6th Year Student Lectures in Pediatric Cardiology

Cyanotic Congenital Heart Disease

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Introduction

- **Congenital heart disease (CHD) is present in about 9 of every 1,000 liveborn children.**
- **Data from the UJH suggest that our incidence is higher, and it is about 13 to 15 per 1000 live births.**
- **About one half will need an intervention either interventional catheterization or surgery**
- **About 25 to 30% will need congenital heart surgery.**

- **Congenital Heart disease presents with one of three pathophysiological models:**
- **1. Acyanotic Volume Overload Lesions eg: VSDs PDAs AVSDs**
- **2. Acyanotic Pressure Overload Lesions: AS, PS Aortic Coarctation**
- **3. Cyanotic Heart Lesions which is the scope of this lecture: The 5 Ts**

Presentations of CHD in the Newborn

- 1. Asymptomatic Newborn with a Heart Murmur
- 2. Cyanotic Newborn
- 3. Decreased Peripheral pulses, differential cyanosis and shock
- 4. Congestive Heart Failure

Cyanotic Heart Disease

- What is cyanosis?
- The presence of at least 4 g/dL of deoxygenated blood in the systemic circulation.
- This corresponds to a pulse ox level of 80% or below.

- What are the causes of cyanosis?
- 1. Respiratory Disease
- 2. Cardiac Disease
- 3. Methemoglobinemia

Cyanotic Congenital Heart Disease

- Having de oxygenated blood shunted to the systemic circulation results from two main pathophysiologic models
- 1. Normal Cardiac Connections with a communication and a right sided obstruction
- 2. Abnormal Cardiac Connections

Normal Cardiac Connections with a Shunt and Right sided obstruction

- Normal Cardiac Connections Mean:
- We have atria receiving their respective veins.
- We have ventricles with their corresponding inlet Valves
- We have outflow tracts emerging from their respective ventricles

- Examples of Cyanotic Heart Disease with normal connections
- 1. Tetralogy of Fallot
- 2. Pulmonary Atresia VSD
- 3. Tricuspid Atresia
- 4. Pulmonary atresia with intact ventricular septum
- 5. Ebstein s anomaly.

Neonate with a cyanotic CHD

- The 5 T's
- **Tetralogy of Fallot**
- **Tricuspid atresia / Ebstein's anomaly**
- **Transposition of the great arteries**
- **Truncus arteriosus**
- **TAPVR**

Approach to a Child with Cyanosis

- Accurate History Prenatal and Natal.
- Vital Signs and UL : LL pulse oximetry
- Accurate Physical Examination.
- Hyperoxia testing
- Chest Xray
- Determine your DDX!

Severe Cyanosis: Pulse ox less than 70% with or without Acidosis

- Cyanosis can be subtle
- SEVERE cyanosis in the first few hours of life is usually : DDX:
- TGA (increased or normal pulmonary blood flow)
- PA with or without a VSD (decreased PBF)
- Severe Ebstein's anomaly (decreased PBF)

Mild cyanosis: Pulse oximetry above 70% and no Acidosis

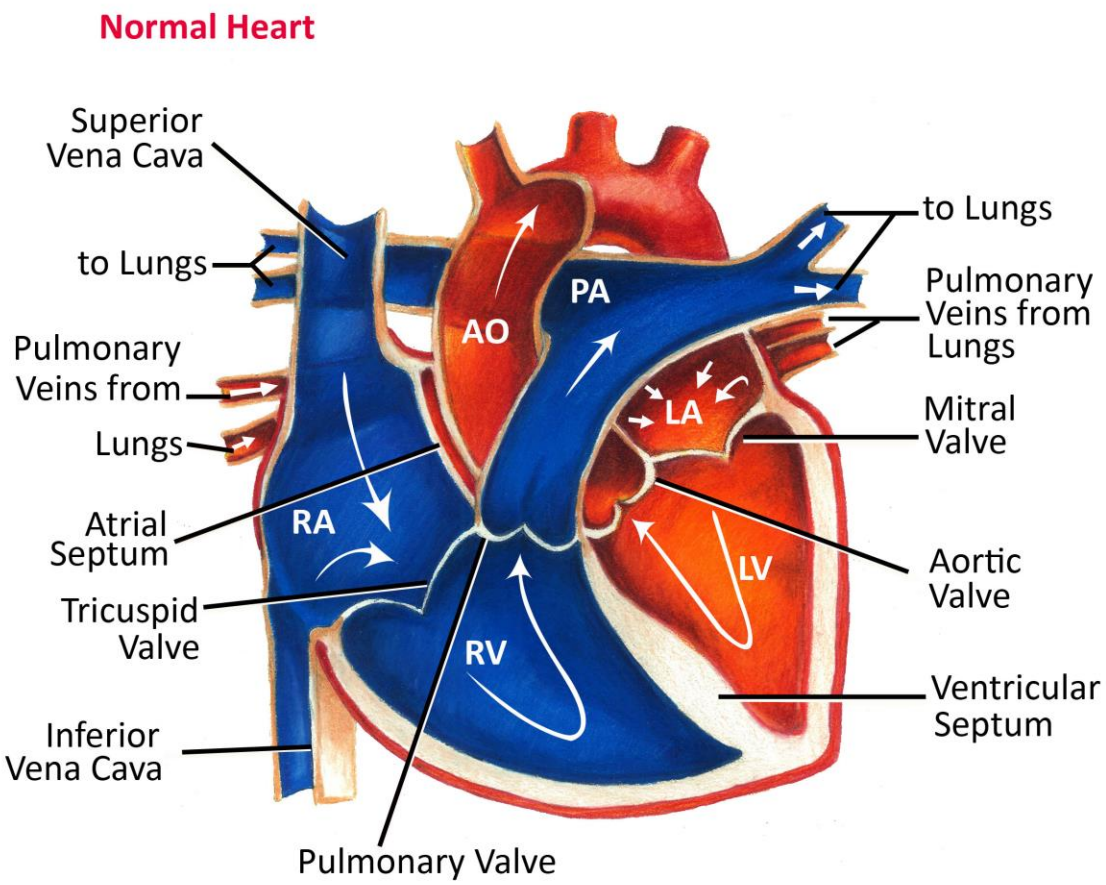
With increased PBF:

- Pink Tetralogy of Fallot
- Truncus arteriosus
- Tricuspid atresia with a large VSD
- TAPVR

Mild Cyanosis with decreased PBF

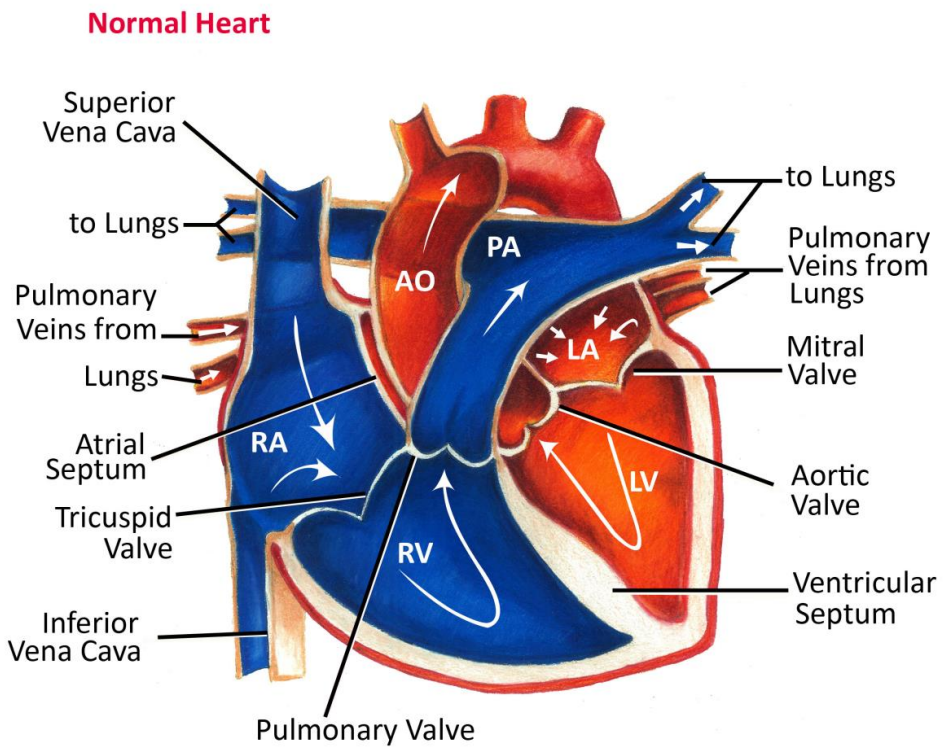
- TOF with mild cyanosis and moderate pulmonary valve stenosis.

Normal Heart

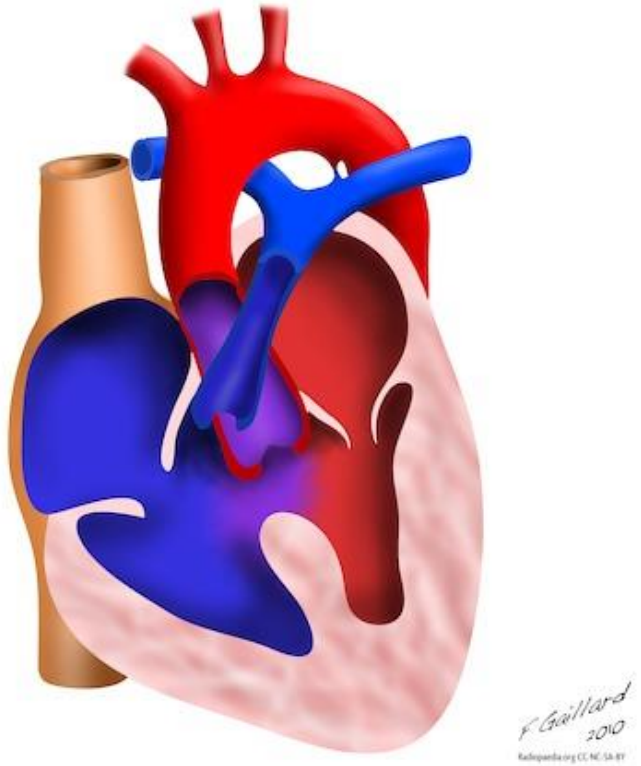


Tetralogy of Fallot

Normal Heart

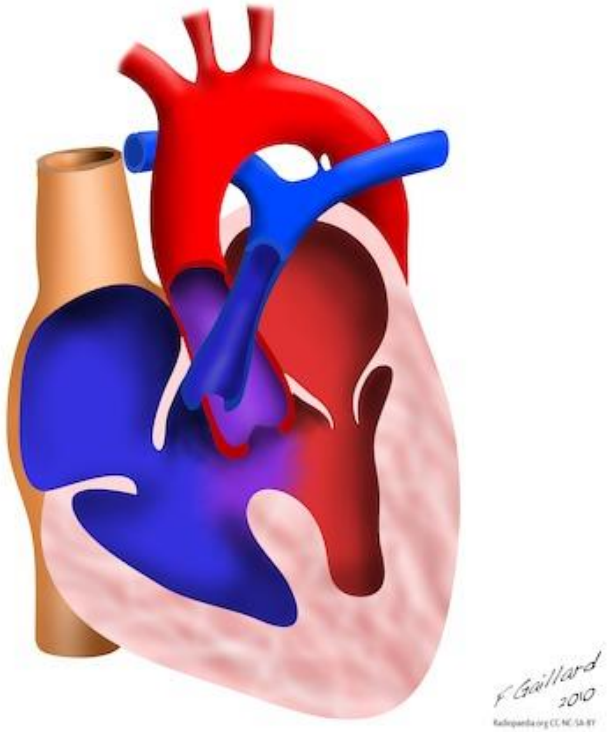


Tetralogy of Fallot

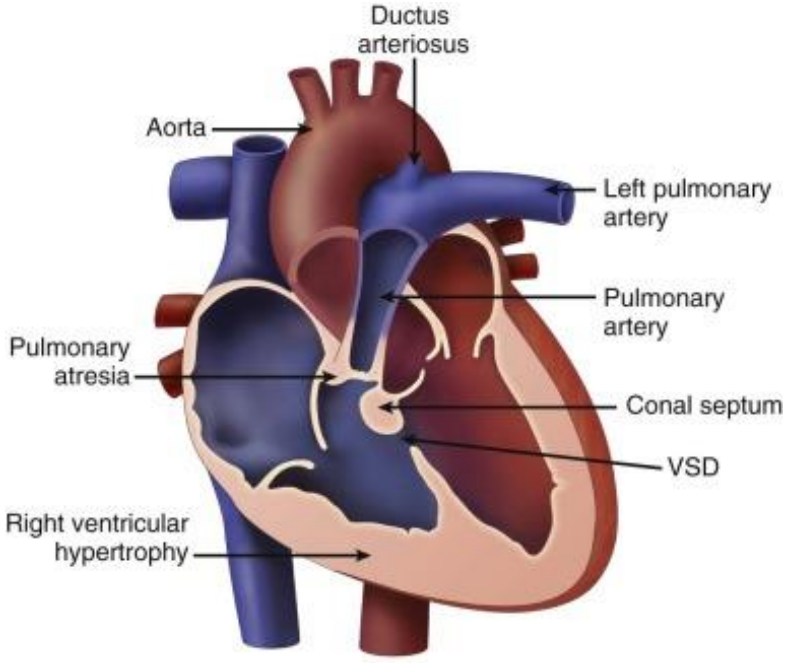


Spectrum of TOF

TOF



Pulmonary Atresia VSD



CYANOTIC CHD

- Tetralogy of Fallot
- Tetralogy of Fallot is the most common cyanotic CHD, accounting for 5% of all CHD.
- It develops from the anterior malalignment of the interventricular septum, which leads to a VSD, as well as overriding of the VSD by the aorta And narrowing of the pulmonary outflow tract due to the septal deviation, and this causes RV outflow (infundibular) obstruction and consequent RV hypertrophy

- Monology of Fallot for the pediatric cardiologist with the main pathology being the anterior deviation of the IV septum creating a VSD behind it and sub PS/PS anterior to it with subsequent development of the RVH.

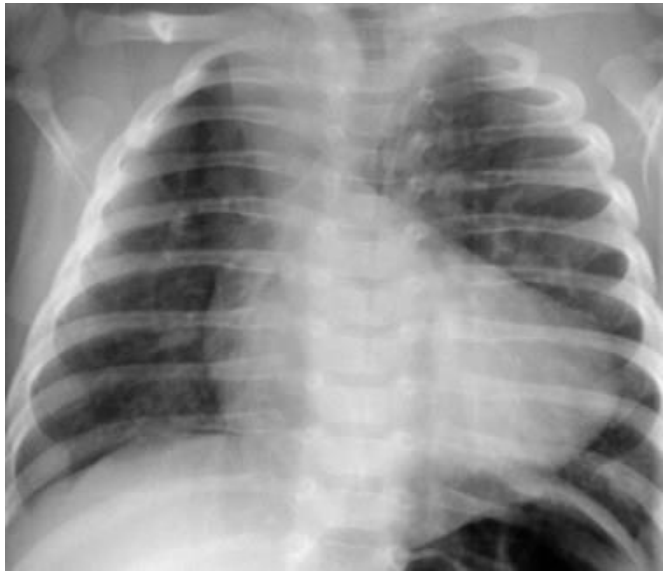
TOF Presentation

- Patients with tetralogy of Fallot have a harsh ejection systolic murmur heard over the pulmonic area, indicating pulmonic stenosis.
- There may be a single second sound.
- Higher saturations Indicate less RV outflow obstruction

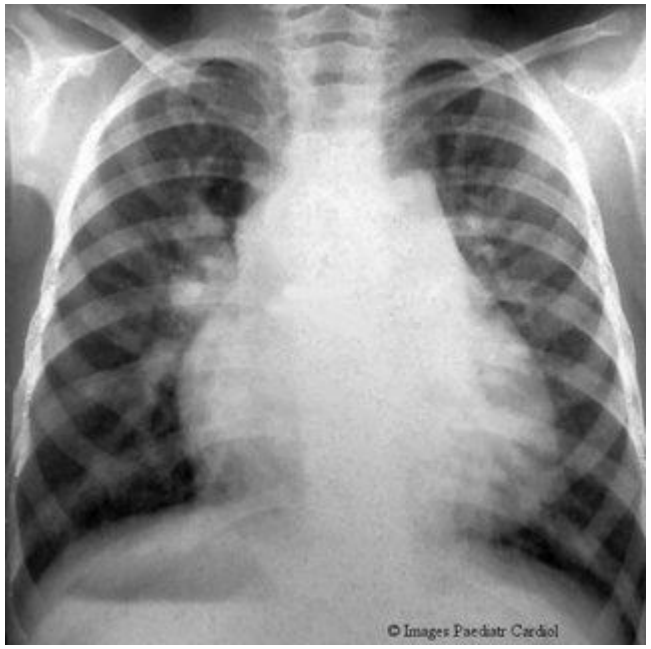
- **Management**.
- The time of presentation and intervention
- for tetralogy of Fallot is determined by the degree of RV Out flow obstruction and the limitation of pulmonary blood flow.
- These clinical indicators are the oxygen saturation levels and the development of hypercyanotic spells.
- The Doppler echocardiographic indicators are the pressure gradients observed across the RV outflow tract.
- Children with saturations less than 80% or those having hypercyanotic spells are scheduled for surgery.

TOF Chest Xray Appearance

Boot Shaped Heart and mild PS



Boot Shaped Heart with severe PS or PA



TOF Management

Pink TOF

Cyanotic TOF

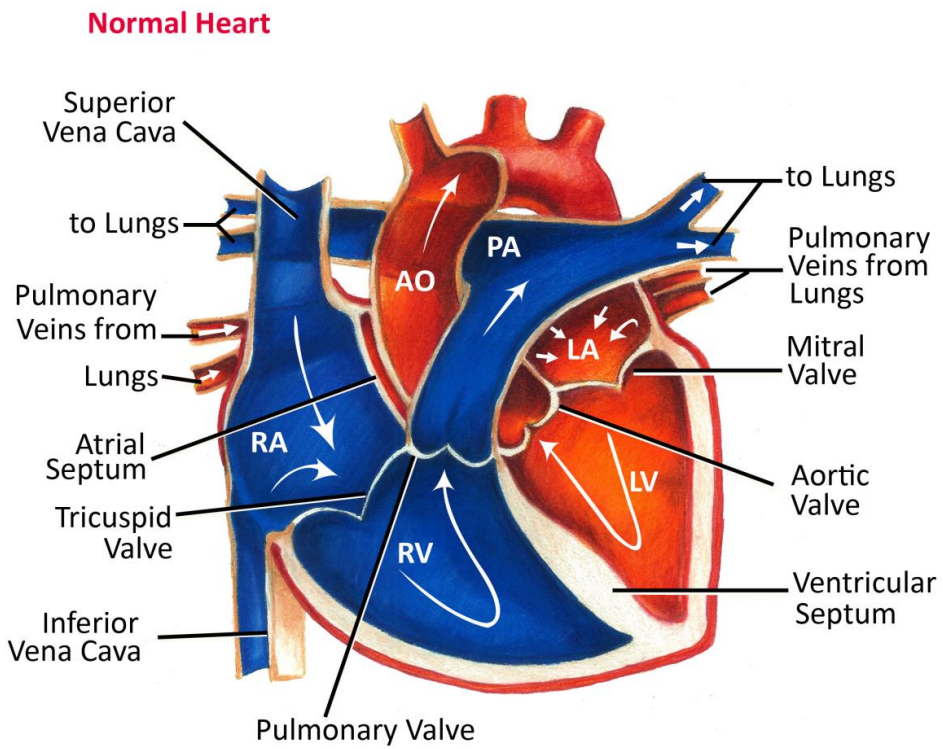
TET Spells

Tet Spells

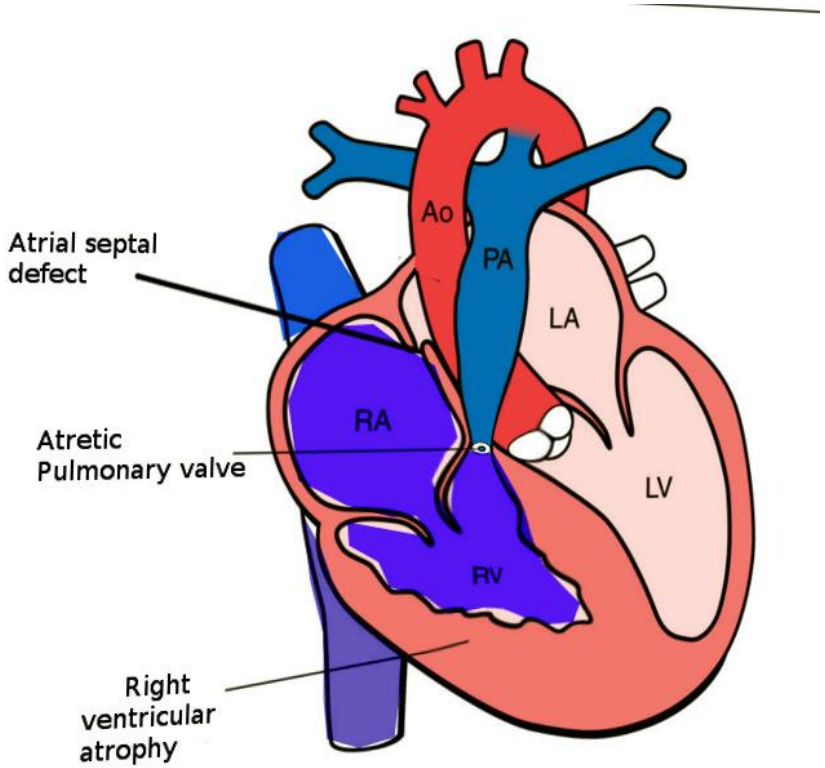
- **Acute “Tet spells” are managed by first helping to calm the patient (eg, handing the child to the parent to hold, allowing the child to feed, or**
- **administering sedation if needed),**
- **placing the child in a knees-to-chest position (to increase systemic vascular resistance this may be achieved by having the parent hold the baby in her arms and cradling the knees and chest together),**
- **initiating oxygen (preferably through the least noxious route for the child, for example, blow-by oxygen or use of a face mask),**
- **administering a bolus of intravenous fluids, and,**
- **finally, intravenous metoprolol (to slow the heart rate) and**
- **phenylephrine (to increase systemic vascular resistance).**
- **Some patients will require anesthesia (on the way to the**
- **operating room).**

Pulmonary Atresia Intact Septum

Normal Heart

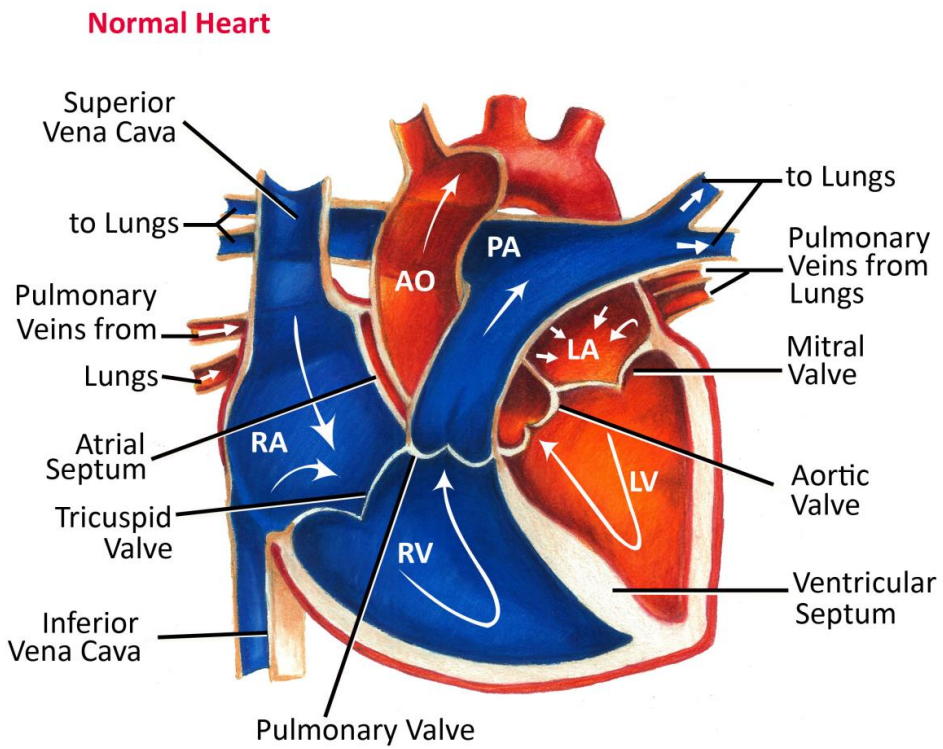


Pulmonary Atresia Intact Septum

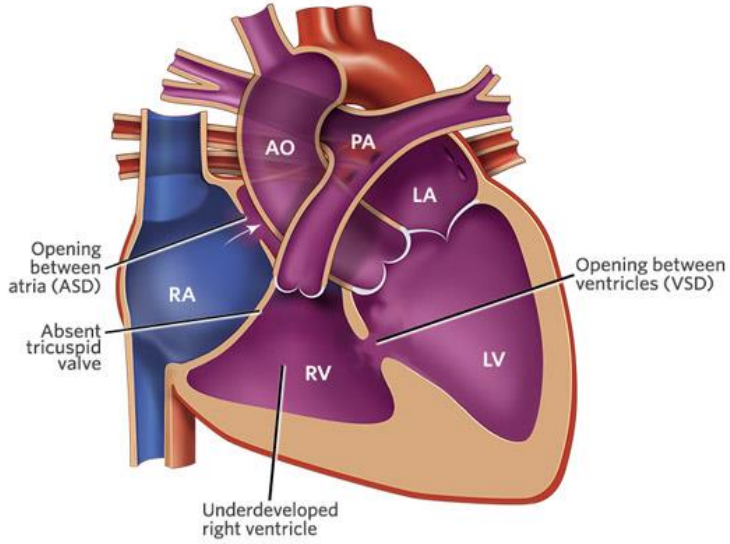


Tricuspid Atresia

Normal Heart

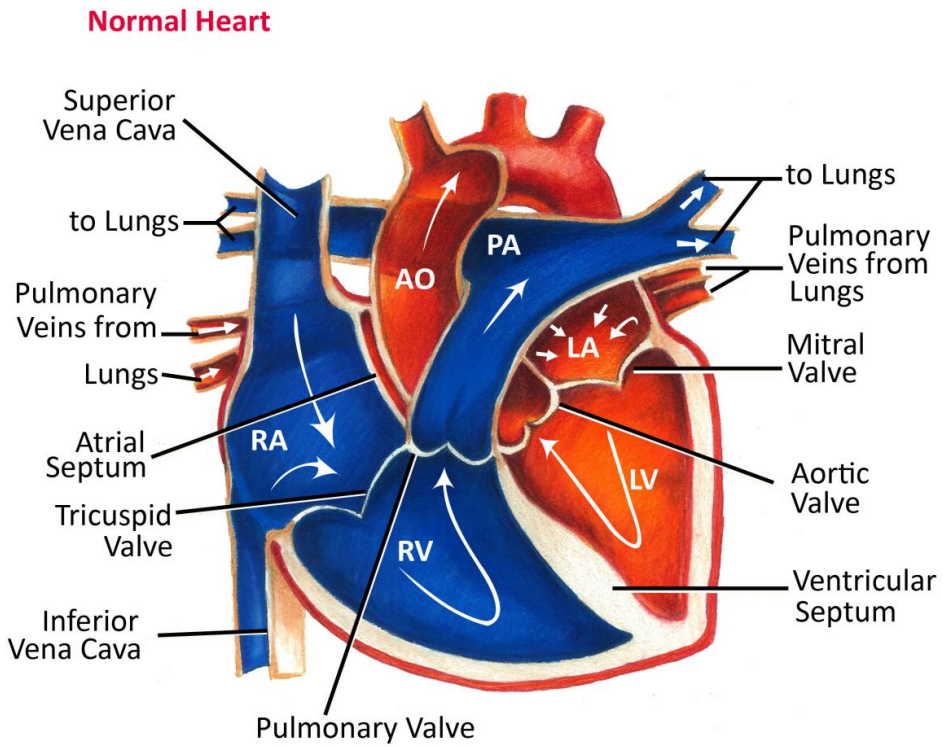


Tricuspid Atresia

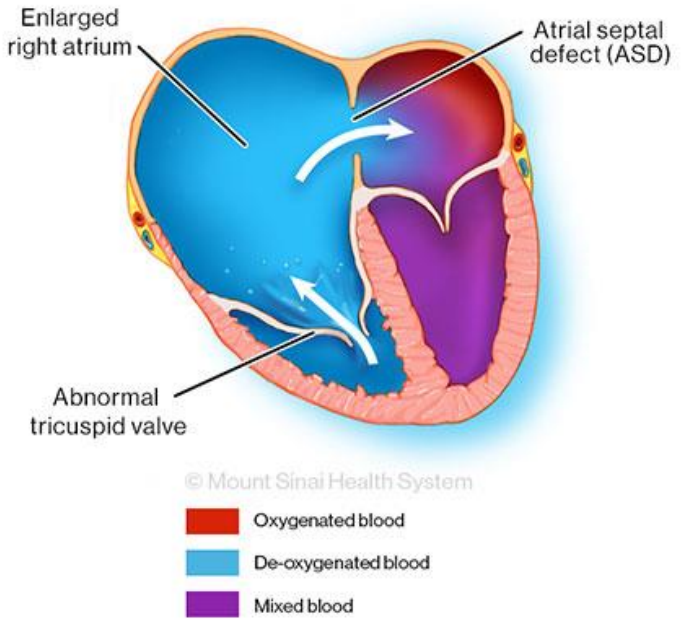


Ebsteins Anomaly

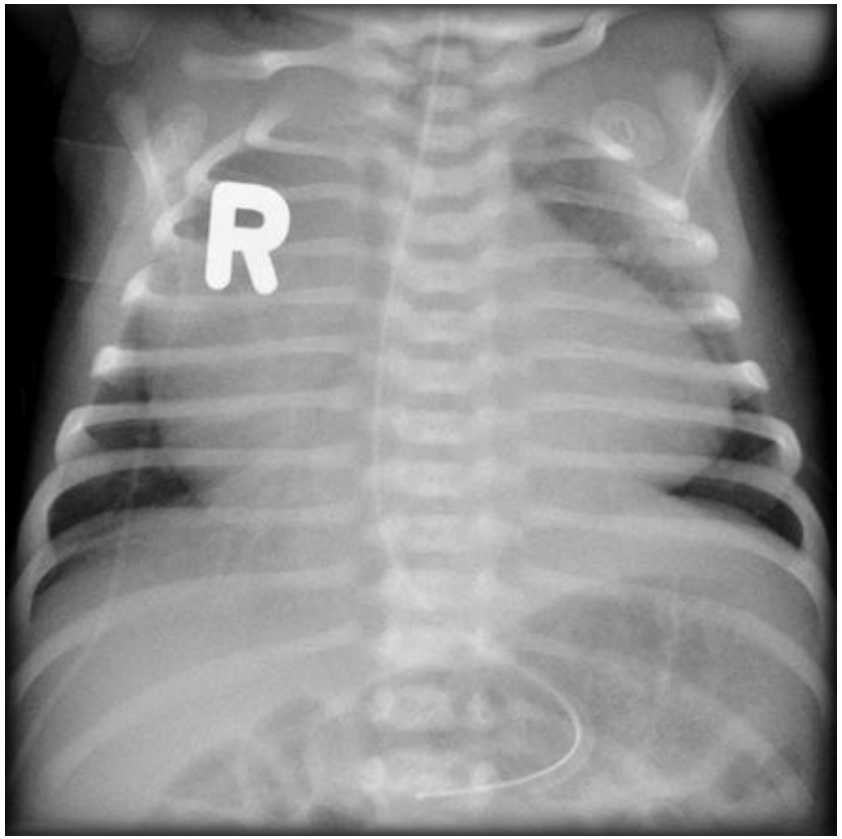
Normal Heart

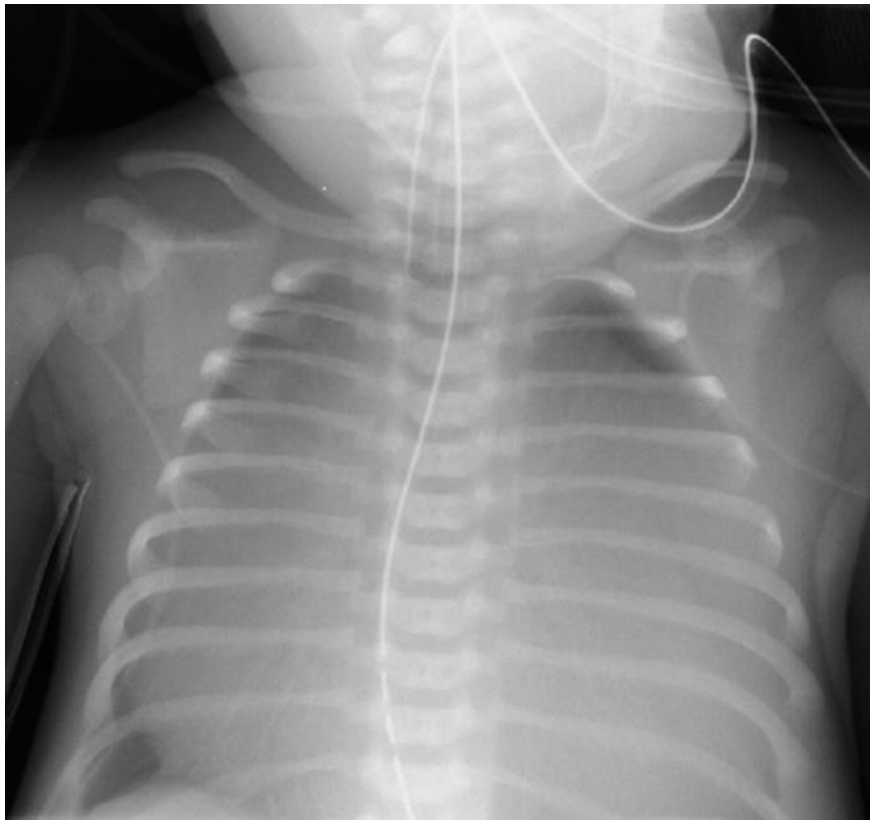


Ebstein s Anomaly with or without PS



Ebstein s Anomaly Chest X Ray





Cyanotic Heart Disease with Abnormal Connections TGA, TA and TAPVR

Abnormal Outflow Connections

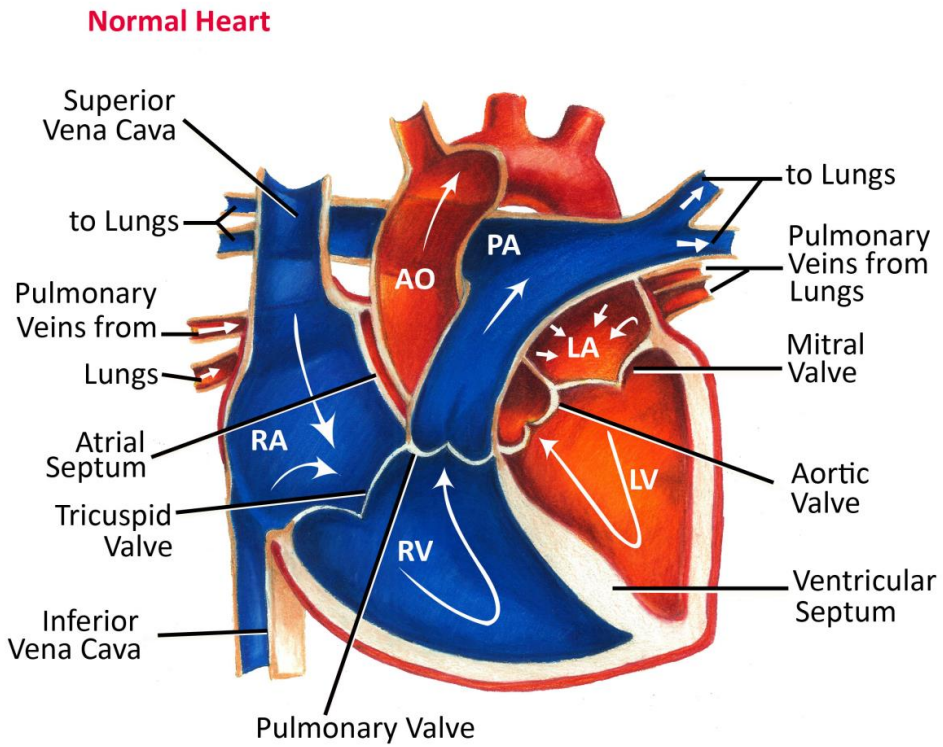
- D-TGA
- Truncus Arteriosus

Abnormal Inflow Connections

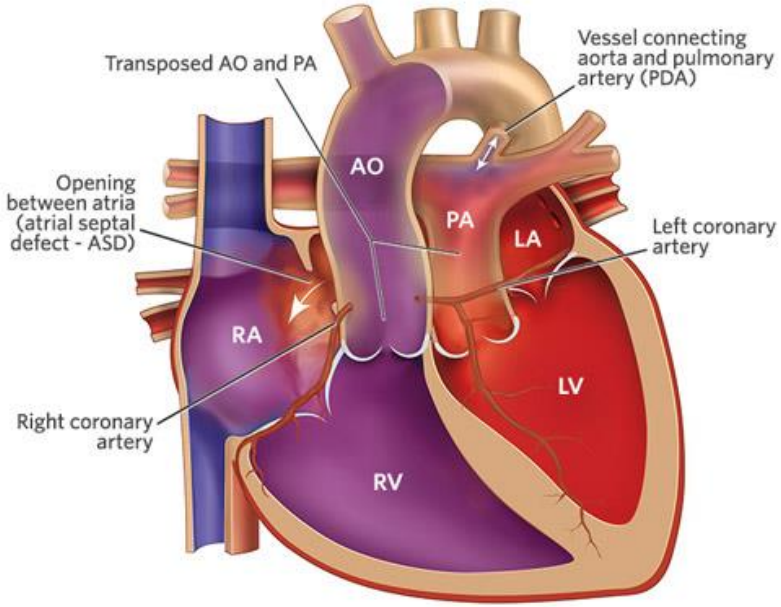
- TAPVR

Transposition of the Great Vessels

Normal Heart



D-TGA



Transposition of the Great Arteries

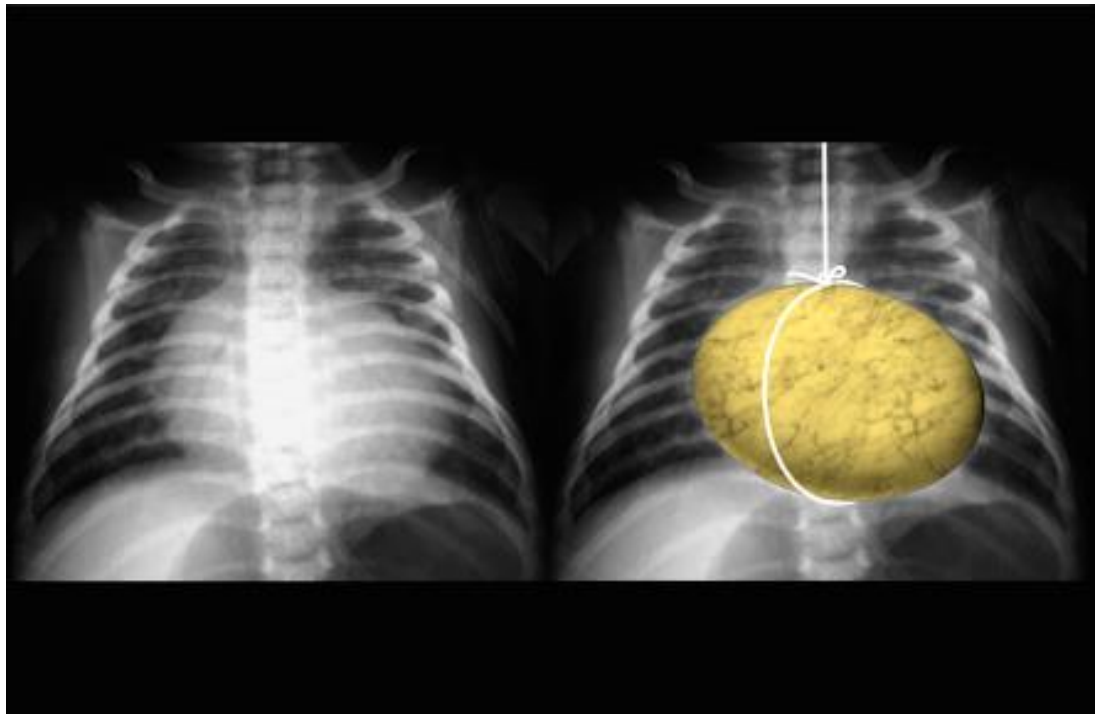
- **Transposition of the great arteries is the second most common cyanotic CHD, accounting for about 2% of all**
- **CHD.**
- **It is the most common cyanotic heart disease manifesting in the first week after birth.**
- **There is ventriculoarterial discordance, with the aorta arising from the RV (usually anterior to the pulmonary artery) and the pulmonary artery arising from the LV. Hence, the systemic**
- **and pulmonary circulations are in parallel, with systemic venous (deoxygenated) blood returning to the right atrium, the RV, and going out the aorta again.**
- **There is mixing of blood at the atrial level through a patent foramen ovale or an ASD or at the ventricular level through a VSD (35% to 40% of transposition of the great arteries).**

TGA

Presentation

- Severe Cyanosis
- Acidosis
- No respiratory Distress

Chest Xray

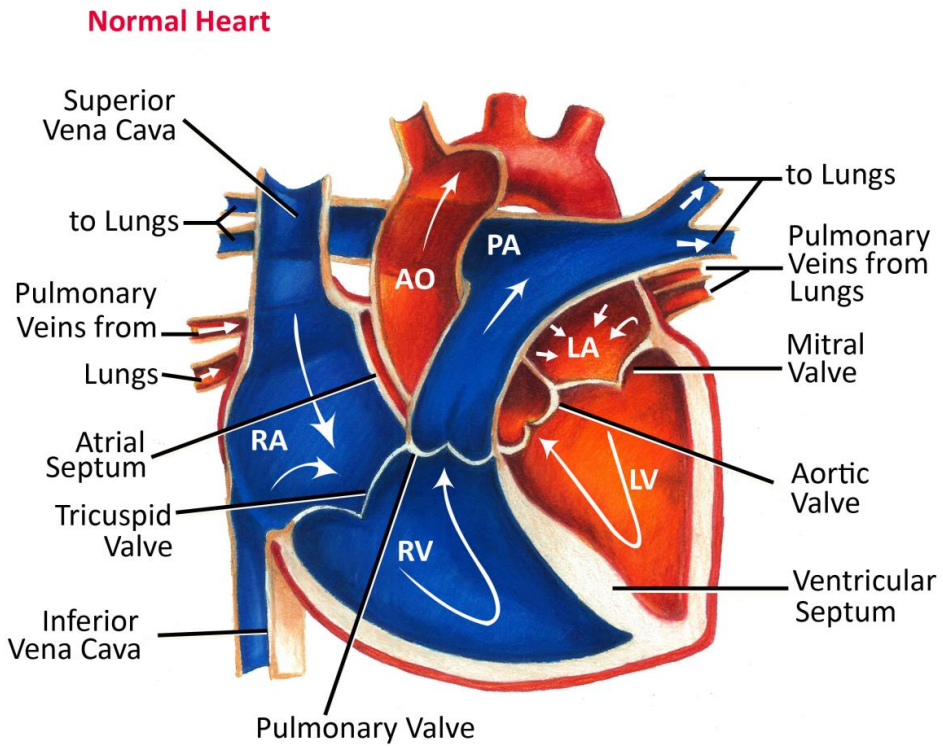


- Clinical Manifestations and Diagnosis. Newborns with transposition of the great arteries present with cyanosis within the first 12 hours after birth and are not responsive to oxygen or mechanical ventilation.
- The presence of a VSD may delay presentation.
- Chest radiography may show a narrow mediastinal silhouette due to the orientation of the
- great vessels and thymic regression, and EKGresults may b normal or show RV hypertrophy.
- There is usually no murmur at examination; however, there may be a single S2 with a
- loud aortic component due to the orientation of the great vessels.

- Management. Once the diagnosis is established, these
- patients require reparative surgery to switch the great vessels
- to the appropriate ventricles, known as the arterial switch
- procedure.
- They may need respiratory support with oxygen, mechanical ventilation, and initiation of prostaglandin

Truncus Arteriosus

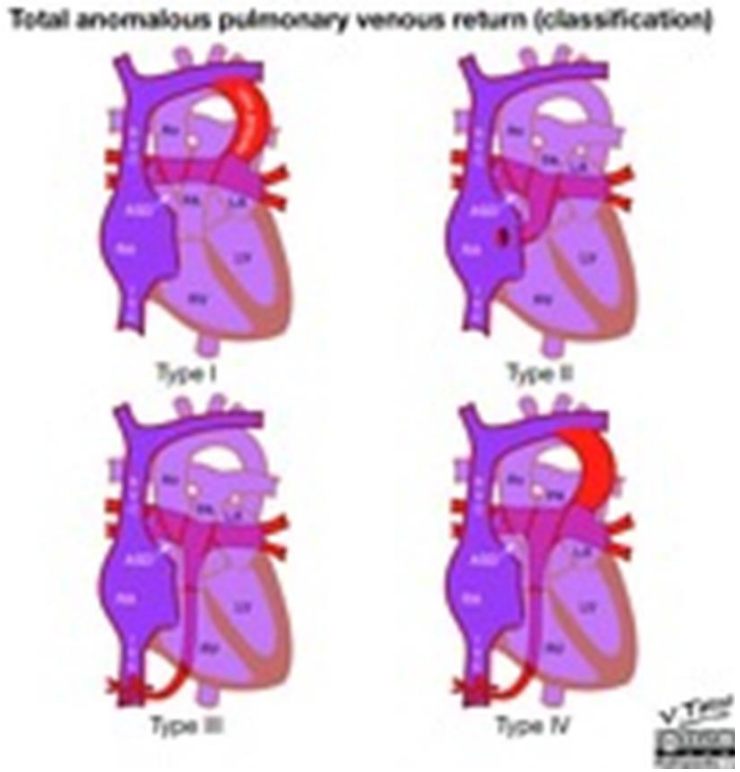
Normal Heart



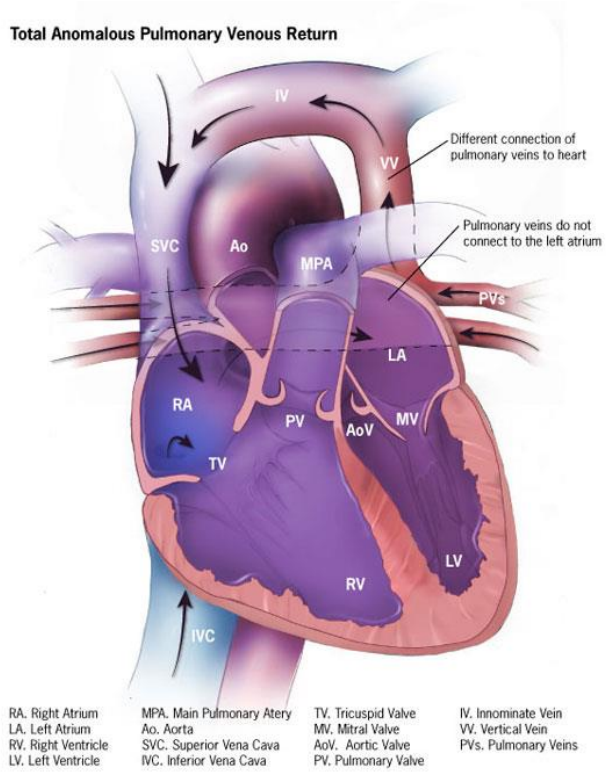
Truncus Arteriosus

Total Anomalous Pulmonary Venous Return

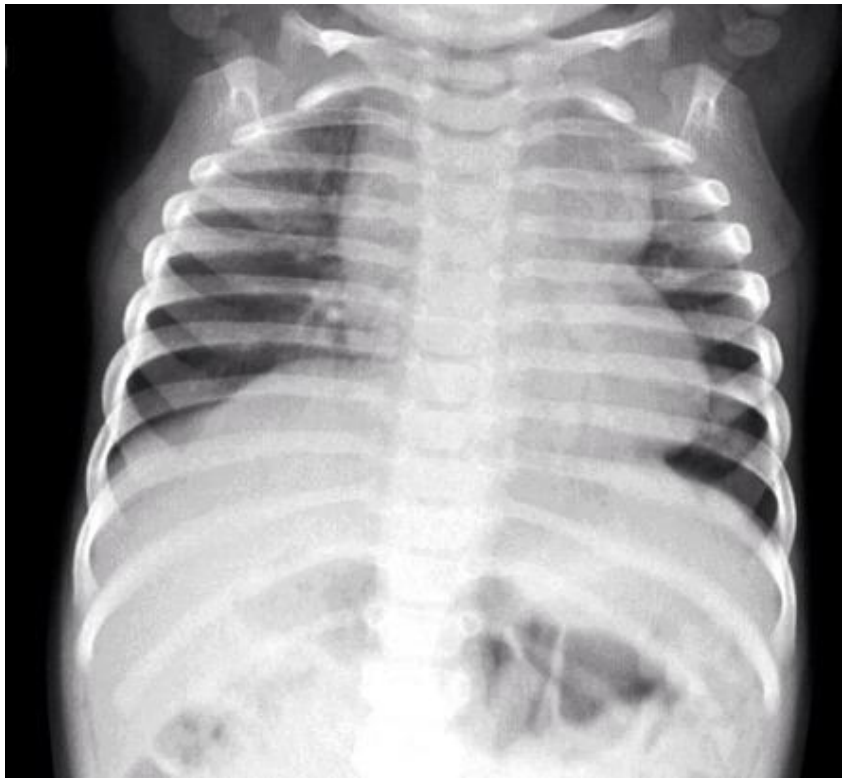
Types of TAPVR



Supra Cardiac TAPVR



Supra Cardiac TAPVR Xray



Presentation

Management of Cyantoic Heart Disease

- Severe Cyanosis
- Acidosis
- PDA dependant lesion
- Start PGE1
- All will improve except infracardiac obstructive TAPVR

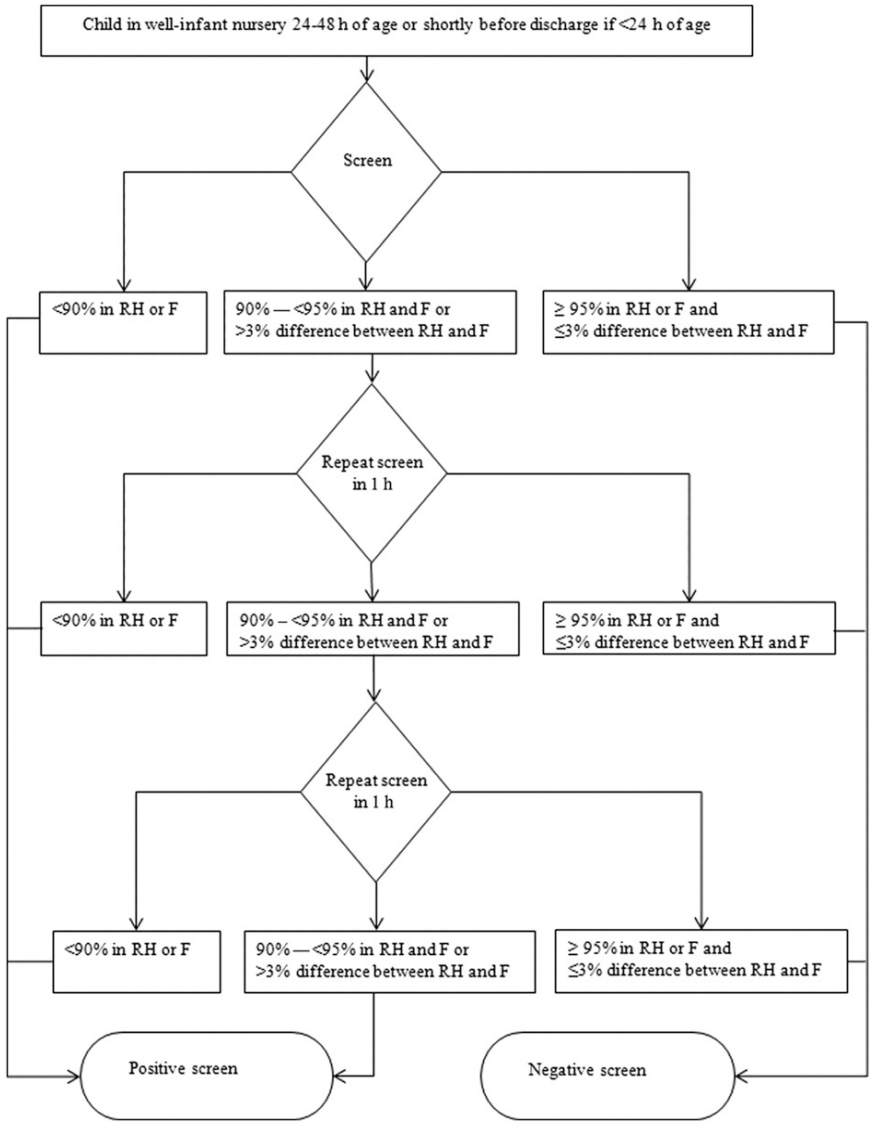
Approach to a Cyanotic New Born

- It may be challenging to distinguish cardiac disease from pulmonary disease or sepsis in a newborn with cyanosis and/or respiratory distress
- History and Physical Examination and Degree of Cyanosis and Acidosis
- Assessment and Chest radiography
- Response to oxygen and/or positive pressure ventilation
- Lack of response to 100% oxygen for at least 10 minutes (hyperoxygenation) indicates that the etiologic origin for the clinical picture is likely cardiac, and further cardiac workup is indicated.

- chest radiography and echocardiography performed to rule out cyanotic CHD are indicated in a cyanotic infant who is not responsive to hyperoxygenation.

- The pulse oximetry screen for critical CHD in newborns was approved as a part of the Routine Universal Screening Program in 2011 and has been adopted in 46 states and the District of Columbia.

Pulse ox screen



- History
- Physical Examination.
- Hyperoxia Test and UL LL Saturations
- Chest Xray failed screening result is an indication for prompt evaluation, including chest radiography, EKG, and echocardiography (supervised and evaluated by a pediatric cardiologist).
- Determine if the degree of cyanosis is severe
- Determine if you have metabolic acidosis
- Severe cyanosis and metabolic acidosis initiate PGE1 with or without bicarbonate
- If cardiac investigations or a pediatric cardiologist are not easily available in such a situation, initiation of prostaglandin infusion with close monitoring of the airway (due to risk of apnea with prostaglandin initiation) and prompt transport to a higher-level center are recommended.

TOF Presentation

History

Physical Examination

- E1 until the time of surgery. Prostaglandin allows the PDA to remain open, which shunts blood from the aorta into the pulmonary circulation. This increases the amount of blood
- returning to the left atrium and mixing at the atrial level, as
- long as the foramen is open. Most patients undergo a
- catheter-based procedure called balloon atrial septostomy
- to help create or enlarge the ASD to allow more mixing
- while awaiting surgery. If cyanosis persists despite an open
- PDA, a fluid bolus or inotropic support may help improve
- mixing and systemic arterial saturations. After repair, these
- patients require regular cardiology follow-up for life. The
- first arterial switch procedure was performed in 1975, and
- since the patients undergoing this procedure are followed
- up for progressively longer periods of time, data about the
- long-term complications are emerging. Depending on the
- study, 5% to 30% of these patients have been reported to
- require reintervention at 25-year follow-up for a variety of
- reasons, most commonly regurgitation of the neoaortic
- valve (more than 75%), supravalvar pulmonary stenosis
- (more than 75%), and coronary artery disease (5% to 8%).
- (28)(29)