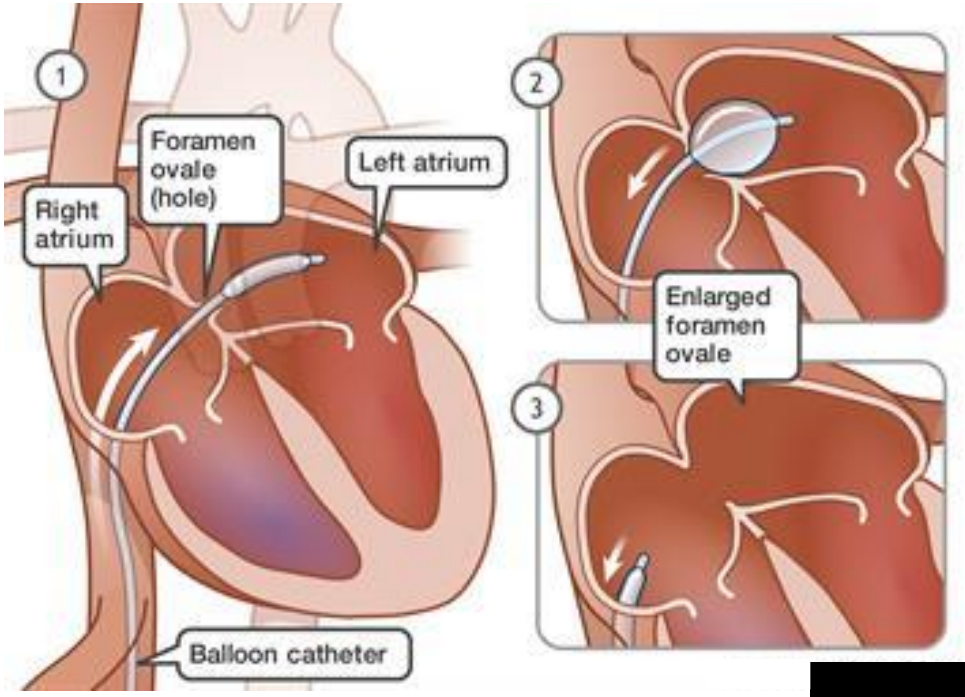


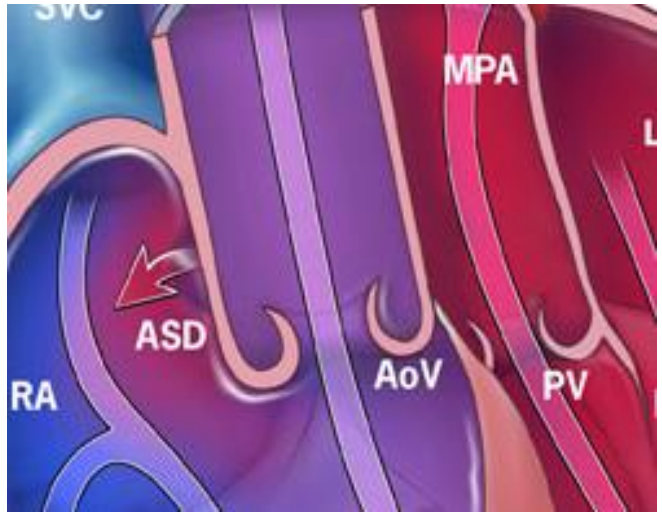
Transposition of the Great Arteries

- CXR: narrowed mediastinum, cardiomegaly
  - “egg on a string”
- Exam: single, loud s2, murmur may be absent
- O2 sat goal: >75%
- Requires PGE to maintain PDA
- May need mechanical ventilation for BAS
  - Supplemental O2 acceptable but will not significantly improve sats

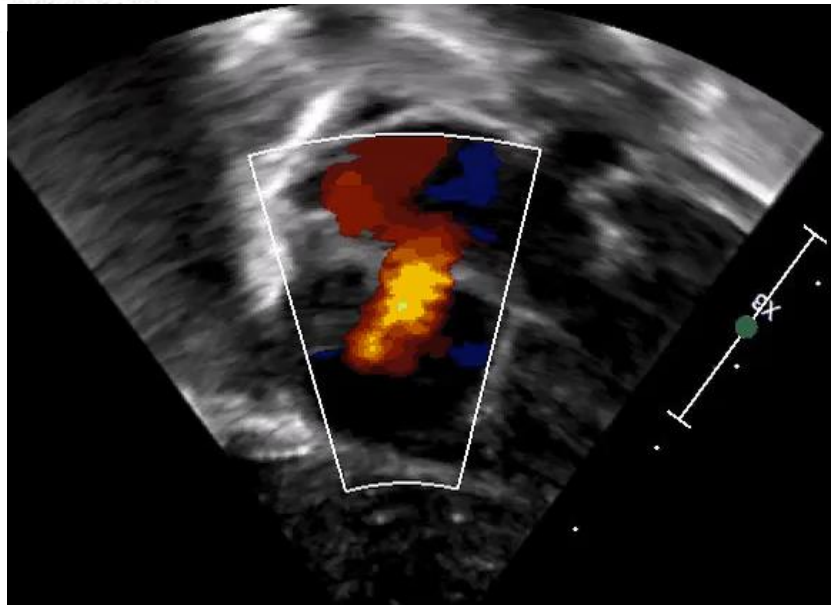


Balloon Atrial Septostomy





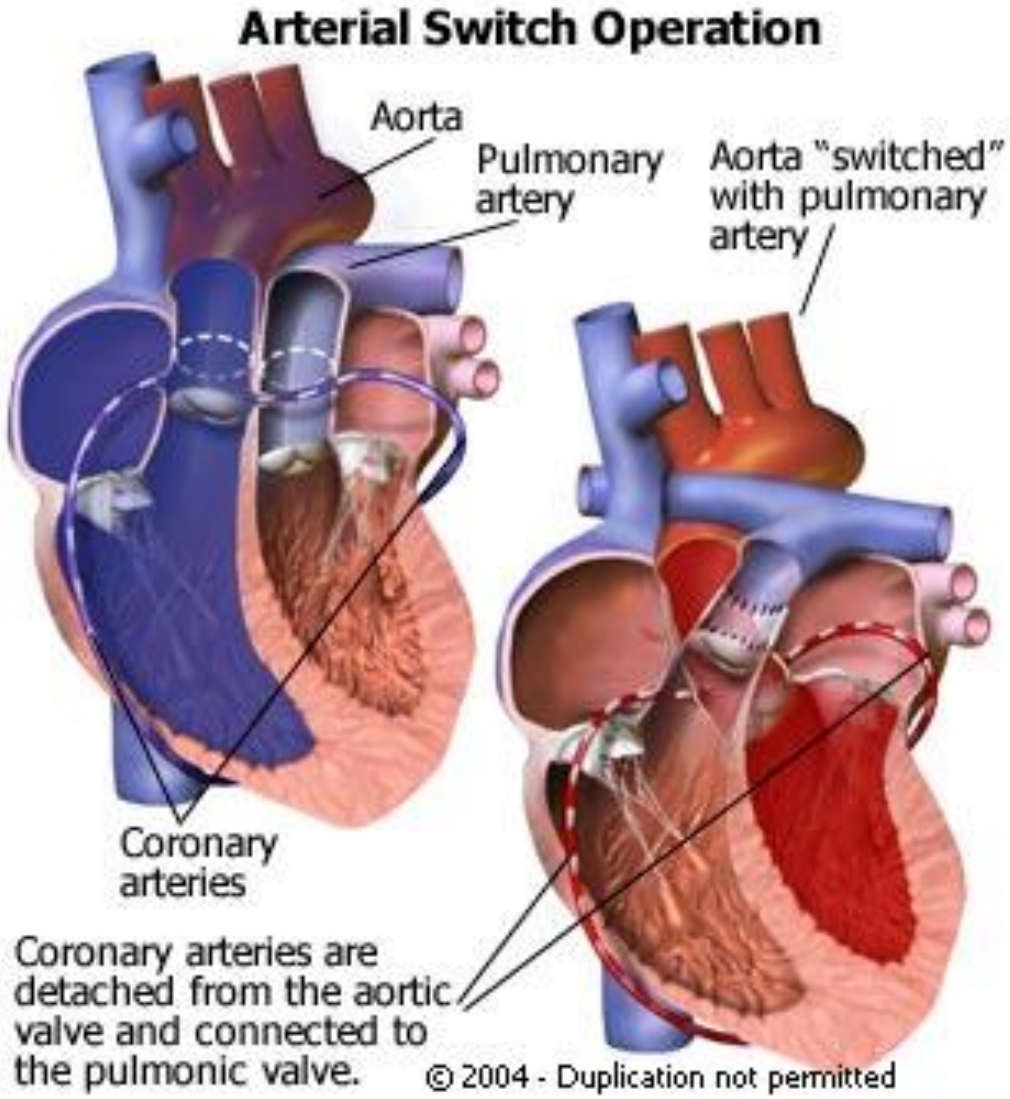
Goal: increase left to right atrial shunting



Phoenix Children's

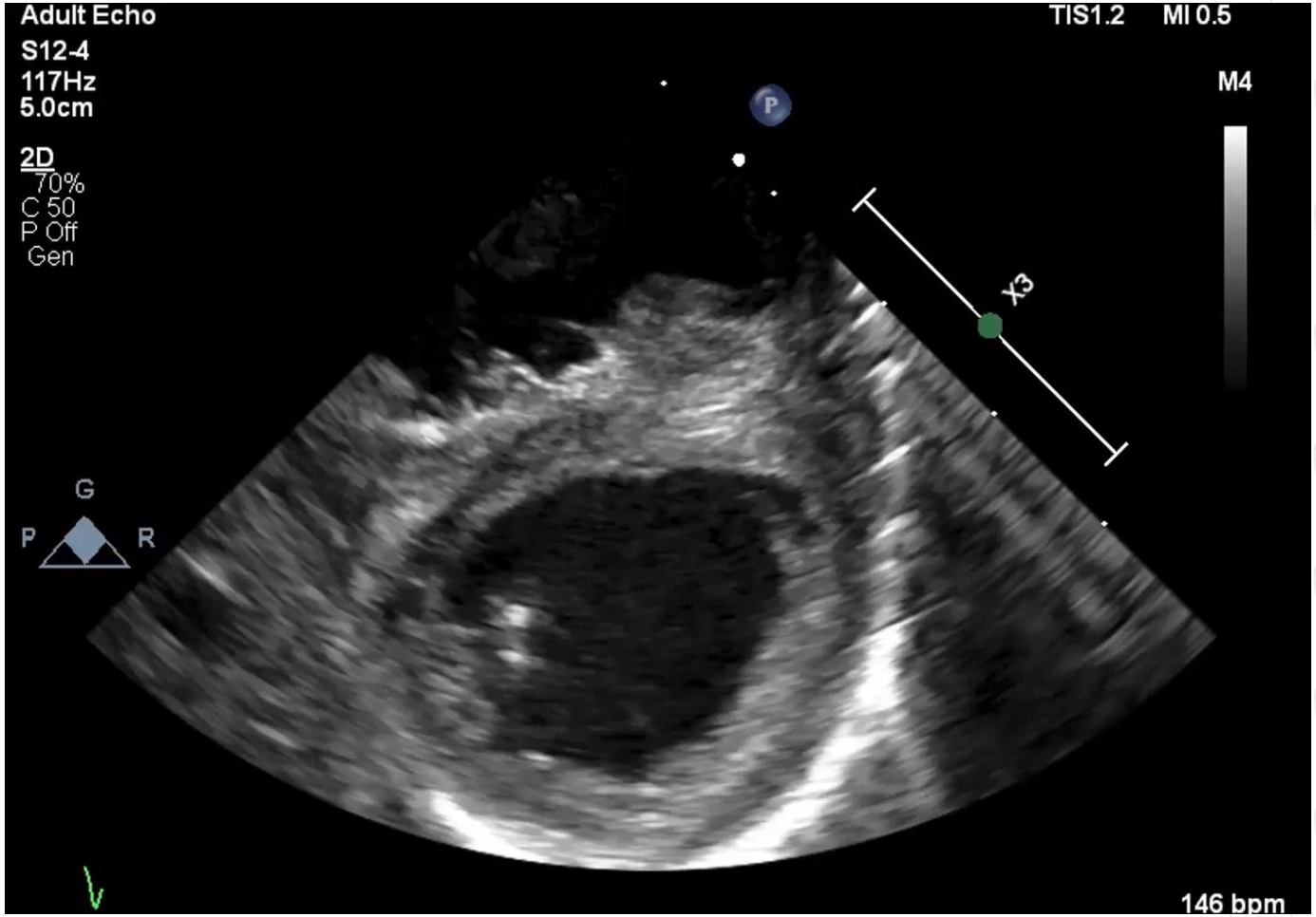
D-TGA: Surgical Repair

- Transect the PA and the aorta above the level of the coronary arteries
- Remove coronary arteries with button of tissue around the ostia
- Switch the arteries and suture in place
  - LeCompte – branch pulmonary arteries drape over the aorta anteriorly
- Suture in coronary arteries into the new aorta



D-TGA: Post Surgical Emergencies

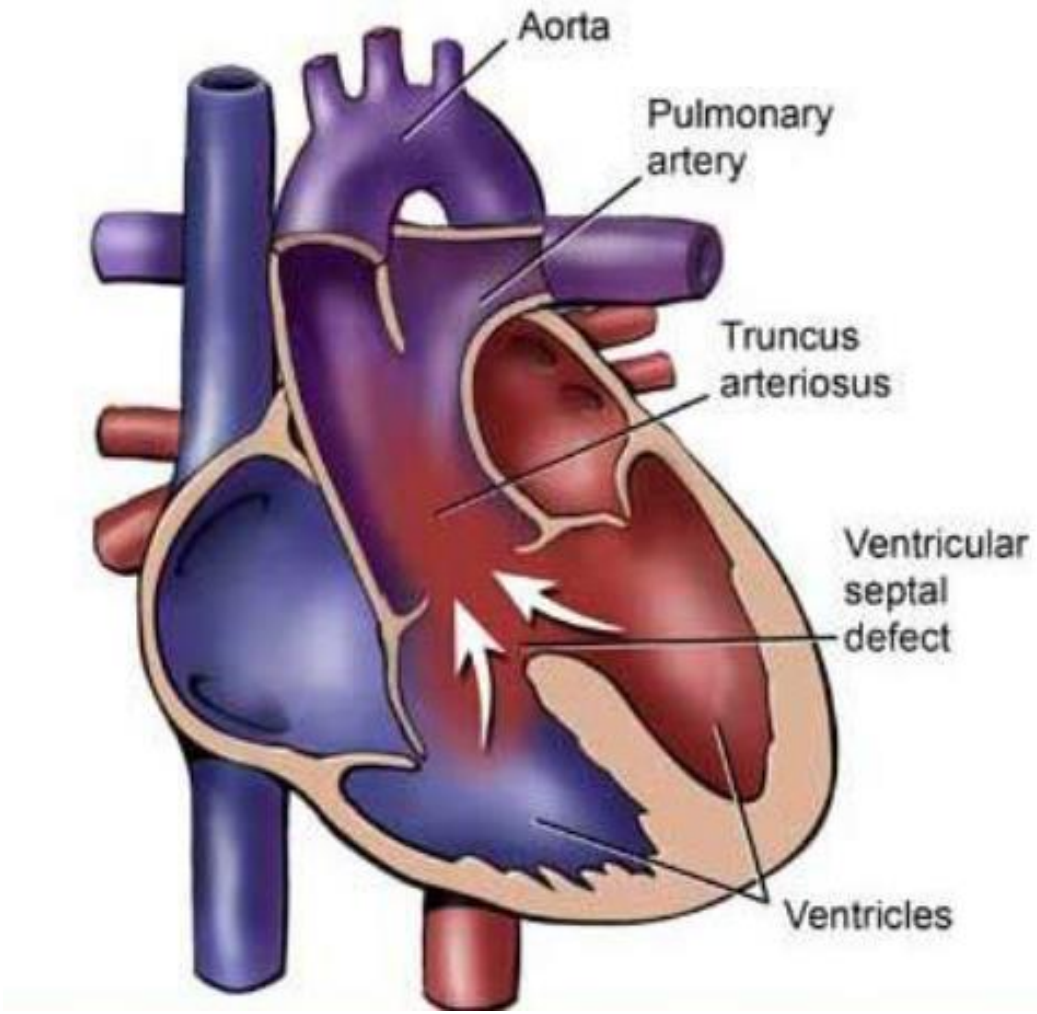
- Coronary ischemia
- Ventricular arrhythmias
- Intervention
  - Oxygen
  - Intubation with mechanical ventilation
  - Decrease metabolic demand
  - Lidocaine or amiodarone



5 day old with D-TGA post arterial switch operation

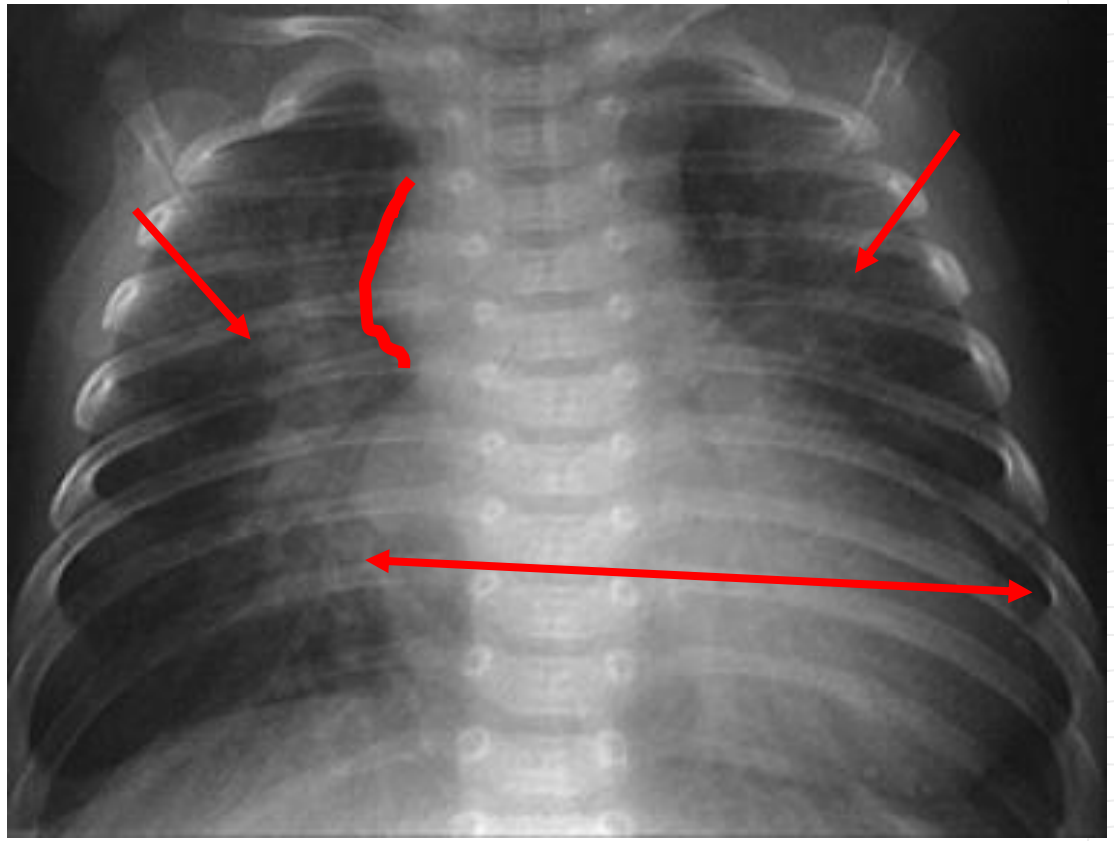
Truncus Arteriosus

- Common artery supplying systemic, pulmonary, and coronary artery circulations
- One semilunar (truncal) valve
  - Stenotic and regurgitant
- Malalignment VSD with overriding common arterial trunk
- 20-25% with right aortic arch
- 20-25% with DiGeorge syndrome



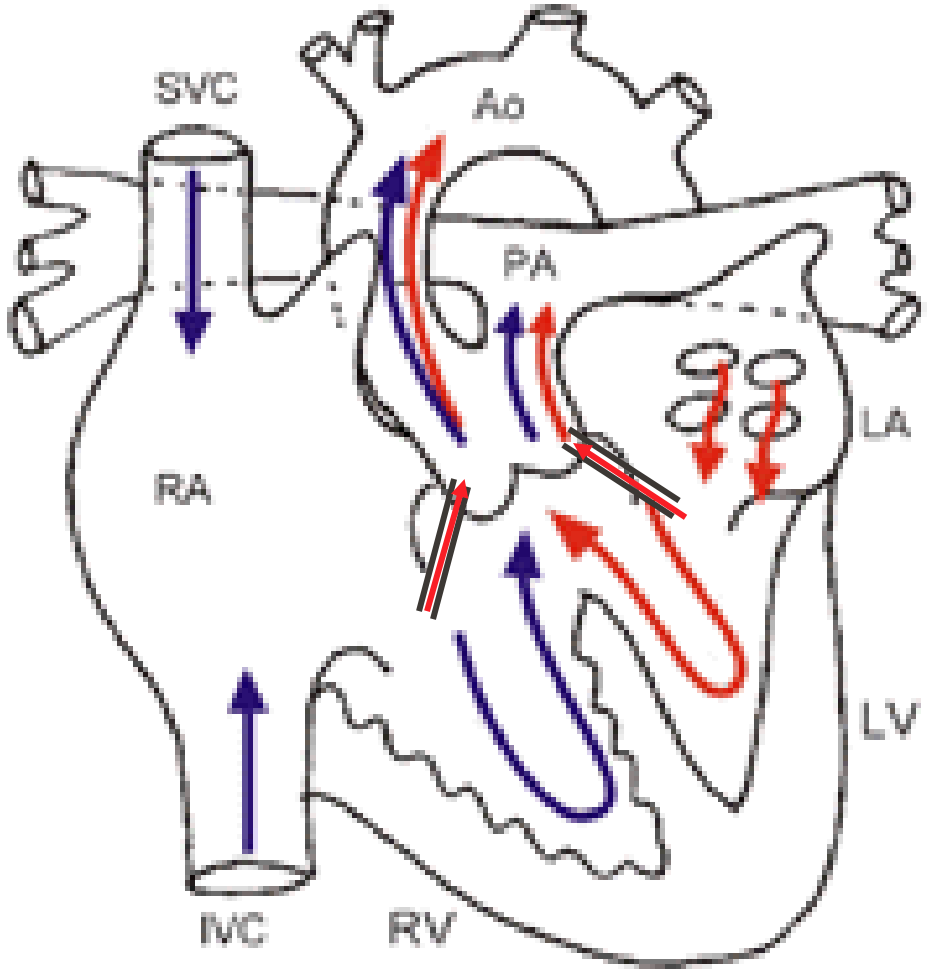
Truncus Arteriosus

- CXR: increased pulmonary vascular markings, cardiomegaly, right aortic arch
- EKG: biventricular hypertrophy
- Exam: holosystolic murmur, diastolic murmur, loud single S2, bounding pulses
- O2 sat goal: 75-85%



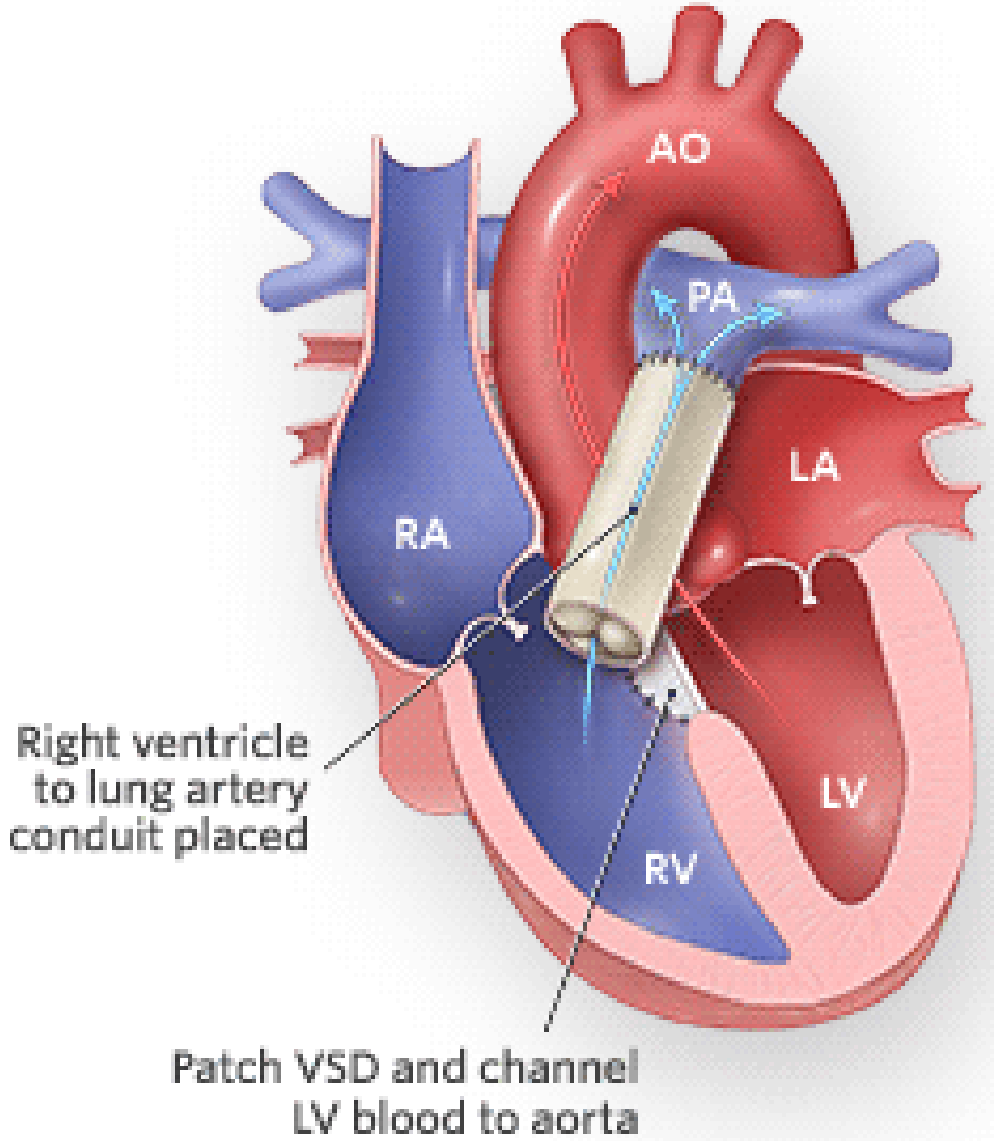
Truncus Arteriosus: Physiology

- At birth, high pulmonary vascular resistance keeps pulmonary and systemic flow balanced
- At 2-6 weeks, the resistance falls resulting in overcirculation to the pulmonary arteries leading to pulmonary edema
  - Bounding pulses, tachycardia
  - Tachypnea, sweating, poor feeding
  - Decreased coronary artery perfusion leading to ischemia!



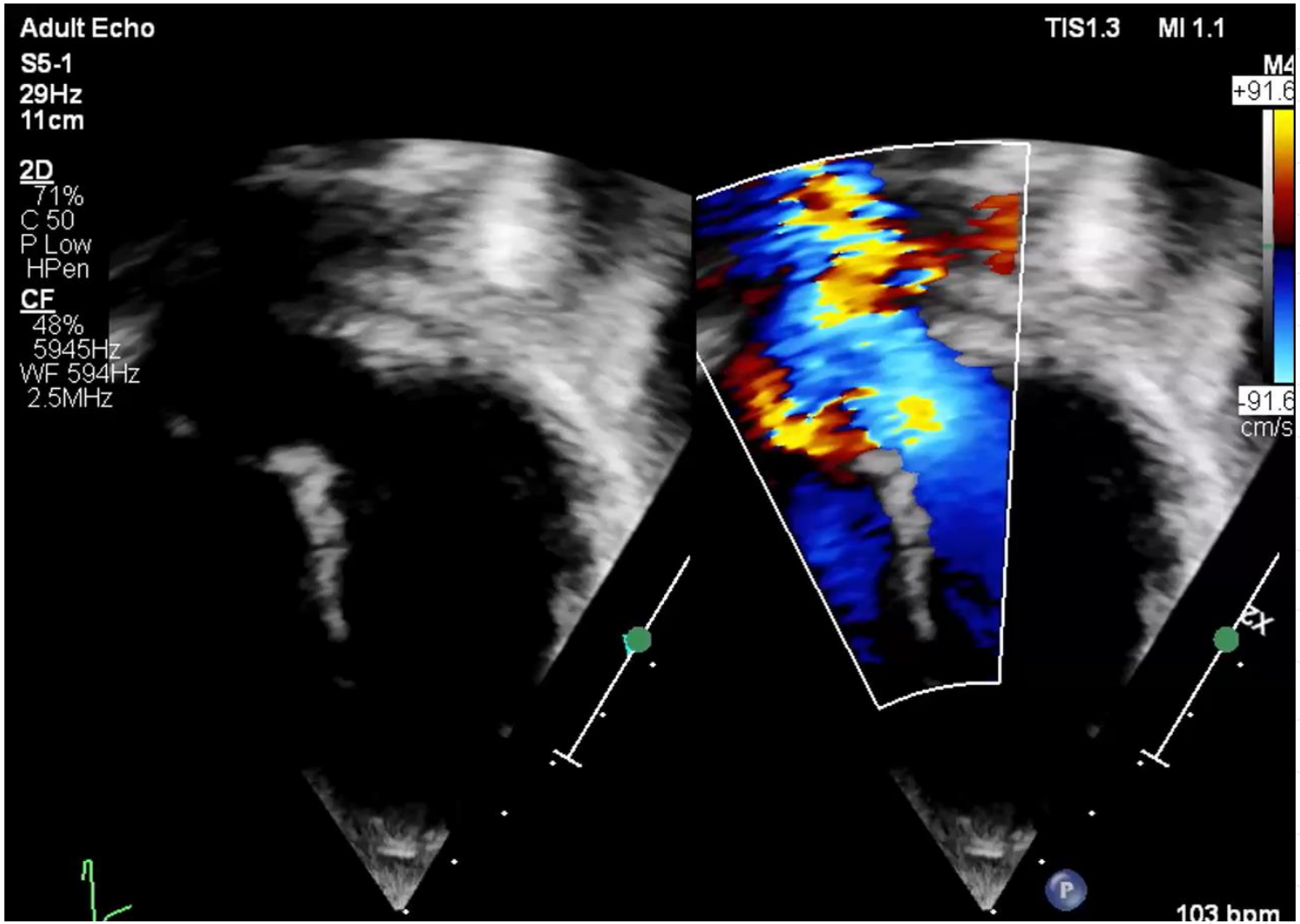
Truncus Arteriosus: Surgical Repair

- Close the VSD
- Separate the branch pulmonary arteries from common trunk
- Place RV-PA conduit
- Closure of ASD/PFO
- Possibly repair the truncal valve



Truncus Arteriosus: Post Surgical Emergencies

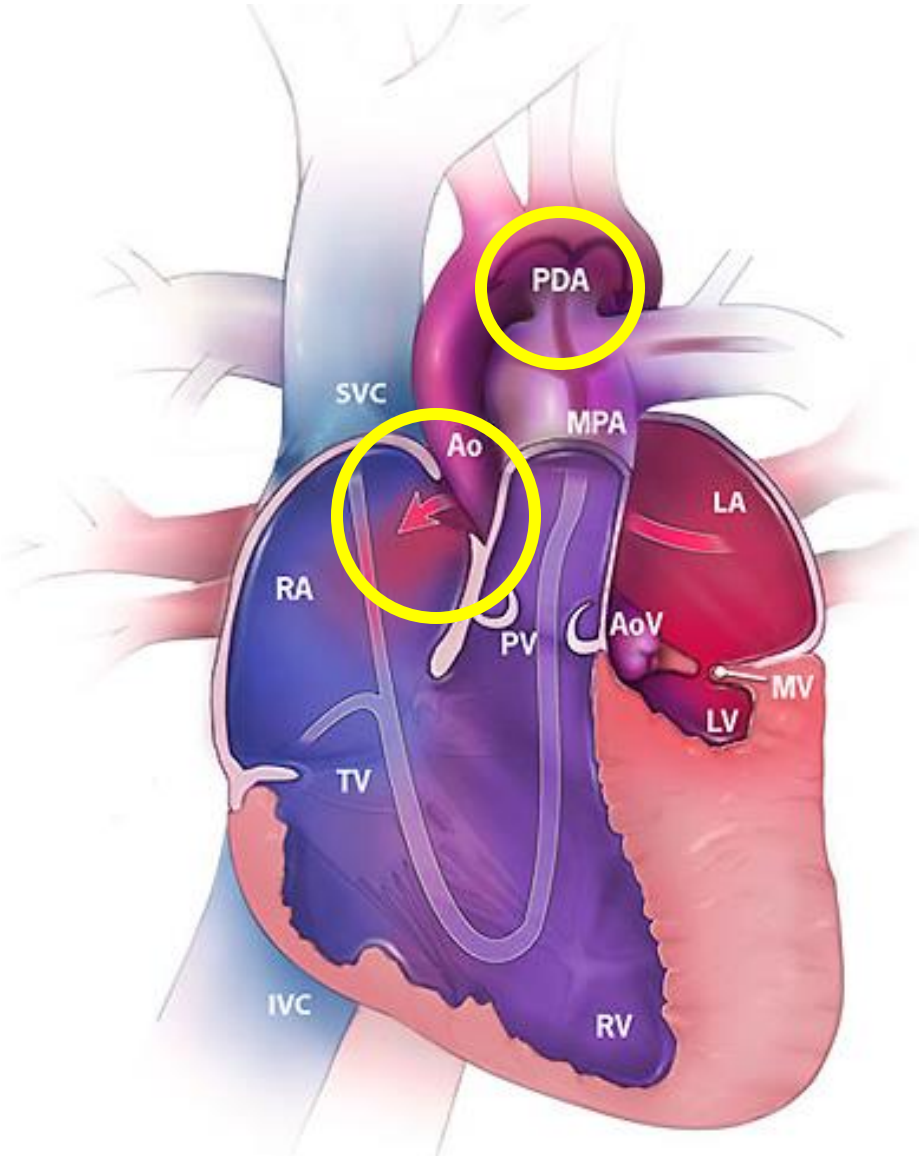
- Ventricular arrhythmias
- Coronary ischemia (with truncal valve regurgitation)
- Pseudoaneurysm rupture
- Intervention
  - Antiarrhythmics
  - Fluid resuscitation
  - Intubation
  - Pressors



3 year old post truncus arteriosus repair now with severe truncal valve regurgitation

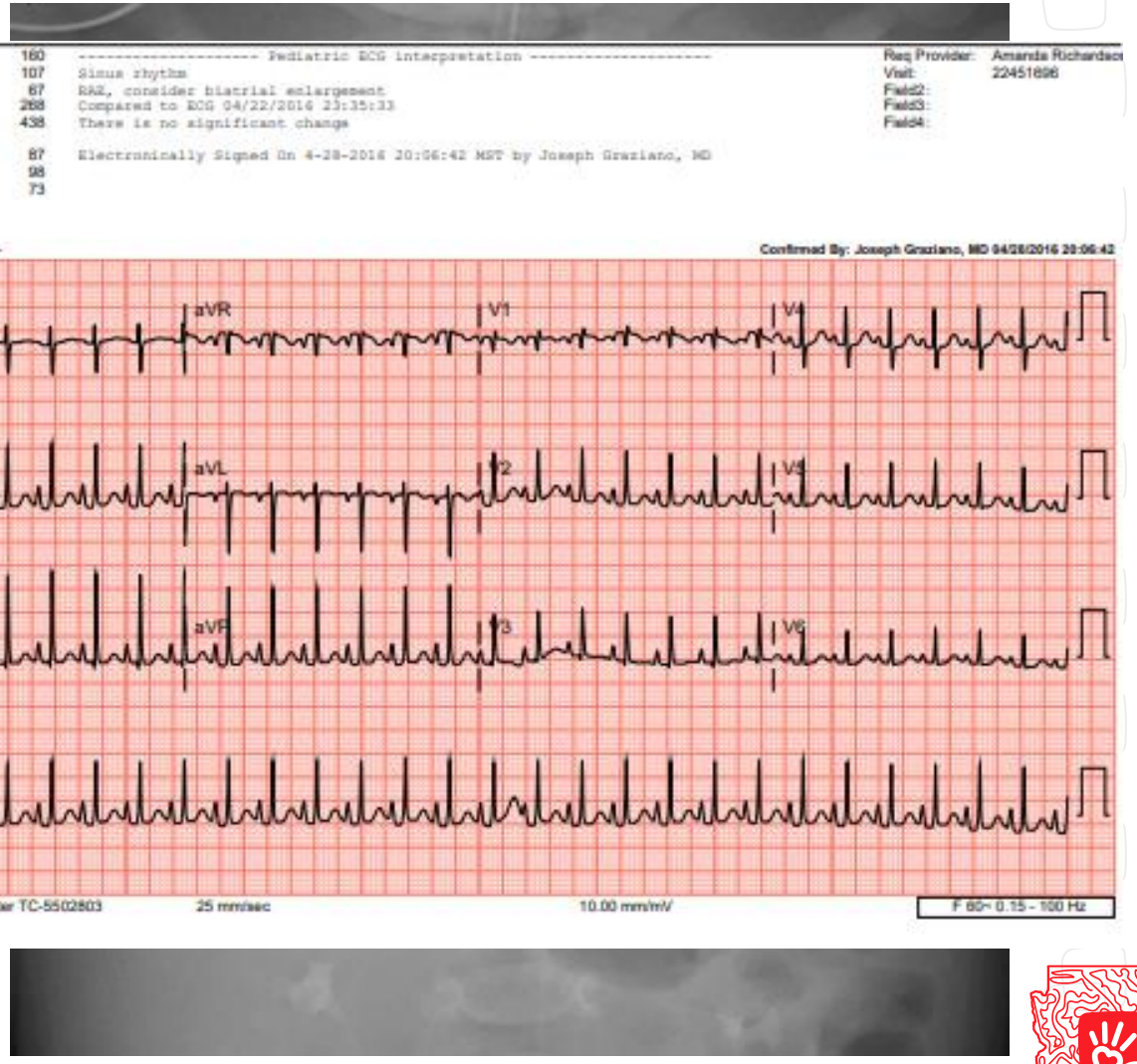
Hypoplastic Left Heart Syndrome

- Small left heart structures (mitral/aortic valves, LV, aorta)
- Ductal dependent for systemic circulation
- Foramen ovale might be the only egress of the pulmonary venous flow from the left atrium
  - Restrictive foramen ovale associated with profound cyanosis and worse outcomes
  - If detected prenatally, intervention may be possible



Hypoplastic Left Heart Syndrome

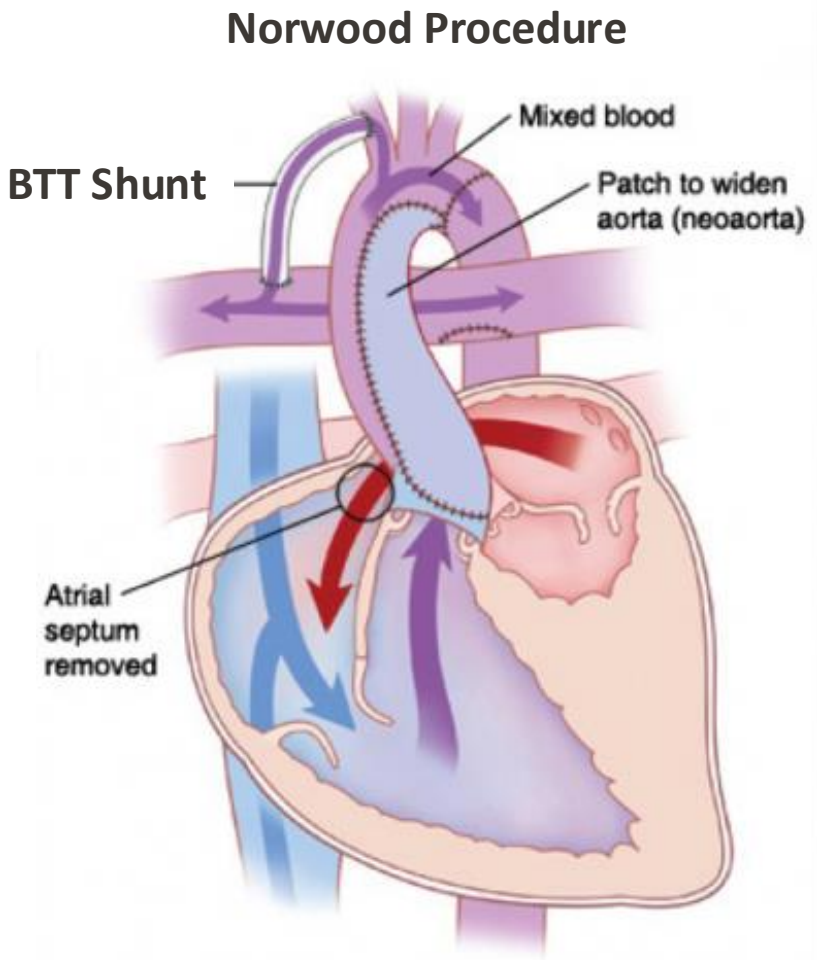
- CXR: prominent RA border, increased pulmonary vascular markings (with restrictive atrial septum)
- EKG: RVH, RAE
- Exam: systolic ejection murmur, loud S1, cool extremities with dampened pulses, cyanosis, respiratory distress
- O2 saturation: 75-85% (goal)

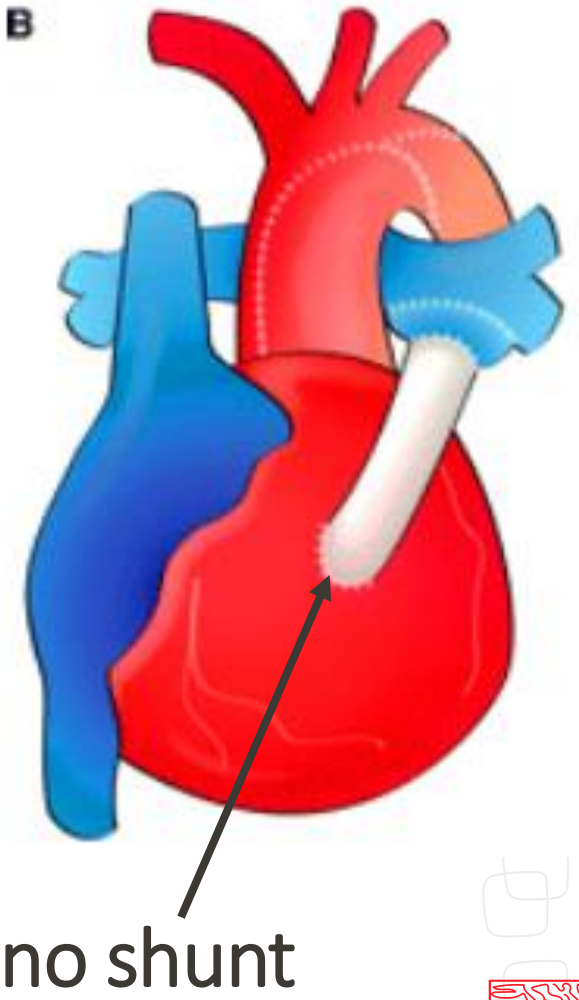


HLHS: Staged Palliative Surgery

Norwood (Stage 1) operation in the neonatal period

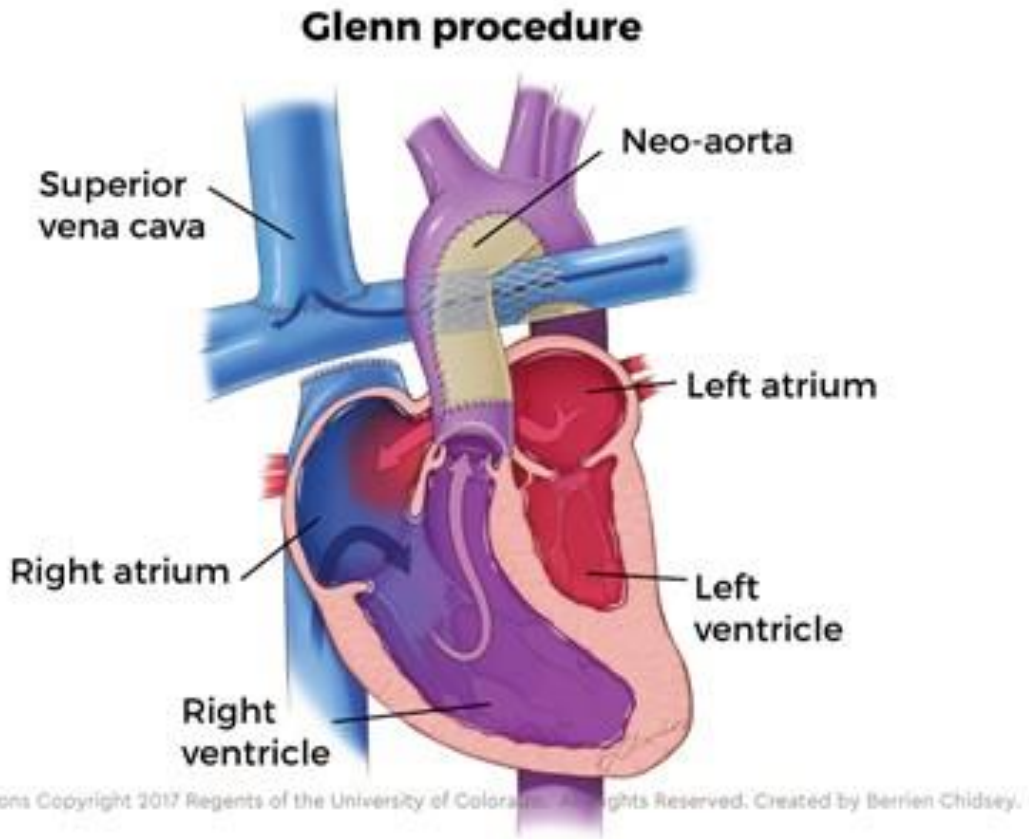
- Aortic arch reconstruction
- Atrial septectomy
- DKS anastomosis (connect aorta and pulmonary artery)
- Place BTT shunt



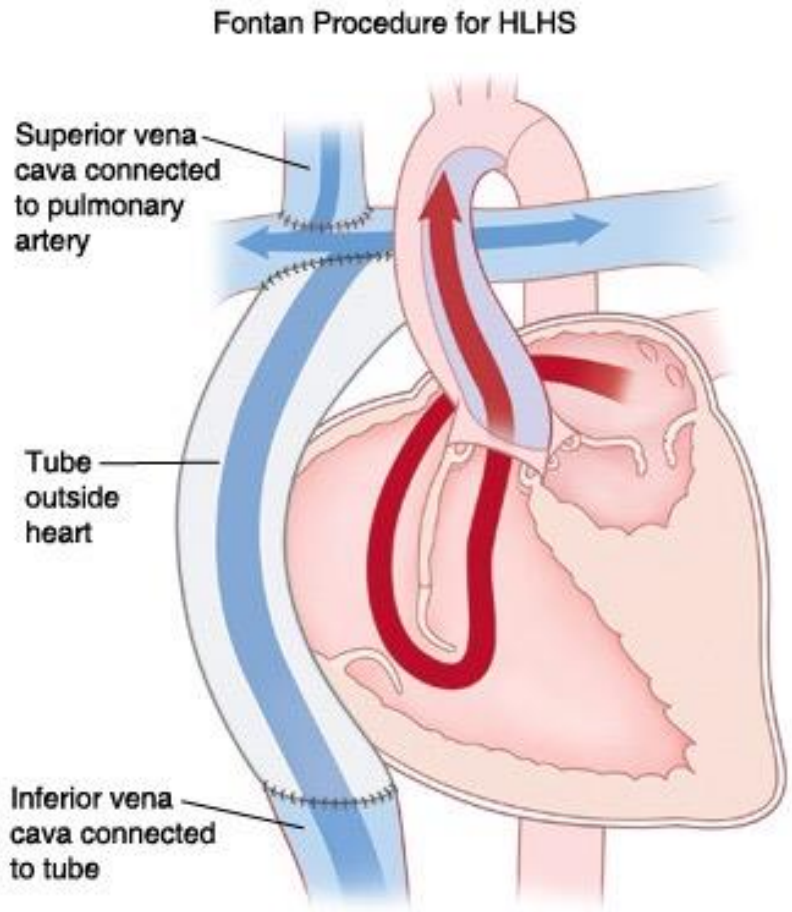


HLHS: Staged Palliative Surgery

Glenn at 4-6 months of age



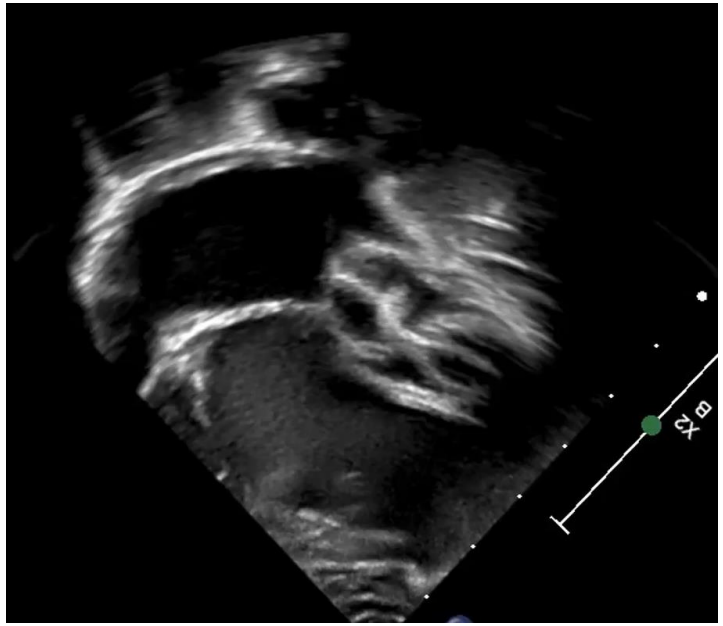
Fontan completion at 3-4 years of age



HLHS: Post Surgical Emergencies

- Shunt occlusion
- Imbalance in systemic and pulmonary blood flows
- Aortic arch obstruction
- Restrictive atrial septum
- Pulmonary artery obstruction
- AV regurgitation
- Cardiac dysfunction
- Arrhythmia

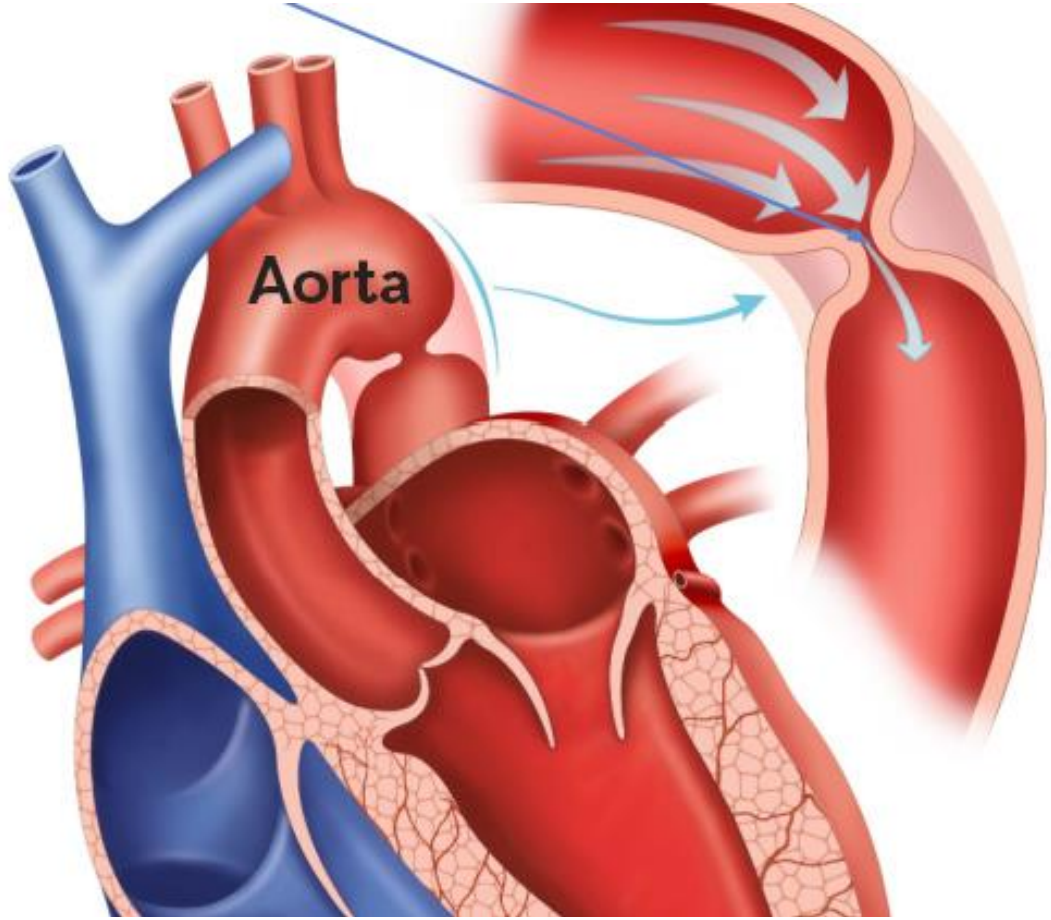
- Intervention
  - Intubation
  - Use oxygen cautiously (goal sats 75-85% for stage 1)
  - Inotropic support
  - Fluid resuscitation
  - Heparin



3 month old with HLHS post flow restrictor placement.

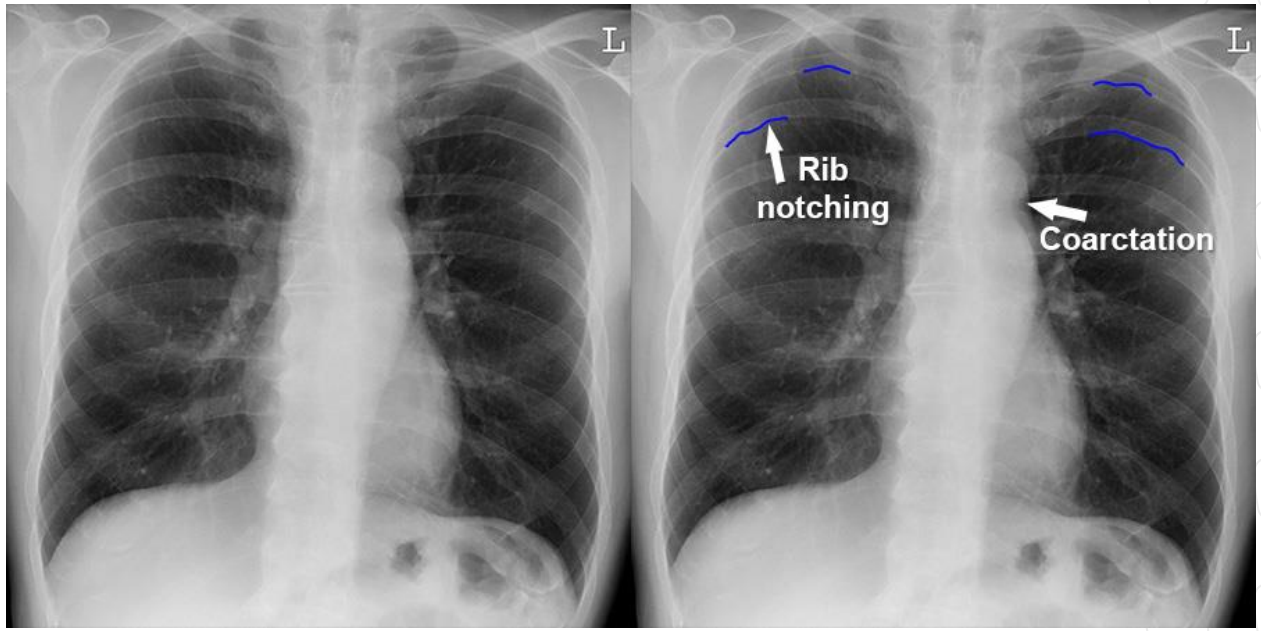
Coarctation of the Aorta

- Narrowing of the aorta near the left subclavian artery
- High BP in UE and lower BP in LE
- Stabilized by PGE infusion to open PDA
- May be associated with aortic valve stenosis



Coarctation of the Aorta

- CXR: “figure 3” appearance of aorta, rib notching (collaterals)
- EKG: LVH
- Exam: systolic ejection murmur RUSB and left scapular area, continuous murmur throughout precordium and back, cool LE with decreased pulses, UE to LE BP pressure gradient
- O2 saturations: normal

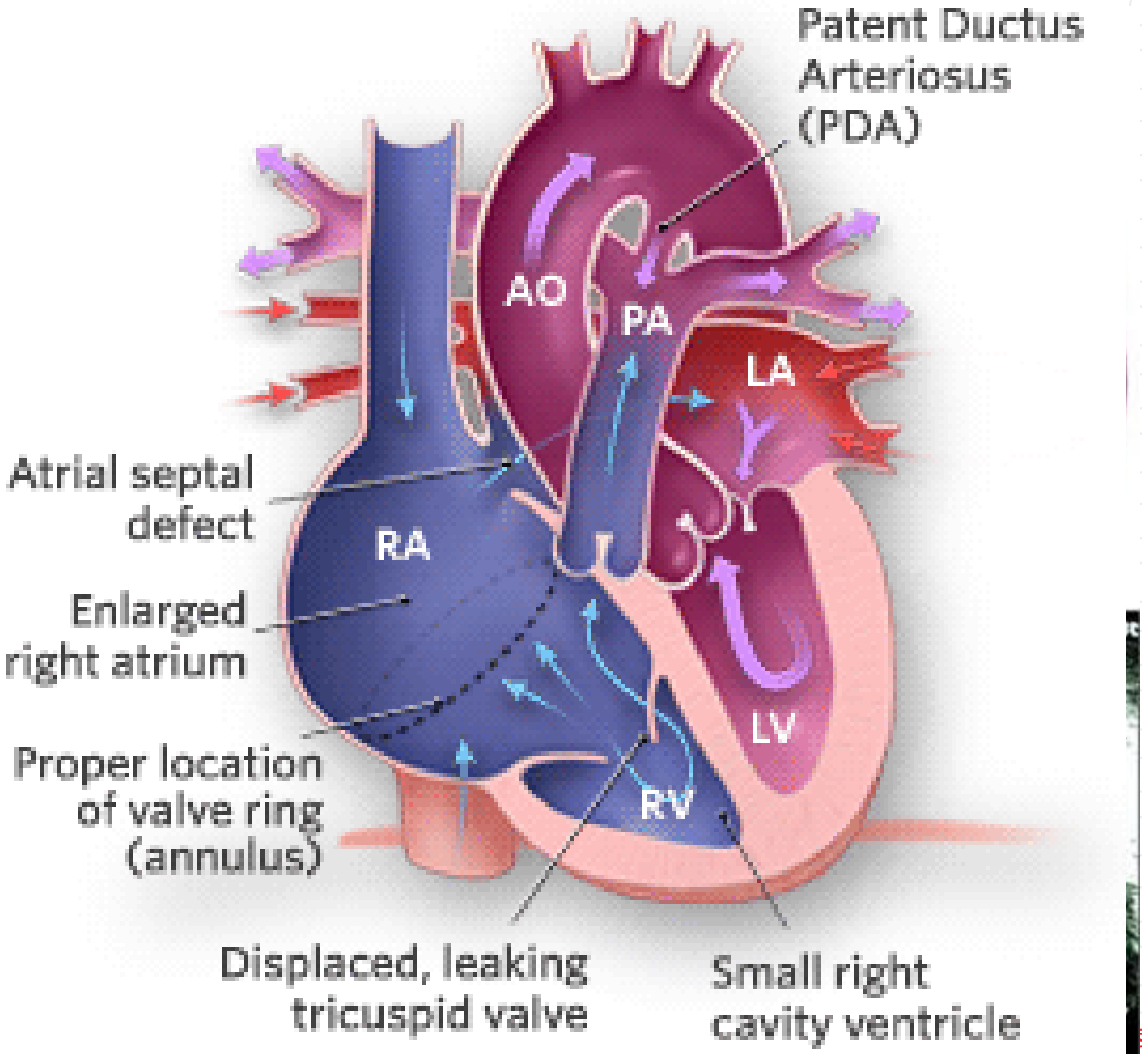


Coarctation of the Aorta – Intervention

- Surgical repair – multiple options
- Left subclavian flap repair results in decreased LUE BP
- Catheter intervention possible in adolescents and adults
- Emergencies
  - Pseudoaneurysm rupture
  - Dissection
  - Hypertensive emergency

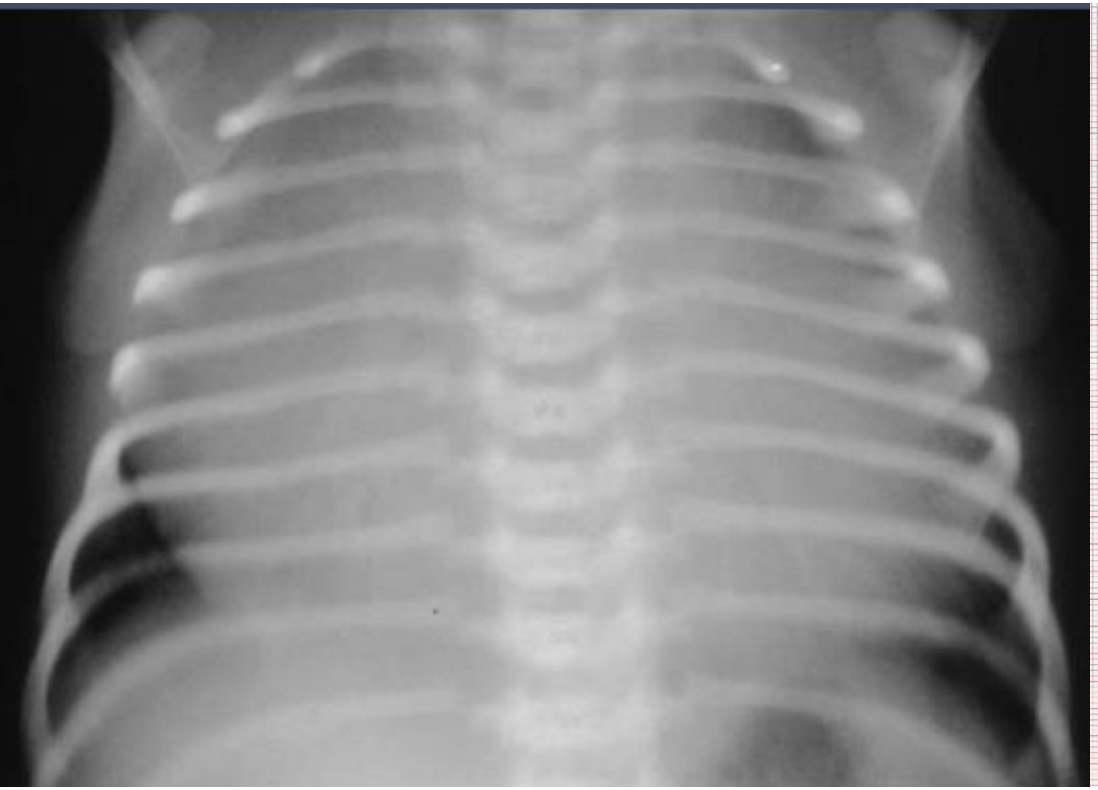
Ebstein Anomaly

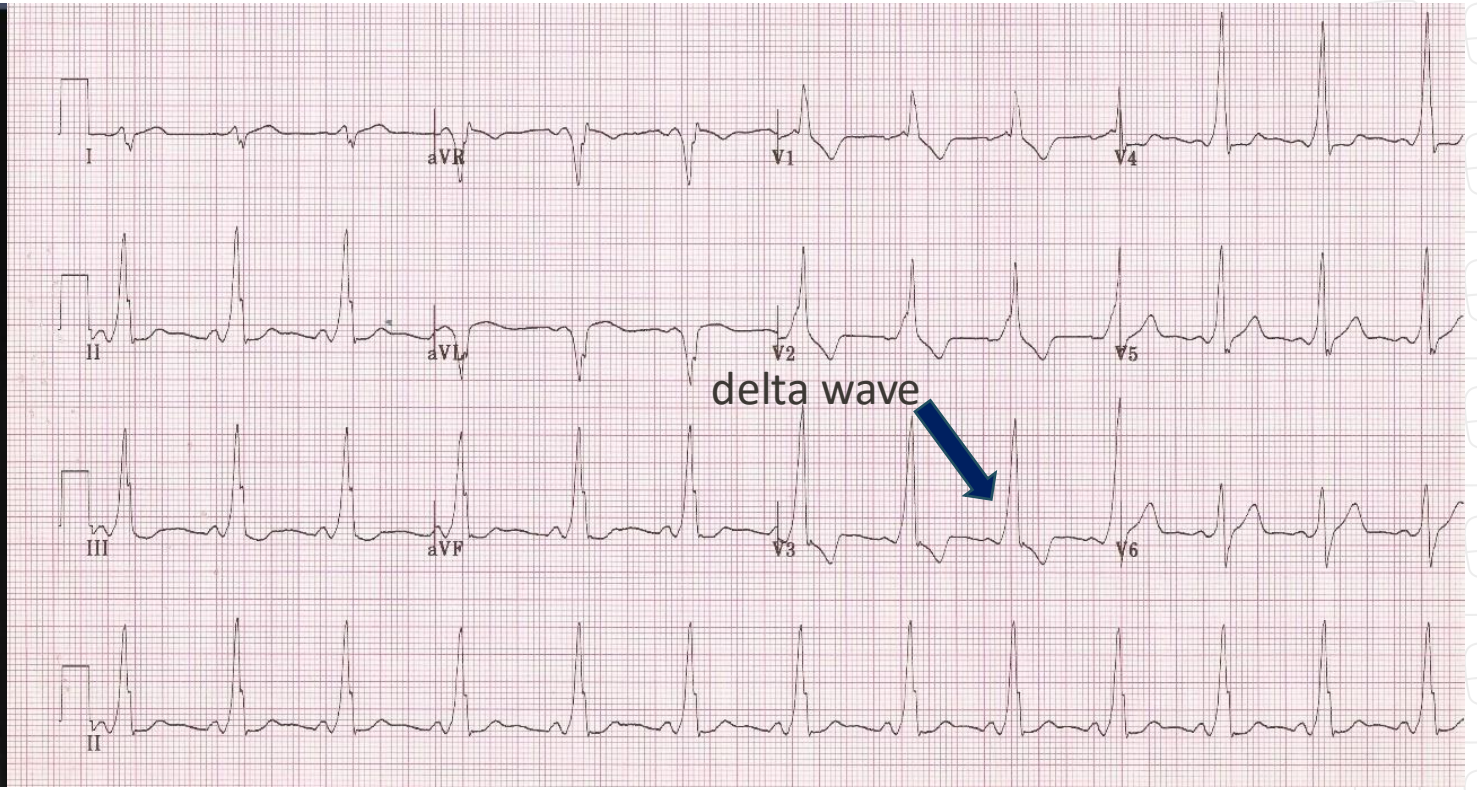
- Apical displacement of tricuspid valve
- Failure of delamination of the TV leaflets
  - Tricuspid regurgitation
  - Right atrial/ventricular enlargement
  - Right ventricular dysfunction
  - “Functional pulmonary atresia”
  - Pulmonary regurgitation



seen with Ebstein’s Malformation

Ebstein Anomaly – CXR/EKG

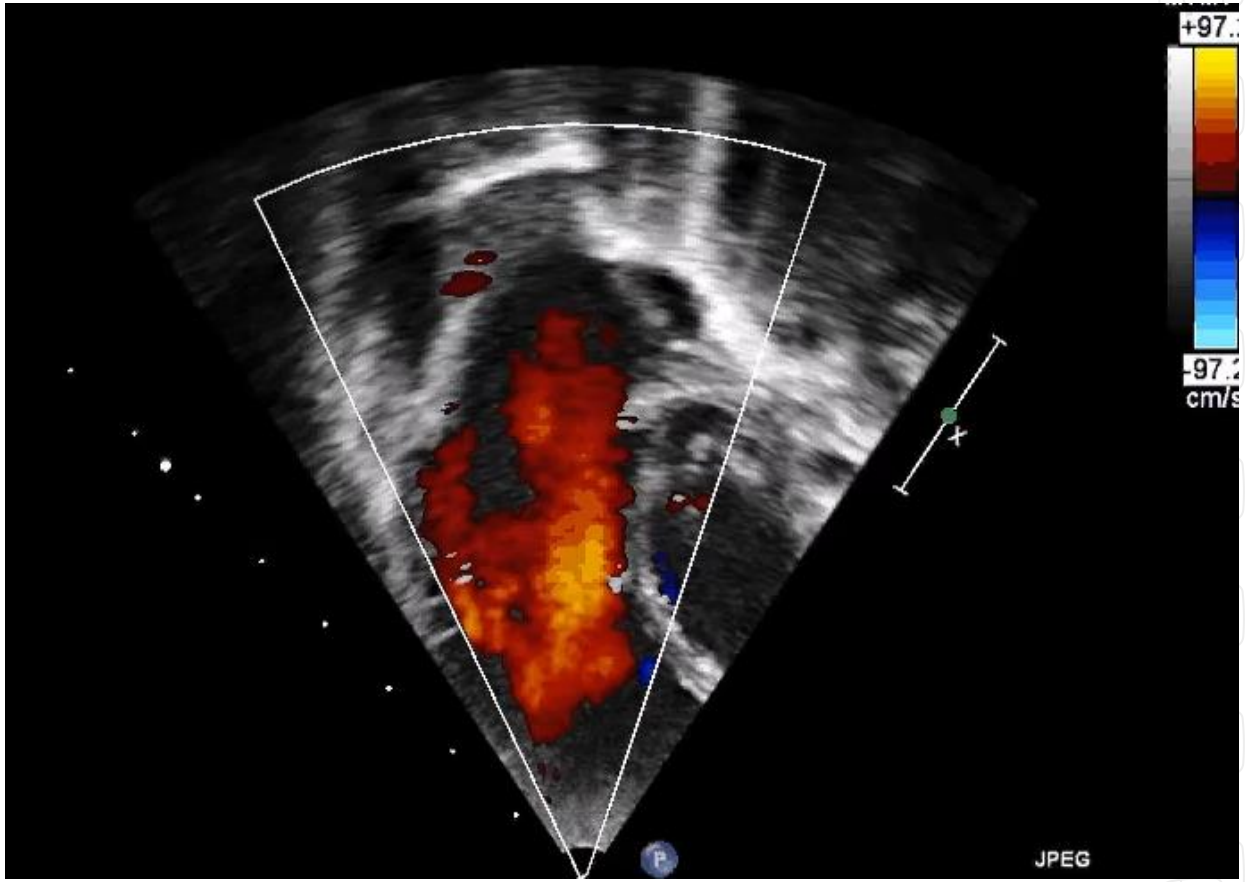




Lung Hypoplasia
Only lesion with massive cardiomegaly and clear lung fields

20% have Wolf-Parkinson-White Syndrome with risk for SVT

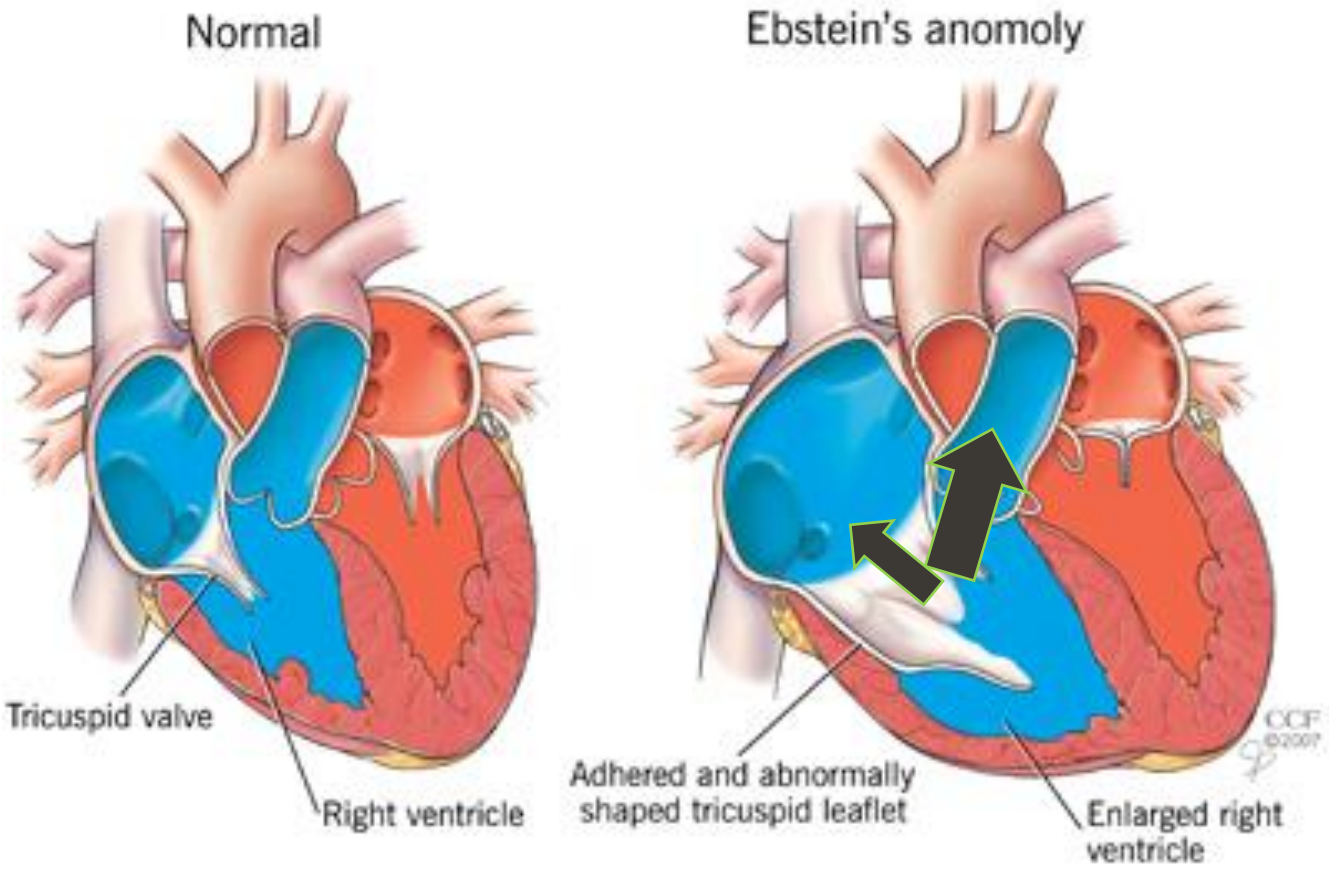
Ebstein Anomaly: Echo Findings



Severe Tricuspid Regurgitation
Right Atrial Enlargement

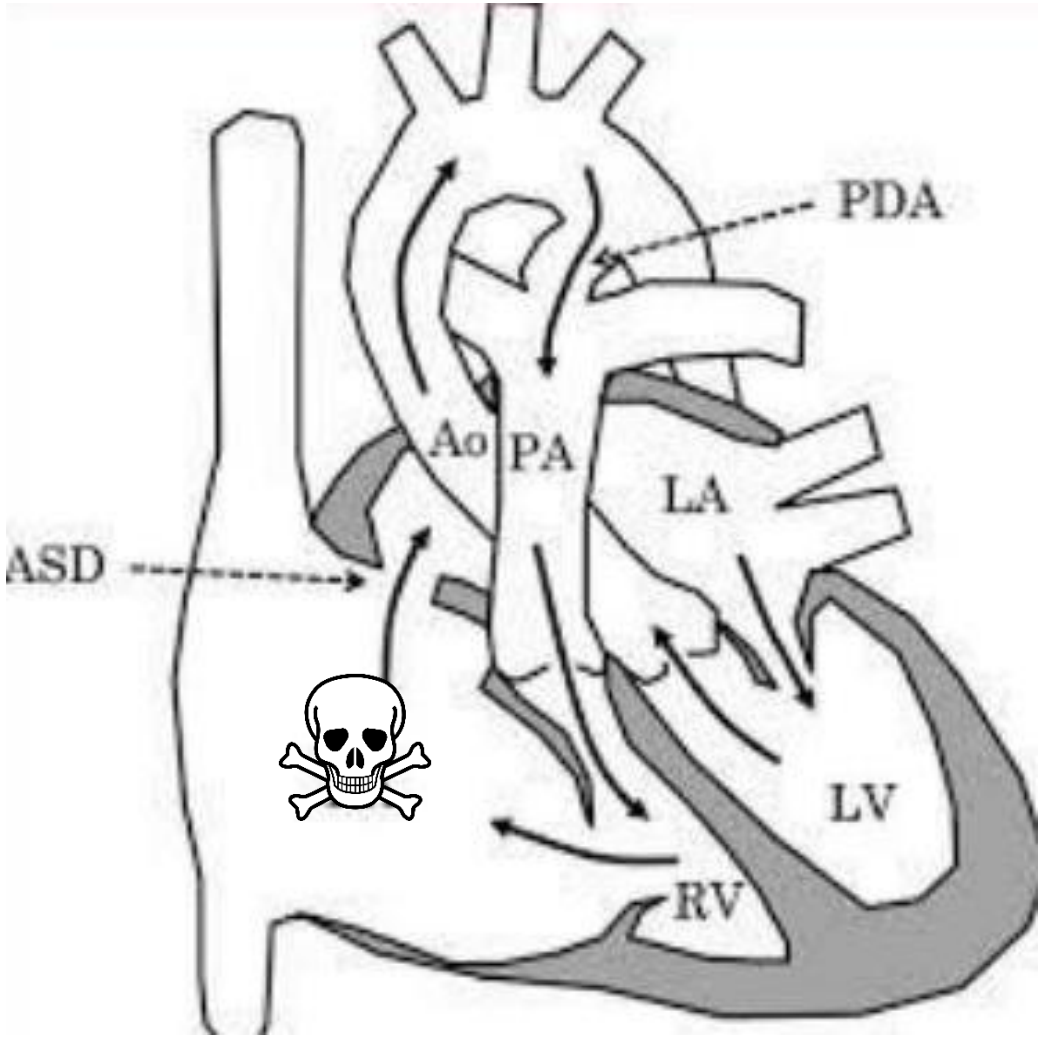
Ebstein Anomaly – Presentation

- Exam: tachypnea, holosystolic murmur, hyperactive precordium, mild cyanosis
- O2 saturation >90%



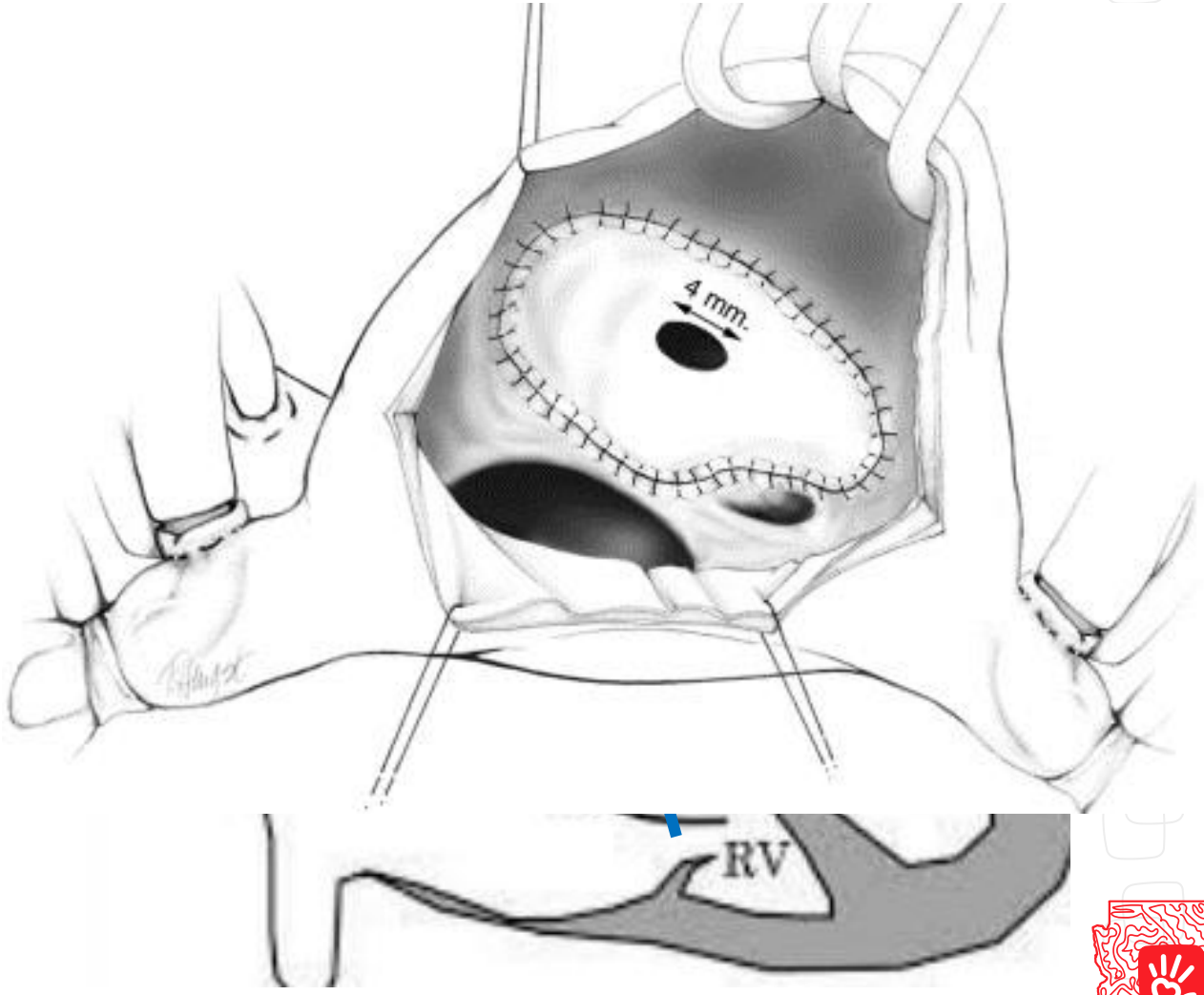
Ebstein Anomaly – Presentation

- Exam: tachypnea, holosystolic murmur, hyperactive precordium, mild cyanosis
- O2 saturation >90%
- Potential for cardiogenic shock with the “Circle of Death”



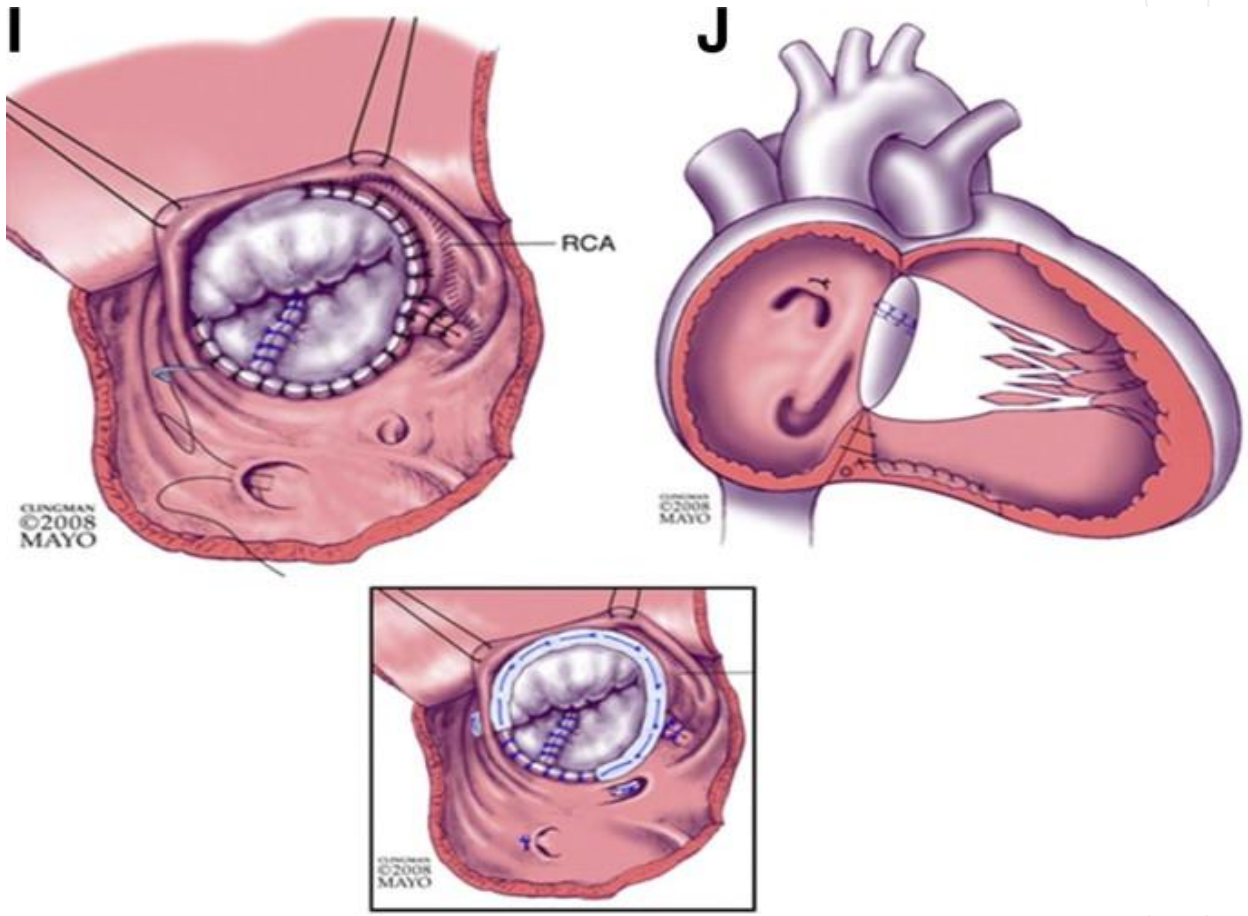
Ebstein Anomaly - Surgery

- No neonatal surgery with mild presentation
- Consider balloon pulmonary valvuloplasty if RV can generate adequate pressure
- Starnes procedure
  - Oversew TV with a fenestration
  - Place BTT shunt
  - May ligate the MPA
  - Eventually Fontan procedure



Ebstein Anomaly – Corrective Surgery with Two Ventricles

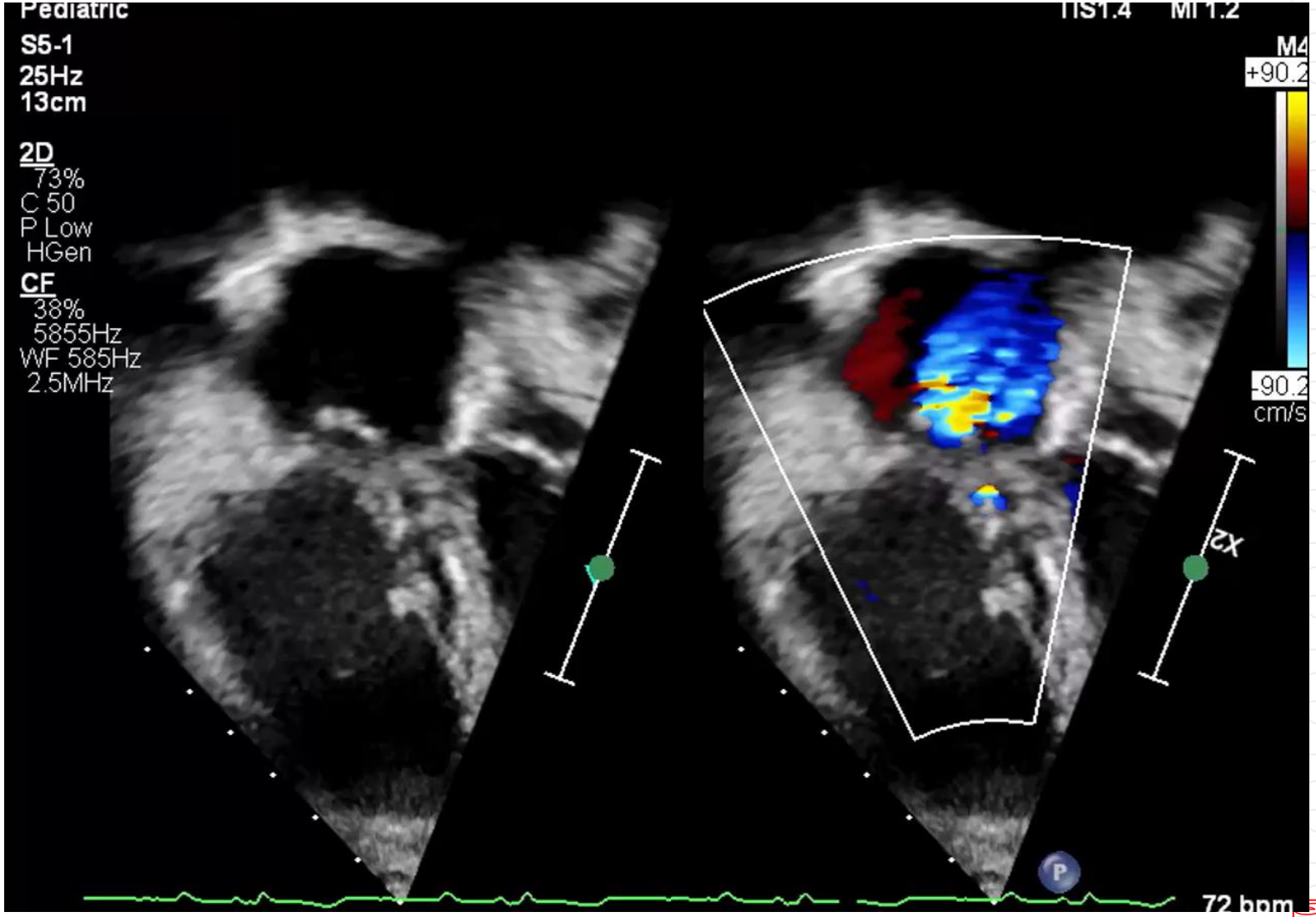
- Cone Reconstruction
  - Anatomic repair creating a new TV annulus
  - Surgical delamination of the TV leaflets
  - Right atrial excision and reduction
  - Plication of TV leaflet and annulus with resuspension
- Typically done 4-5 years of age
- Maze procedure of right atrium to control arrhythmias



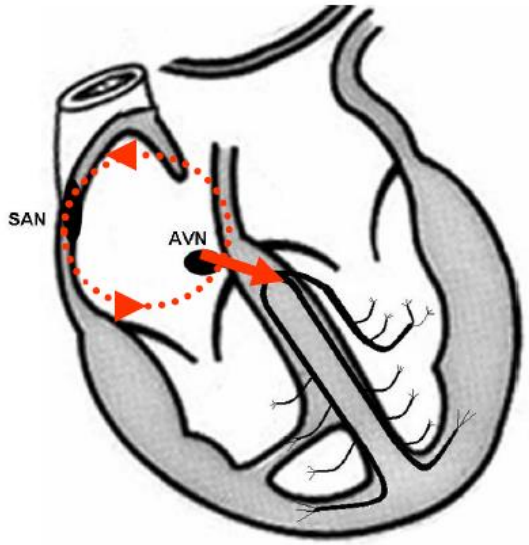
Ebstein Anomaly – Post Surgical Emergencies

- Atrial arrhythmias
- Thromboembolism
- RV failure
- Cardiogenic shock

- Intervention
  - Antiarrhythmics
  - Heparin
  - Fluid resuscitation
  - Epinephrine



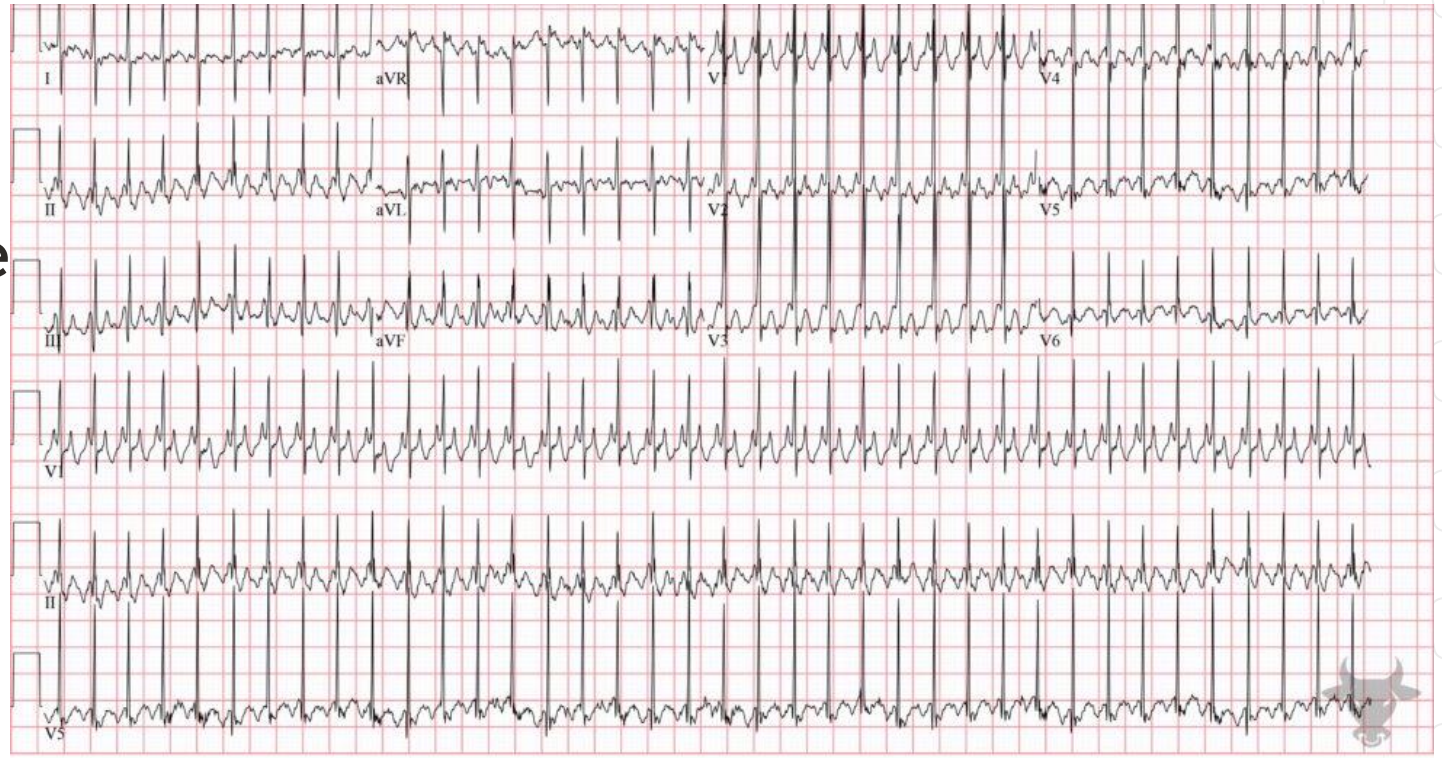
3 year old post Cone procedure to repair the Ebstenoid tricuspid valve

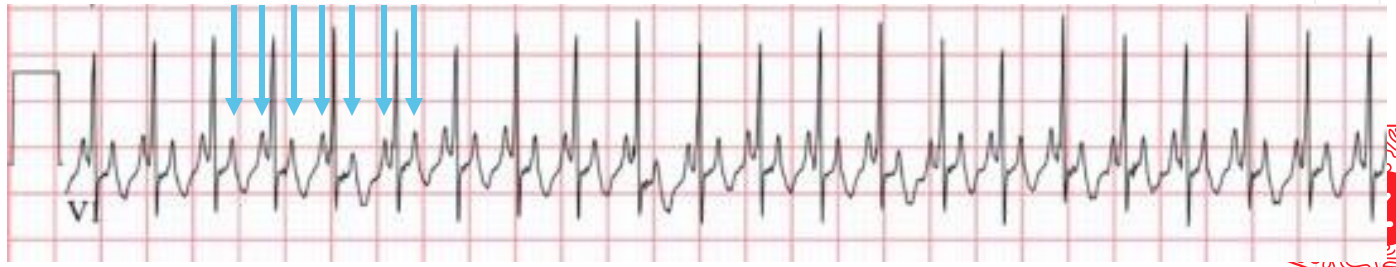


history of fetal
al heart rate 240
the 3rd trimester
term and had the
What is the

A. Atrial flutter
B. Ventricular tachycardia
C. Supraventricular tachycardia
D. Premature atrial contractions

Atrial rate: 480 bpm Ventricular rate: 240 bpm





Question 2

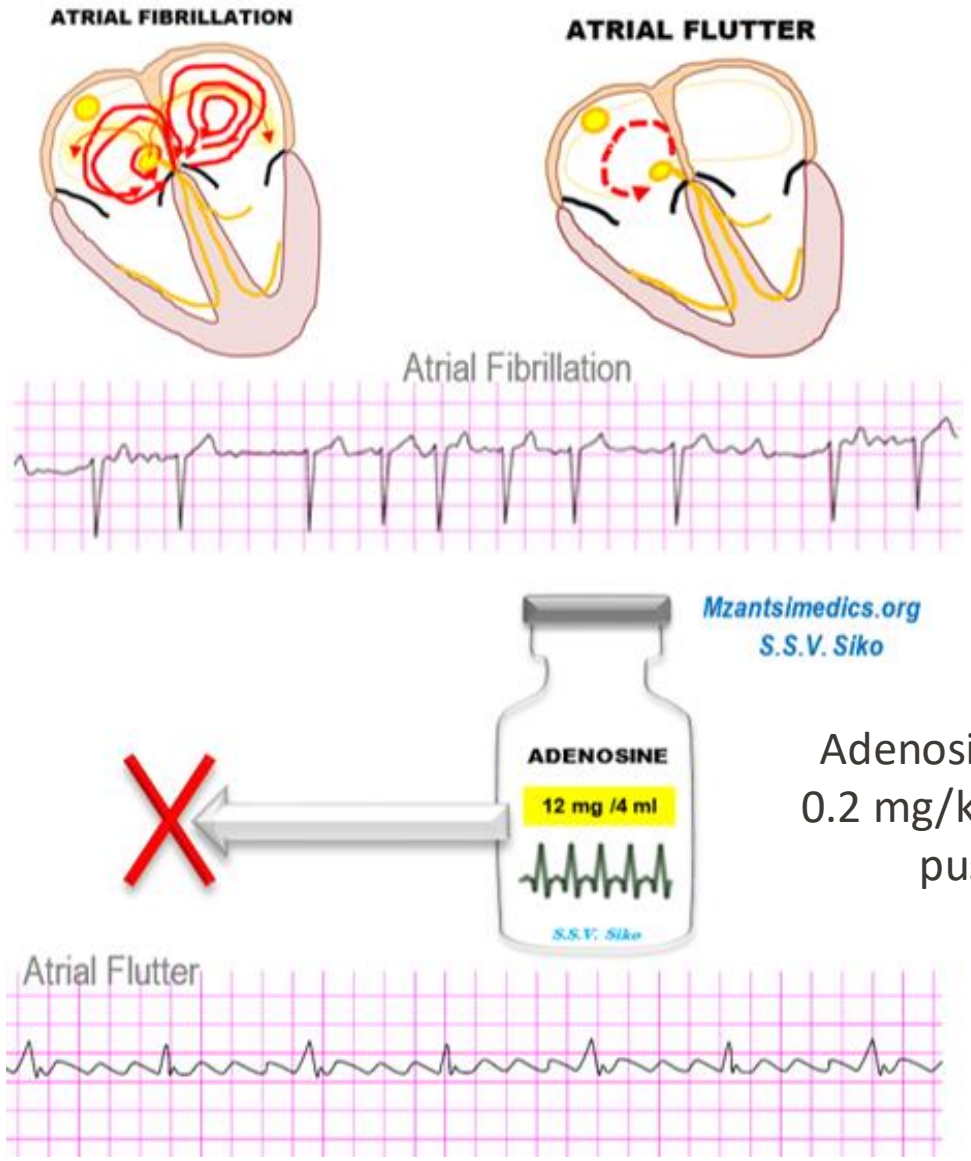
What intervention can be performed to confirm the diagnosis of atrial flutter?

A. Electrical cardioversion

B. Administer adenosine

C. Intubate and start mechanical ventilation

D. Push on the abdomen



Adenosine 0.1-0.2 mg/kg IV fast push

Question 3

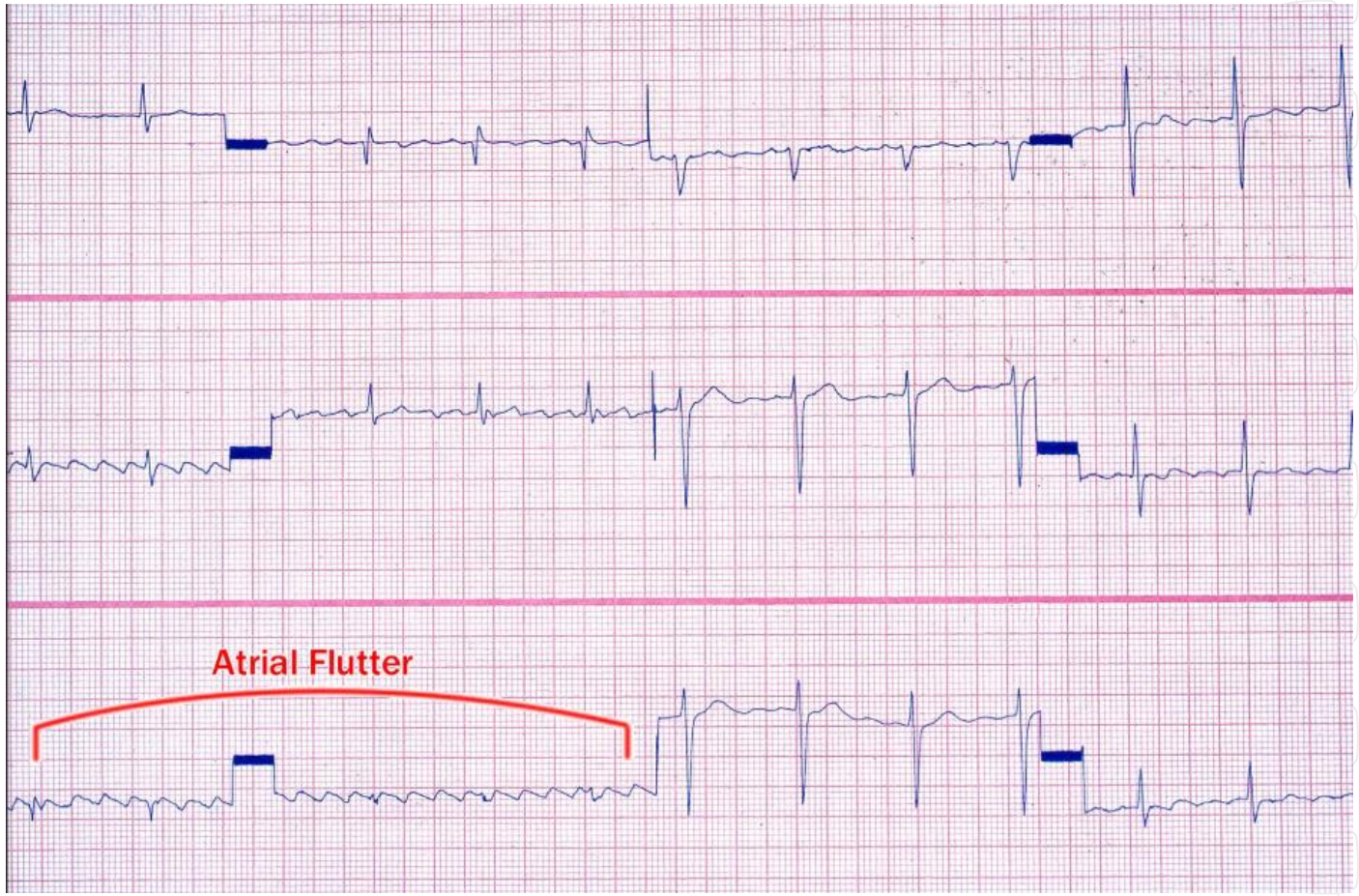
Our newborn with atrial flutter is warm and well perfused with mild tachypnea. What is the next best step?

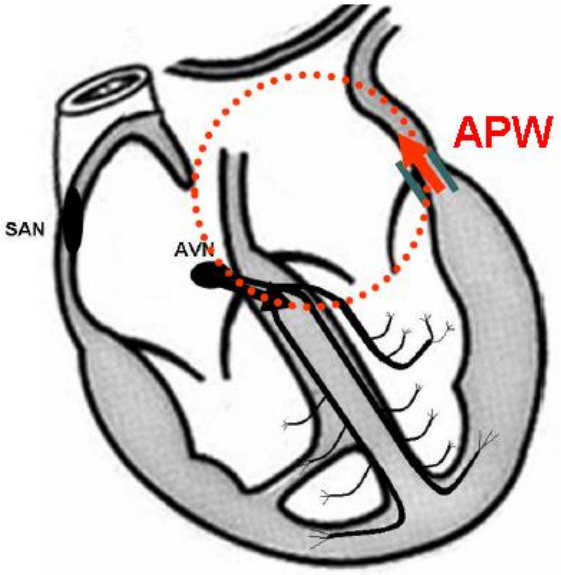
A. Observation in the NICU

B. Discharge to home and allow the baby to get bigger

C. Synchronized electrical cardioversion

D. Start amiodarone





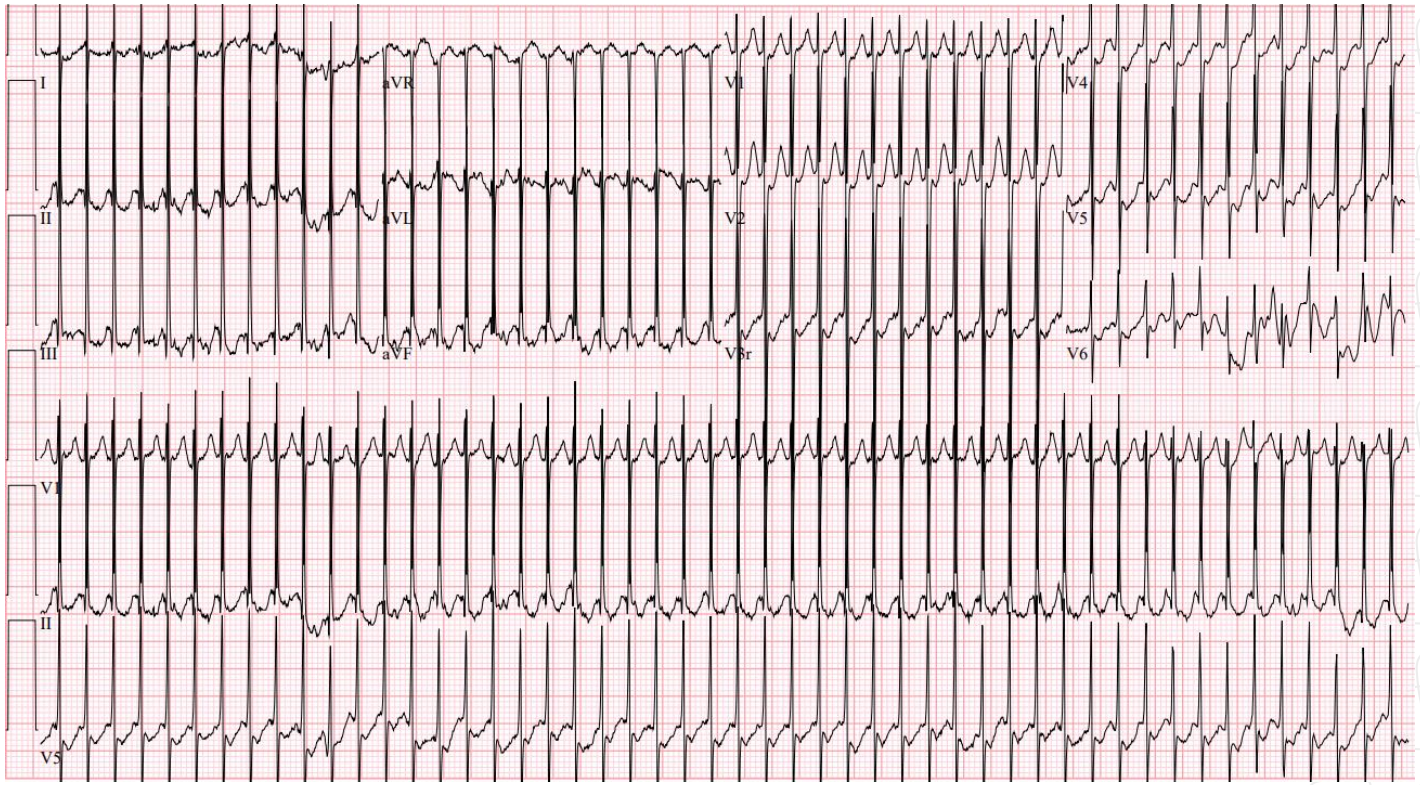
presents to the
y distress,
efill time 4
art rate 290 bpm.
ed. What is the
t best step?

d bolus

B. SVT and electrical cardioversion

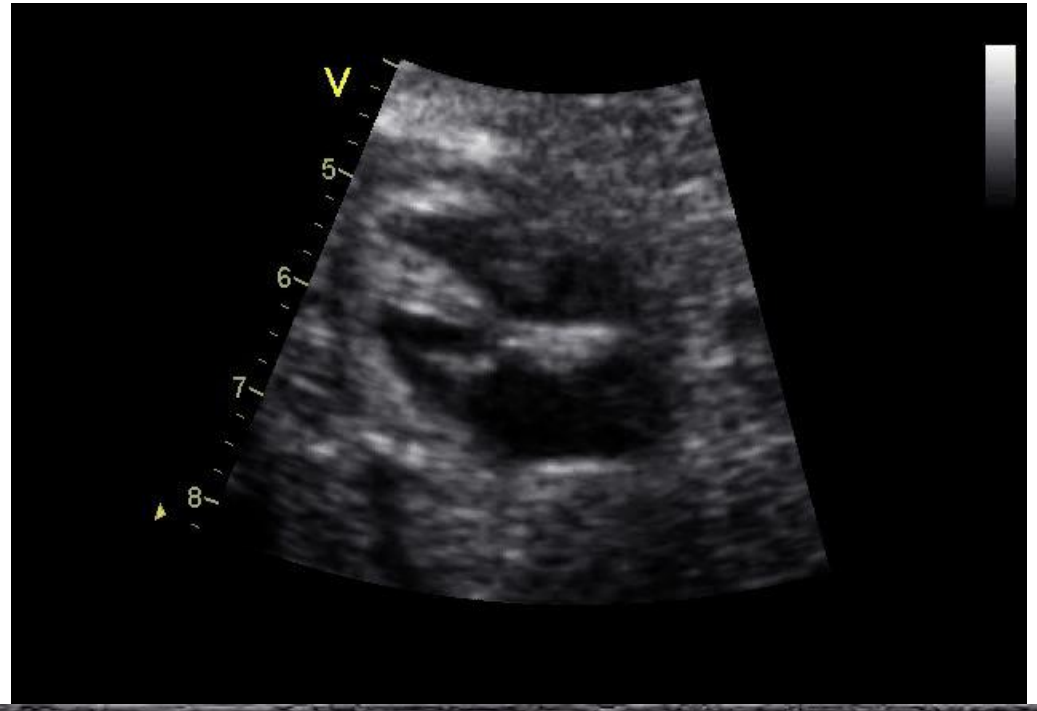
C. Atrial fibrillation and IV adenosine

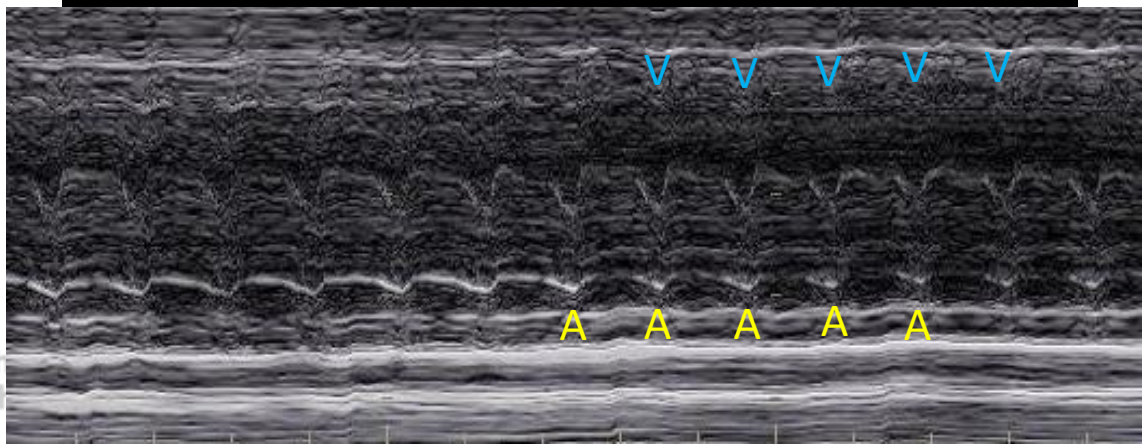
D. SVT and IV adenosine



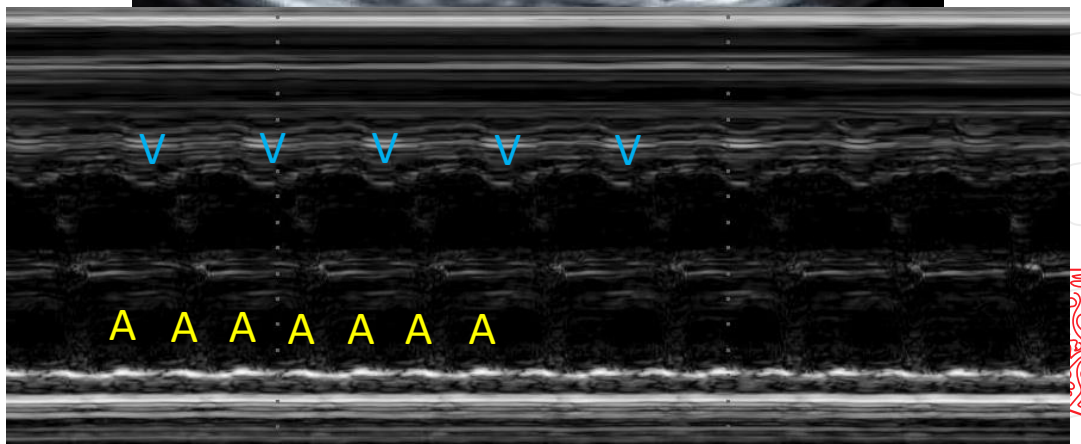
Comparison of Fetal SVT and Atrial Flutter

Supraventricular Tachycardia





Atrial Flutter



Conclusions

- Evaluation and stabilization of critical heart disease requires understanding anatomy and physiology
- Think of the 5 “T”s and HLHS and Ebstein Anomaly in your differential diagnosis when failing the CCHD Pulsox Screening Test
- Although echo is the gold standard for diagnosis, CXR and EKG may be very helpful in making the correct diagnosis
- PCH Pediatric Cardiology is always here to help!
- PCH Center for Heart Care – 602-933-3366

Phoenix Children's®

Thank you!

Questions

What three cardiac lesions cause a holosystolic murmur?

A. Truncus arteriosus

B. VSD

C. Mitral regurgitation

D. Tricuspid regurgitation

E. Hypoplastic left heart syndrome