

# Prehospital Recognition and Management of Fetal, Neonatal, and Pediatric Congestive Heart Failure

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## Objectives:

1. How do we define “congestive heart failure” in infants and children?
2. What is the incidence of congestive heart failure in pediatric patients?
3. Should we do anything differently when we assess the child?
4. How are we going to treat these children?
5. How will our pre-hospital interventions (or lack thereof) impact the child's overall medical situation?

## Congestive Heart Failure: Overview

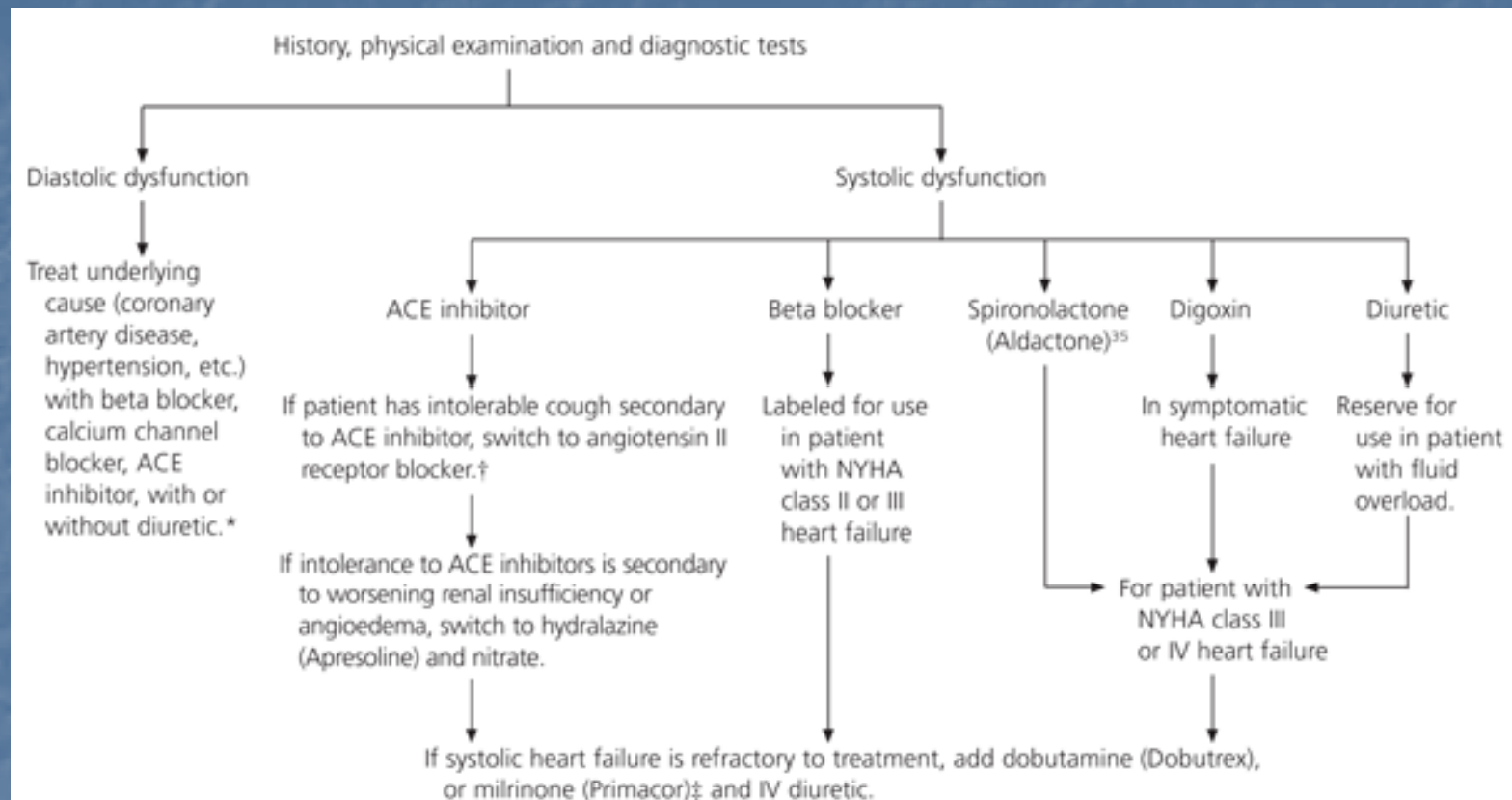
- “A clinical state of systemic and pulmonary congestion resulting from the inability of the heart to pump as much blood as is required to meet the metabolic demands of the body at normal physiologic venous pressures.”
- Causes:
  - Volume overload and/or pressure overload on a normal myocardium:
    - Left-to-right shunts, aortic stenosis
  - Primary myocardial abnormality:
    - Myocarditis, cardiomyopathy
  - Arrhythmias
    - Supraventricular/ventricular tachyarrhythmias, atrial fibrillation (controlled or uncontrolled)
  - Pericardial diseases
    - Pericardial effusion



## Congestive Heart Failure: Overview

- The heart can respond to increased demands by means of one of the following:
  - Tachycardia (relative)
    - (Controlled by neural and humoral input.)
  - Ventricular hypercontractility
    - (Circulating catecholamines and autonomic input.)
  - Preload augmentation
    - (Mediated by constriction of the venous capacitance vessels and the renal preservation of intravascular volume.)
- As the demands on the heart outstrip the normal range of physiologic compensatory mechanisms, signs of CHF occur.
  - Tachycardia (excessive)
  - Systemic venous congestion
  - High catecholamine levels
  - Insufficient cardiac output with poor perfusion and end-organ compromise

# Systolic Dysfunction



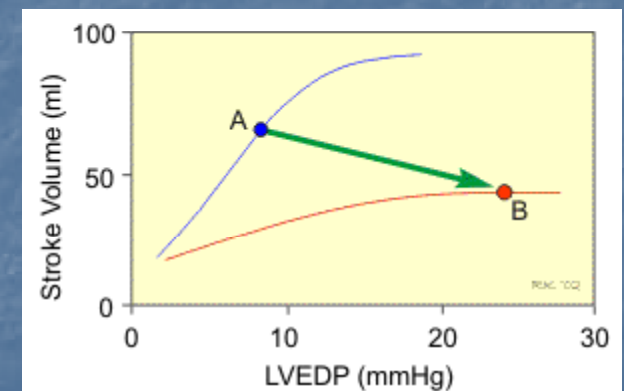
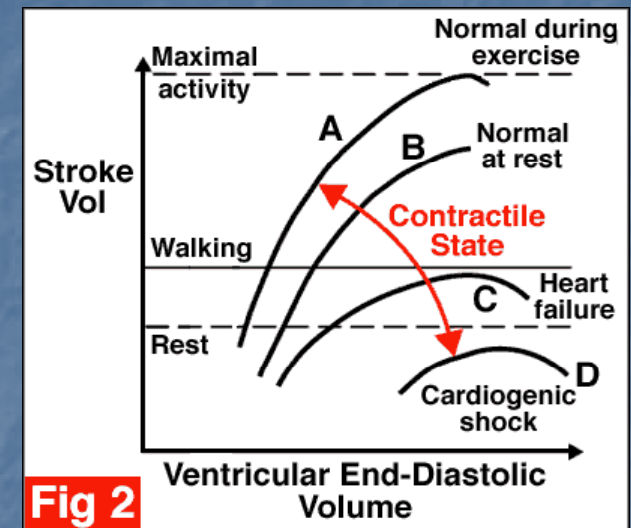
\*—Diuretics are best used to treat acute congestive heart failure and as adjunctive therapy for hypertension.

†—Note that the likelihood of angioedema and renal insufficiency is increased with ACE inhibitors and angiotensin-receptor blockers. Watch for late-breaking results from clinical trials on the efficacy of angiotensin-receptor blockers alone and in combination with ACE inhibitors compared with ACE inhibitors alone.

‡—The addition of milrinone is preferred in patients already receiving a beta blocker.

## Systolic Dysfunction

- Characterized by diminished ventricular contractility that results in an impaired ability to increase the stroke volume to meet systemic demands.
- Decreased contractility causes a downward shift in the Frank-Starling curve.
- This results in a decreased stroke volume and a compensatory rise in preload.
- Contributing factors
  - Anatomic stresses (eg, coarctation of the aorta)
    - → Increased afterload (end-systolic wall stress)
  - Neurohormonal factors
    - → Increased SVR (systolic dysfunction)

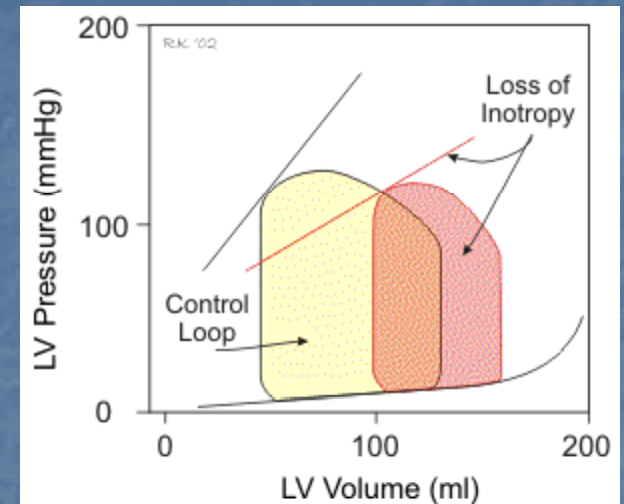


**Figure 1.** The Frank-Starling relationship showing the effects of heart failure (ventricular systolic dysfunction, loss of inotropy) on stroke volume and ventricular preload (left ventricular end-diastolic pressure, LVEDP). Point A, control point; point B, ventricular failure.

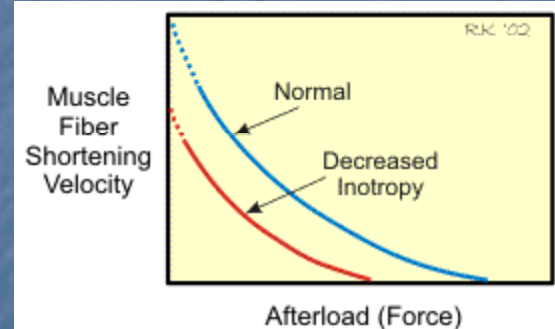


## Systolic Dysfunction

- The rise in preload is considered compensatory because it activates the Frank-Starling mechanism to help maintain SV despite the loss of inotropy.
- The net effect of systolic dysfunction is a decrease in stroke volume.
- Because stroke volume decreases and end-diastolic volume increases, there is a substantial reduction in EF.
- Reduction in EF occurs because a loss of inotropy yields a decrease in the shortening velocity of cardiac fibers.
- Treatment for systolic dysfunction involves the use of inotropic drugs, afterload reducing drugs, venous dilators, and diuretics.



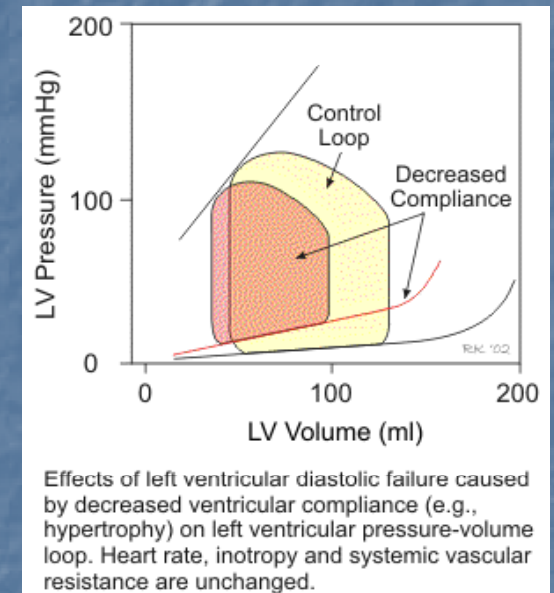
**Figure 2.** Effects of acute left ventricular failure (loss of inotropy) on left ventricular pressure-volume loop. Heart rate unchanged.



**Figure 3.** Effects of ventricular failure (decreased inotropy) on the force-velocity relationship. Decreased inotropy decreases velocity of fiber shortening at any given afterload.

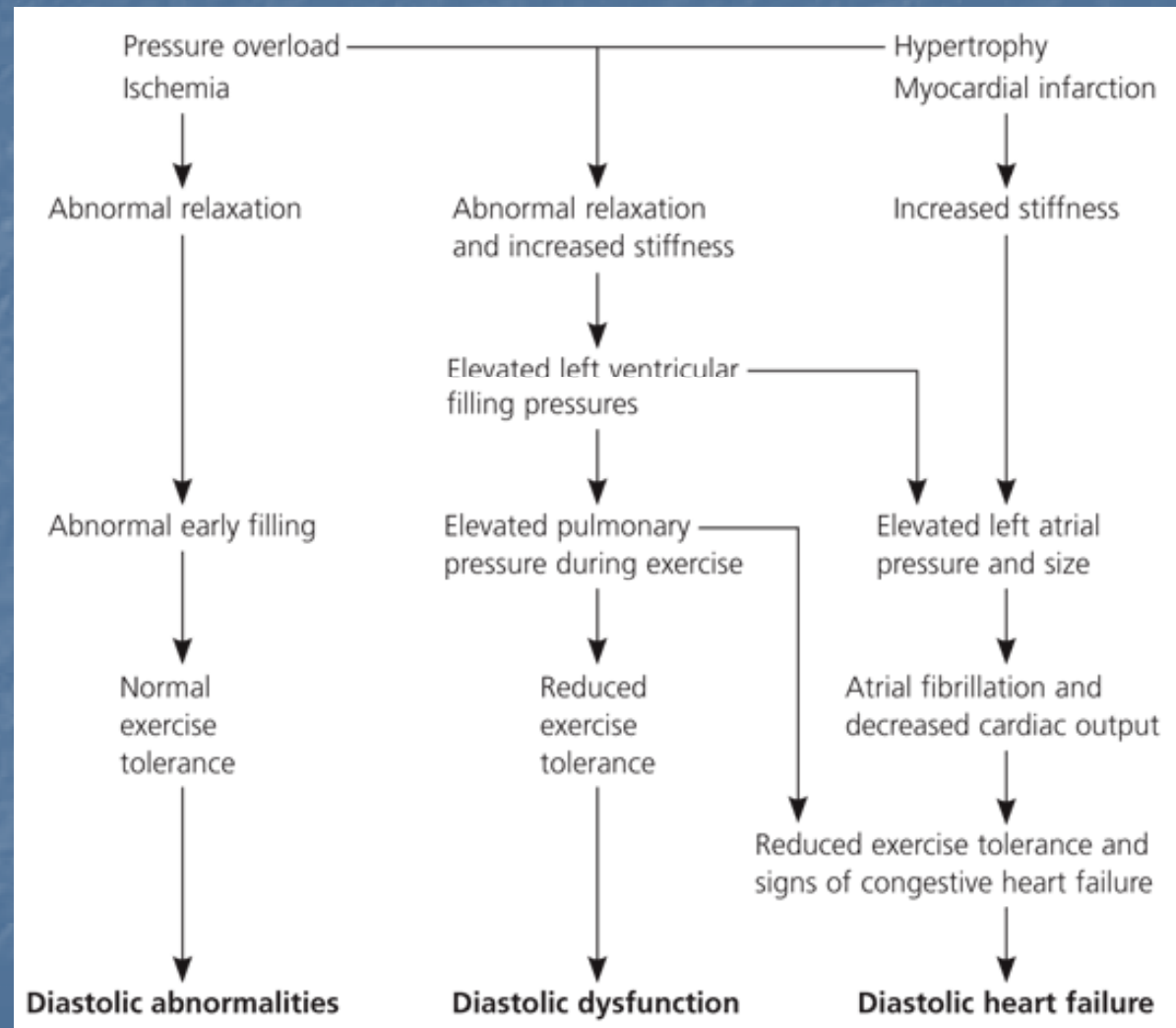
# Diastolic Dysfunction

- Causes of primary diastolic dysfunction include:
  - An anatomic obstruction that prevents ventricular filling
    - Pulmonary venous obstruction
  - A primary reduction in ventricular compliance
    - Cardiomyopathy, transplant rejection
  - External constraints
    - Pericardial effusion
  - Poor hemodynamics after the Fontan procedure
    - Elevated pulmonary vascular resistance
- Changes in ventricular diastolic properties have an adverse effect on stroke volume.
- LVEDV depends upon the venous return and compliance of the ventricle during diastole.



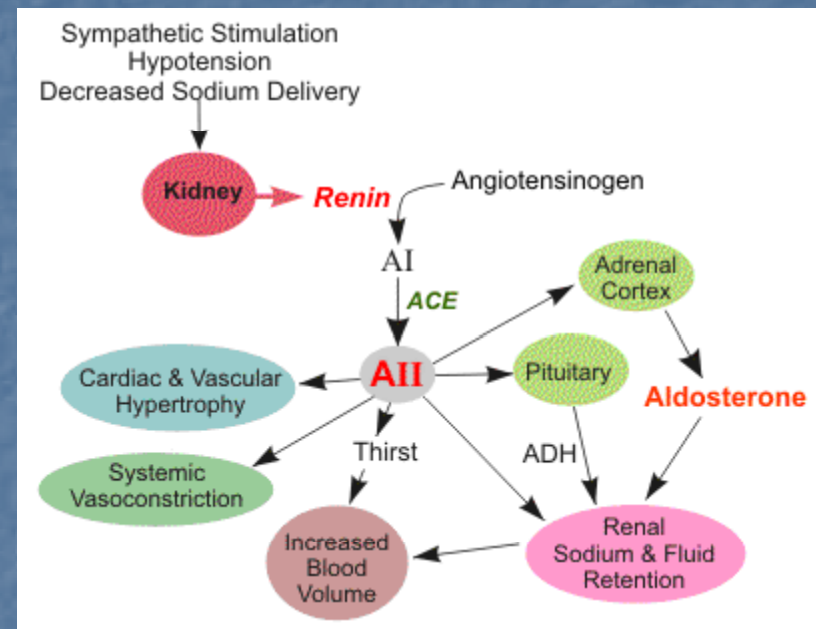


## Diastolic Dysfunction



## Diastolic Dysfunction

- LV Diastolic failure → Decreased LVEDV  
→ Increased LVEDP → Decreased EF
- RV Diastolic failure → Increased LVEDP  
→ Systemic congestion/ascites
- Depending on the change in SV and LVEDV,  
there may or may not be a decrease in EF.
- Increased venous pressures also occur because  
of an increase in blood volume due to  
activation of the RAAS, which causes renal  
retention of sodium and water.
- Treatment for diastolic dysfunction involves  
the use of diuretics and ACE inhibitors.



(Figure 1. Renin-Angiotensin-Aldosterone System)

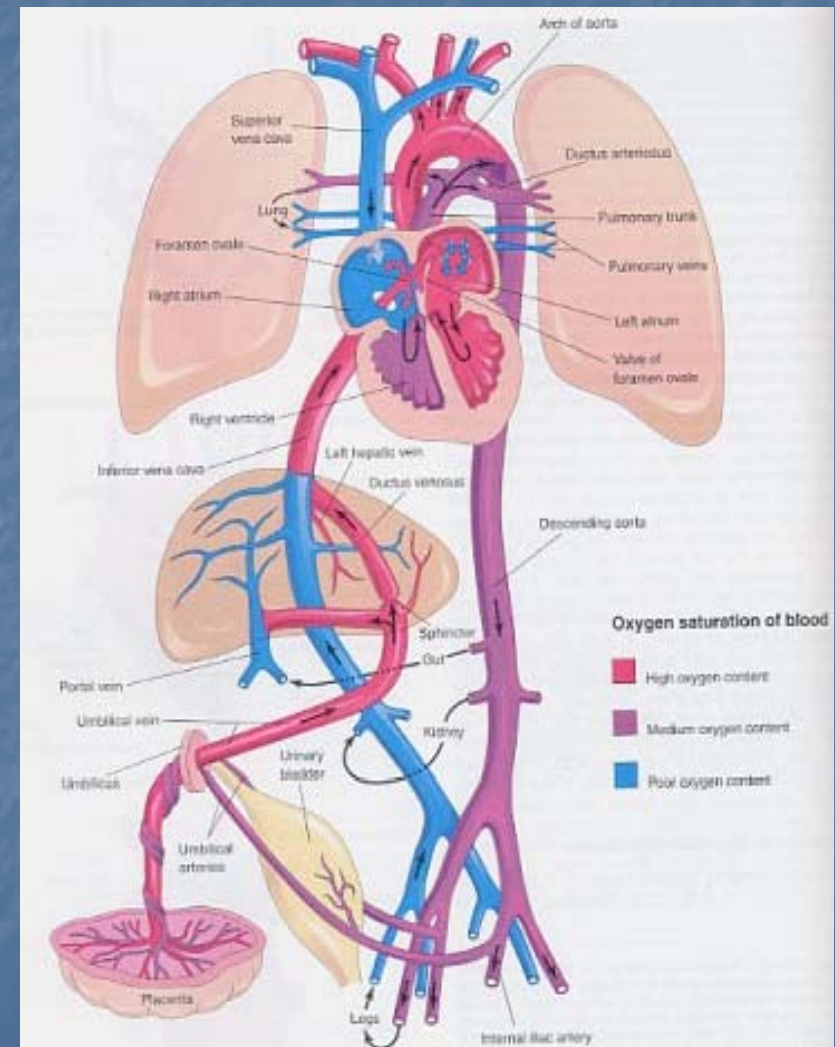
# Fetal Congestive Heart Failure





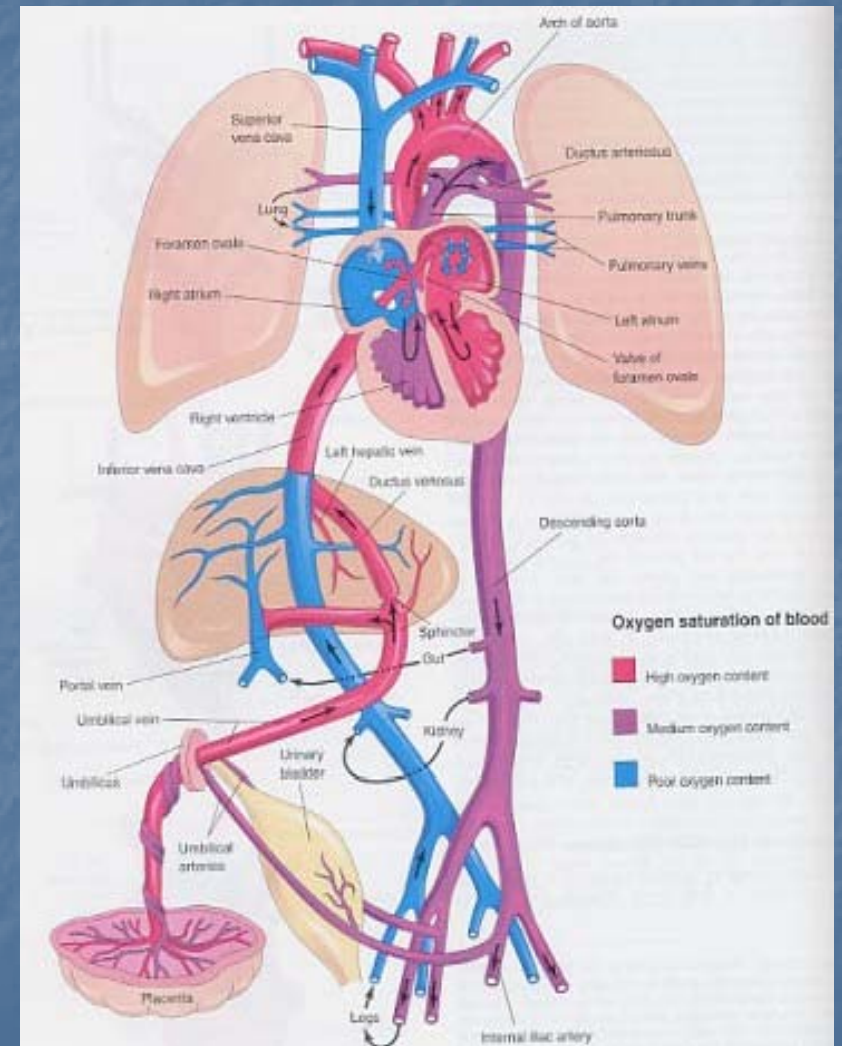
## Fetal Circulation in Review:

- In the fetus, the ventricles pump blood in parallel rather than in series.
  - LV → aorta, upper body
  - RV → ductus arteriosus, lower body, placenta
- The lungs have a high resistance in utero and the placenta fulfills the role of oxygenating the blood and ridding the body of waste.
- Highly oxygenated blood from the placenta passes to the ductus venosus where a portion bypasses the liver and passes predominantly to the left atrium.
- The relatively de-oxygenated blood from the upper body passes to the tricuspid valve and then to the ductus arteriosus and lungs.



## Fetal Circulation in Review:

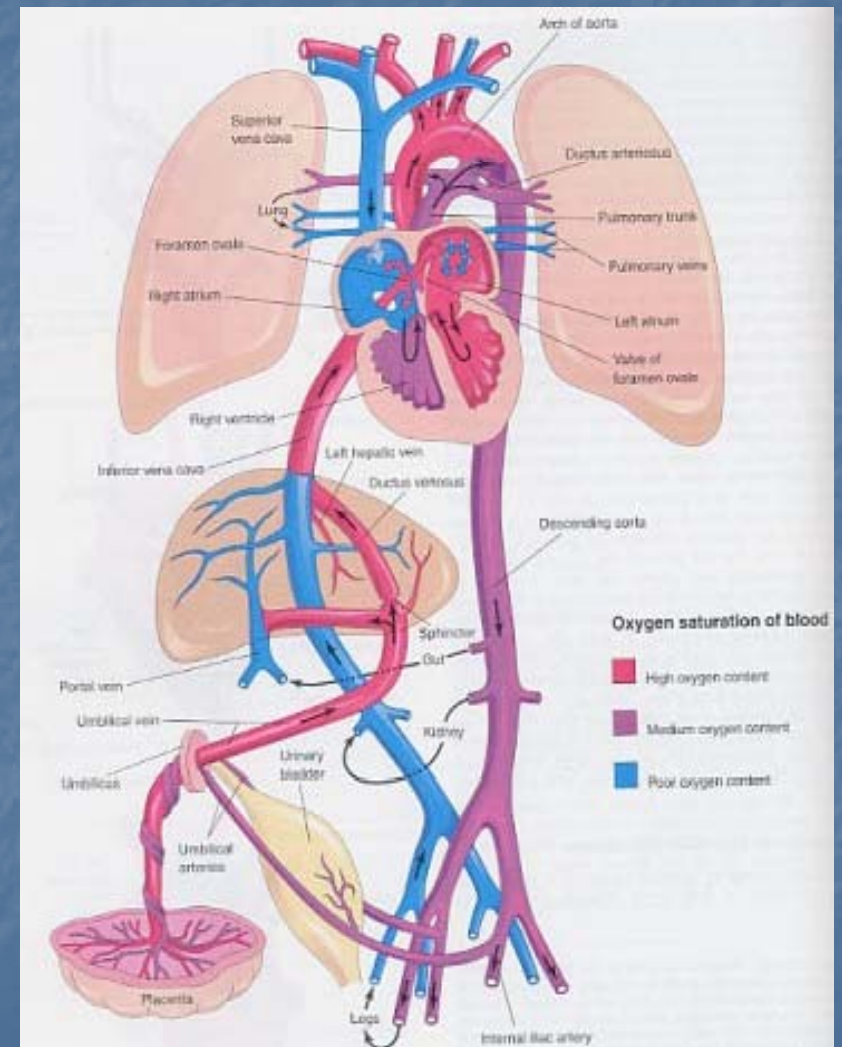
- The de-oxygenated blood from the IVC and the right hepatic veins is directed to the RA and predominantly to the tricuspid valve.
- This distribution of lower body flow is accomplished by the posterior portion of the IVC connecting directly to the foramen ovale, and the superior portion of the atrial septum, which overlies the IVC, effectively dividing it into two streams.
- Presence of three shunts allows the fetal heart to work with two parallel circulations rather than one series circulation.
  - *ductus venosus*
  - *foramen ovale*
  - *ductus arteriosus*





## Fetal Circulation in Review:

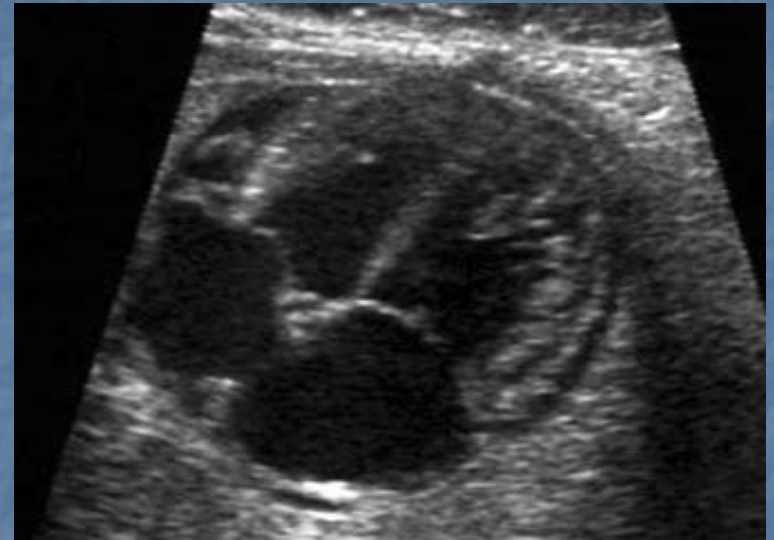
- Right and left atrial pressures are almost equal because of the presence of the foramen ovale, and right and left ventricular pressures are equal due to the ductus arteriosus.
- LV → upper body, cerebral circulation
- RV → pulmonary arteries, lower body, placental circulation via ductus arteriosus
- As a further consequence of the parallel circulations, ventricular outputs can be different.
- In the case of obstruction on one side of the heart, the other side is able to increase its work or even to supply the whole circulation alone.





## Fetal Congestive Heart Failure: Overview

- Disorders that are fatal in the immediate neonatal period are often well tolerated in the fetus due to the pattern of fetal blood flow
  - Presence of three shunts allows the fetal heart to work with two parallel circulations rather than one series circulation.
    - *ductus venosus*
    - *foramen ovale*
    - *ductus arteriosus*
- Nonimmune hydrops fetalis
  - Produced by severe CHF in the fetus
  - Characterized by accumulation of edema in at least 2 compartments
    - Ascites
    - Pleural and pericardial effusions
    - Anasarca (extreme generalized edema)



# Fetal Congestive Heart Failure: Overview

## Incidence of Fetal CHF:

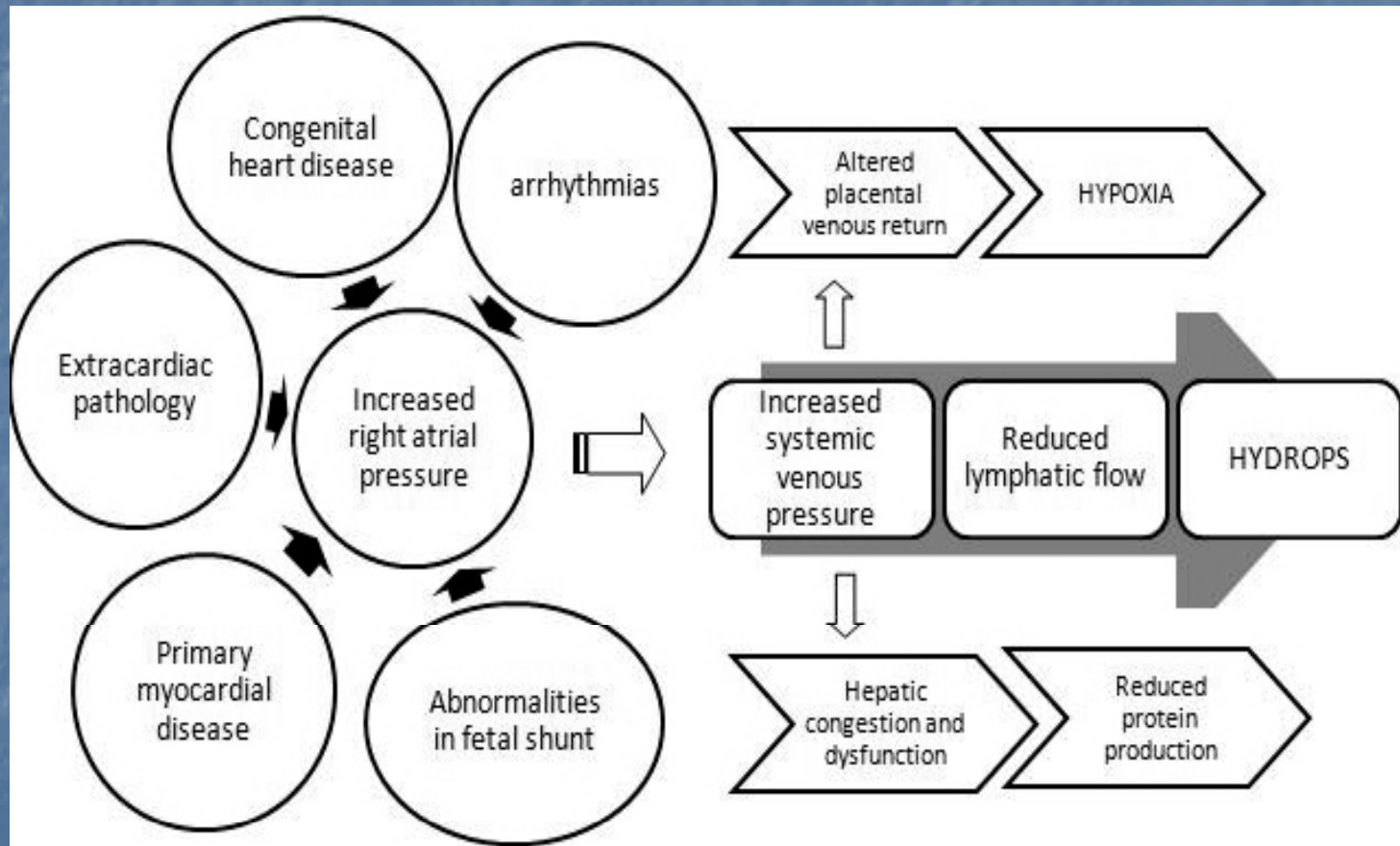
- Approximately 8 out of every 1,000 pregnancies.

## Causes of Fetal CHF:

- SVT, VT, 3rd degree AV block or other fetal arrhythmias
- Anemia
- Myocarditis
- Congenital heart disease with valvular regurgitation
  - Severe tricuspid regurgitation due to Ebstein's anomaly of the tricuspid valve
- Mitral regurgitation due to AV canal defect
- AV fistula with high cardiac output
- Non-cardiac malformations
  - Diaphragmatic hernia or cystic hygroma
- Twin-twin transfusion recipient volume and pressure overload

# Fetal Congestive Heart Failure: Pathophysiology

## Pathophysiology of Fetal Congestive Heart Failure:





# Fetal Congestive Heart Failure: Pathophysiology

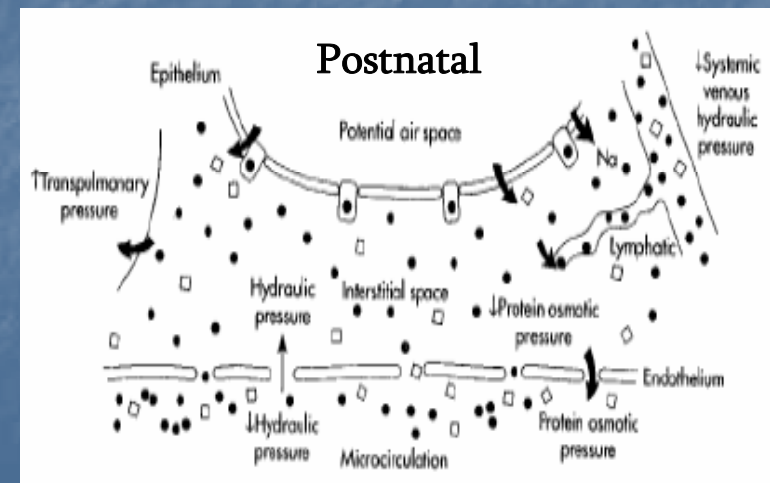
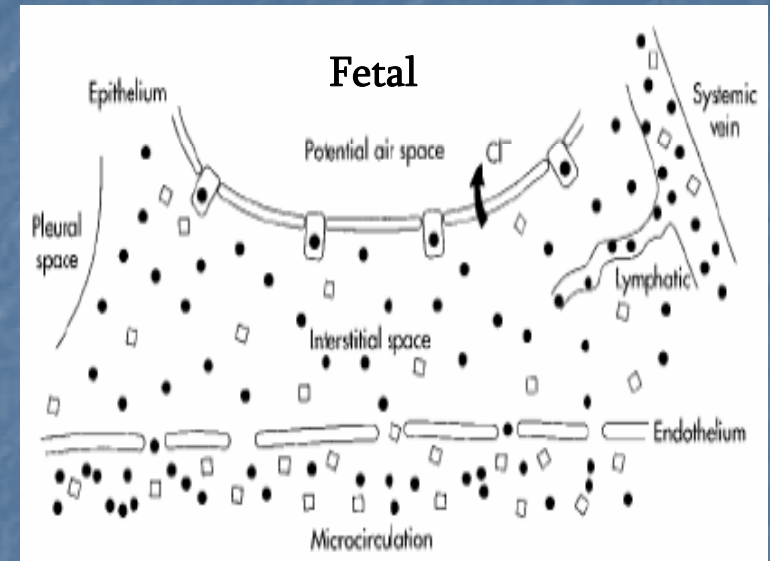
## Pathophysiology of Fetal Congestive Heart Failure:

- End-stage fetal heart failure results in hydrops fetalis.
- Causes of to decreased cardiac reserve in response to stress and to higher susceptibility of the fetus for CHF:
  - Reduced ability of the fetal heart to contract and generate force
  - Lower myocardial compliance
  - Diminished Frank-Starling mechanism
- With increasing atrial pressure, the output of the heart plateaus at a much lower pressure in utero than postnatally.

# Fetal Congestive Heart Failure: Pathophysiology

## Pathophysiology of Fetal Congestive Heart Failure:

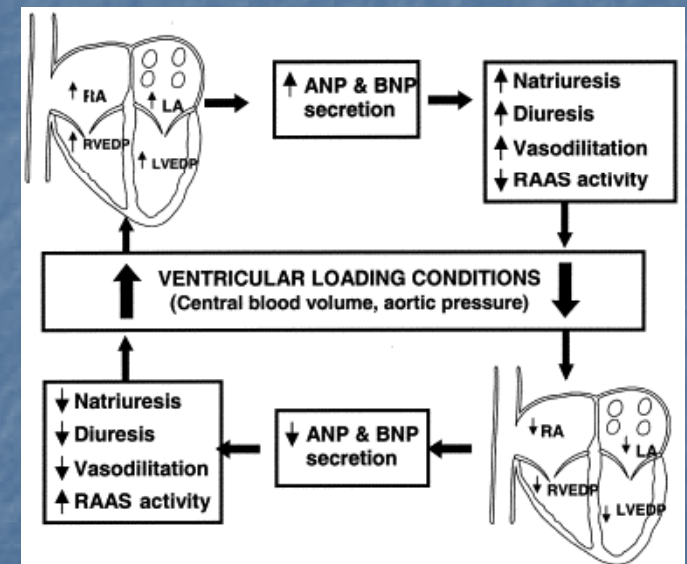
- In the fetus, even small increases in venous pressure have been shown to alter fetal organ function.
- Factors favoring fluid movement out of capillaries into the surrounding tissues:
  - The younger the fetus, extracellular water content increases and tissue pressure decreases.
  - In the fetus, a more permeable capillary membrane permits fluid and protein movement between intravascular and extravascular spaces.
  - Albumin concentration, largely responsible for oncotic pressure, is lower in the fetus and increases with gestational age.



# Fetal Congestive Heart Failure: Pathophysiology

## Pathophysiology of Fetal Congestive Heart Failure:

- Elevated venous pressure may reduce lymphatic flow, further favoring the development of hydrops.
- Decreased arterial blood pressure and elevated filling pressures also trigger hormonal responses :
  - Production of plasma arginine vasopressin[ADH]; decreasing urinary production
  - Production of angiotensin II; increasing fluid accumulation
  - Production of atrial natriuretic peptide[ANP]; increasing capillary permeability

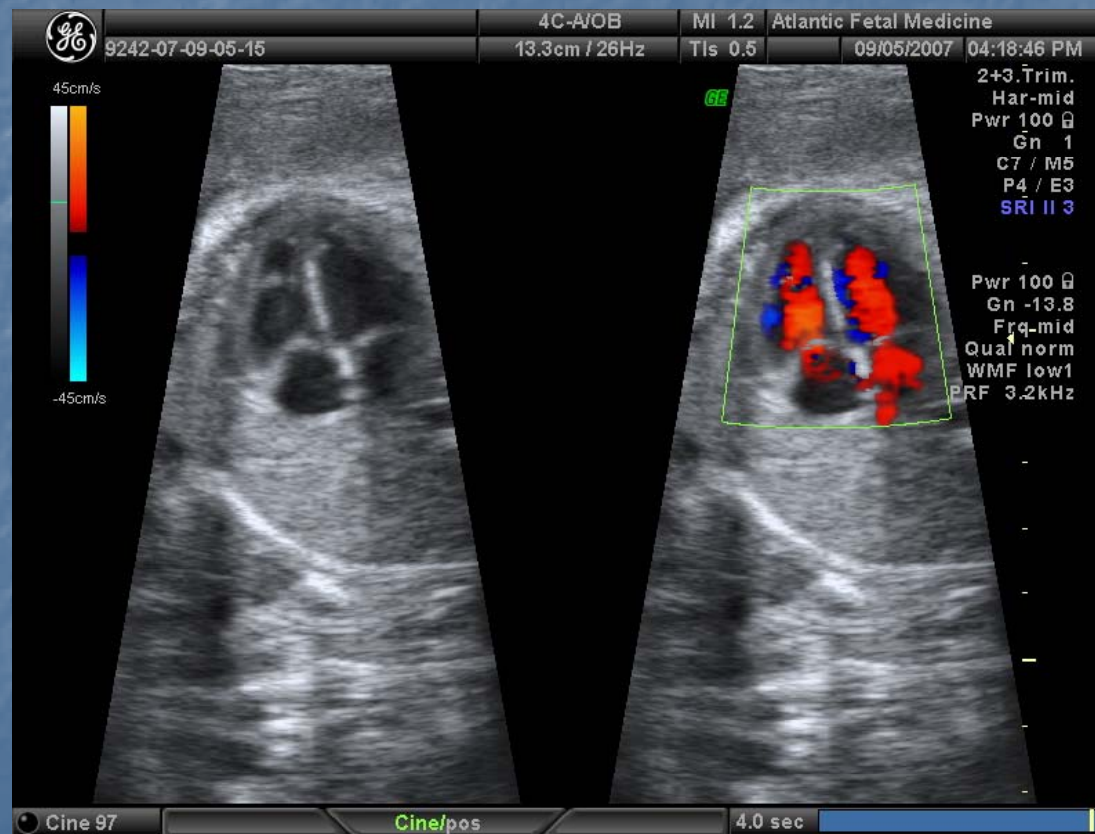




# Fetal Congestive Heart Failure: Assessment

## Assessment of Fetal Congestive Heart Failure:

- Initial data about the fetus is collected using echocardiography.



# Fetal Congestive Heart Failure: Assessment

## Assessment of Fetal Congestive Heart Failure:

- Initial data about the fetus is collected using echocardiography.
- Cardiac size/thoracic size:
  - Cardiac area : Thoracic area (C/T) Ratio (normal 0.25-0.35)  
-or -
  - C/T circumference ratio (normal <0.5).
- Venous Doppler:
  - Inferior caval (or hepatic venous) (increased atrial reversal) and umbilical cord vein (pulsations).
- Four-valve Doppler:
  - Any valvular leaks should be evaluated further.
- If there are abnormalities in any of these measurements, a cardiac cause or associated physiological problem may be present and detailed study is indicated to rule out serious cardiovascular involvement.

# Fetal Congestive Heart Failure: Management

## Management of Fetal Congestive Heart Failure:

- Usual treatment of placental dysfunction is designed to improve the vascular impedance of the placenta and to increase the flow of oxygenated blood to the fetus.
  - Bedrest
  - Improved nutrition
  - Maternal oxygen (NC is more than sufficient, unless mother is overtly hypoxic/cyanotic)
    - May lead to an improvement in placental function.
  - Tocolytics (terbutaline, nifedipine, salbutamol, ritodrine, magnesium sulfate, indomethacin)
    - May relax the placenta and improve its function.
- Advanced heart failure in this setting with severely decreased arterial pO<sub>2</sub> and poor nutrition is manifested by non-specific signs of increased right ventricle and right atrium size, as well as atrial reversal in the venous Doppler pattern, and altered forward flow velocities.



# Fetal Congestive Heart Failure: Management

## Management of Fetal Congestive Heart Failure:

- Oxygen
  - Maternal oxygen
    - (NC is more than sufficient, unless mother is overtly hypoxic/cyanotic)
  - May lead to an improvement in placental function.
    - **Oxygen Flow Rate: 1-4 L/min via NC.**
- Corticosteroids (Dexamethasone):
  - Early use of this medication may prevent progression of fetal/antenatal heart block and myocardial injury later in life.
  - In pregnancies where the mother has SLE and/or Sjögrens syndrome, dexamethasone has been shown to be beneficial if there are signs of valvular regurgitation, heart block, valvulitis, myocardial dysfunction, or effusion.
    - **Dexamethasone Dosage: 4 mg PO QD given to the mother.**
    - **\*\*\*IV dexamethasone is commonly given PO, in a small volume of water or juice.**



# Fetal Congestive Heart Failure: Management

## Management of Fetal Congestive Heart Failure:

- **Maternal Digitalization (Digoxin)**
  - When cardiomegaly is present, treatment of the fetus is reasonable if the pregnancy will be continuing for long enough for medication levels to reach therapeutic levels.
  - Limits the progression of CHF in fetuses with an abnormal cardiovascular system, but is not effective once fetal hydrops is present.
  - Digoxin has been used in such circumstances due to its antiadrenergic benefits and the significant experience that has been gained about its safety in pregnancy.
    - **Digoxin Dosage: 0.5 mg PO x 1 (\*\*IN CONSULTATION WITH M.D.\*\*)**
      - » After initial dose: 0.5 mg BID, followed by 0.25 – 0.75 mg/day.
      - » Fetal/maternal ratio: 0.3 – 1.3
      - » Based on maternal serum levels, with a trough level of 1.0 - 2.5 ng/mL to avoid any maternal side effects.

# Fetal Congestive Heart Failure: Management

## Management of Fetal Congestive Heart Failure:

- Tocolytics (Terbutaline/Magnesium/Nifedipine):
  - Tocolytics are designed to relax the placenta and improve function.
  - Nifedipine is a CCB that increases cardiac output and oxygen delivery during rest and exercise in PPH, and provides acute pulmonary vasodilation by decreasing pulmonary VR.
- Terbutaline Sulfate Dosage: 0.25 mg SQ/IM/IV(preferred route) q20 min
  - » Alt. Dosage : 0.25 mg(250mcg) IV x 1, followed by 5 mcg/min drip.
- Magnesium Sulfate Dosage: 4 gram load IV over 30 min, followed by 1 gram/hr drip.
- Nifedipine Dosage: 10 mg PO q15 min up to 4 doses (40mg)
  - » \*\*\*DISCUSS WITH MD PRIOR TO ADMINISTERING\*\*\*
  - » \*\*\*SHOULD NOT BE COMBINED WITH OTHER TOCOLYTICS\*\*\*

# Fetal Congestive Heart Failure: Management

## Management of Fetal Congestive Heart Failure:

- In-Hospital Advanced Care for Fetal Congestive Heart Failure:
  - Intravascular Transfusion (via umbilical vein)
    - Hydrops or fetal anemia (Hct level < 30%) is an indication for umbilical vein transfusion in infants with pulmonary immaturity.
    - Maternal sedation → diazepam; Fetal sedation/paralysis → diazepam/pancuronium.
    - PRBCs given by slow-push infusion after cross matching with the mother.
      - » CMV-negative, leukopoor following irradiation to minimize risk of GVHD.
    - Post-transfusion Hct goal of 45–55% and PRBC's can be repeated every 3–5 weeks.
    - The survival rate for intrauterine transfusions is 89%; the complication rate is 3%.
      - » Potential Complications: Membrane rupture/preterm delivery, infection, fetal distress requiring emergency cesarean delivery, and perinatal death.



# Neonatal Congestive Heart Failure



# Neonatal Congestive Heart Failure: Overview

## Incidence of Neonatal CHF:

- Approximately 30% of the roughly 32,000 infants with CHD born each year in the United States.

## Causes of Neonatal CHF:

- Neonates and infants younger than age 2 months are the most likely group to present with congestive heart failure related to structural heart disease.
- The systemic or pulmonary circulation may depend on the patency of the ductus arteriosus, especially in patients presenting in the first few days of life.
- In these patients, prompt cardiac evaluation is mandatory!
- Myocardial disease due to primary myopathic abnormalities or inborn errors of metabolism must be investigated.
- Consider and treat all potentially reversible causes from the outset:
  - Respiratory illnesses
  - Anemia
  - Known or suspected infection (septicemia) must be considered and appropriately managed.
- The diagnosis of CHF in older children is often straightforward, but it may be difficult to distinguish CHF from pulmonary disease or sepsis in the neonate.

# Neonatal Congestive Heart Failure: Overview

## Overview of Neonatal CHF:

- In the past, the most sensitive and specific variables for the presence of CHF were:
  - A history of less than 3.5 oz/feed
  - Abnormal respiratory pattern with a resting respiratory rate greater than 50/min
  - Diastolic filling sounds and hepatomegaly
- Moderate to severe CHF was considered to be present when:
  - A history of less than 3 oz/feed or greater than 40 min/feed
  - Abnormal respiratory pattern with a resting respiratory rate greater than 60/min
  - Diastolic filling sounds and moderate hepatomegaly
- Severe CHF was accompanied by a resting heart rate greater than 170/min, decreased perfusion, and severe hepatomegaly.
- The grading of the severity of CHF in infants should include an accurate description of these historical and clinical variables.



# Neonatal Congestive Heart Failure: Overview

## Overview of Neonatal CHF:

- Usual presenting features:
  - Feeding difficulties
  - Excessive/inappropriate diaphoresis
  - Excessive/inappropriate tachycardia
  - Respiratory distress
  - Irritability
  - Weak cry
- Grading the severity of CHF in infants is difficult and is not standardized.
- Resting heart rates(in a calm infant) in excess of 180/min are abnormal even in the setting of respiratory distress and suggest CHF.

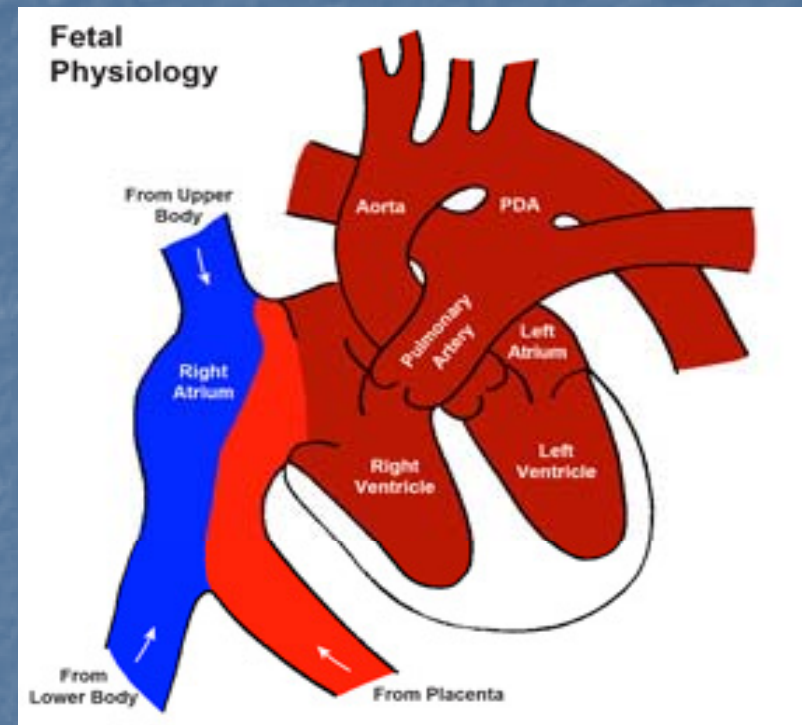


*9 day old with CHF 2<sup>o</sup> undiagnosed HLHS; precipitated by circumcision. During circumcision, SpO<sub>2</sub> dropped to 75%; pt. received emergent PGE-1 infusion, and heart surgery to perform Norwood Procedure and creation of a Blalock-Taussig shunt. (Subclavian artery → to pulmonary circulation.)*

# Neonatal Congestive Heart Failure: Pathophysiology

## Pathophysiology of Neonatal CHF:

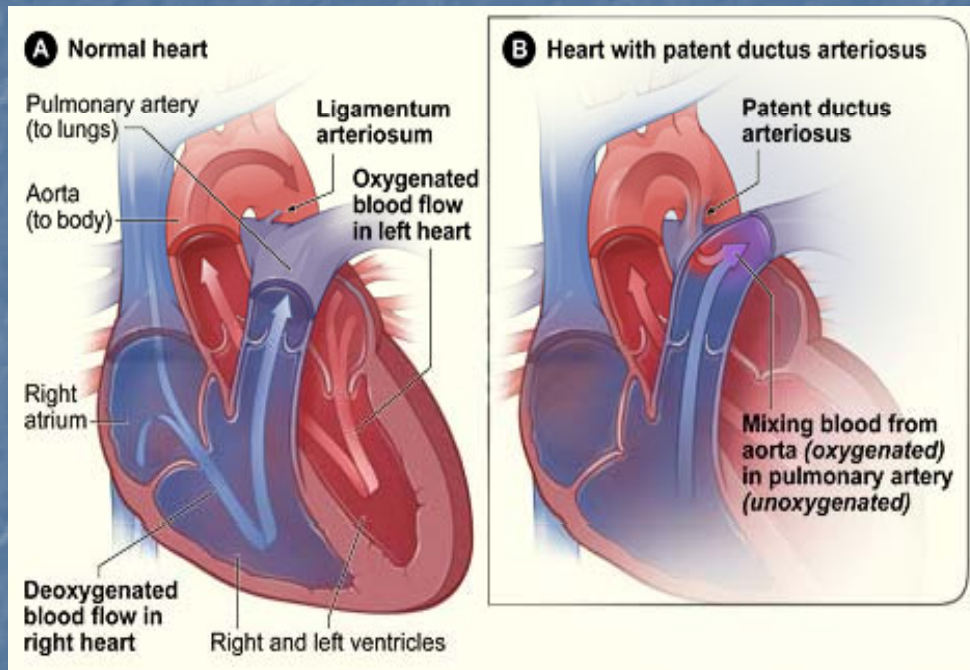
- Most structural heart defects do not cause CHF within hours of birth.
- Instead, myocardial dysfunction secondary to asphyxia, hypoglycemia, hypocalcemia or sepsis are usually responsible for CHF on the first day.
- Tricuspid regurgitation secondary to hypoxia induced papillary muscle dysfunction or Ebstein's anomaly of the valve is also recognised.
- This improves as the pulmonary artery pressure falls over the next few days.



*Fetal/Neonatal heart at birth; prior to PDA closure.*



# Neonatal Congestive Heart Failure: Pathophysiology



## Pathophysiology of Neonatal CHF:

- Serious cardiac disorders which are potentially curable but carry a high mortality if untreated, often present with CHF in the first week of life.
- The closure of the ductus arteriosus is often the precipitating event leading to catastrophic deterioration in a seemingly healthy neonate.
- Prostaglandin E1 (PGE-1), should be utilized in babies with ductal-dependent disorders.



# Neonatal Congestive Heart Failure: Pathophysiology

## Pathophysiology of Neonatal CHF:

- The most common cause of CHF in infants is a ventricular septal defect (VSD) that presents around 6-8 weeks of age.
  - This is because the volume of the left to right shunt increases as the pulmonary resistance falls.
- Although a murmur of VSD is apparent by one week, the full blown picture of CHF occurs around 6-8 weeks.
- Other left to right shunts like PDA present similarly.
- Medical management of CHF is perhaps most important in this age group, since the VSD may close on follow up.
- It is equally important to understand that spontaneous improvement in CHF could result from development of obstructive pulmonary arterial hypertension, even in early childhood.

# Neonatal Congestive Heart Failure: Assessment

## Assessment of Neonatal CHF:

- A fresh scoring system has recently been developed to assess the clinical status of infants with congestive heart failure.
  - Heart failure scores for infants with congestive heart failure.

Symptoms (score)	Frequent (2)	Occasional (1)	None (0)
Breathing difficulty			
Interrupted feeds			
Vomiting			
Sweating			
Poor activity			
Irritability			
Edema			

Maximum score = 14

# Neonatal Congestive Heart Failure: Assessment

## Assessment of Neonatal CHF:

- Modified Ross Heart Failure Classification for Children

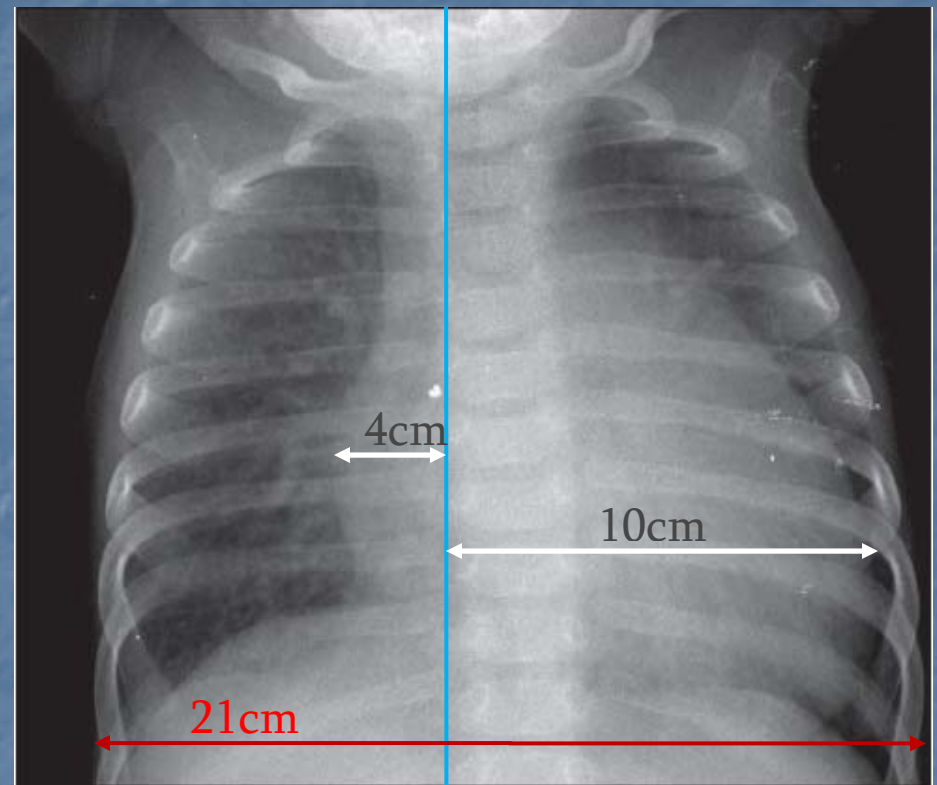
Severity	Symptomatology
Class I	Asymptomatic
Class II	Mild tachypnea or diaphoresis with feeding in infants Dyspnea on exertion in older children
Class III	Marked tachypnea or diaphoresis with feeding in infants Marked dyspnea on exertion Prolonged feeding times with growth failure
Class IV	Tachypnea, retractions, grunting, or diaphoresis at rest



# Neonatal Congestive Heart Failure: Assessment

## Assessment of Neonatal CHF:

- **Abnormal assessment findings:**
  - Heart rates > 220/min indicate supraventricular tachycardia.
  - Heart rates > 300/min with an irregular R-R indicate atrial fibrillation with Wolff-Parkinson-White syndrome.
  - Tachypnea with respiratory rate >60/min in a sleeping neonate is abnormal.
  - Cardiothoracic ratio of > 60% in the newborn and > 55% in older infants with CHF is the rule. However, an expiratory film could often be misinterpreted as showing cardiac enlargement.



$$14/21 = 0.\overline{666} \times 100 = CT \text{ ratio of } 66\%$$

# Neonatal Congestive Heart Failure: Assessment

## Assessment of Neonatal CHF:

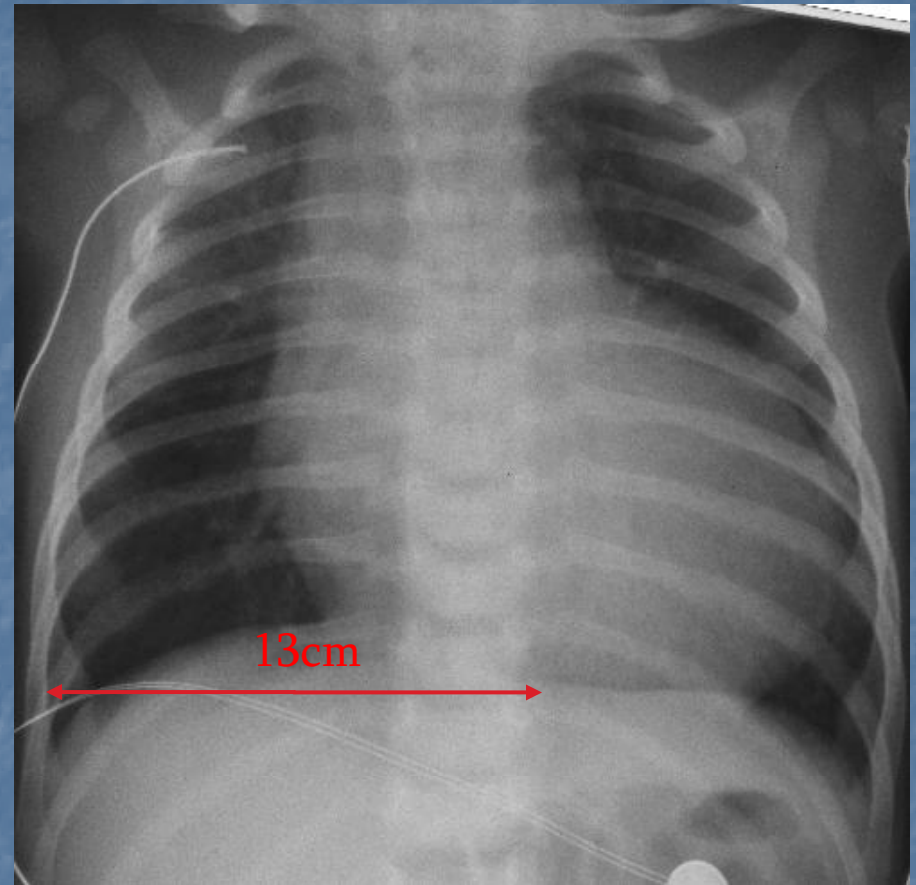
- Abnormal assessment findings:
  - Hepatomegaly
    - Hepatic enlargement regresses quickly in response to therapy and is thus a useful indicator of treatment.
  - Gallop rhythms are the most helpful sign in the diagnosis of CHF.
    - Gallop rhythms are uncommon in neonates and young children.
    - Since S3 and S4 are rarely heard in the neonatal period, their presence denotes a pathologic process.



*S3 gallop*



*S4 gallop*

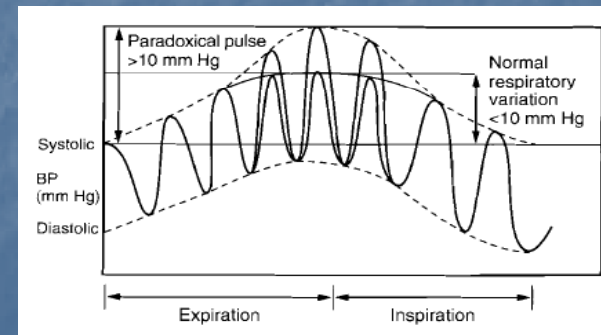
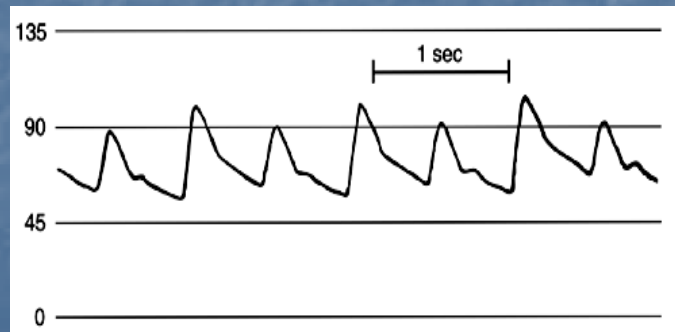


*Hepatomegaly in the neonate.*

# Neonatal Congestive Heart Failure: Assessment

## Assessment of Neonatal CHF:

- **Abnormal assessment findings:**
  - Wheezing may occur with left ventricular failure and may be confused with bronchiolitis, but rales are uncommon and suggest associated pneumonia or a severe CHF.
  - Cold extremity, low blood pressure, skin mottling are signs of impending shock.
- **Pulsus alternans**  
(alternate strong and weak contractions of a failing myocardium)
- **Pulsus paradoxus**  
(decrease in pulse volume and BP with inspiration)





# Neonatal Congestive Heart Failure: Assessment

## Assessment of Neonatal CHF:

- Regarding CHF in this age group a few points need to be emphasized:
  - Peripheral pulses and oxygen saturation (by a pulse oximeter) should be checked in both the RIGHT WRIST and LEFT FOOT in ALL NEONATES AND INFANTS.
    - A lower saturation in the lower limbs means right to left ductal shunting and occurs due to pulmonary hypertension, coarctation of aorta or aortic arch interruption.
  - ASD's/VSD's do not lead to CHF in the first two weeks of life.
    - Therefore, an additional cause must be sought (eg. coarctation of aorta or TAPVC).
  - Premature infants have a poor myocardial reserve and a PDA may result in CHF in the first week.
  - Adrenal insufficiency due to enzyme deficiencies or neonatal thyrotoxicosis could present with CHF in the first few days of life.
  - 4-Extremity BP's should be obtained in all neonates and infants as part of your primary assessment.
    - Anyone who says "you don't need to obtain a BP on an infant or child is WRONG.
    - Would you take a 65-year-old male to the hospital without taking a BP?
    - Would you treat a well-appearing newborn any differently if their BP was 38/18?

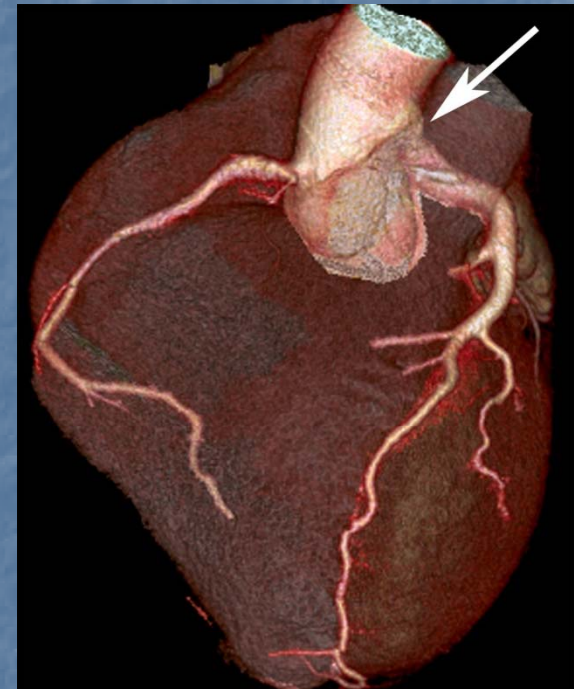
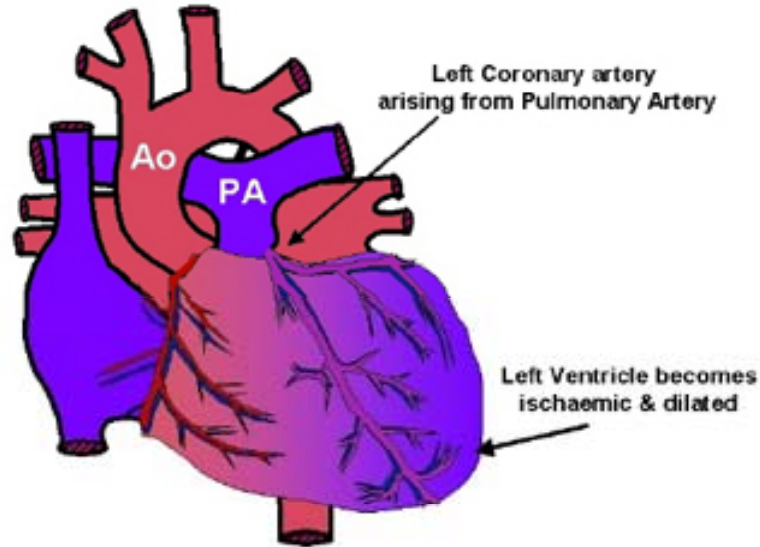
## Neonatal Congestive Heart Failure: ALCAPA

- Anomalous Left Coronary Artery arising from the Pulmonary Artery (ALCAPA):
  - Anomalous left coronary artery arising from the pulmonary artery (ALCAPA), a rare disease in this age group merits separate mention, since it is curable and often missed.
  - As the pulmonary artery pressure decreases in the neonatal period, these babies suffer from episodes of angina and myocardial infarction.
  - Marked distress during feedings, diaphoresis, increased respiratory effort, tachycardia, systolic murmur and crepitations are all common findings in patients with ALCAPA.
  - The 12-Lead EKG shows pathologic Q waves or LVH.
  - These infants are often misdiagnosed as having “dilated cardiomyopathy.”

## Neonatal Congestive Heart Failure: ALCAPA

- Anomalous Left Coronary Artery arising from the Pulmonary Artery (ALCAPA):

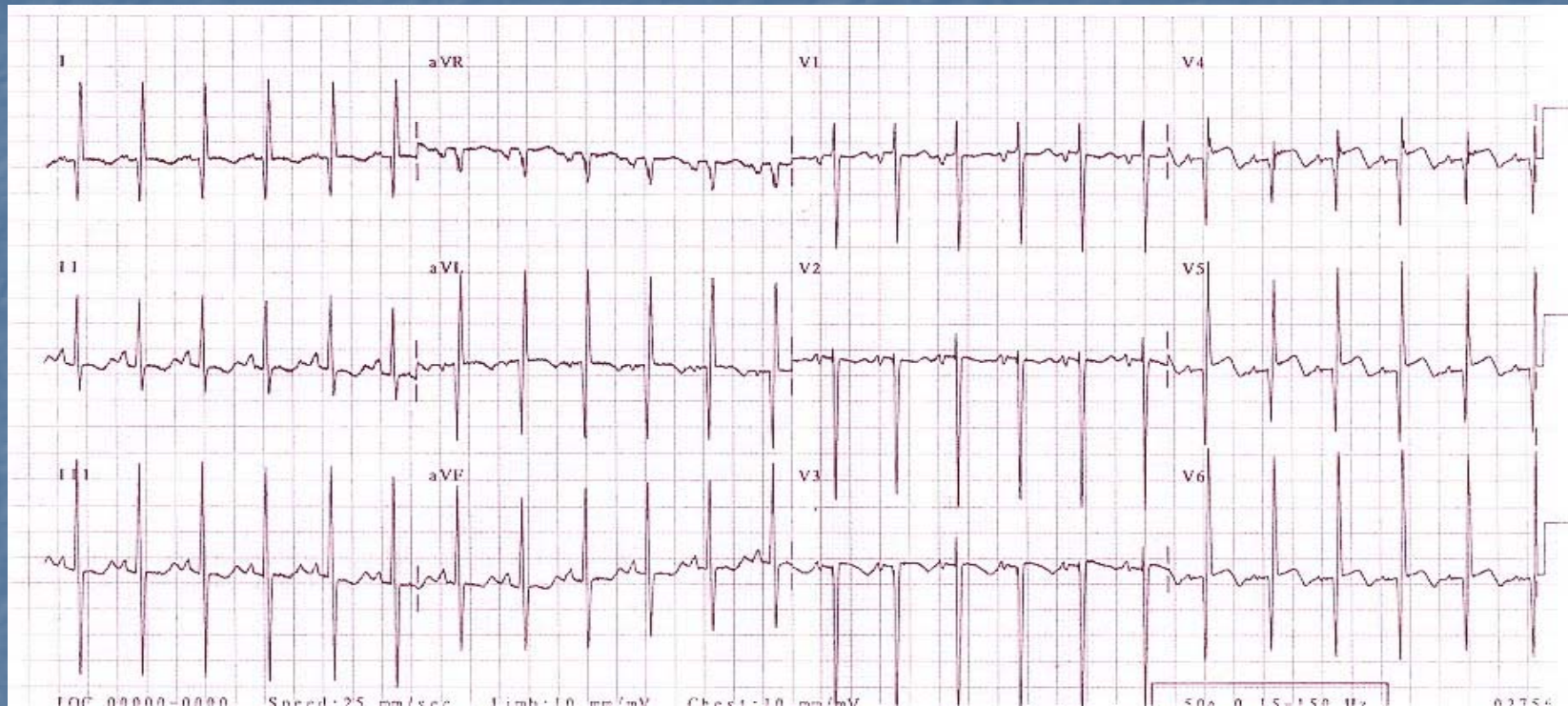
### Anomalous Left Coronary Artery from Pulmonary Artery (ALCAPA)





## Neonatal Congestive Heart Failure: ALCAPA

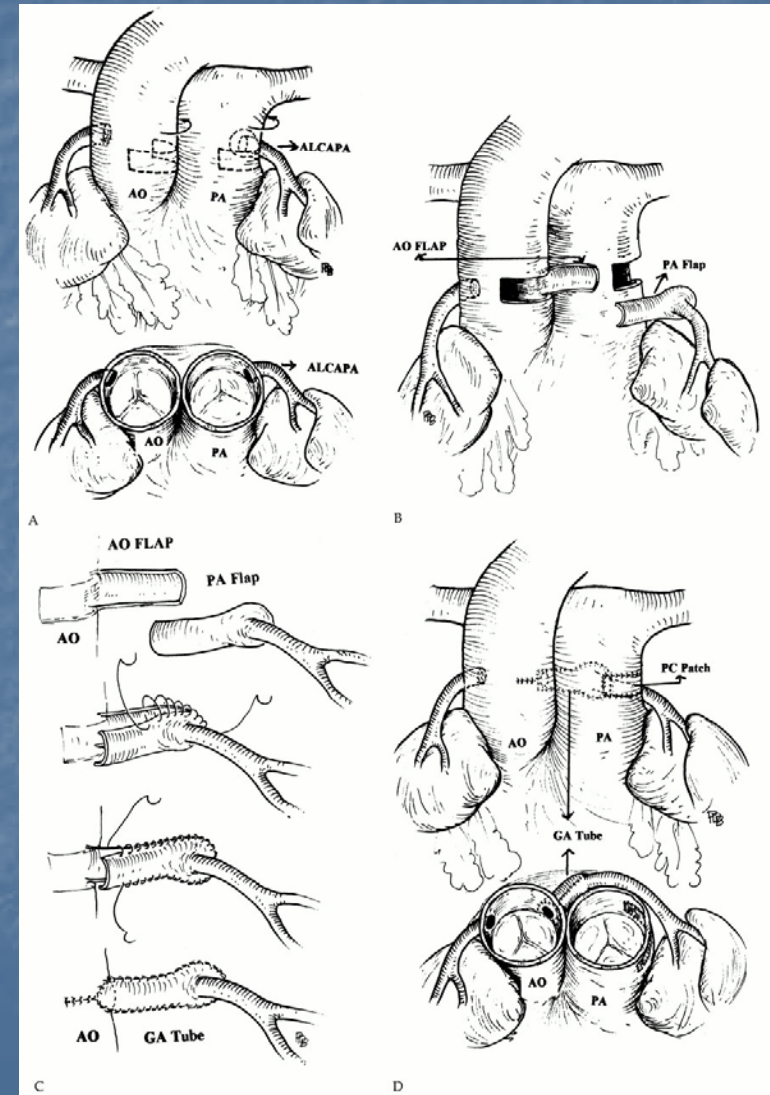
- *12-Lead Electrocardiogram that was obtained from a neonate with ALCAPA.*



- \*\*\*This baby is having an Acute Myocardial Infarction. Treat accordingly.\*\*\*
  - ECG has a anterolateral infarct pattern; deep Q wave in I, aVL and Lead V3-V6.
  - This ECG pattern in newborn is very suggestive of ALCAPA.
  - Q waves deeper than 3 mm with an inverted T wave in aVL is specific to ALCAPA and differentiates this disorder from dilated cardiomyopathy.
  - Irritability and sweating during feeding is a sign of heart failure in the infant, so we should also consider the ischemia on the ECG and the anterolateral infarction; this is a rare diagnosis in someone so young.

# Neonatal Congestive Heart Failure: ALCAPA

- Management of ALCAPA:
  - Surgery is required to fix ALCAPA.
  - There are a number of surgical procedures that can be used, including:
    - Detaching the LCA from the PA and suturing it into the correct position on the AO.
    - Creating a tunnel from the AO to the ALCAPA, and then closing the connection between the LCA and the PA.
    - Removing the faulty LCA and then using saphenous vein, to create a new LCA.
    - Creating a connection between the left SCA and the LCA, so that the very oxygen-rich blood from the SCA feeds the LCA.





# Neonatal Congestive Heart Failure: Management

- Management of Neonatal Congestive Heart Failure:
  - Intravenous Access
    - Establish IV (e.g. 22-24G) for diuresis, vasodilatory/vasopressor agents, pain management, and potential antibiotic therapy.
      - » These patients often have lots of scar tissue at common venipuncture sites, and smaller(than usual) IV catheters may need to be utilized (e.g. 22-24G).
      - » IV sites commonly hands, feet, and/or saphenous veins. Avoid antecubital veins if possible, as these sites may be needed for cardiac catheterization.
      - » If antecubital access is deemed necessary, preferred access would be on the left side unless arrhythmias are present.
  - Intraosseous Access
    - Establish IO (e.g. 15-18G) for those neonates deemed to be *in extremis*.
      - » Commonly used for pediatric patients in cardiac arrest, intraosseous access should be reserved for neonates experiencing life-threatening arrhythmias, requiring immediate vasopressors, having an NSTEMI/STEMI, or are so profoundly unstable that vascular access should not be deferred.



# Neonatal Congestive Heart Failure: Management

- Management of Neonatal Congestive Heart Failure:
  - Continuous Positive Airway Pressure(CPAP)
    - Standard of care for all patients with acute pulmonary edema.
    - Nasal CPAP and/or mask CPAP is commonly used on neonates with pulmonary edema/CHF by neonatal/pediatric transport teams.
    - Perfectly acceptable, and very beneficial prehospital therapy if supplies can be procured and appropriate training is conducted by experienced personnel.
      - » **Settings:**
        - **Pressure: 5 cm/H<sub>2</sub>O.**
        - **FiO<sub>2</sub>: Start at 24%(or lowest setting); titrate up/down to SpO<sub>2</sub> > 95%.**
  - Oxygen
    - Oxygen should be placed on all patients with presumed chest pain or ischemia of cardiac nature.
      - » **Oxygen Flow Rate: 0.5 - 2 L/min via NC.**



# Neonatal Congestive Heart Failure: Management

- Management of Neonatal Congestive Heart Failure:
  - Diuretics (Furosemide/Bumetanide):
    - Diuretics may be helpful if frank pulmonary edema is present.
      - » Furosemide Dosage: 0.5 - 1 mg/kg IV (Greatly preferred in neonates)
      - » Bumetanide Dosage: 0.02 - 0.1 mg/kg IV
  - Aspirin (ASA):
    - Platelet aggregation inhibitor for patients presenting with NSTEMI/STEMI.
      - » Aspirin Dosage: 15 mg/kg PO (rounded to the nearest ¼ tab)
        - (e.g. ½ - 81mg ASA tab, ¾ 81mg ASA tab, etc...)
  - Opiates (Morphine/Hydromorphone/Fentanyl):
    - Recommended treatment for neonates with pain or ischemia of cardiac nature, and/or for vasodilation in NSTEMI/STEMI/CHF.
      - » Morphine Dosage: 0.05 - 0.15 mg/kg IV (\*\*Preferred drug of choice)
      - » Hydromorphone Dosage: 0.015 - 0.02 mg/kg IV
      - » Fentanyl Dosage: 0.5 - 1 mcg/kg IV
        - (\*\*SLOW PUSH; otherwise may cause chest wall rigidity. \*\*)

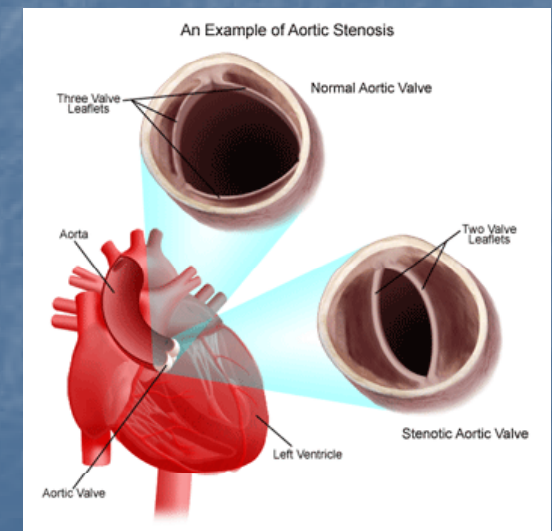
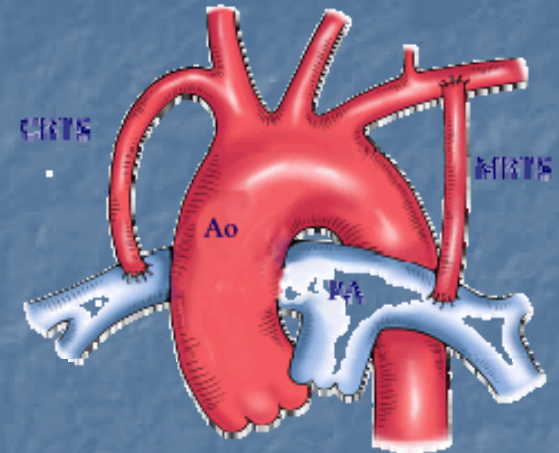
# Neonatal Congestive Heart Failure: Management

- Management of Neonatal Congestive Heart Failure:
  - Dopamine Drip
    - Particularly useful if cardiogenic shock is present in the setting of CHF.
      - » Dopamine Dosage: 5-20 mcg/kg/min IV
        - Calculation:  $6 \times (\text{wt in kg}) = \text{mg Dopamine to add to 100 cc bag of NS.}$
        - \*\*\*At this concentration, 1 cc/hr = 1 mcg/kg/min.
  - Dobutamine Drip
    - Particularly useful if cardiogenic shock is present in the setting of CHF.
      - » Dobutamine Dosage: 5-20 mcg/kg/min IV
        - Calculation:  $6 \times (\text{wt in kg}) = \text{mg Dopamine to add to 100 cc bag of NS.}$
        - \*\*\*At this concentration, 1 cc/hr = 1 mcg/kg/min.
  - Epinephrine Drip
    - Indicated for cardiogenic shock in the setting of CHF, if refractory to dopamine or dobutamine.
      - » Epinephrine Drip Dosage: 0.1-1.0 mcg/kg/min IV
        - Calculation:  $0.6 \times (\text{wt in kg}) = \text{mg Dopamine to add to 100 cc bag of NS.}$
        - \*\*\*At this concentration, 1 cc/hr = 0.1 mcg/kg/min.



# Neonatal Congestive Heart Failure: CHF Beyond Infancy

- CHF Beyond Infancy:
  - Onset of CHF beyond infancy is unusual in patients with congenital heart disease and suggests a complicating factor like valvular regurgitation, infective endocarditis, myocarditis, or anemia.
  - Complicating factors must be treated aggressively and without delay.
  - In a surgically palliated patient(e.g. after a Blalock-Taussig shunt), continued volume or pressure overload may be the primary cause of the new-onset or worsening CHF.
  - Uncommonly, rapid disease progression such as worsening of aortic or pulmonary stenosis may cause CHF in childhood.
  - Acquired diseases such as respiratory illnesses, septicemia, CMV, EBV, and/or HSV are common causes of CHF in children.



# Neonatal Congestive Heart Failure: Long Term Expectations

- Long-term prognosis of neonatal patients:
  - Treatment of acute and reversible causes hold the greatest prognosis for these patients.
  - Babies with defects amenable to surgical repair, typically experience a good quality of life.
  - Some will have to remain on medicine, but many will be able to stop taking medicines if a surgical correction can be made.
    - In this case, the occurrence of CHF is often greatly reduced or eliminated.
  - Numerous appointments with a cardiologist will be required after the surgery, to make sure the child's heart is working properly.
  - As the child grows, a cardiologist will continue to monitor the heart; typically every few months.



# Neonatal Congestive Heart Failure: Long Term Expectations

- Long-term prognosis of neonatal patients:
  - Many children who were born with congenital heart diseases do very well and don't have any restrictions to school, activity or sports.
  - However, children who were born with CHD, and/or those that have undergone surgical palliation will require life-long care by a cardiologist.
  - Because they are at higher risk for abnormal heart rhythms and because they were born with a heart defect, annual (or more frequent) visits with a cardiologist will be required.
  - Once children are older than 18 years, they will transfer to an adult cardiologist.





# Pediatric Congestive Heart Failure



# Pediatric Congestive Heart Failure: Overview

## Incidence of Pediatric CHF:

- Incidence of heart failure caused by congenital heart disease and cardiomyopathy affects 12,000 – 35,000 children below age 19 in the United States each year.

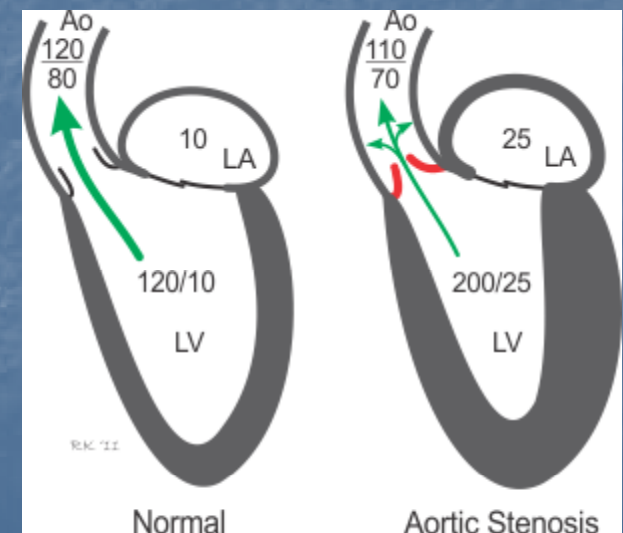
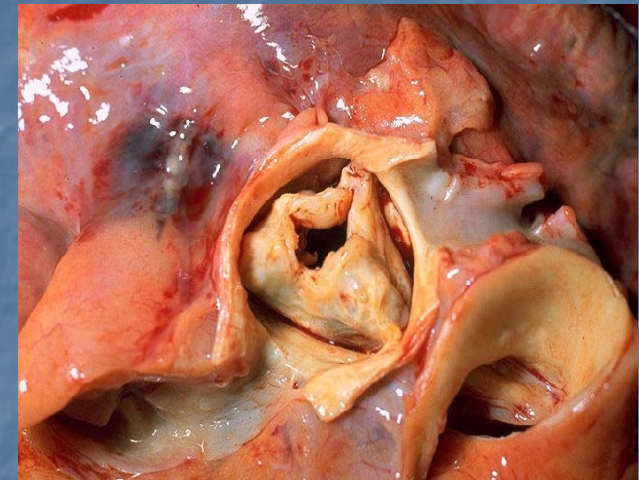
## Causes of Pediatric CHF:

- Left-sided obstructive disease
  - Valvular /subvalvular aortic stenosis
  - Coarctation of the aorta
- Myocardial dysfunction
  - Myocarditis
  - Cardiomyopathy
- Hypertension
- Renal failure
- Arrhythmogenic or ischemic etiology

# Pediatric Congestive Heart Failure: Pathophysiology

## Pathophysiology of Pediatric CHF:

- Left-sided obstructive disease
  - Aortic stenosis
    - Resistance to systolic ejection occurs and a systolic pressure gradient develops between LV and AO.
    - Increased LVEDP causes decreased CO 2° diastolic dysfunction.
    - Impaired contractility → decreased CO 2° systolic dysfunction, resulting in heart failure.
    - Diastolic filling of the left ventricle depends on effective atrial contraction. AF → HF 2° loss of CO.
    - Hypertrophic myocardium → increased oxygen requirement with diminished oxygen delivery 2° diminished coronary flow, decreased diastolic perfusion, and myocardial ischemia.

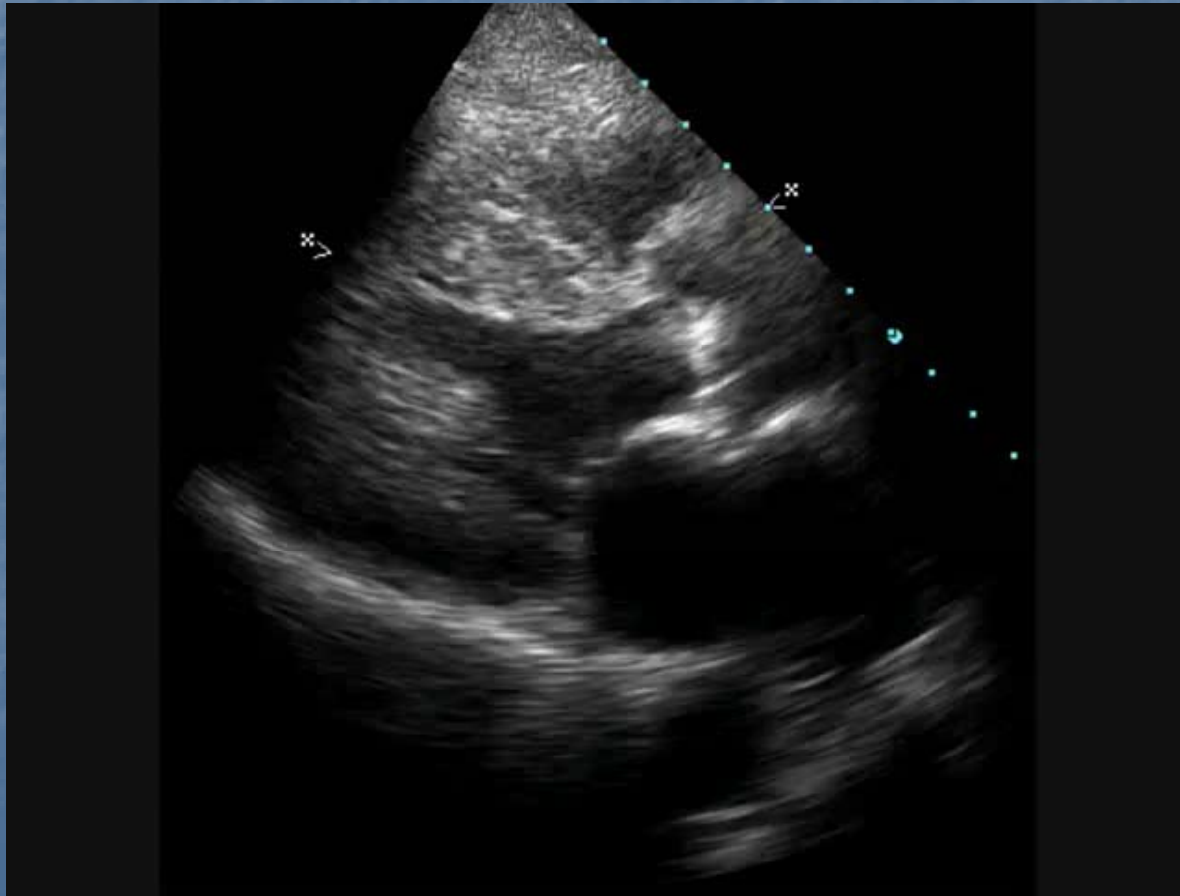




# Pediatric Congestive Heart Failure: Pathophysiology

## Pathophysiology of Pediatric CHF:

- Left-sided obstructive disease
  - Aortic stenosis (Echocardiographic profile)



# Pediatric Congestive Heart Failure: Pathophysiology

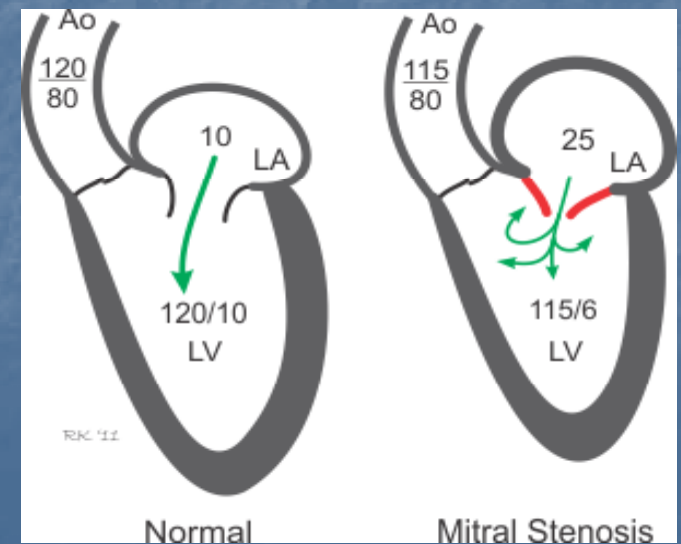
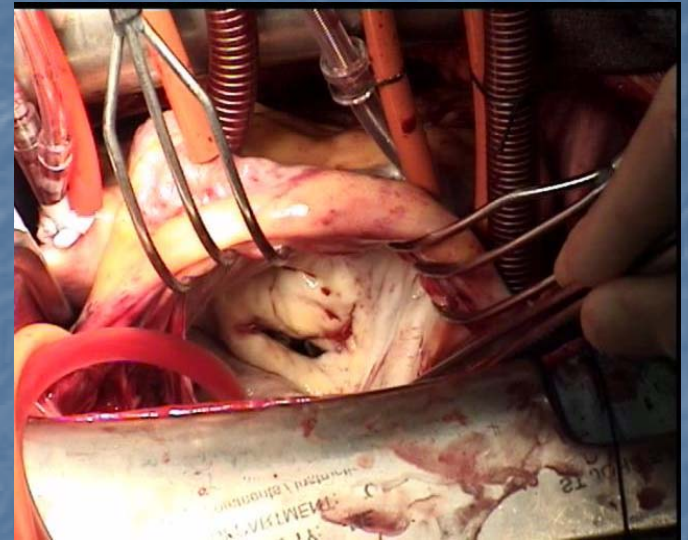
## Pathophysiology of Pediatric CHF:

- Left-sided obstructive disease
  - Sub-aortic stenosis
    - Subvalvular stenosis occurs in about 10% of aortic stenosis, is more common in males and can be caused by either a discrete membrane or a diffuse fibrous ring in the LVOT.
    - The obstructions can create turbulence thickening the aortic valve leaflets and predisposing to endocarditis.
    - Varying severities of obstruction occur.
    - Degrees of genetic predisposition are unclear but a pedigree of multiple family members with discrete subaortic membrane has been reported.

# Pediatric Congestive Heart Failure: Pathophysiology

## Pathophysiology of Pediatric CHF:

- Left-sided obstructive disease
  - Mitral stenosis
    - Obstruction to left ventricular inflow due to structural abnormality of the mitral valve, which prevents proper filling of the left ventricle during diastole.
    - Patients are generally asymptomatic at rest during the early stage of the disease.
    - Acute insult leads to formation of multiple inflammatory foci in the endocardium and myocardium.
    - With time, the valve apparatus becomes thickened, calcified, and contracted, and adhesion occurs.
    - Stenosis typically occurs decades after the episode of acute rheumatic carditis.
    - **The most common cause of mitral stenosis is rheumatic fever.**

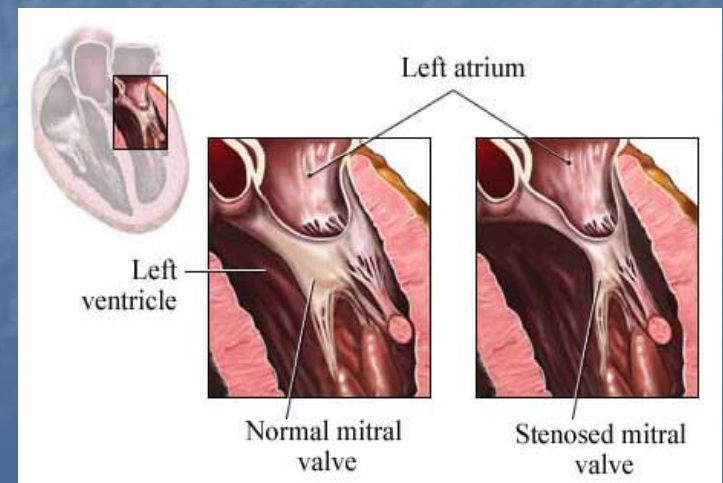
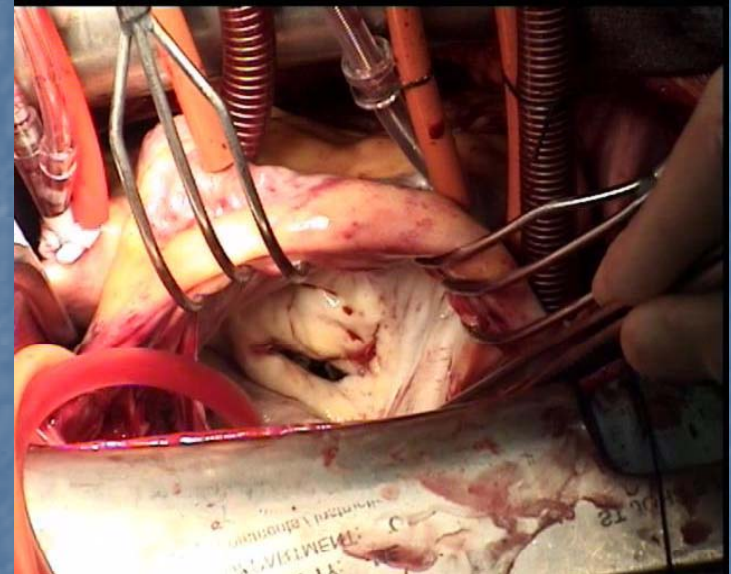




# Pediatric Congestive Heart Failure: Pathophysiology

## Pathophysiology of Pediatric CHF:

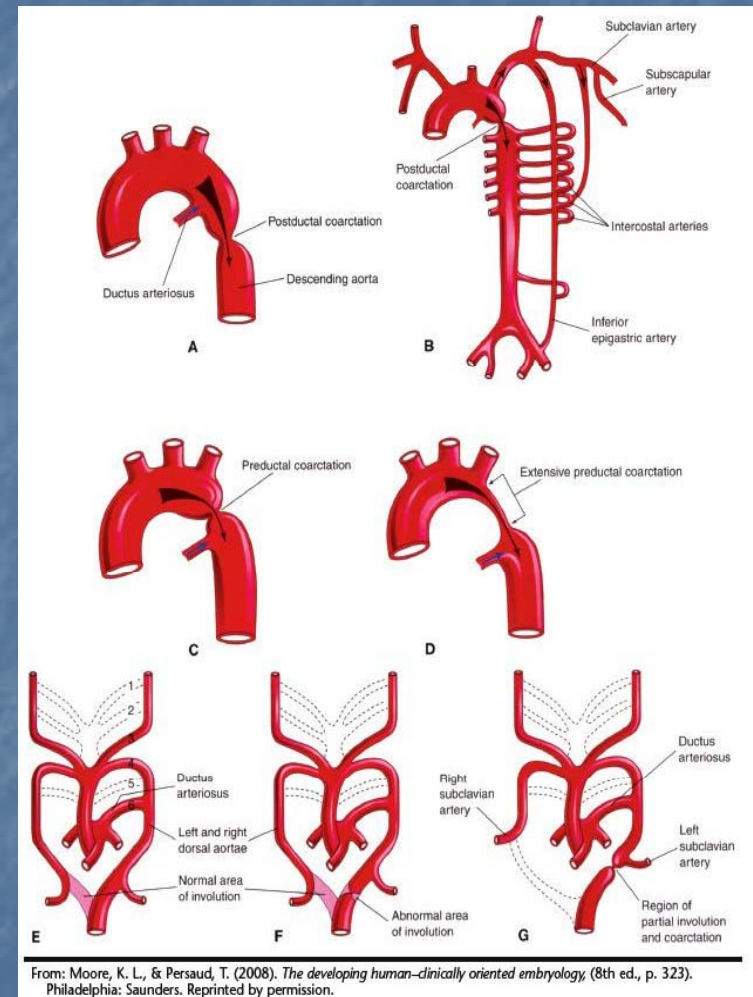
- Left-sided obstructive disease
  - Mitral stenosis
    - 15% of patients develop embolic episodes usually associated with atrial fibrillation.
    - Embolic episodes may occur even in the patient with sinus rhythm.
    - Pregnant women with mild mitral stenosis may become symptomatic during their second trimester because of the increase in blood volume and cardiac output.
    - Hoarseness can develop from compression of the left recurrent laryngeal nerve against the pulmonary artery by the enlarged left atrium.



# Pediatric Congestive Heart Failure: Pathophysiology

## Pathophysiology of Pediatric CHF:

- Left-sided obstructive disease
  - Coarctation of the aorta
    - Congenital narrowing of the aorta, usually distal to the origin of the left subclavian artery, opposite the area of the ductus arteriosus at the aortic isthmus.
    - Relatively common defect accounting for 5-8% of all CHD's.
      - May occur in association with various other lesions, most commonly bicuspid aortic valve and VSD.
    - Presentation may vary from acute decompensation at the time of ductal closure in the infant, to upper extremity hypertension in the older child or adult.
    - Symptoms may be subtle initially, and patients may make repeated trips to the physician before finally presenting in extremis.

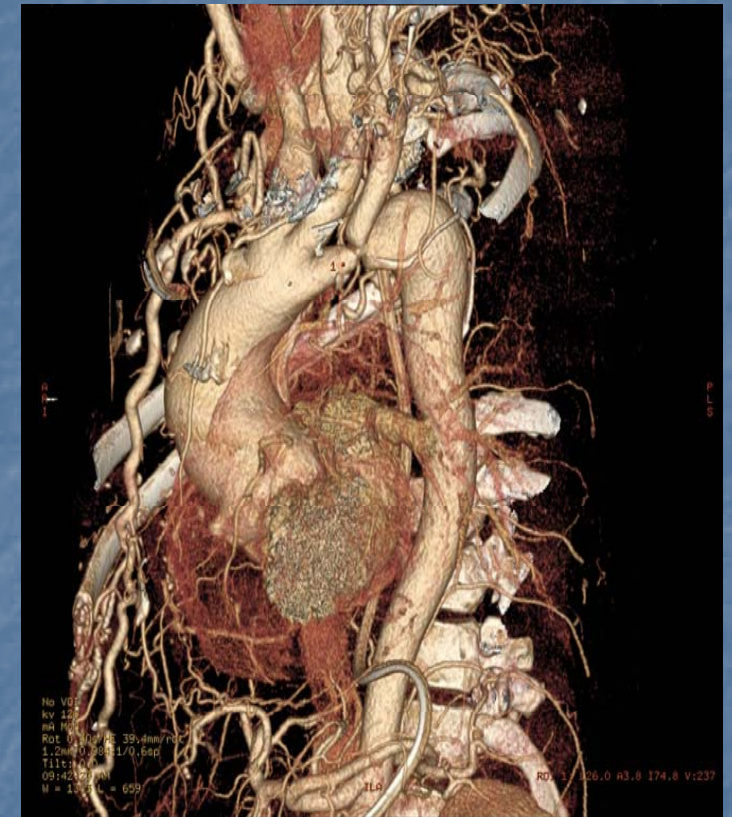




# Pediatric Congestive Heart Failure: Pathophysiology

## Pathophysiology of Pediatric CHF:

- Left-sided obstructive disease
  - Coarctation of the aorta
    - In adolescents, coarctation of the aorta is best diagnosed clinically based on simultaneous palpation of femoral and brachial pulses.
    - These patients often have not developed overt CHF because of the presence of arterial collateral vessels.
    - Four-extremity blood pressures must be obtained. A pressure gradient of more than 20 mmHg in favor of the arms may be considered evidence of CoA.
      - e.g. RA:126/84, LA:130/88,  
RL: 92/68, LL: 94/66
    - CoA may occur as an isolated defect or in association with various other lesions, most commonly bicuspid aortic valve and VSD.
    - CoA is the most common congenital heart defect in patients with Turner Syndrome (45,XO) with as many as 20% affected.





# Pediatric Congestive Heart Failure: Pathophysiology

## Pathophysiology of Pediatric CHF:

- Myocardial dysfunction
  - Pediatric myocarditis
    - Viral myocarditis
      - » Coxsackievirus types A and B (especially type B)
      - » Adenovirus (most commonly types 2 and 5)
      - » Cytomegalovirus (CMV) [\*\*\*EXTREME RISK to pregnant women\*\*\*]
      - » Echovirus
      - » Epstein-Barr virus (EBV)
      - » Hepatitis C virus (HCV)
      - » Herpes virus (HSV-1/HSV-2)
      - » Human immunodeficiency virus (HIV)
      - » Influenza and parainfluenza
      - » Measles
      - » Mumps, associated with endocardial fibroelastosis (EFE)
      - » Parvovirus B19
      - » Poliomyelitis virus
      - » Rubella
      - » Varicella

# Pediatric Congestive Heart Failure: Pathophysiology

## Pathophysiology of Pediatric CHF:

- Myocardial dysfunction
  - Pediatric myocarditis
    - Viral myocarditis
      - » Complications of myocarditis may include the following:
        - Arrhythmias (specifically AV conduction disturbances)
        - Congestive heart failure
        - Thromboembolism
        - Further decrease in ventricular function
        - Dilated cardiomyopathy
    - Nonviral myocarditis
      - » Protozoal infections (Chagas' disease)
      - » Giant cell myocarditis
      - » Systemic lupus erythematosus (SLE)
      - » Kawasaki disease
      - » Dermatomyositis
      - » Sarcoidosis
      - » Scleroderma
      - » Drugs (Cyclophosphamide, phenylbutazone, acetazolamide, amphotericin B, indomethacin, tetracycline, isoniazid, methyldopa, phenytoin, penicillin, and sulfonamides.)

# Pediatric Congestive Heart Failure: Pathophysiology

## Pathophysiology of Pediatric CHF:

- Myocardial dysfunction
  - Pediatric myocarditis
    - Nonviral myocarditis
      - » The general principles of therapy for CHF are applicable to patients with myocarditis.
      - » These principles include the manipulation of preload, afterload, and contractility.
      - » Important potential interventions for CHF caused by myocarditis of a nonviral origin:
        - Fluid restriction (strict I/O's, cognizance of volumes infused)
        - Diuretics (furosemide, bumetanide)
        - Continuous intravenous (IV) inotropic agents (dopamine, dobutamine)
        - IV vasodilator agents important potential interventions for CHF caused by myocarditis of a nonviral origin.



# Pediatric Congestive Heart Failure: Pathophysiology

## Pathophysiology of Pediatric CHF:

- Myocardial dysfunction
  - Cardiomyopathy
    - Hypertrophic cardiomyopathy (HCM)
      - » Most common pediatric cardiomyopathy.
      - » Autosomal dominant trait attributed to mutations in one of a number of genes that encode for one of the sarcomere proteins.
      - » An insertion/deletion polymorphism in the gene encoding for ACE alters the clinical phenotype of the disease.
      - » The D/D genotype of ACE is associated with more marked hypertrophy of the left ventricle and may be associated with higher risk of adverse outcomes.
    - Dilated cardiomyopathy (DCM)
    - Arrhythmogenic right ventricular dysplasia (ARVD)
      - Epsilon waves - most common finding on 12-lead EKG.



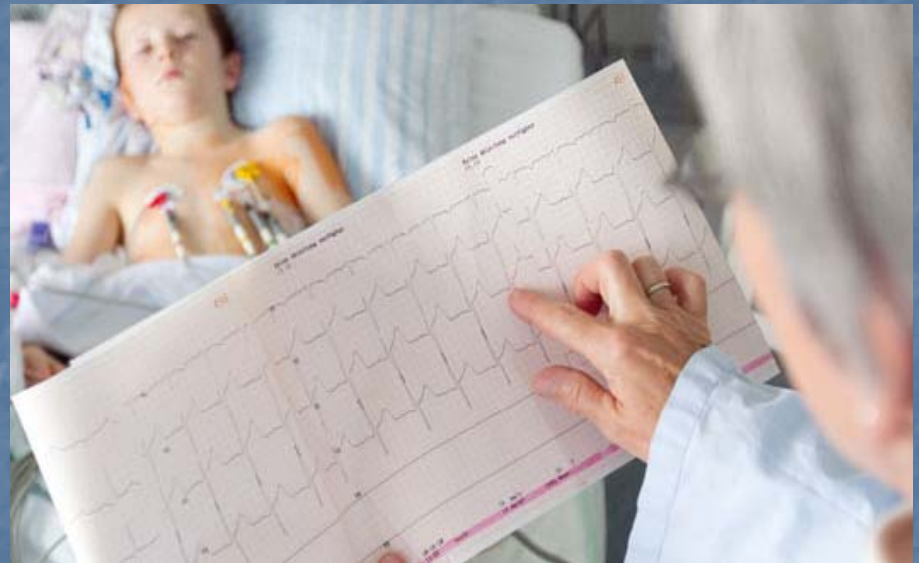
*Epsilon waves as indicated by red arrows in lead V3.*

## Pediatric Congestive Heart Failure: Assessment

- **Assessment of Pediatric Congestive Heart Failure:**
  - Regardless of etiology, the first manifestation of CHF is usually tachycardia.
    - An obvious exception is CHF 2° a primary bradyarrhythmia or complete heart block.
  - Patients can present with any type of dysrhythmia, including AV conduction disturbances. \*\*\*Perform a 12-lead EKG!\*\*\*
    - Sinus tachycardia is typical, and the rate is faster than expected for the degree of fever present, which is typically low-grade.
    - Junctional tachycardia is also seen and can be difficult to control medically.
  - Chest pain may be the initial presentation for older children.
    - May be due to myocardial ischemia or concurrent pericarditis.
      - » Signs of left-sided congestion: Tachypnea, respiratory distress, and rales/wheezing.
      - » Signs of right-sided congestion: Hepatosplenomegaly, JVD, edema, ascites, pleural effusions.
  - Older children with uncompensated CHF may have “lower than usual” energy levels.

# Pediatric Congestive Heart Failure: Assessment

- Assessment of Pediatric Congestive Heart Failure:
  - Clinical manifestations of poor cardiac output:
    - Cool extremities
    - Diaphoresis
    - Pallor
    - Abdominal pain
    - Nausea/vomiting
    - Exercise intolerance
    - Dizziness/syncope
    - Poor growth
    - AMS/ALOC
  - Growth parameters
    - » \*\*\*Poor weight gain is a key indication of poorly compensated heart failure.\*\*\*





# Pediatric Congestive Heart Failure: Assessment

- Assessment of Pediatric Congestive Heart Failure:
  - Abnormal assessment findings:
    - General appearance:
      - » “Ill-appearing child”
      - » Perspiration, cyanosis
    - Vital signs:
      - » Tachycardia (>120 beats per minute in the child)
      - » Tachypnea (>30 breaths per minute in the school age child)
      - » Blood pressure. Do 4 limb blood pressures if aortic coarctation is suspected
      - » Oxygen saturation may or may not be abnormal in children with congenital heart diseases.
      - » Listen for S1, S2. Abnormal S1/S2 may be a clue to valvular disease.
      - » A loud P2 is in strong indication of pulmonary overload.
    - Palpate for thrills, right and left sided heaves; listen for gallop rhythms (S3, S4) and murmurs
    - Respiratory Exam:
      - » Increased work of breathing, tachypnea, indrawing, tracheal tugging
      - » Auscultation; listening for signs of pulmonary edema

# Pediatric Congestive Heart Failure: Management

- Management of Pediatric Congestive Heart Failure:
  - Intravenous Access
    - Establish medium-bore IV (e.g. 20-22G) for diuresis, vasodilatory/vasopressor agents, pain management, and potential antibiotic therapy.
      - » IV sites are essentially the same as in adults.
        - Hands, wrists, forearms, saphenous veins, EJ if peripheral collapse or in extremis.
        - Avoid antecubital veins if possible, as these sites may be needed for cardiac catheterization.
        - If antecubital access is deemed necessary, preferred access would be on the left side unless arrhythmias are present.
  - Intraosseous Access
    - Establish IO (e.g. 15G EZ-IO) for those deemed to be in extremis.
      - » Particularly in situations where at least 1 peripheral attempt has failed, and jugular stick was unsuccessful.
      - » Commonly used for pediatric patients in cardiac arrest, intraosseous access should be reserved for patients experiencing life-threatening arrhythmias, requiring immediate vasopressors, having an ACTIVE STEMI, or are so profoundly unstable that vascular access should not be deferred.

# Pediatric Congestive Heart Failure: Management

- Management of Pediatric Congestive Heart Failure:
  - Oxygen
    - Oxygen should be placed on all pediatric patients with cardiac complaints.
      - » Dosage: 1-4 L/min via NC.
        - (Modify based on hx of lung disease, etc...)
  - Continuous Positive Airway Pressure(CPAP)
    - Standard of care for all patients with acute pulmonary edema.
      - » CPAP Settings:
        - Pressure: 5-15 cm/H<sub>2</sub>O; titrated in stepwise fashion.
        - FiO<sub>2</sub>: Start at 24%(or lowest setting); titrate up/down to SpO<sub>2</sub> > 95%.
  - Aspirin (ASA):
    - Platelet aggregation inhibitor for patients presenting with NSTEMI/STEMI.
      - » Aspirin Dosage: 15 mg/kg PO for NSTEMI/STEMI (rounded to the nearest ¼ tab)
        - (e.g. ½ - 81mg ASA tab, ¾ 81mg ASA tab, etc...)





# Pediatric Congestive Heart Failure: Management

- Management of Pediatric Congestive Heart Failure:
  - Diuretics (Furosemide/Bumetanide):
    - Diuretics may be helpful if frank pulmonary edema is present.
      - » Furosemide Dosage: 0.5 - 1 mg/kg IV
      - » Bumetanide Dosage: 0.02 - 0.1 mg/kg IV
  - Nitrates (Nitroglycerin):
    - Particularly useful if evidence of myocardial ischemia is present.
    - If myocardial ischemia without ST segment elevation is present, the patient should still receive nitroglycerin.
      - » Dosage: For > 5 years old: 0.2 mg SL q5min  
For > 8 years old: 0.4 mg SL q5min
  - Opiates (Morphine/Hydromorphone/Fentanyl):
    - Narcotics may be helpful if relief cannot be obtained with nitroglycerin.
      - » Morphine Dosage: 0.1 mg/kg IV
      - » Hydromorphone Dosage: 0.015 – 0.02 mg/kg IV
      - » Fentanyl Dosage: 0.5 – 1 mcg/kg IV
        - (\*\*SLOW PUSH; otherwise may cause chest wall rigidity.\*\*)

# Pediatric Congestive Heart Failure: Management

- Management of Pediatric Congestive Heart Failure:
  - ACE Inhibitors (Captopril/Enalapril/Enalaprilat):
    - Treatment of choice for HF/CHF with HTN emergency 2<sup>o</sup> presumed nephropathy.
      - » Captopril Dosage: 6.25 – 12.5 mg SL/PO (0.3 – 0.5 mg/kg/dose)
      - » Enalapril Dosage: 0.1 mg/kg PO
      - » Enalaprilat Dosage: 0.01 mg/kg IV
  - Bronchodilators (Albuterol/Ipratropium):
    - Consider trial of bronchodilators, especially if patient has pulmonary history or wheezing on exam, and findings do not appear to be of cardiac origin.
      - » Albuterol Dosage: 2.5 - 5 mg/dose via HHN
      - » Ipratropium Dosage: 0.5 mg via HHN x 1 dose

# Pediatric Congestive Heart Failure: Management

- Management of Pediatric Congestive Heart Failure:
  - Dopamine Drip
    - Particularly useful if cardiogenic shock is present in the setting of CHF.
      - » Dosage: 5-20 mcg/kg/min IV
        - Calculation:  $6 \times (\text{wt in kg}) = \text{mg Dopamine to add to 100 cc bag of NS.}$
        - \*\*\*At this concentration, 1 cc/hr = 1 mcg/kg/min.
  - Dobutamine Drip
    - Particularly useful if cardiogenic shock is present in the setting of CHF.
      - » Dosage: 5-20 mcg/kg/min IV
        - Calculation:  $6 \times (\text{wt in kg}) = \text{mg Dopamine to add to 100 cc bag of NS.}$
        - \*\*\*At this concentration, 1 cc/hr = 1 mcg/kg/min.
  - Epinephrine Drip (IF REFRACTORY TO DOPAMINE OR DOBUTAMINE)
    - Indicated for SEVERE cardiogenic shock in the setting of CHF.
      - » Dosage: 0.1-1.0 mcg/kg/min IV
        - Calculation:  $0.6 \times (\text{wt in kg}) = \text{mg Dopamine to add to 100 cc bag of NS.}$
        - \*\*\*At this concentration, 1 cc/hr = 0.1 mcg/kg/min.



# Congestive Heart Failure in the Adolescent



# CHF in the Adolescent: Overview

## Incidence of CHF in the Adolescent:

- Incidence of heart failure caused by congenital heart disease and cardiomyopathy affects 12,000 – 35,000 children below age 19 in the United States each year.

## Causes of CHF in the Adolescent:

- Congestive heart failure in the adolescent can have numerous etiologies:
  - Viral diseases
    - Coxsackie B virus
    - Rubella
    - HSV
    - Influenza
  - Muscular diseases
    - Duchenne muscular dystrophy
    - Becker muscular dystrophy
    - Friedreich's ataxia
  - Leukemia
    - Secondary to administration of Gleevec(imatinib)



# CHF in the Adolescent: Overview

## Causes of CHF in the Adolescent:

- Congestive heart failure in the adolescent can have numerous etiologies:
  - Sickle Cell Anemia
    - Cardiomegaly (common radiological finding in SCD patients)
    - Fluid overload during crisis
    - LA dysfunction
  - Cocaine
    - Cocaine-induced cardiomyopathies/tachyarrhythmias
  - Cystic Fibrosis
    - RV enlargement secondary to lung disease via hypoxic vasoconstriction and pulmonary vascular remodeling.
    - Extremely poor prognosis once RV failure is evident, particularly if not treated early.



# CHF in the Adolescent: Overview

## Causes of CHF in the Adolescent:

- Congestive heart failure in the adolescent can have numerous etiologies:
  - Chronic arrhythmias
    - Supraventricular tachycardia (SVT)
    - Ventricular tachycardia (VT)
  - Cardiomyopathies
    - Tachycardia-induced cardiomyopathy (TIC)
    - Hypertrophic cardiomyopathy (HCM)
    - Dilated cardiomyopathy (DCM)
    - Restrictive cardiomyopathy (RCM)
    - Arrhythmogenic right ventricular cardiomyopathy (ARVC)

# CHF in the Adolescent: Overview

## Overview of CHF in the Adolescent:

- Adolescents have signs and symptoms of heart failure that are similar to those of adults.
  - Dyspnea that is exaggerated by exercise
  - Chronic cough secondary to pulmonary congestion
  - Warm/dry + wheezing = Asthma. Cold/diaphoretic + wheezing = CHF.
- Symptoms of CHF in adolescents might be subtle, but often an illness will be enough to exacerbate underlying hemodynamic abnormalities and allow CHF to become apparent.
- Fatigue and weakness are late findings.
  - Adolescents with mild to moderate HF may not appear in distress.
  - Those with more severe HF may demonstrate dyspnea and tachypnea at rest.
- \*\*\*An adolescent who cannot speak in full sentences, is on the verge of cardiorespiratory failure and should be aggressively treated immediately.

# CHF in the Adolescent: Overview

## Assessment of CHF in the Adolescent:

- It is not as common for CHF to present in adolescence, compared to younger ages.
- A murmur may be detected in adolescence because it is louder, or it may actually be new.
  - Additionally, findings of an ASD, pulmonary flow murmur and widely split S2 may be easier to detect in adolescence.
- A bicuspid aortic valve may develop a leak; presenting as a new early diastolic murmur. or become stenotic, and cause a systolic ejection murmur presenting at the right upper sternal border, with or without a click.
- Hypertrophic cardiomyopathy may develop in an adolescent, and initial presentation may be syncope or cardiac arrest. ARVD/HCM should be suspected in cases of cardiac arrest.
- Symptoms of CHF in adolescents might be subtle, but often an illness will be enough to exacerbate underlying hemodynamic abnormalities and allow CHF to become apparent.
- CHF is usually from sudden ventricular dysfunction or arrhythmias.
  - Bradycardia and/or syncope can be a sign of third degree heart block.



## CHF in the Adolescent: Pathophysiology of SCD-Related HF

- Pathophysiology of Sickle Cell Disease-Related Heart Failure:
  - CHF primarily results from high output secondary to anemia.
  - Chronic anaemia increases cardiac output and may cause left ventricular enlargement and cardiac insufficiency.
  - Older patients and those who receive frequent transfusions may develop cardiac disease which may lead to heart failure.
  - Chronic severe anemia and hypoxemia impose sustained demands on the heart.
  - At the same time, sickle cell patients are in a continuous state of hyperdynamic circulation and therefore, frequently exhibit systolic murmurs.
  - \*\*\* A 12-lead EKG should be performed on ALL SCD patients complaining of chest pain and/or shortness of breath. \*\*\*
    - Myocardial perfusion abnormalities have been discovered in children with SCD, and vaso-occlusive crises have been found to cause myocardial damage in children.

## CHF in the Adolescent: Assessment of SCD-Related HF

- **Assessment of Sickle Cell Disease-Related Heart Failure:**
  - Abnormal assessment findings (typically 2<sup>o</sup> the anemia)
    - Dyspnea on exertion, weakness
    - Chest pain, palpitations → \*\*\*Perform a 12-lead EKG!\*\*\*
    - Myocardial perfusion abnormalities have been discovered in children with SCD, and vaso-occlusive crises have been found to cause myocardial damage in children.
    - Murmurs, third heart sounds, and enlarged heart
  - Ask the patient (or parent) if they still have their spleen, or if they have undergone a splenectomy.
    - The hemolytic process in SCD is accelerated by the spleen.
    - The spleen is usually infarcted by the end of childhood in SCD patients.
  - Splenic sequestration crises:
    - Acute, painful enlargements of the spleen, caused by sudden pooling of the blood into the spleen 2<sup>o</sup> circulatory defect leading to sudden hypovolaemia.
    - Splenic sequestration crises are considered an emergency. If not treated, patients may die within 1–2 hours due to circulatory failure.
    - Management is supportive, sometimes with blood transfusions.
    - These crises are transient, they continue for 3–4 hours and may last for one day.

## CHF in the Adolescent: Management of SCD-Related HF

- Management of Sickle Cell Disease-Related Heart Failure:
  - The treatment of choice for CHF in SCD patients is transfusion, not diuretics.
  - Incidence of nephropathy increases during adolescence; leading to HTN.
  - Treatment guidelines for ACE inhibitor use in SCD patients are needed to guide timing of treatment or dosing, and involvement of a nephrologist with experience in treating SCD nephropathy is recommended.
- Oxygen
  - Not routinely recommended for SCD patients unless there is a specific indication (e.g. ACS, pneumonia, inability to maintain a “reasonable” SpO<sub>2</sub>.)
    - » Dosage: 0.5-4 L/min via NC.



## CHF in the Adolescent: Management of SCD-Related HF

- Management of Sickle Cell Disease-Related Heart Failure:
  - Intravenous Access
    - Establish medium-bore IV (e.g. 20G) for volume replacement, pain management, and potential transfusion.
      - » These patients often have lots of scar tissue at common venipuncture sites, and smaller(than usual) IV catheters may need to be utilized (e.g. 22-24G).
      - » IV sites commonly include fingers, lateral wrists, forearms, elbows, feet, and/or saphenous veins. Avoid antecubital veins if possible.
      - » \*\*\*Blood products CANNOT be given through an IV in which Lactated Ringer's solution has been administered.\*\*\*
  - ACE Inhibitors:
    - Treatment of choice for SCD-related HF/HTN 2° presumed nephropathy.
    - **\*\*\*DO NOT administer to SCD patients without consulting M.D.**
      - » Captopril Dosage: 25 mg SL/PO
      - » Enalapril Dosage: 0.1 mg/kg PO
      - » Enalaprilat Dosage: 0.01 mg/kg IV

## CHF in the Adolescent: Management of SCD-Related HF

- Management of Sickle Cell Disease-Related Heart Failure:
  - Continuous Positive Airway Pressure(CPAP)
    - Standard of care for all patients with acute pulmonary edema.
    - SCD patients with acute chest syndrome and poor respiratory effort may benefit from CPAP.
      - » **Settings:**
        - **Pressure: 5-15 cm/H<sub>2</sub>O; titrated in stepwise fashion.**
        - **FiO<sub>2</sub>: Start at 24%(or lowest setting); titrate up/down to SpO<sub>2</sub> > 95%.**
  - Furosemide(Lasix)/Bumetanide(Bumex):
    - Diuretics may be helpful if frank pulmonary edema is present.
      - » **Furosemide Dosage: 0.5 - 1 mg/kg IV**
      - » **Bumetanide Dosage: 0.02 - 0.1 mg/kg IV**

## CHF in the Adolescent: Management of SCD-Related HF

- Management of Sickle Cell Disease-Related Heart Failure:
  - Opiates (Morphine/Hydromorphone/Fentanyl):
    - Recommended treatment for SCD-related pain crisis.
      - » Morphine Dosage: 0.05 - 0.15 mg/kg IV
      - » Hydromorphone Dosage: 0.015 – 0.02 mg/kg IV
      - » Fentanyl Dosage: 0.5 – 1 mcg/kg IV
        - (\*\*SLOW PUSH; rapid administration causes chest wall rigidity.\*\*)
  - NSAID Pain Management (Ketorolac):
    - Alternative treatment for SCD-related pain crisis; may be combined with opiates for unresolved pain.
      - » Ketorolac Dosage: 0.5 mg/kg IV (Max: 30 mg)
  - Bronchodilators (Albuterol/Ipratropium):
    - Consider trial of bronchodilators, especially if patient has history of reactive airway disease or wheezing on exam.
      - » Albuterol Dosage: 2.5 - 5 mg/dose via HHN
      - » Ipratropium Dosage: 0.5 mg via HHN x 1 dose



## CHF in the Adolescent: Management of SCD-Related HF

- Management of Sickle Cell Disease-Related Heart Failure:
  - Dopamine Drip
    - Particularly useful if cardiogenic shock is present in the setting of CHF.
      - » Dosage: 5-20 mcg/kg/min IV
        - Calculation:  $6 \times (\text{wt in kg}) = \text{mg Dopamine to add to 100 cc bag of NS.}$
        - \*\*\*At this concentration, 1 cc/hr = 1 mcg/kg/min.
  - Dobutamine Drip
    - Particularly useful if cardiogenic shock is present in the setting of CHF.
      - » Dosage: 5-20 mcg/kg/min IV
        - Calculation:  $6 \times (\text{wt in kg}) = \text{mg Dopamine to add to 100 cc bag of NS.}$
        - \*\*\*At this concentration, 1 cc/hr = 1 mcg/kg/min.
  - Epinephrine Drip (IF REFRACTORY TO DOPAMINE OR DOBUTAMINE)
    - Indicated for SEVERE cardiogenic shock in the setting of CHF.
      - » Dosage: 0.1-1.0 mcg/kg/min IV
        - Calculation:  $0.6 \times (\text{wt in kg}) = \text{mg Dopamine to add to 100 cc bag of NS.}$
        - \*\*\*At this concentration, 1 cc/hr = 0.1 mcg/kg/min.

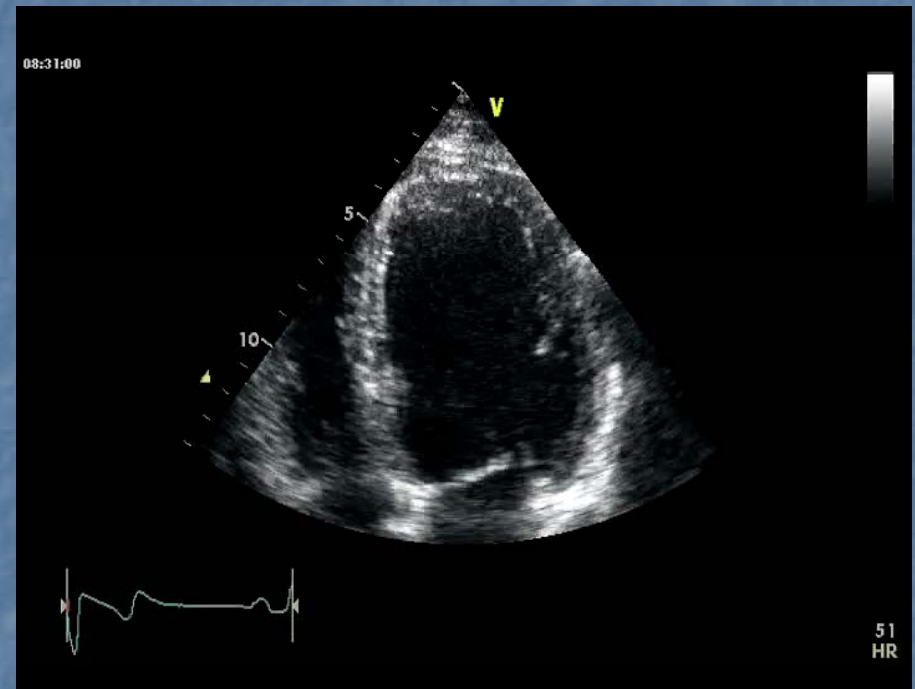
# CHF in the Adolescent: Pathophysiology of Cocaine-Related HF

- Pathophysiology of Cocaine-Related Heart Failure:
  - CHF primarily results from high output secondary to anemia.
  - Regarding the subacute and chronic cardiomyopathies, a clear association has been made between ischemic cardiomyopathy and cocaine use.
    - Regional wall motion abnormalities can be observed, even in patients with no history of myocardial infarction.
  - This syndrome is characterized by evidence of multiple infarcts with normal coronary arteries upon catheterization.
    - This is presumed to be present because of vasospasm or thrombosis.
  - Chronic cocaine use has been estimated to increase left ventricular muscle mass by up to approximately 70%
  - Chronic adrenergic stimulation may play a role in the development of cocaine-related cardiomyopathy and non-fatal/lethal arrhythmias.

# CHF in the Adolescent: Pathophysiology of Cocaine-Related HF

- Pathophysiology of Cocaine-Related Heart Failure:

- Adulterants such as arsenic and magnesium, have been suggested as contributing to cocaine-related cardiomyopathy.
- Long-term cocaine use has been associated with regional left ventricular diastolic dysfunction when analyzed by MRI.
- A history of MI may be present but often is absent.
- Symptoms of chronic CHF usually are absent, but a history of prior CHF related to cocaine use may be present.





## CHF in the Adolescent: Assessment of Cocaine-Related HF

- **Assessment of Cocaine-Related Heart Failure:**
  - Classic presentation of a patient under the influence of cocaine is tachycardia and hypertension.
    - Although the initial increase in heart rate and blood pressure are dose dependent, plateau of heart rate and BP may occur.
  - The patient's skin may be cool and clammy though the patient is hyperthermic; therefore, obtaining a temperature is advisable.
  - Myocardial ischemia/infarction may occur, and can be a cause of CHF in the acute setting.
    - \*\*\*Perform a 12-lead EKG!\*\*\*
  - Unstable tachyarrhythmias may occur; all patients should be placed on a cardiac monitor.
  - Chronic cocaine use may result in CHF 2<sup>o</sup> cardiomyopathy.

## CHF in the Adolescent: Assessment of Cocaine-Related HF

- **Assessment of Cocaine-Related Heart Failure:**
  - The symptoms of cocaine-related HF are the same as symptoms for other forms of congestive heart failure. The onset may be very sudden and of short duration.
  - \*\*\*Symptoms of chest pain may appear to be of muscular origin but may represent ischemia or infarct.\*\*\*
  - With acute binge use of cocaine, the patient may present with acute CHF and pulmonary edema.
  - Hypotension, rather than hypertension, may predominate, making the diagnosis and treatment more difficult.
  - EKG may show evidence of arrhythmias, acute ischemia, infarction, LVH and nonspecific ST-T wave changes.

## CHF in the Adolescent: Management of Cocaine-Related HF

- Management of Cocaine-Related Heart Failure:
  - Treatment consists of standard therapy for congestive heart failure.
  - Diuretics and vasodilators as tolerated. If shock is present, inotropic agents and vasopressors are indicated.
  - If evidence of ongoing ischemia is present, aggressive use of agents directed at relieving vasospasm (nitrates and calcium channel blocking drugs) are indicated.
  - \*\*\*If arrhythmias are present and are felt to be compromising the clinical situation, they should be treated aggressively.\*\*\*
  - \*\*\*The use of beta-blocking drugs is contraindicated, as are IA and IC sodium-channel blockers. Data on the use of magnesium and/or amiodarone are not supportive either.



## CHF in the Adolescent: Management of Cocaine-Related HF

- Management of Cocaine-Related Heart Failure:
  - Oxygen
    - Oxygen should be placed on all patients with cocaine-related cardiac complaints.
    - If the NC agitates the patient, consider deferring O2 via NC.
      - » Rationale: Lower myocardial O2 demand in a non-agitated patient.
      - » Dosage: 2-4 L/min via NC as per ACS guidelines.
  - Lorazepam(Ativan)
    - Patients with HTN may require occasional use of benzodiazepines for sedation.
      - » Ativan Dosage: 0.1 mg/kg IV
  - Continuous Positive Airway Pressure(CPAP)
    - Standard of care for all patients with acute pulmonary edema.
      - » Settings:
        - Pressure: 5-15 cm/H2O; titrated in stepwise fashion.
        - FiO2: Start at 50%; titrate up/down to SpO2 > 95%.

## CHF in the Adolescent: Management of Cocaine-Related HF

- Management of Cocaine-Related Heart Failure:
  - Nitroglycerin
    - Particularly useful if evidence of myocardial ischemia is present.
    - If myocardial ischemia without ST segment elevation is present, the patient should still receive nitroglycerin.
      - » Dosage: 0.4 mg SL q5min
  - Morphine/Hydromorphone/Fentanyl
    - Narcotics may be helpful if relief cannot be obtained with nitroglycerin.
      - » Morphine Dosage: 0.1 mg/kg IV
      - » Hydromorphone Dosage: 0.015 – 0.02 mg/kg IV
      - » Fentanyl Dosage: 1 mcg/kg IV
  - Furosemide/Bumetanide:
    - Diuretics may be helpful if frank pulmonary edema is present.
      - » Furosemide Dosage: 1 mg/kg IV
      - » Bumetanide Dosage: 0.02 - 0.1 mg/kg IV

# CHF in the Adolescent: Management of Cocaine-Related HF

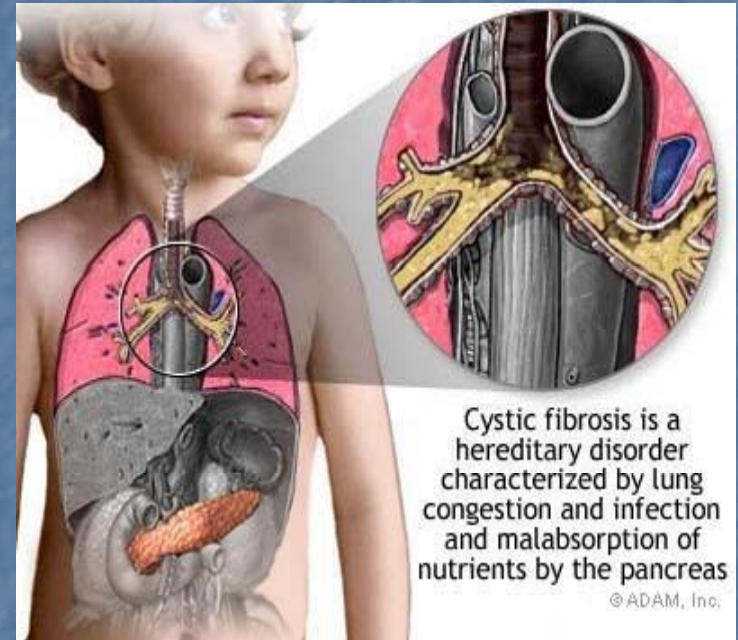
- Management of Cocaine-Related Heart Failure:
  - Dopamine Drip
    - Particularly useful if cardiogenic shock is present in the setting of CHF.
      - » Dosage: 5-20 mcg/kg/min IV
        - Calculation:  $6 \times (\text{wt in kg}) = \text{mg Dopamine to add to 100 cc bag of NS.}$
        - \*\*\*At this concentration, 1 cc/hr = 1 mcg/kg/min.
  - Dobutamine Drip
    - Particularly useful if cardiogenic shock is present in the setting of CHF.
      - » Dosage: 5-20 mcg/kg/min IV
        - Calculation:  $6 \times (\text{wt in kg}) = \text{mg Dopamine to add to 100 cc bag of NS.}$
        - \*\*\*At this concentration, 1 cc/hr = 1 mcg/kg/min.
  - Vasopressin (FOR CARDIAC ARREST ONLY)
    - Useful for cardiac arrest 2° cocaine; increases coronary blood flow.
      - » \*\*\*DO NOT use Epinephrine for cardiac arrest 2° cocaine ingestion.\*\*\*
        - Dosage: 40 units IV x 1
        - \*\*\*Epinephrine has the same effect on the heart as cocaine; increases myocardial demand.\*\*\*
        - Vasopressin increases coronary blood flow and myocardial oxygen availability.
        - Cocaine toxicity causes acidosis. Epinephrine is ineffective in acidosis. Vasopressin demonstrates vasoconstricting efficacy even with severe acidosis.



## CHF in the Adolescent: Pathophysiology of CF-Related HF

- Pathophysiology of Cystic Fibrosis-Related Heart Failure:

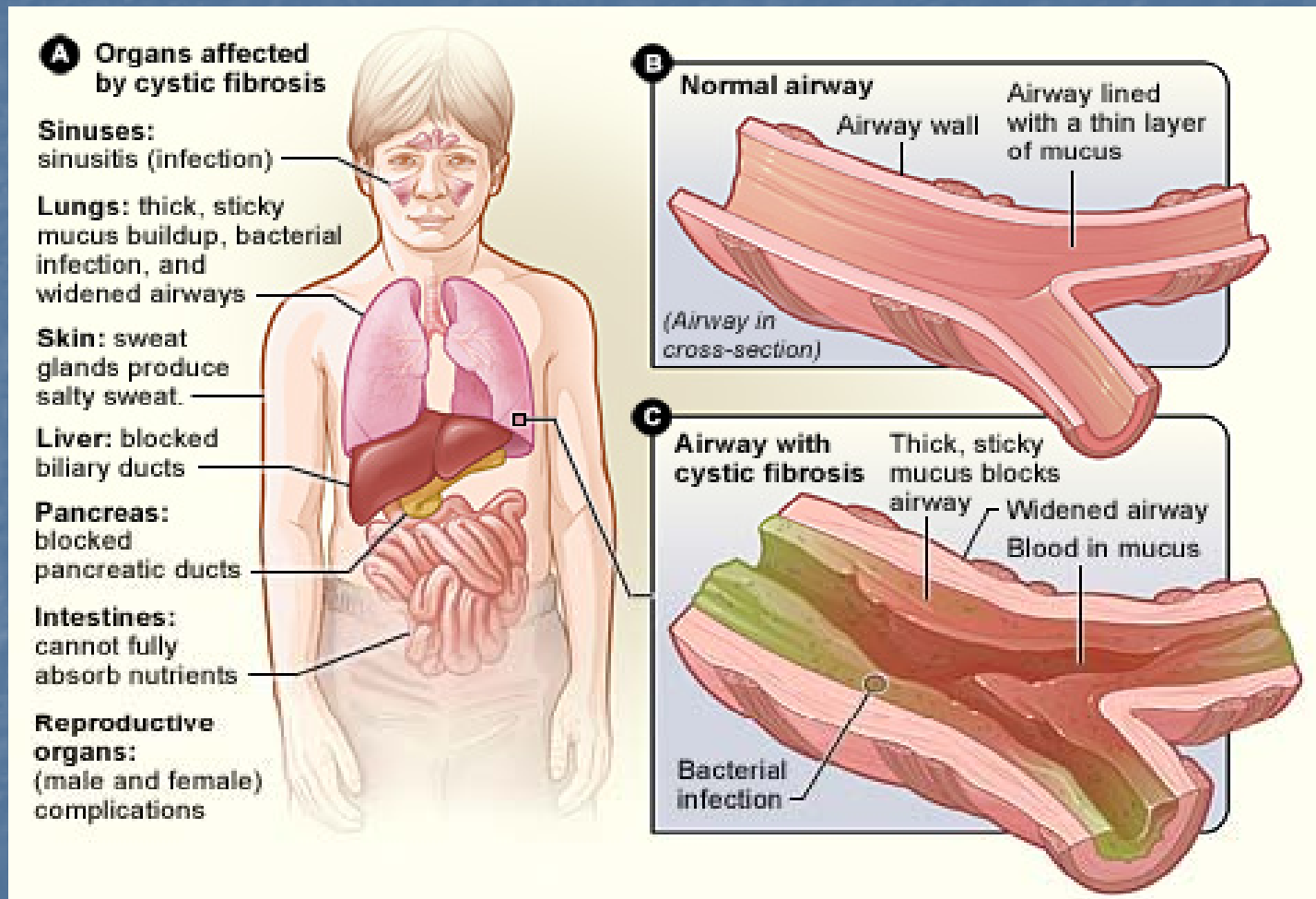
- In cystic fibrosis (CF) precapillary injury is the main pathology leading to pulmonary heart disease according to the WHO definition of cor pulmonale.
- CHF primarily results from raised pulmonary artery pressure (PAP) secondary to chronic or acute-on-chronic hypoxia from the progressive lung destruction.
- The pulmonary vascular changes in CF are similar to that of other hypoxic lung diseases.



Cystic fibrosis is a hereditary disorder characterized by lung congestion and infection and malabsorption of nutrients by the pancreas

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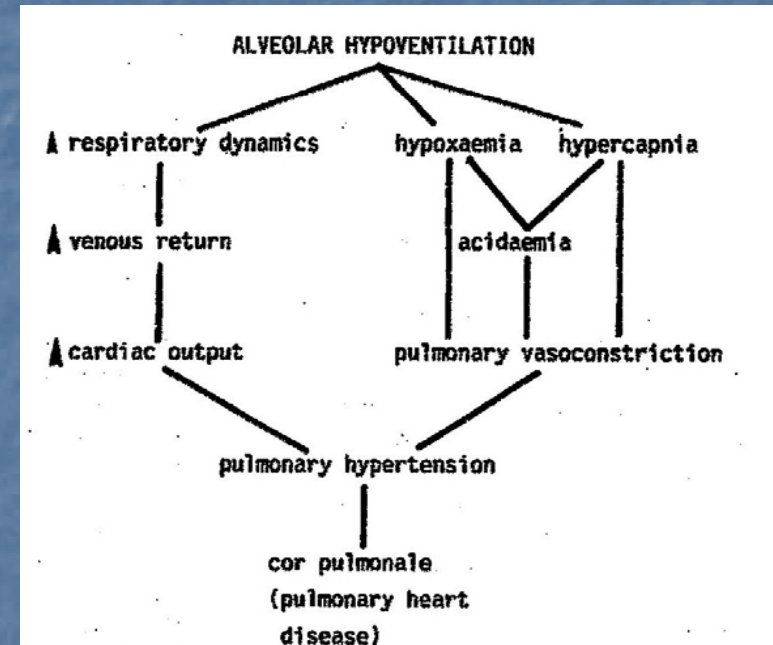
## CHF in the Adolescent: Pathophysiology of CF-Related HF



## CHF in the Adolescent: Pathophysiology of CF-Related HF

- Pathophysiology of Cystic Fibrosis-Related Heart Failure:

- In CF, V/Q abnormalities are the rule and this mismatch leads to the redistribution of blood flow.
- Hypoxic pulmonary vasoconstriction results in pulmonary HTN, and increases energy demands on the RV.
- Small pulmonary arterioles thicken and once these changes occur, acute responses to oxygen cannot be expected.
- Even though PAP may decrease, so will C/O with no alteration in PVR.



*Figure 1. Pathophysiology of cor pulmonale in cystic fibrosis*

*Table 1. Mechanisms of pulmonary artery pressure increase*

<b>Hypoxic vasoconstriction</b>
<b>Mediator release</b>
<b>Chronic pulmonary inflammation</b>
PGF2alpha
thromboxane
<b>Vascular tone</b>
<b>Bronchopulmonary vascular anastomoses</b>
<b>Peripheral blood vessel loss</b>



## CHF in the Adolescent: Assessment of CF-Related HF

- Assessment of Cystic Fibrosis-Related Heart Failure:

- Ultimately, overall assessment of the CF patient in cor pulmonale will have to be done in the hospital, due to equipment limitations:

- EKG
  - » May or may not be useful as respiratory vs. cardiac cause is obvious in this case, but should be done to evaluate for AMI.
- Chest x-ray
- Echocardiography
- Pulmonary function testing
- MUGA scan

**Table 2. Symptoms and signs of pulmonary heart disease in cystic fibrosis**

### **Symptoms**

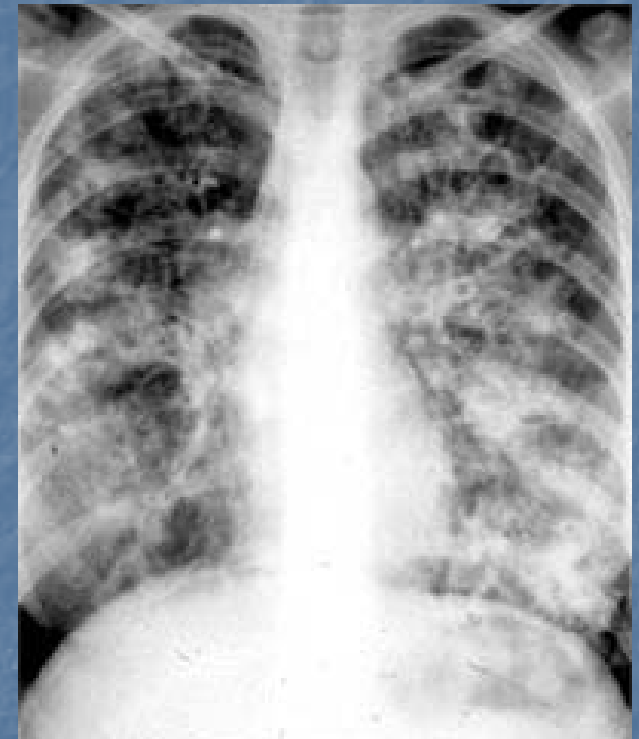
Dyspnoea: at rest on exertion	both occur in patients with chronic respiratory disease such as CF
Syncope (cough), dizziness	failure to increase CO (due to increased intrathoracic pressure)
Chest pain	'right heart angina'
Haemoptysis	may occur in CF (bronchiectasis)
other: palpitations hoarseness epigastric pain nausea, vomiting	

### **Signs**

Chest wall deformities	seen in CF.
Cyanosis	low CO right-left shunting haemoglobin desaturation
Tachypnoea	seen in CF
Cardiovascular examination:	
visible right ventricular impulse	(seen in thin CF patients)
distended neck veins	
enlarged liver	
blurred auscultation	(overlying hyperinflation in CF)
oedema	(check for hypalbuminaemia in CF)
digital clubbing	(common in CF, non-specific)

## CHF in the Adolescent: Management of CF-Related HF

- Management of Cystic Fibrosis-Related Heart Failure:
  - Oxygen
    - Hypoxia is a prerequisite for oxygen therapy in the CF patient.
    - Oxygen to ensure saturations > 90%.
    - Take special care with patients who may retain CO<sub>2</sub>, because their main respiratory drive is hypoxemia and uncontrolled correction can produce worsening hypoventilation.
      - » Dosage: 1 – 1.5 L/min via NC; or Venturi Mask with FiO<sub>2</sub> of 24%; increased in stepwise fashion to SpO<sub>2</sub> ≥ 90%.



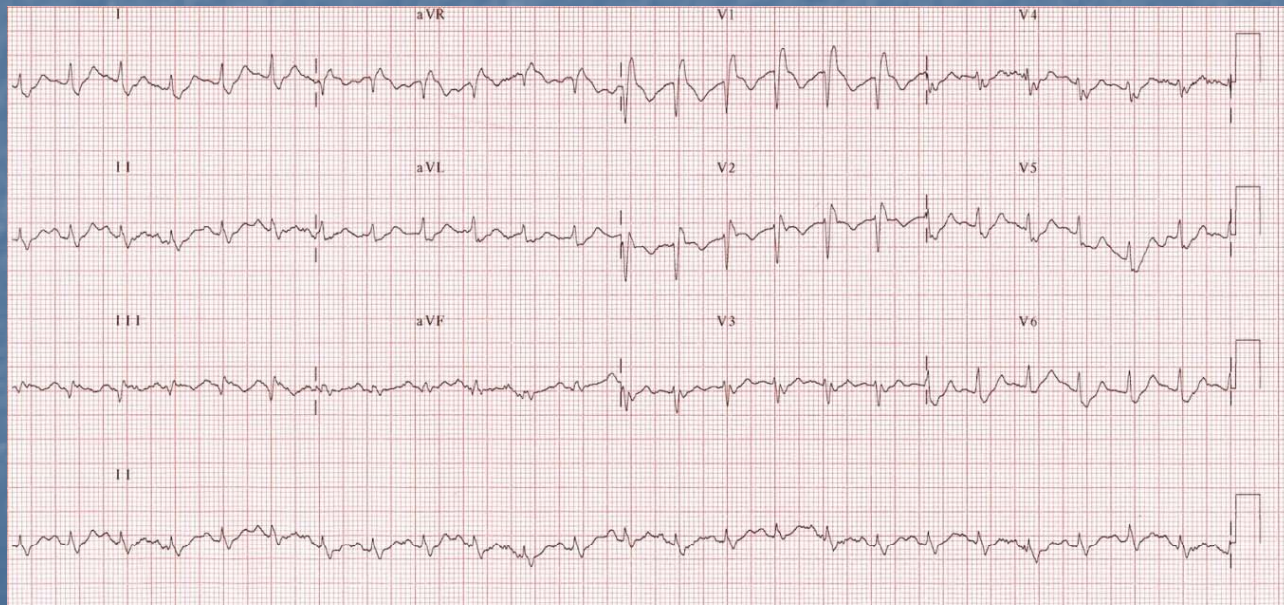
## CHF in the Adolescent: Management of CF-Related HF

- Management of Cystic Fibrosis-Related Heart Failure:
  - Continuous Positive Airway Pressure(CPAP)
    - Standard of care for all patients with acute pulmonary edema.
    - “Sputum clearance and spirometry were not impaired by the short-term administration of either CPAP or BiPAP combined with directed cough.”
    - “According to the benefits of NIPPV on respiratory work and gas exchange and to the comparable spirometry and amount of sputum cleared using the different treatments, we could consider the short-term administration of NIPPV combined with an ACT as a possible airway clearance regimen.” (Placidi et al. )
    - First report on CPAP used to clear bronchial secretions in CF.
      - » **Settings:**
        - **Pressure: 5 cm/H<sub>2</sub>O; titrated in stepwise fashion.**
        - **FiO<sub>2</sub>: Start at 24%(or lowest setting); titrate FiO<sub>2</sub> upward in small increments to achieve an SpO<sub>2</sub> ~90%.**



## CHF in the Adolescent: Management of CF-Related HF

- Management of Cystic Fibrosis-Related Heart Failure:
  - 12-Lead Electrocardiogram
    - In the presence of pulmonary disease, no differentiation between the cardiac and pulmonary origin of EKG alterations can be made.
    - Typical findings may include: RAD, Incomplete RBBB, ST/T-wave changes.
    - 12-Lead EKG should be performed in the usual fashion to evaluate for acute ischemia/infarction.



## CHF in the Adolescent: Management of CF-Related HF

- Management of Cystic Fibrosis-Related Heart Failure:
  - Diuretics (Furosemide/Bumetanide)
    - Diuretics are indicated in frank cardiac failure or if unexplained weight gain is present.
    - Hypoproteinemia occurs in CF and manifests as edema; this can lead to the erroneous use of diuretics. A thorough and meticulous assessment is critical.
      - » Furosemide Dosage: 1 mg/kg IV
      - » Bumetanide Dosage: 0.02 - 0.1 mg/kg IV
  - Bronchodilators (Albuterol/Ipratropium/Terbutaline):
    - Consider trial of bronchodilators, especially if patient has history of reactive airway disease or wheezing on exam.
      - » Albuterol Dosage: 2.5 - 5 mg/dose via HHN
      - » Ipratropium Dosage: 0.5 mg via HHN x 1 dose
      - » Terbutaline Dosage: 0.25 mg SQ/IM/IV(preferred route) q20 min
        - Alt. Dosage : 0.25 mg(250mcg) IV x 1, followed by 5 mcg/min drip.



## CHF in the Adolescent: Management of CF-Related HF

- Management of Cystic Fibrosis-Related Heart Failure:
  - Calcium Channel Blocking Agents (Diltiazem/Nifedipine):
    - Diltiazem HCl(Cardizem):
      - CCB that increases cardiac output and oxygen delivery during rest and exercise in PPH. (Marginally better than nifedipine.)
      - Dosage: 0.25 mg/kg SLOW IV push
      - \*\*\*DISCUSS WITH MD PRIOR TO ADMINISTERING\*\*\*
    - Nifedipine(Procardia):
      - CCB that increases cardiac output and oxygen delivery during rest and exercise in PPH, and provides acute pulmonary vasodilation by decreasing pulmonary VR.
      - Dosage: 10 mg PO (0.25 to 0.5 mg/kg/dose)
      - \*\*\*DISCUSS WITH MD PRIOR TO ADMINISTERING\*\*\*



# CHF in the Adolescent: Management of CF-Related HF

- Management of Cystic Fibrosis-Related Heart Failure
  - Dopamine Drip
    - Particularly useful if cardiogenic shock is present in the setting of CHF.
      - » Dosage: 5-20 mcg/kg/min IV
        - Calculation:  $6 \times (\text{wt in kg}) = \text{mg Dopamine to add to 100 cc bag of NS.}$
        - \*\*\*At this concentration, 1 cc/hr = 1 mcg/kg/min.
  - Dobutamine Drip
    - Particularly useful if cardiogenic shock is present in the setting of CHF.
      - » Dosage: 5-20 mcg/kg/min IV
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  - Epinephrine Drip (IF REFRACTORY TO DOPAMINE OR DOBUTAMINE)
    - Indicated for SEVERE cardiogenic shock in the setting of CHF.
      - » Dosage: 0.1-1.0 mcg/kg/min IV
        - Calculation:  $0.6 \times (\text{wt in kg}) = \text{mg Dopamine to add to 100 cc bag of NS.}$
        - \*\*\*At this concentration, 1 cc/hr = 0.1 mcg/kg/min.

## Fetal/Neonatal/Pediatric CHF in Review: Summary

### Summary:

- CHF in utero is typically a result of congenital defects, arrhythmias, or valvular disorders, and occurs in approximately 8 out of every 1,000 pregnancies.
- CHF in the newborn is typically due to myocardial dysfunction 2° asphyxia, hypoglycemia, hypocalcemia or sepsis. Many cases of CHF are due to ductal-dependent defects that present around day 5 of life when the PDA closes.
- The most common cause of CHF in infants is a ventricular septal defect (VSD) that presents around 6-8 weeks of age; the volume of the left to right shunt increases as the pulmonary resistance falls.
  - Although a murmur of VSD is apparent by one week, the full blown picture of CHF occurs around 6-8 weeks.
- Marked distress during feedings, diaphoresis, increased respiratory effort, tachycardia, systolic murmur and crepitations are all common findings in patients with ALCAPA.
  - As the pulmonary artery pressure decreases in the neonatal period, these babies suffer from episodes of angina and myocardial infarction.

## Fetal/Neonatal/Pediatric CHF in Review: Summary

### Summary:

- CHF in the general pediatric population typically results from left sided obstructive diseases (aortic coarctation, stenosis) infections, uncontrolled hypertension; often 2° renal dysfunction, and arrhythmogenic etiologies.
- Treatment is fairly standard for acute pulmonary edema/CHF in those children, however particular attention is given to attempt to identify and treat the offending cause.
- Poor weight gain in the child and/or adolescent is a hallmark of congenital heart disease, due to chronically increased workload on the heart.
- CHF in adolescents is typically caused by either a cardiomyopathy or arrhythmogenic etiology, an underlying secondary disease process, or by drug use.
- CHF in this age group may be caused due to ischemia/infarction, and a 12-lead EKG should be performed without delay.
- One final thought... These patients and their parents can usually tell you 10 times more than an entire library of medical books.



Comments...Questions...Where is the nearest bathroom...?



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