

Neonatal Grand Rounds – Left Ventricular Outflow Tract Obstruction

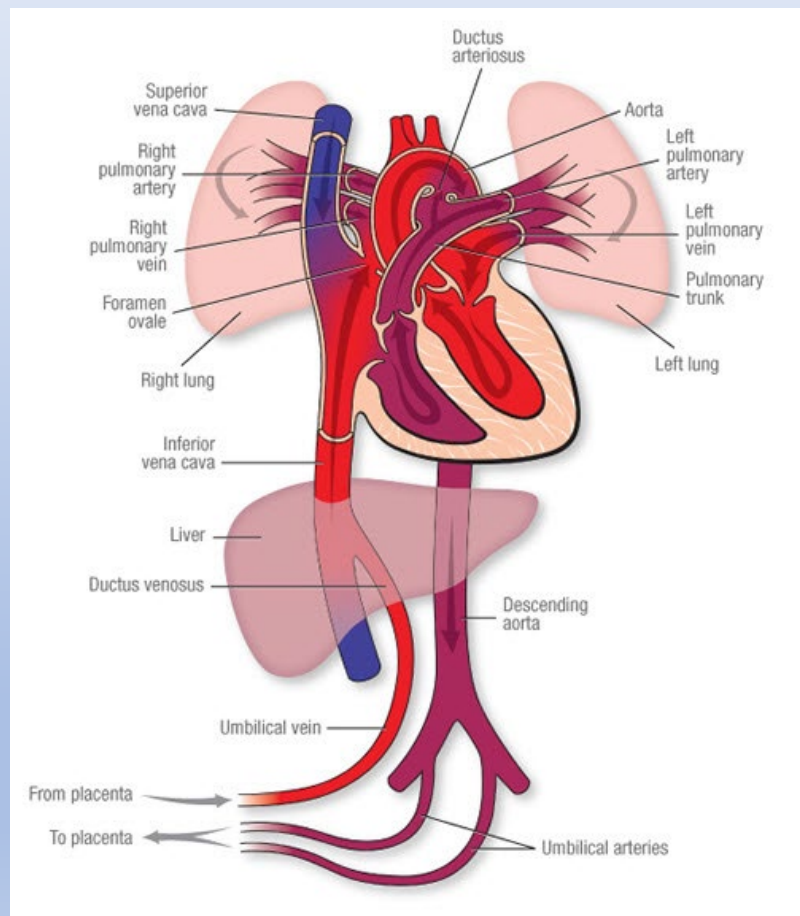
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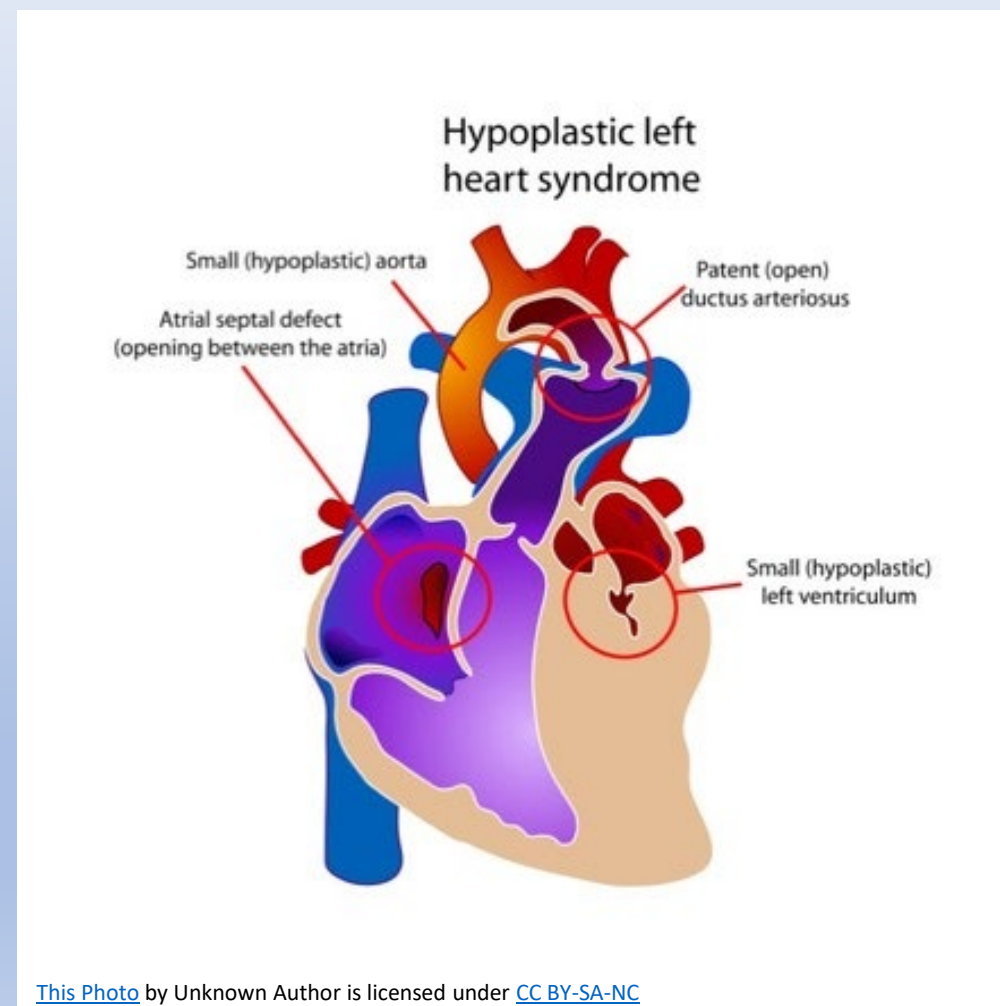
No Disclosures

Fetal Perspectives

Normal Fetal Circulation



Fetal Hypoplastic Left Heart



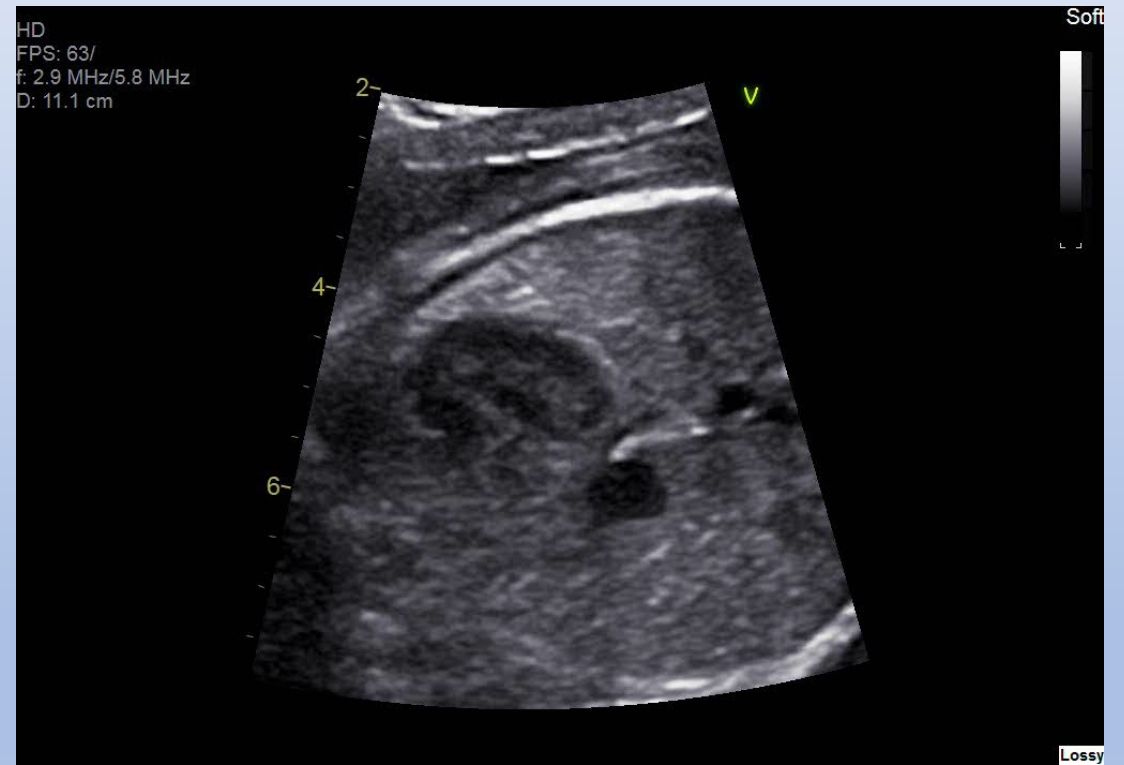
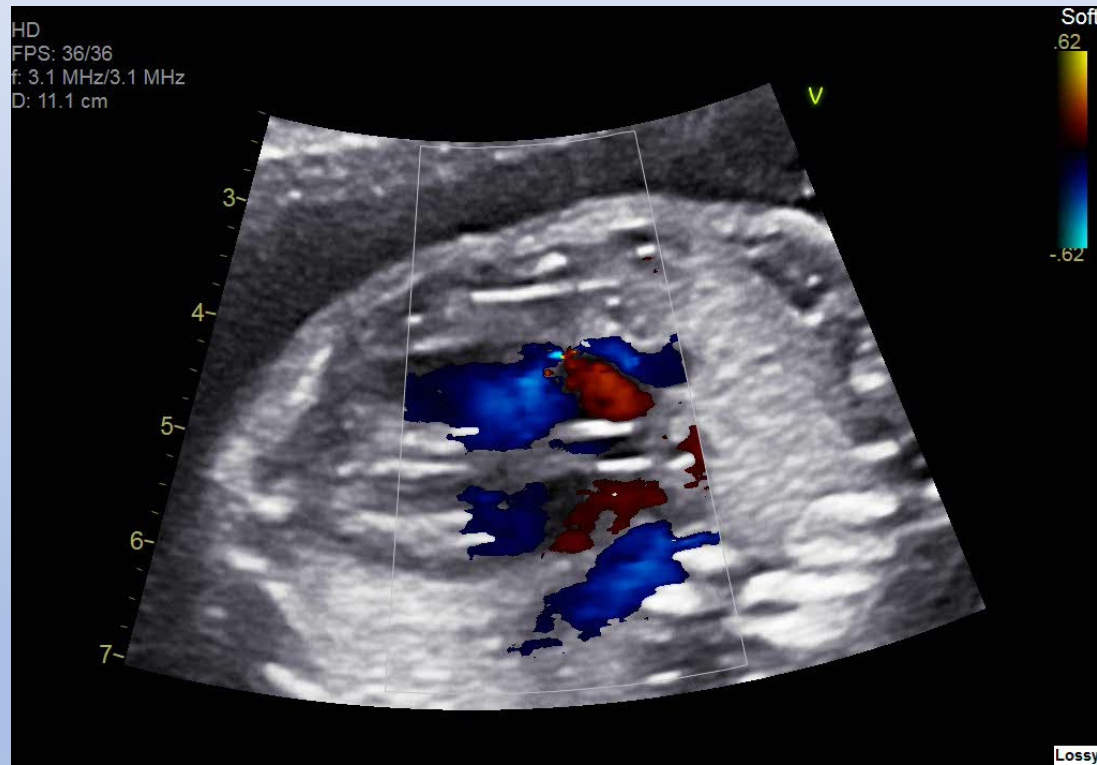
Anatomic Types

- Critical aortic stenosis
- Hypoplastic left heart syndrome
- Other variants of single ventricle with obstruction to systemic output (DILV with TGA, tricuspid atresia with TGA)
- Cardiac Masses (fibroma, rhabdomyomas causing LVOT obstruction)
- Discrete subaortic stenosis
- LVOTO in patients with AV canal defects (chordal tissue)
- HCM, severely affects infants of diabetic mothers

Pathophysiology of Critical Left Heart Obstruction in the Newborn

- Ductus dependent for systemic blood flow
- Oxygenation typically good – may not look cyanotic but depends on having adequate L-R flow across the atrial septum
- Babies may look remarkably well at birth, with ductal closure present with symptoms of poor perfusion – lethargy, poor feeding, appearing grey/ashen, sweating, cold to touch, grunting, eventually shock and death
- Important differential diagnosis of shock/collapse in first month of life (sepsis mimic)

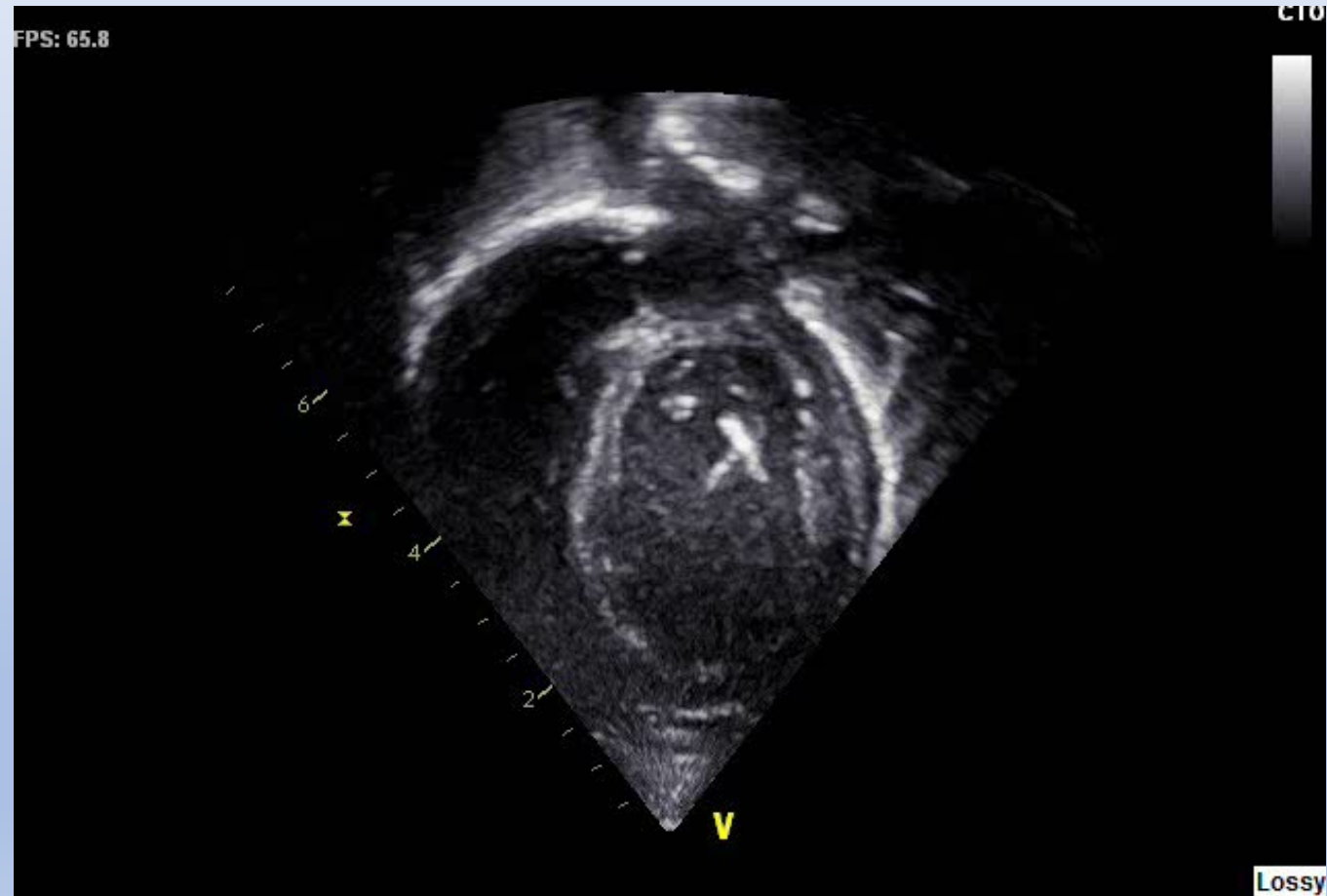
Critical Aortic Stenosis – Fetal Presentation at 24 weeks



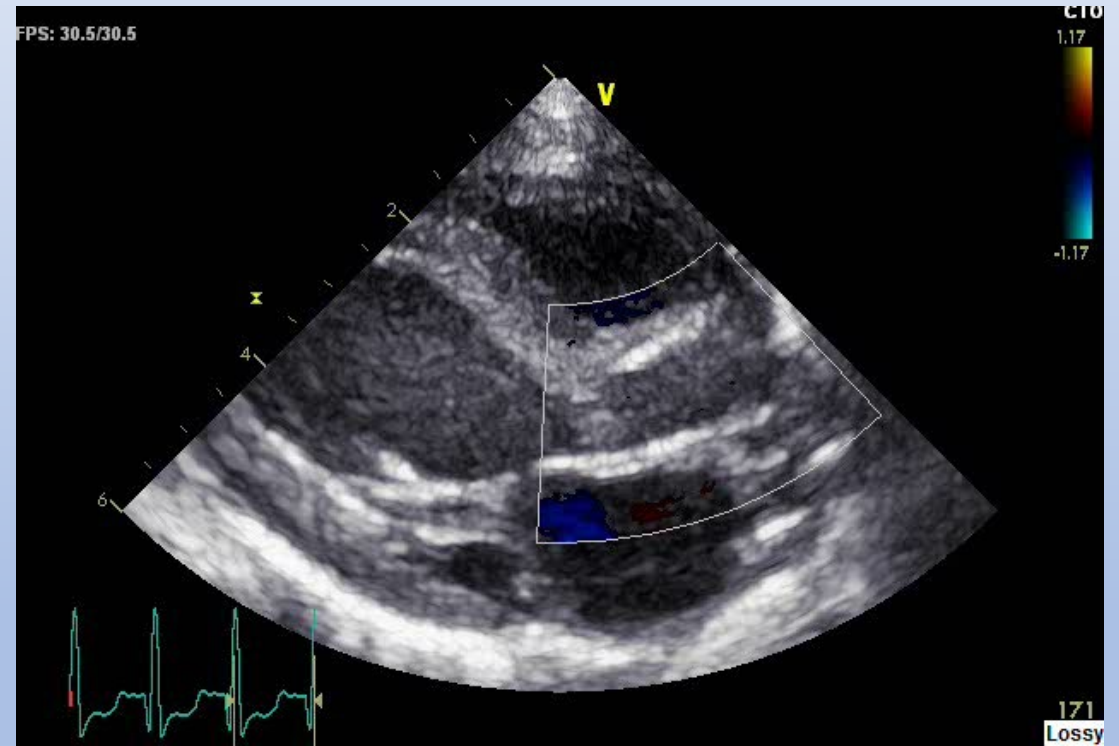
Follow Up at 35 weeks



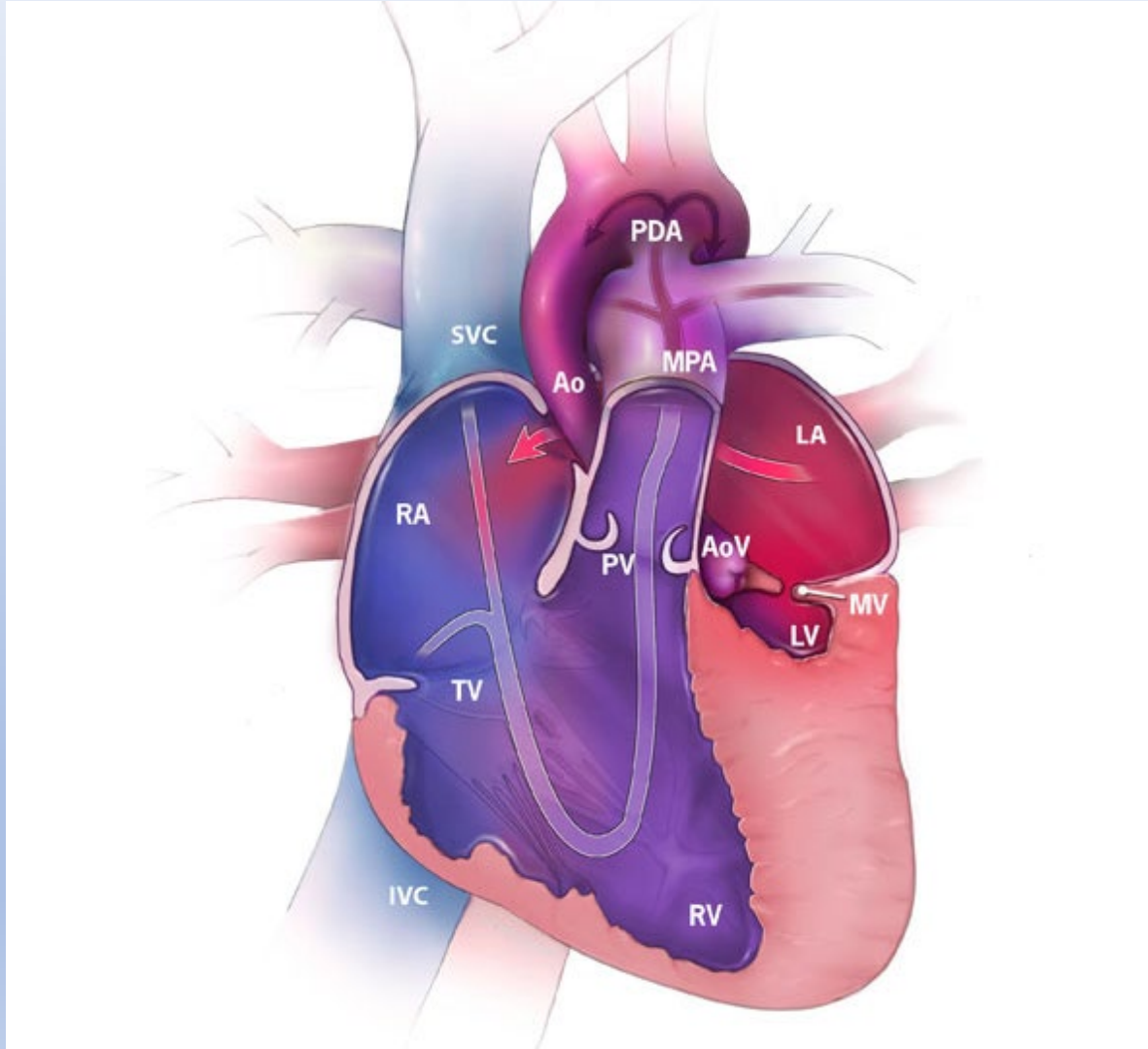
Post Natal Echocardiogram



Severe Neonatal Aortic Stenosis, Severe LV dysfunction.....Management?

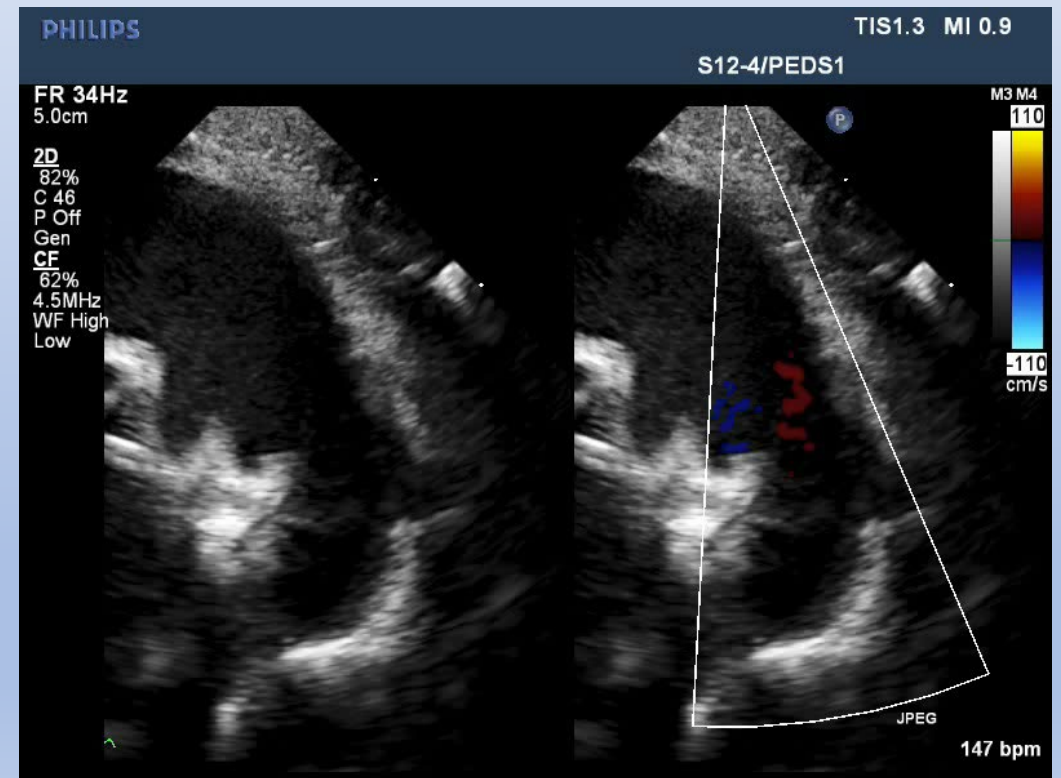
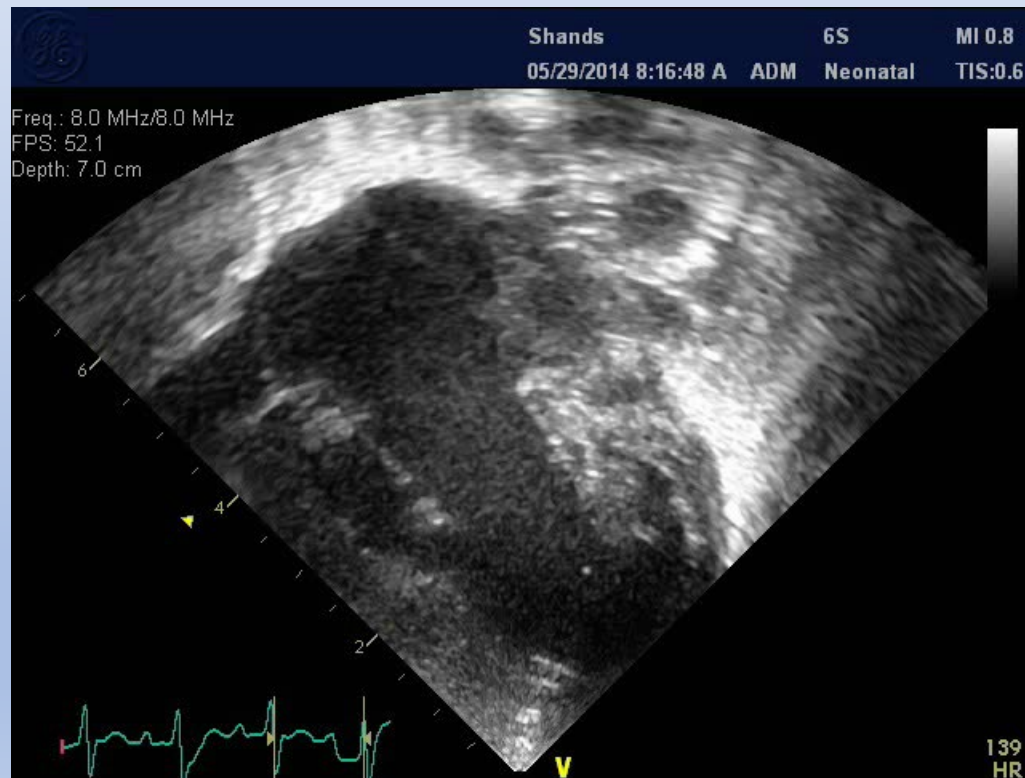


Hypoplastic Left Heart Syndrome

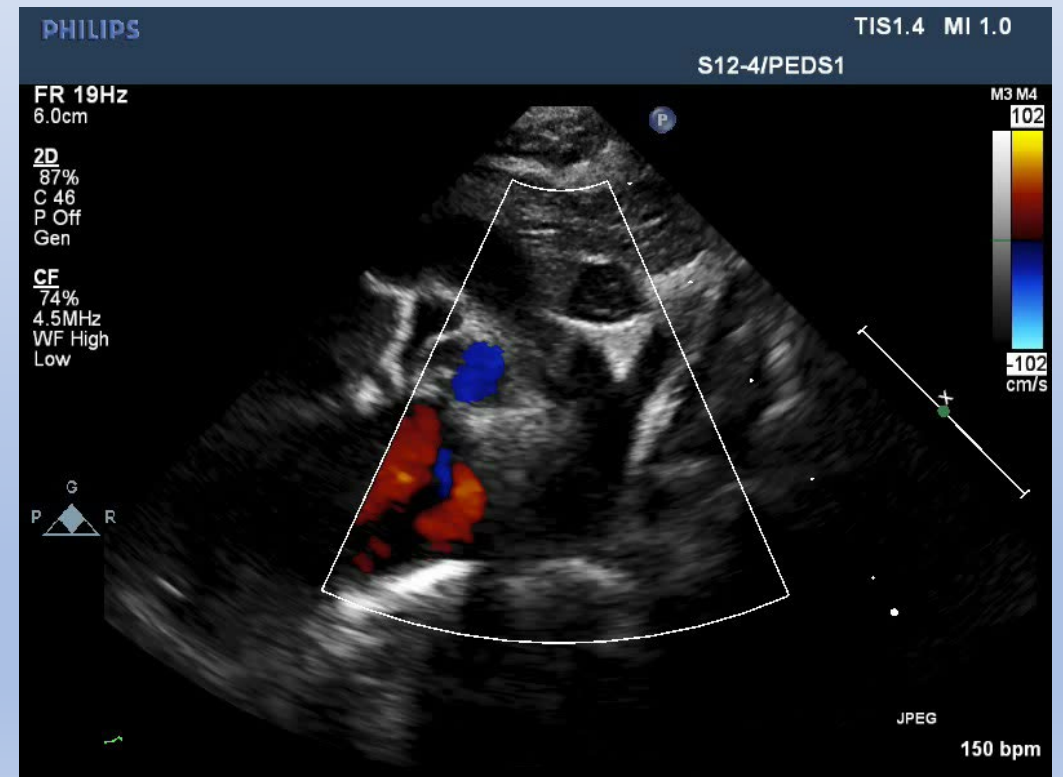


Courtesy: CDC

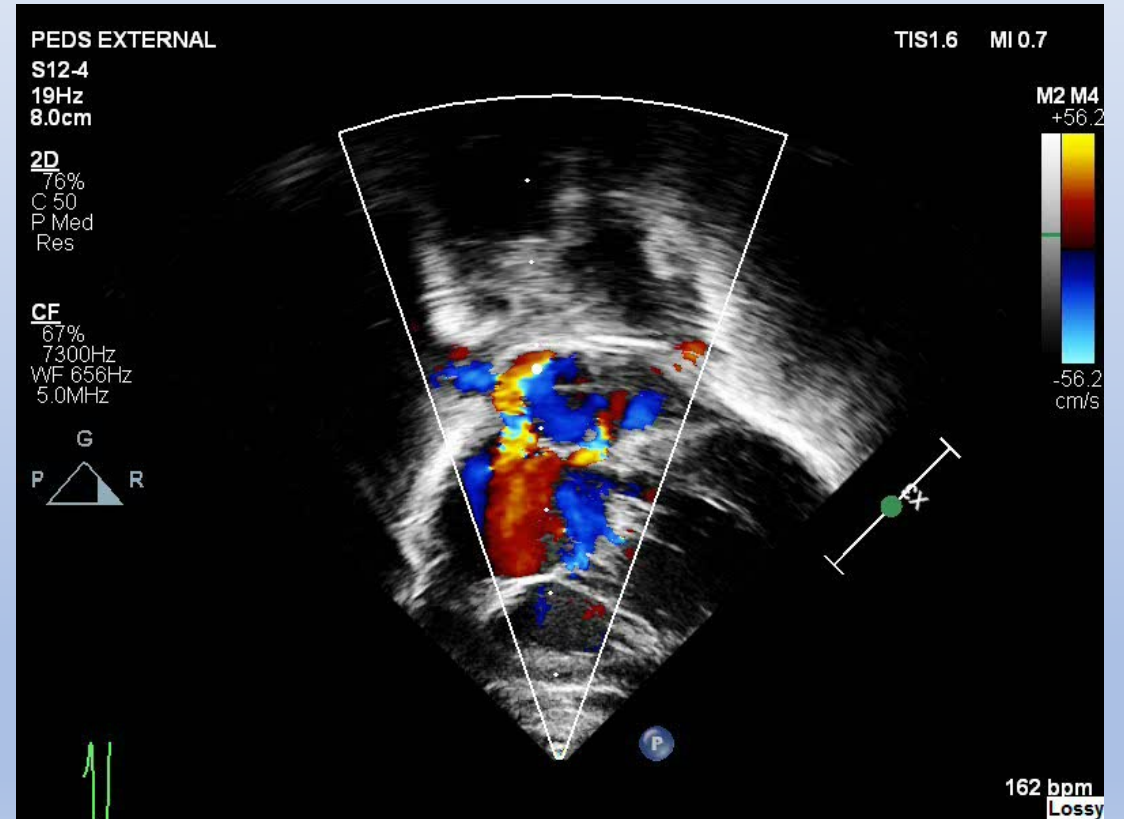
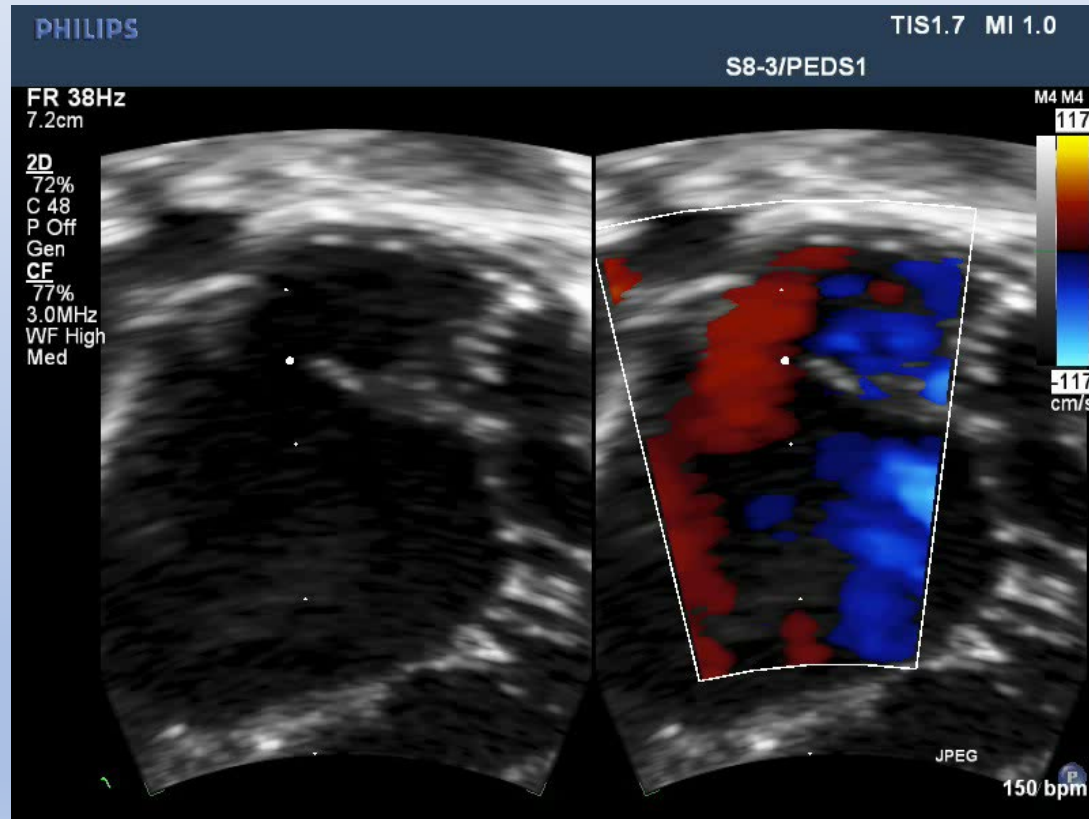
Neonatal HLHS



Neonatal HLHS



Careful Assessment of Atrial Septum



Detection of Critical Left Heart Obstruction in the Newborn

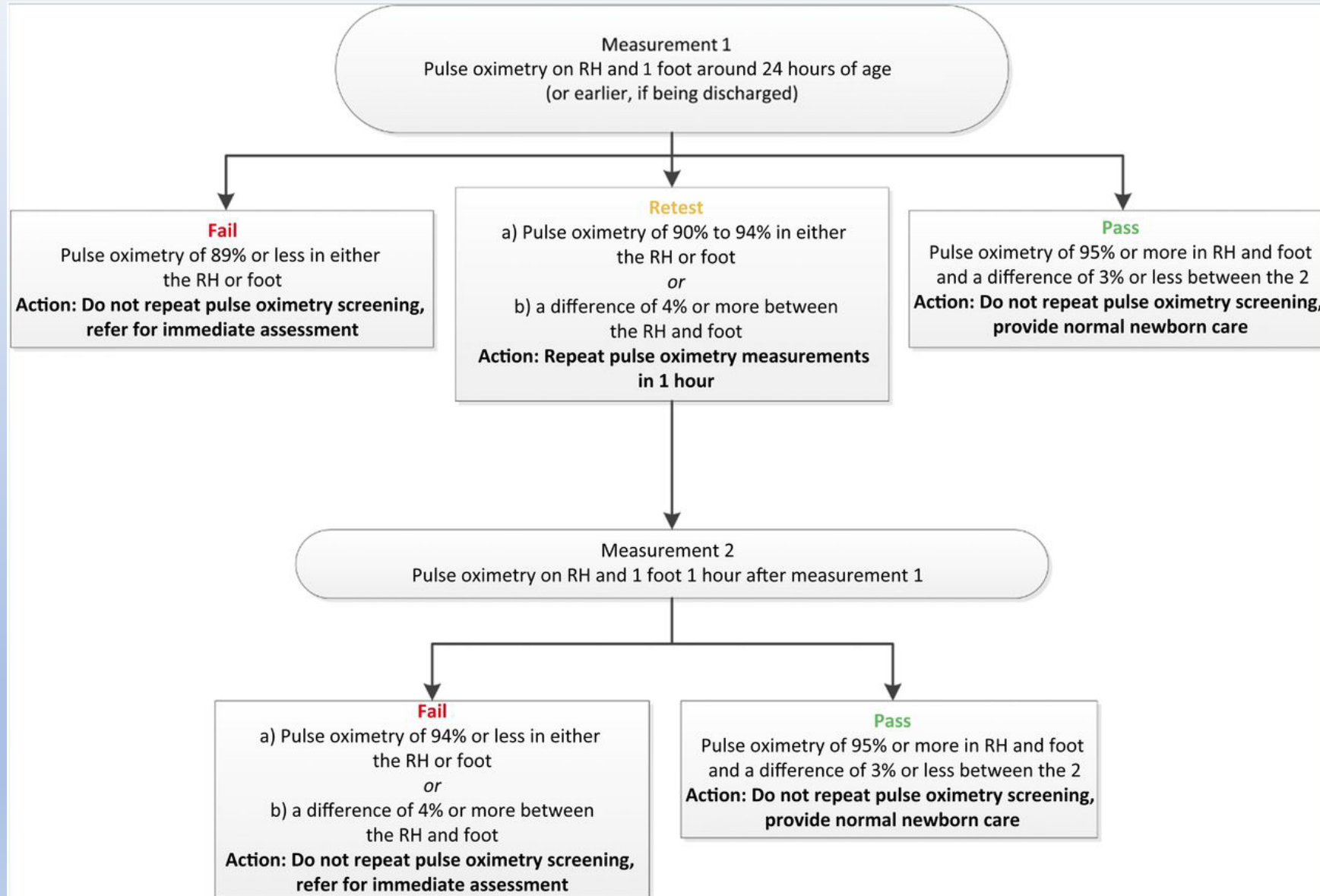
- Prenatal diagnosis
- Clinical Exam is OFTEN inadequate (Why?)
- Pulse oximetry screen

Pulse Oximetry Screening

- Some reports of detection of CHD using pulse ox screening (1995)
- AAP and AHA scientific statements conclude that POS may improve CHD detection (2009)
- US Dept of HHS recommends POS for CCHD be added (2010)
- Individual states (IN, MD, NJ) begin mandating POS (2011)
- 80% states screening for CCHD (2015)
- CCHD screening adopted in all states (2018)



From: Updated Strategies for Pulse Oximetry Screening for Critical Congenital Heart Disease



Expected Sensitivity of POS for CHD

Expected Sensitivity of Pulse Oximetry for Detection of CCHD Screening Targets³⁵

High (>80%)	Medium (60%–80%)	Low (<60%)
Critical pulmonary stenosis	Critical aortic stenosis	Coarctation of the aorta
d-transposition of the great arteries	Double-outlet right ventricle	Ebstein anomaly
Hypoplastic left heart syndrome	Tricuspid atresia	Interrupted aortic arch
Pulmonary atresia		Tetralogy of Fallot
Single ventricle		
Total anomalous pulmonary venous drainage		
Truncus arteriosus		

NICU Management of Baby with critical LVOT obstruction

- Obtain reliable IV access and initiate PGE1 – typical start dose 0.03 mcg/kg/min unless late diagnosis and ductal constriction begun
- If so may need higher doses up to 0.1 or more
- If diagnosis in doubt (availability of echo/cardiology consultation) consider starting PGE in a baby with unexplained cardiovascular collapse even if treating for sepsis
- Be aware of risk of apnea with higher dose PGE or in sick neonates
- Other adverse effects include: hypotension, fever, irritability,
- Periostitis with prolonged use

NICU Management – Monitor for Adequacy of Systemic Output

- Clinical:
 - Are pulses easily palpable
 - Capillary refill
 - Are extremities warm
 - Urine output
- Monitors:
 - NIRS (trend in NIRS very useful as surrogate for tissue perfusion)
- Labs:
 - Lactate
 - ABG (metabolic acidosis)
 - Renal function
 - Liver function (late)
 - Coagulation (Late)
 - SVO2

What if the baby with HLHS is disproportionately hypoxic?

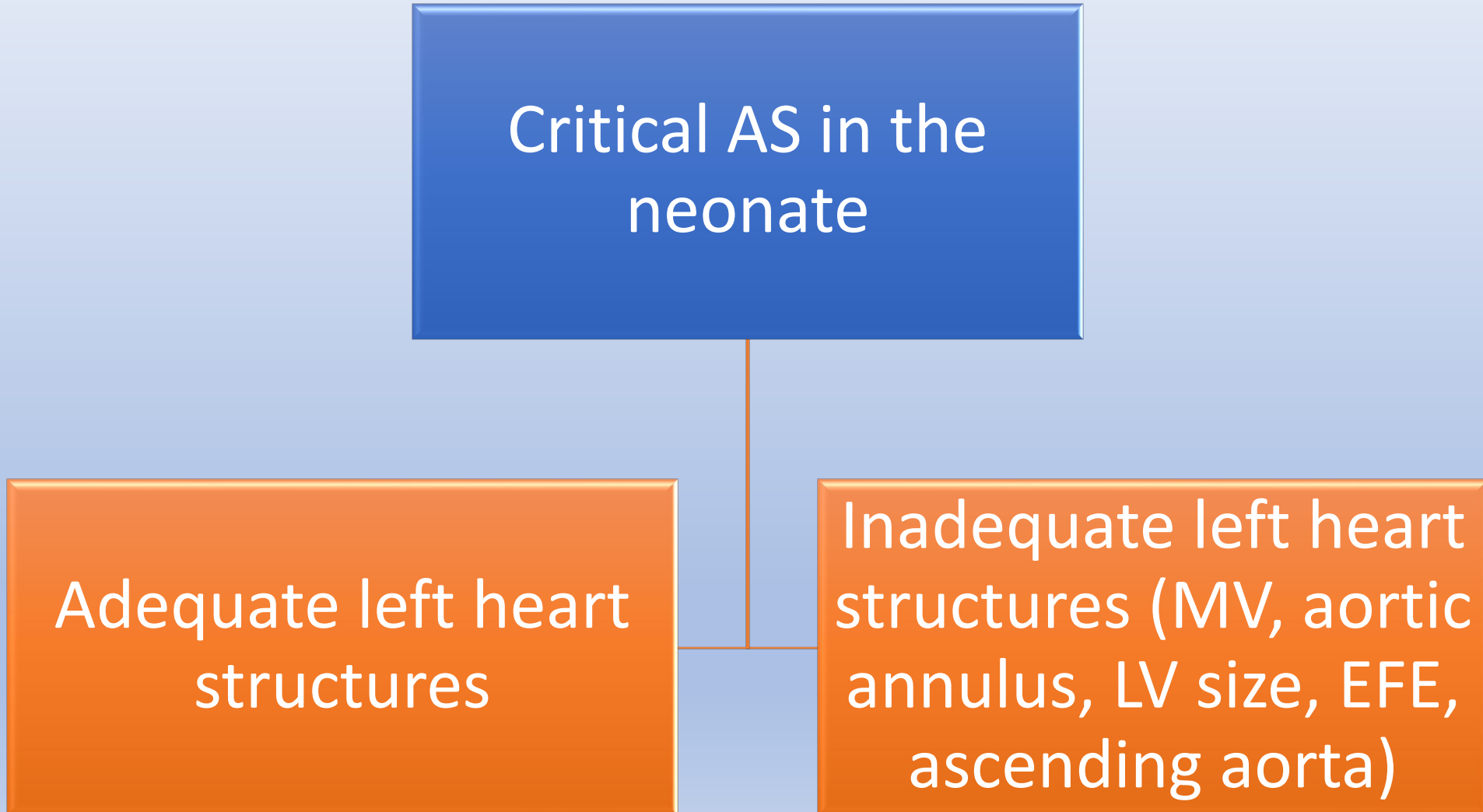
NICU Management – Respiratory Support

- May have high sats high 80-90s
- Patients will have excessive pulmonary blood flow and thus systemic hypoperfusion even with widely patent ductus!
- May develop tachypnea and low BP
- May need HFNC/CPAP for symptomatic benefit
- Avoid supplemental FiO₂ (use 21% FiO₂). Role of sub-ambient oxygen is unclear
- Some neonates will need to be intubated/paralyze for excessive work of breathing. Positive pressure ventilation may help limit PBF
- Permissive hypercapnia (goal: pH 7.30, pCO₂: 55-65) helps raise MAP and protect SBF by causing pulmonary vasoconstriction and reducing overcirculation

NICU Management - Feeding

- Most data tend to support continued feeds in stable patients
- Start very slow and gradually increase
- Avoid fortifying to minimize risk of NEC
- Avoid NJ feeds (residuals and emesis not reliable)
- Monitor carefully for feeding tolerance, increased residuals/emesis, abdominal distension, bloody stool
- Consider partially/fully hydrolyzed formula for feed intolerance

Decision Pathway for Neonates with Critical AS



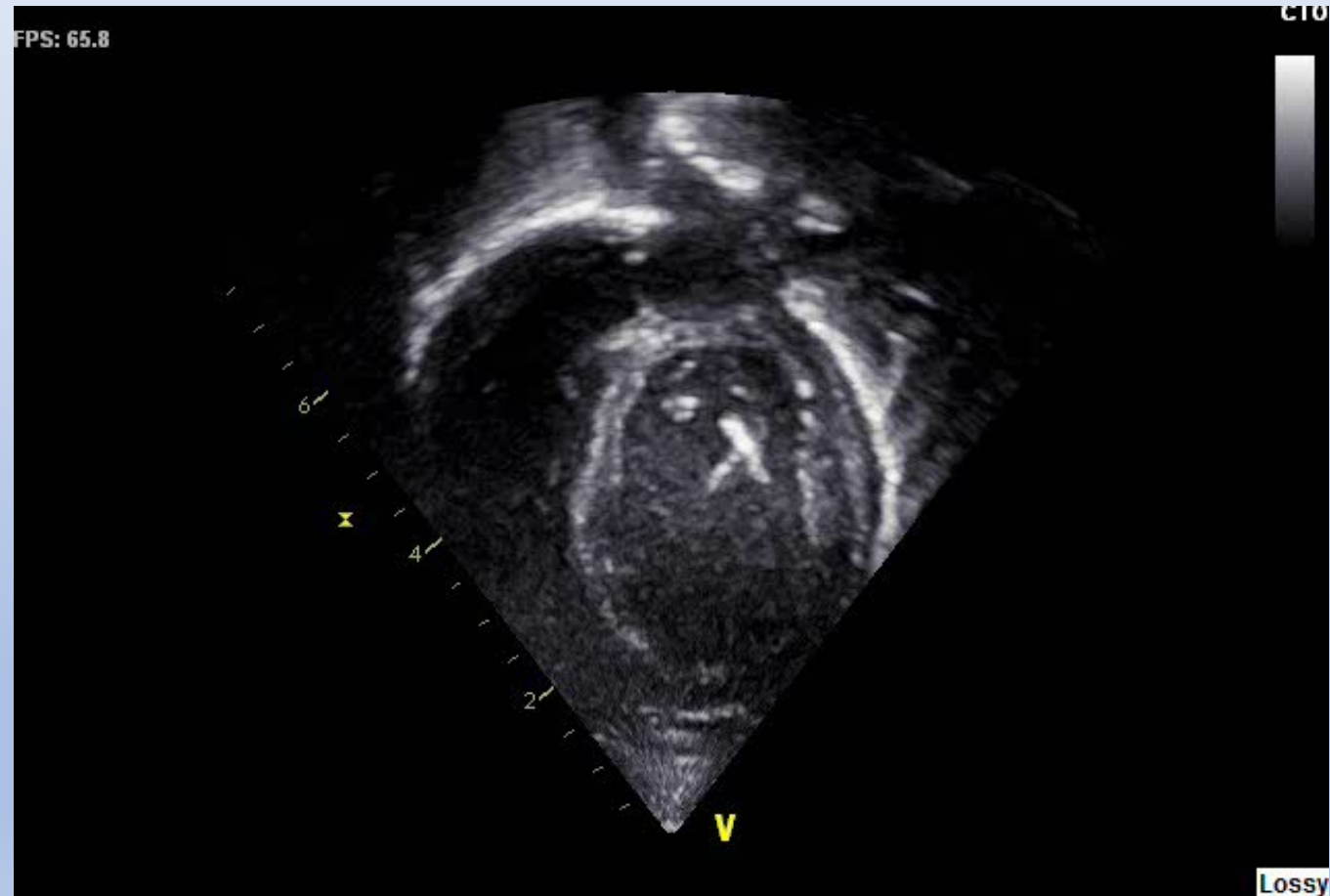
Aortic Stenosis with Adequate Left Heart Structures

- Balloon aortic valvuloplasty
- If inadequate response: consider surgical aortic valvuloplasty
- Ross operation

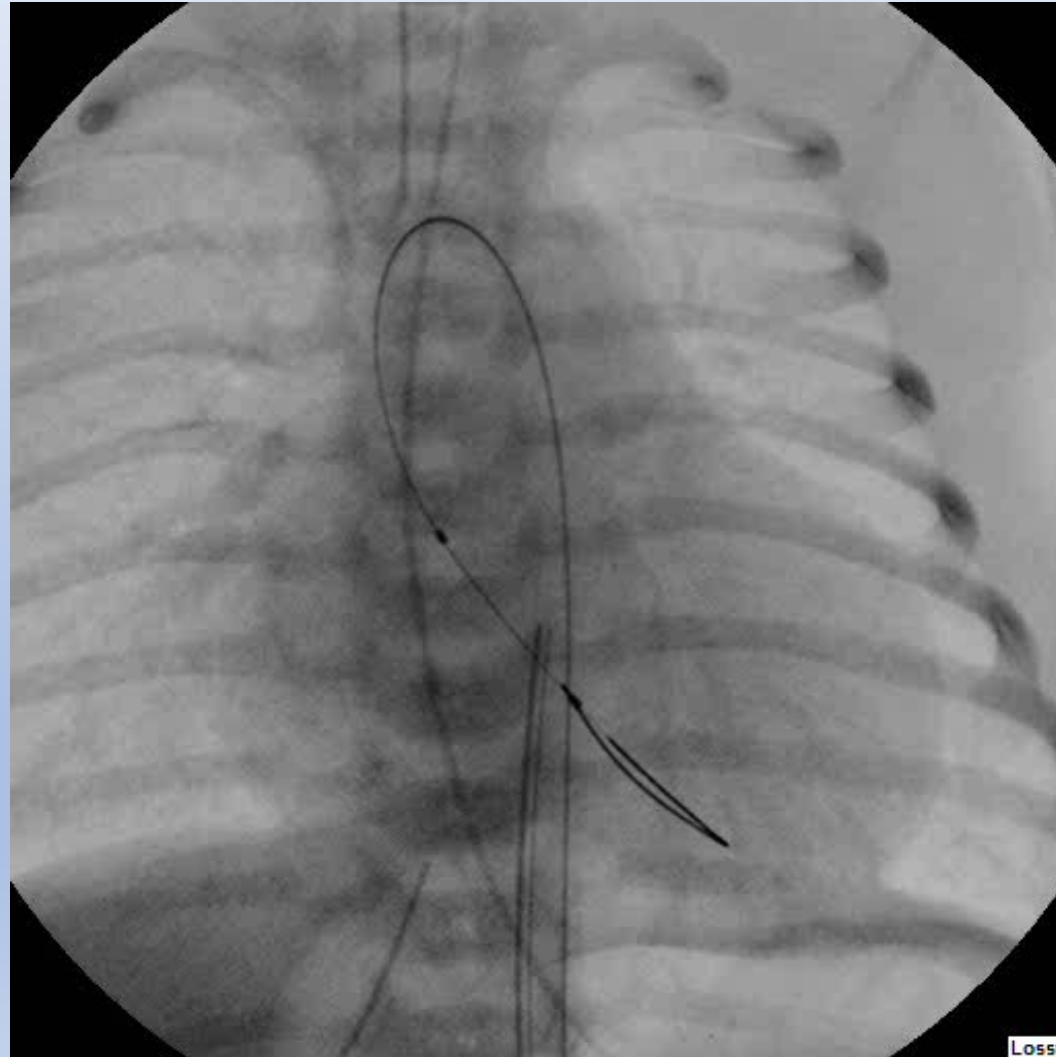
Aortic Stenosis with Inadequate Left Heart Structures

- Single Ventricle algorithm

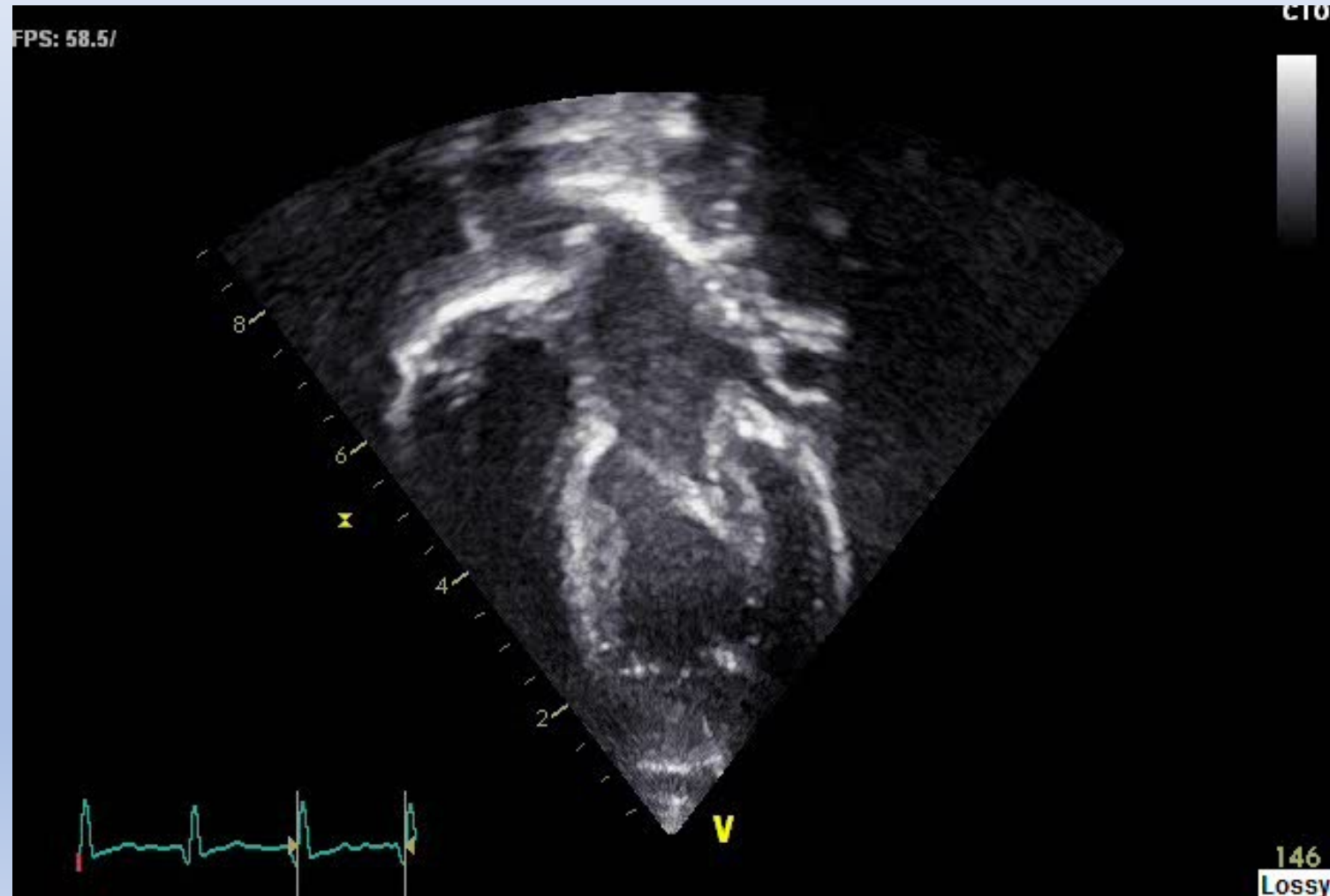
What if the LV Function is Terrible?



Balloon Aortic Valvuloplasty

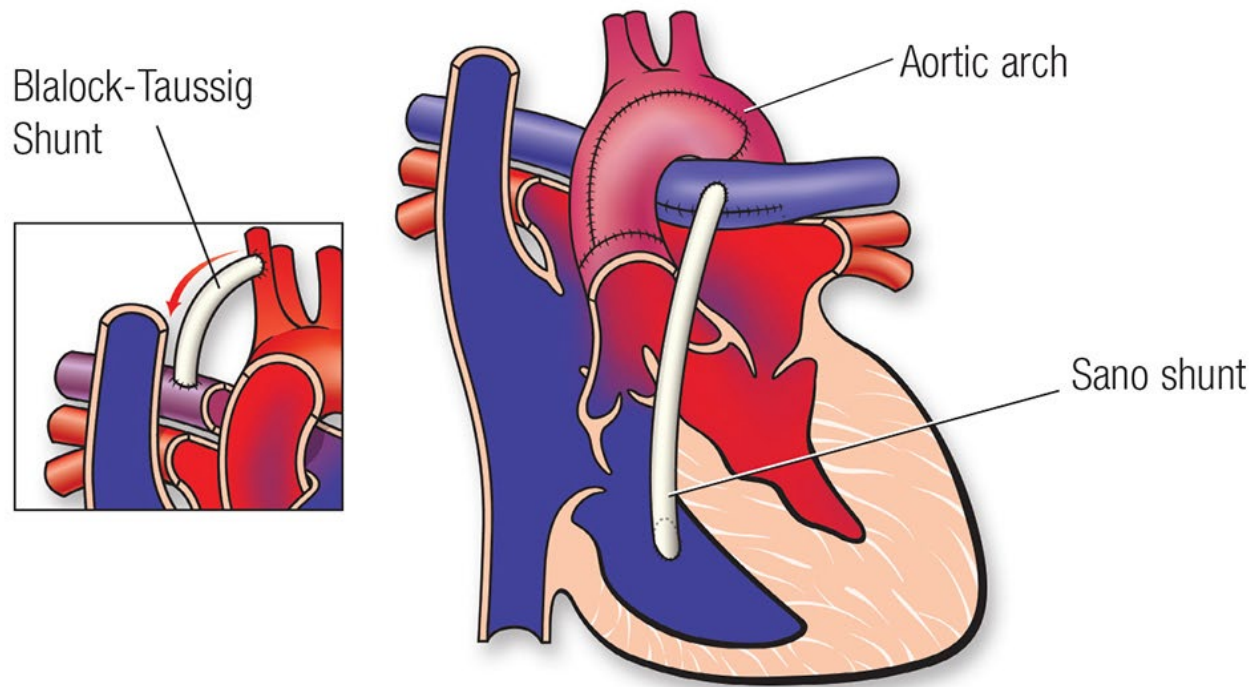


Critical AS infant post balloon -Follow-up at 3 months



HLHS Palliation - Norwood Operation

Aortic Arch Reconstruction



- Principles:

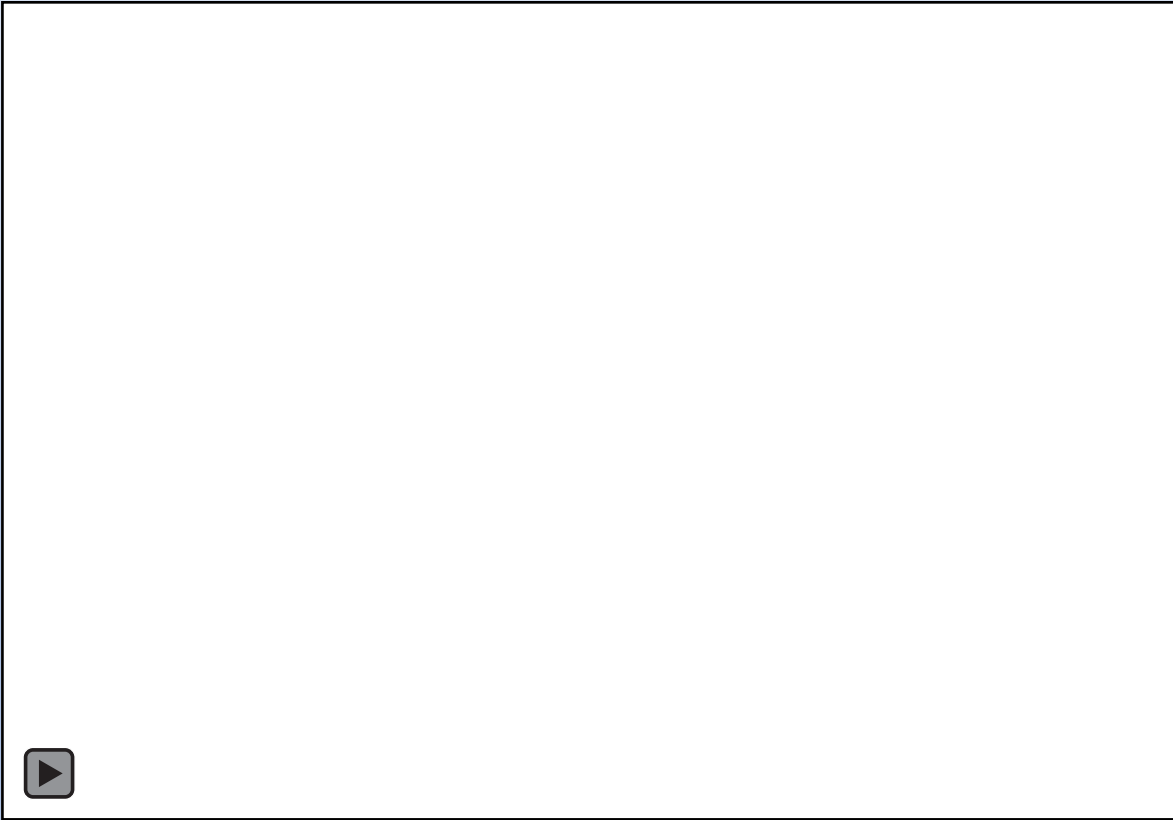
- Unobstructed pulmonary venous return/mixing (atrial septectomy)
- Unobstructed systemic flow (DKS-Norwood anastomosis of Aorta and PA to create neo-aorta, arch repair)
- Appropriate restriction to pulmonary blood flow (5 mm Sano RV-PA conduit)

Source: American Heart Association

<https://www.heart.org/en/health-topics/congenital-heart-defects/about-congenital-heart-defects/single-ventricle-defects>

HLHS with severely restrictive atrial septum is a medical emergency with very high mortality and high long term morbidity

Post Norwood



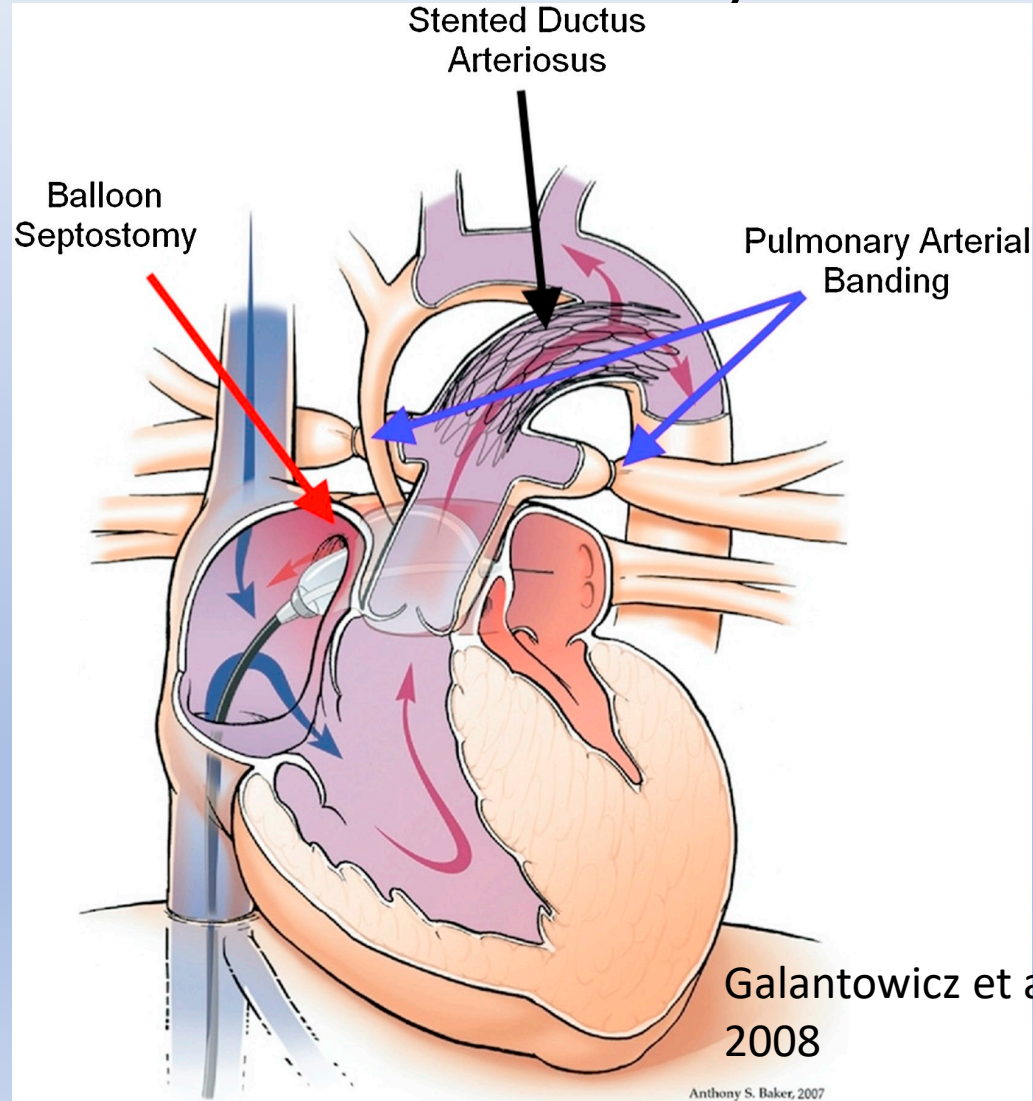
Interstage Monitoring at home

- Family checks daily weight at home
- Family checks pulse ox at home
- Enter data into proprietary program available for review to medical team
- Documented to decrease interstage mortality to under 5% nationwide

Hybrid Palliation for HLHS (reserved for high risk patients at our center)

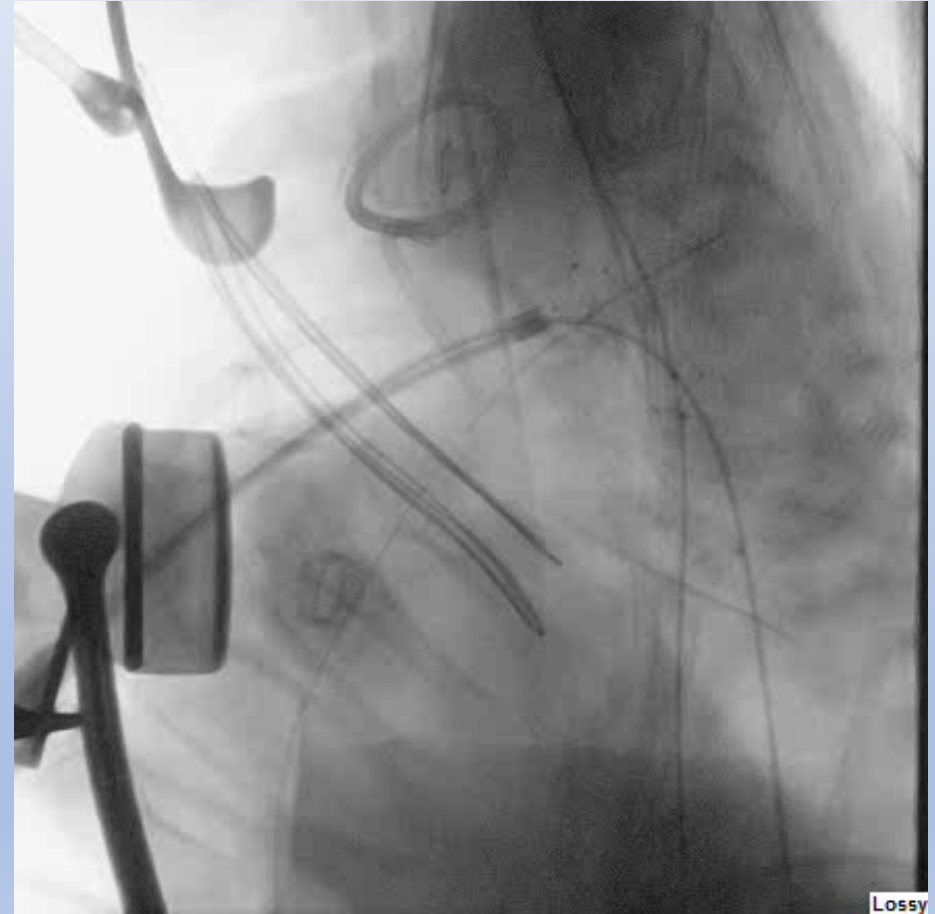
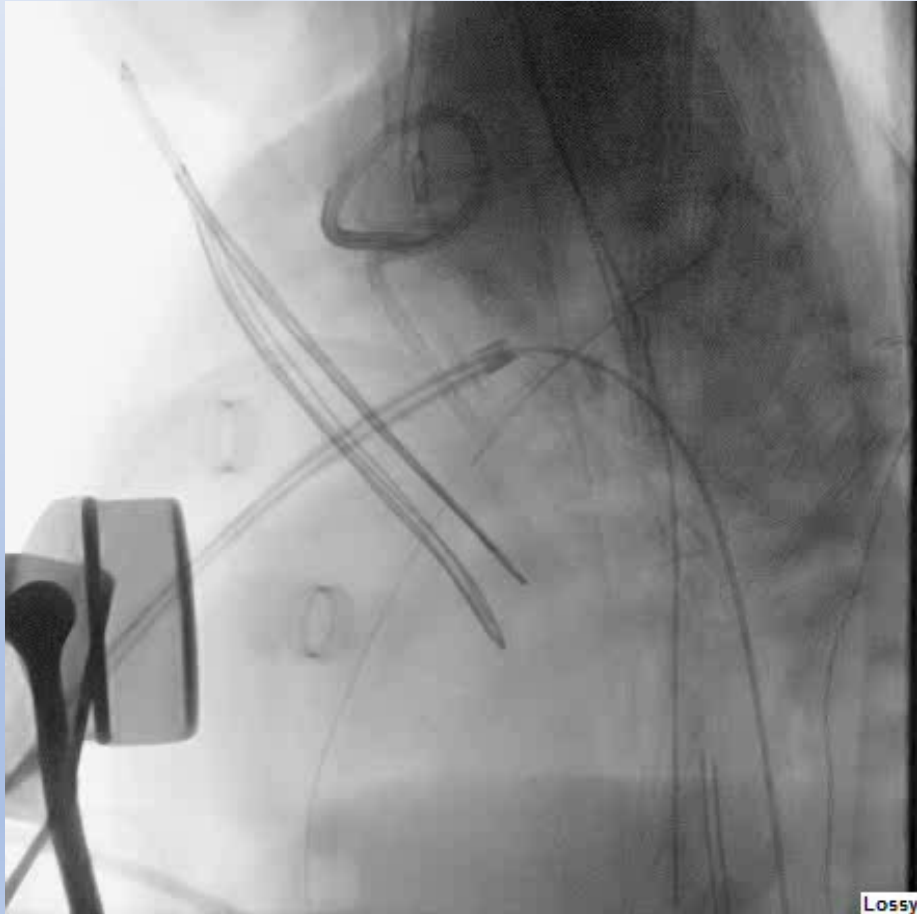
High Risk Features:

- Significant genetic abnormalities
- Significant extracardiac abnormality
- Moderate or worse RV dysfunction
- Moderate or worse TR



Advantage: Can be performed without need for CP bypass

Hybrid Palliation – PDA stent implantation



Conclusions

- Identify clinical features associated with left heart obstructive lesions in newborns
- Manage critical left heart obstruction in the neonate
- Describe the cardiac procedural treatment of the neonate with left heart obstruction

Thank You!