CONGENITAL HEART DISEASE

Kriti Puri, MD

Hugh D. Allen, MD

Athar M. Qureshi, MD

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INTRODUCTION

- Congenital heart disease (CHD) - about 9 out of every 1,000 live-born children
- PRESENTING FEATURES: cyanosis, congestive heart failure (CHF), shock or poor pedal pulses, failed newborn CHD pulse oximetry screen
- PRECIPITATING FACTORS: closure of the patent ductus arteriosus, dropping of pulmonary vascular resistance

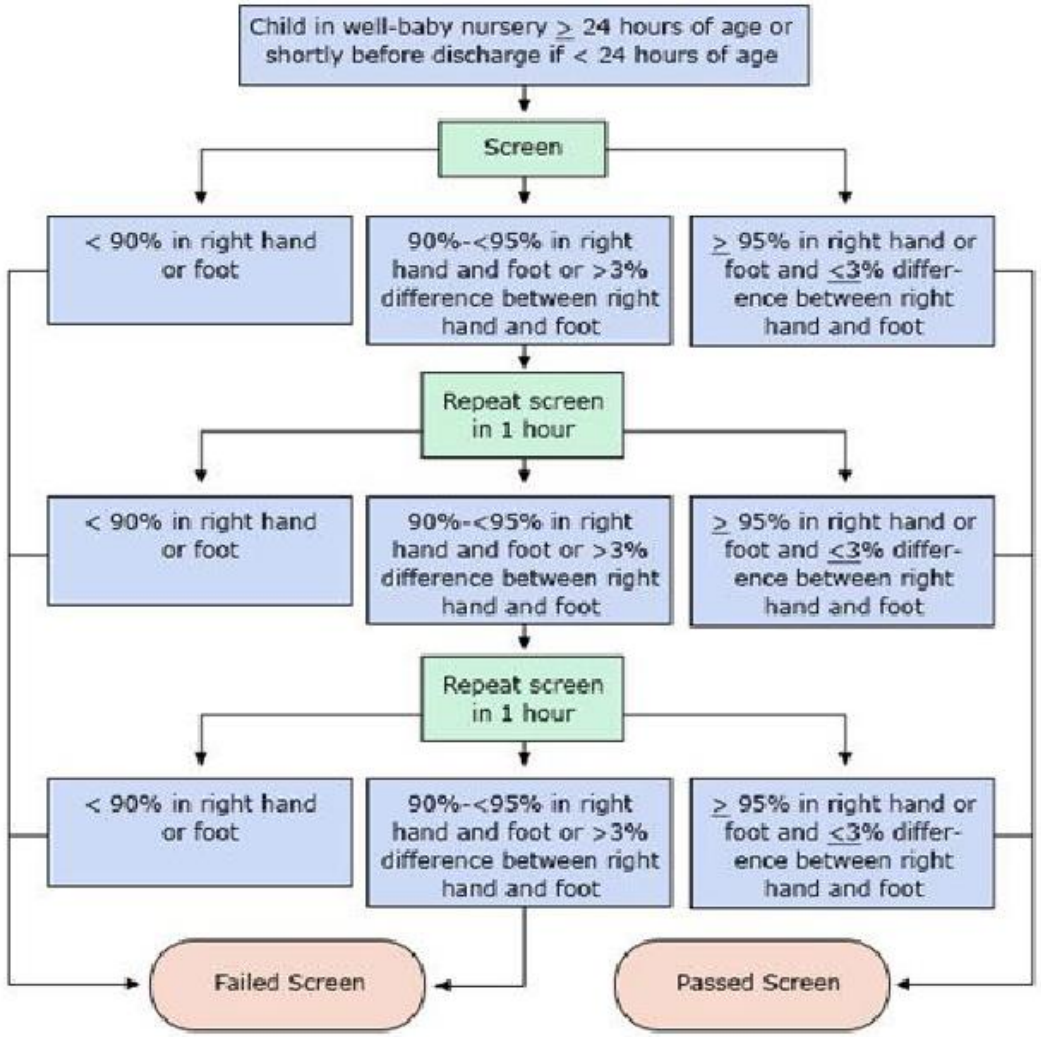
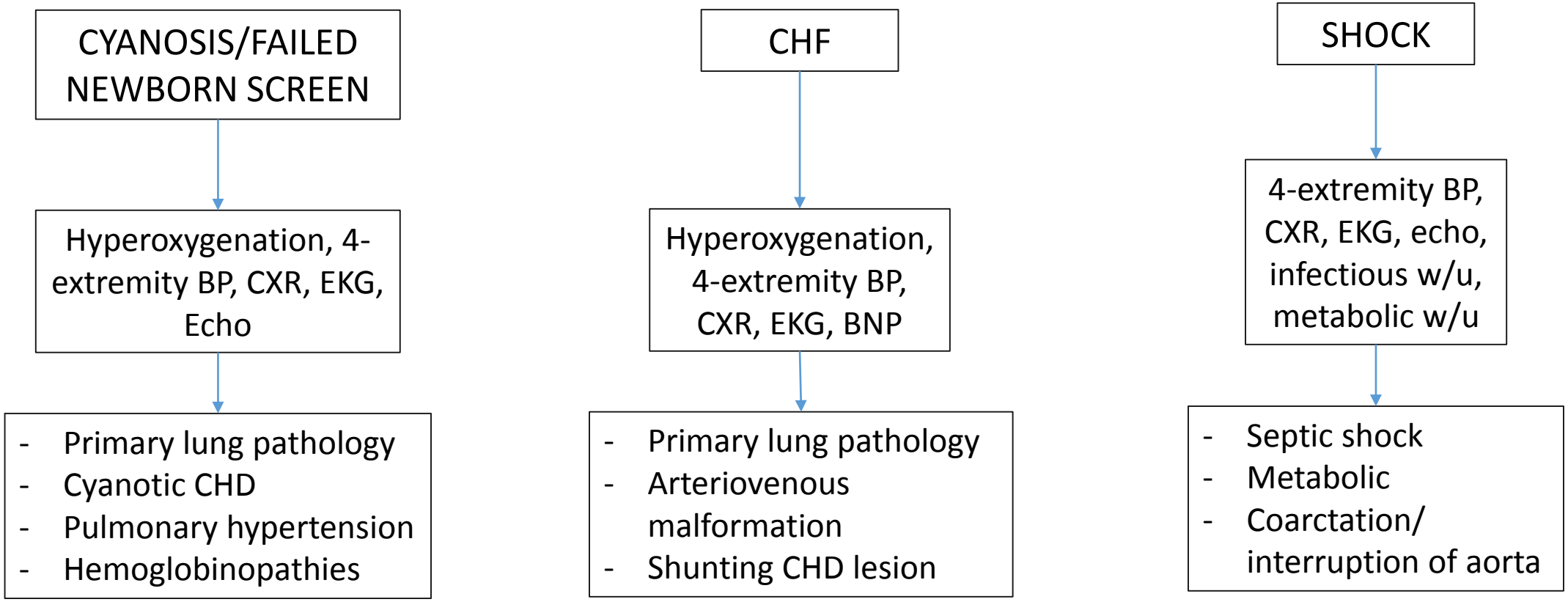


FIGURE 1. PROTOCOL FOR PULSE OXIMETRY SCREENING FOR NEWBORNS FOR CRITICAL CHD

Mahle WT, et al. Pediatrics. 2009;124(2):823–883

NEWBORN PRESENTATIONS OF CHD



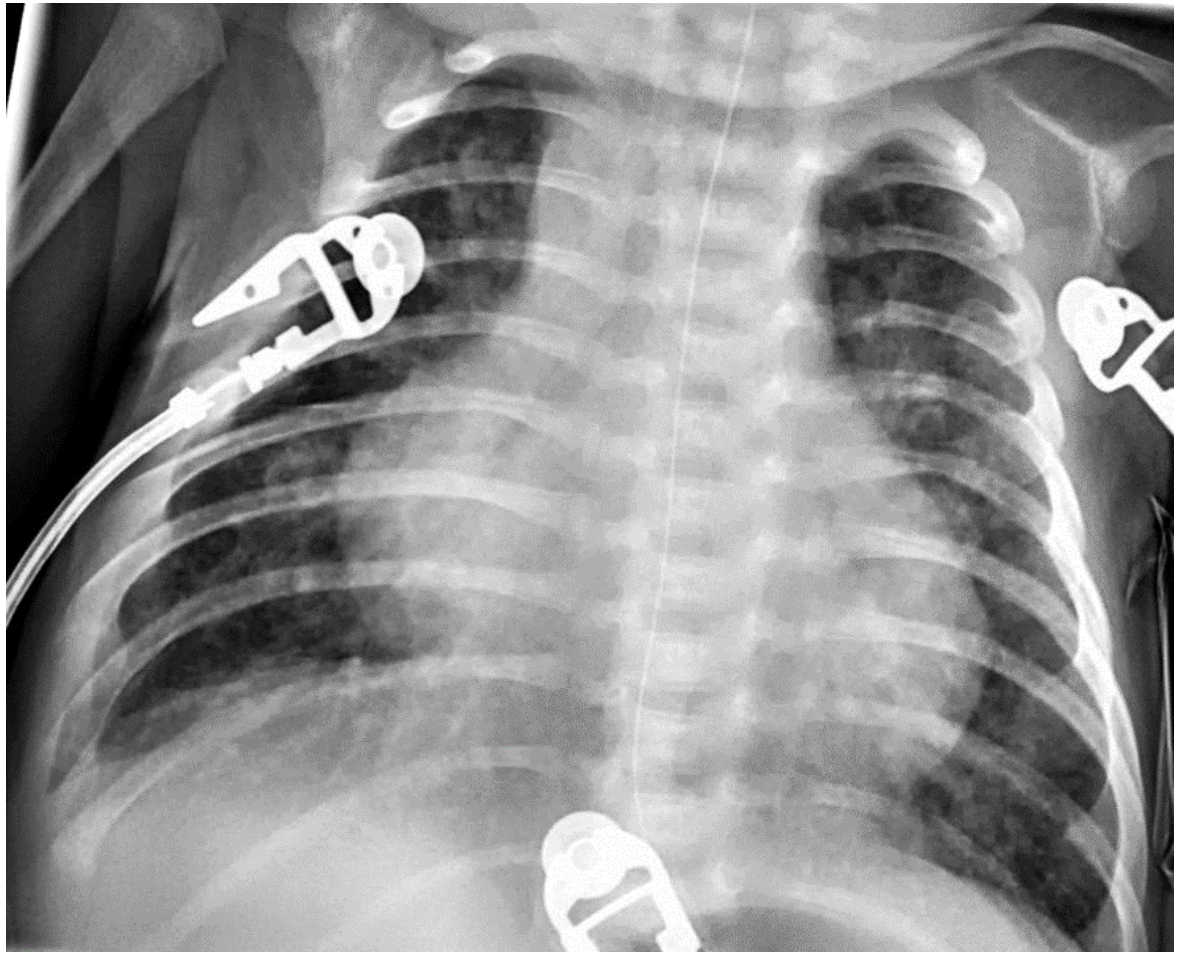


FIGURE 2. CHEST RADIOGRAPH OF A PATIENT SHOWING FEATURES OF CONGESTIVE HEART FAILURE
This chest radiograph of a patient with a VSD shows cardiomegaly, increased pulmonary vascular congestion, and splaying of the bronchi, which are indicative of left atrial enlargement.

ATRIAL SEPTAL DEFECTS

- Left-to-right shunt at ASD -> greater diastolic volume in RV -> right-sided chamber dilation
- EXAM - Systolic murmur over pulmonic region, fixed split S2, left precordial bulge; usually not symptomatic with CHF
- TESTING - Serial echo for RA/RV dilation –> indication for intervention
- MANAGEMENT – Device closure in catheterization laboratory or surgical closure

VENTRICULAR SEPTAL DEFECTS

- Most common CHD – 50%-60%; most common type is perimembranous
- EXAM – newborn with high pulmonary pressures may not have a murmur; older child - S1-coincident pansystolic murmur loudest over the left lower sternal border; diastolic rumble at the apex; other features of CHF
- Perimembranous VSDs can get occluded by aneurysmal tissue, and muscular VSDs become smaller in size with muscular growth
- MANAGEMENT – diuretics, fortified formula, surgical patch repair, device closure in the catheterization lab for muscular VSDs

ATRIOVENTRICULAR SEPTAL DEFECTS

- Also known as *atrioventricular canal defects;* about 50% of patients have Down syndrome
- EXAM - systolic ejection or holosystolic murmur, no murmur if large VSD or pulmonary hypertension, murmur of AV valve regurgitation; features of CHF
- TESTING – EKG with northwest-axis or right-axis deviation; left, right, or bi-ventricular hypertrophy; CXR with cardiomegaly
- MANAGEMENT – diuretics, formula fortification, repair at age 4-8 months; 10%-15% may need repeat intervention within 5 years

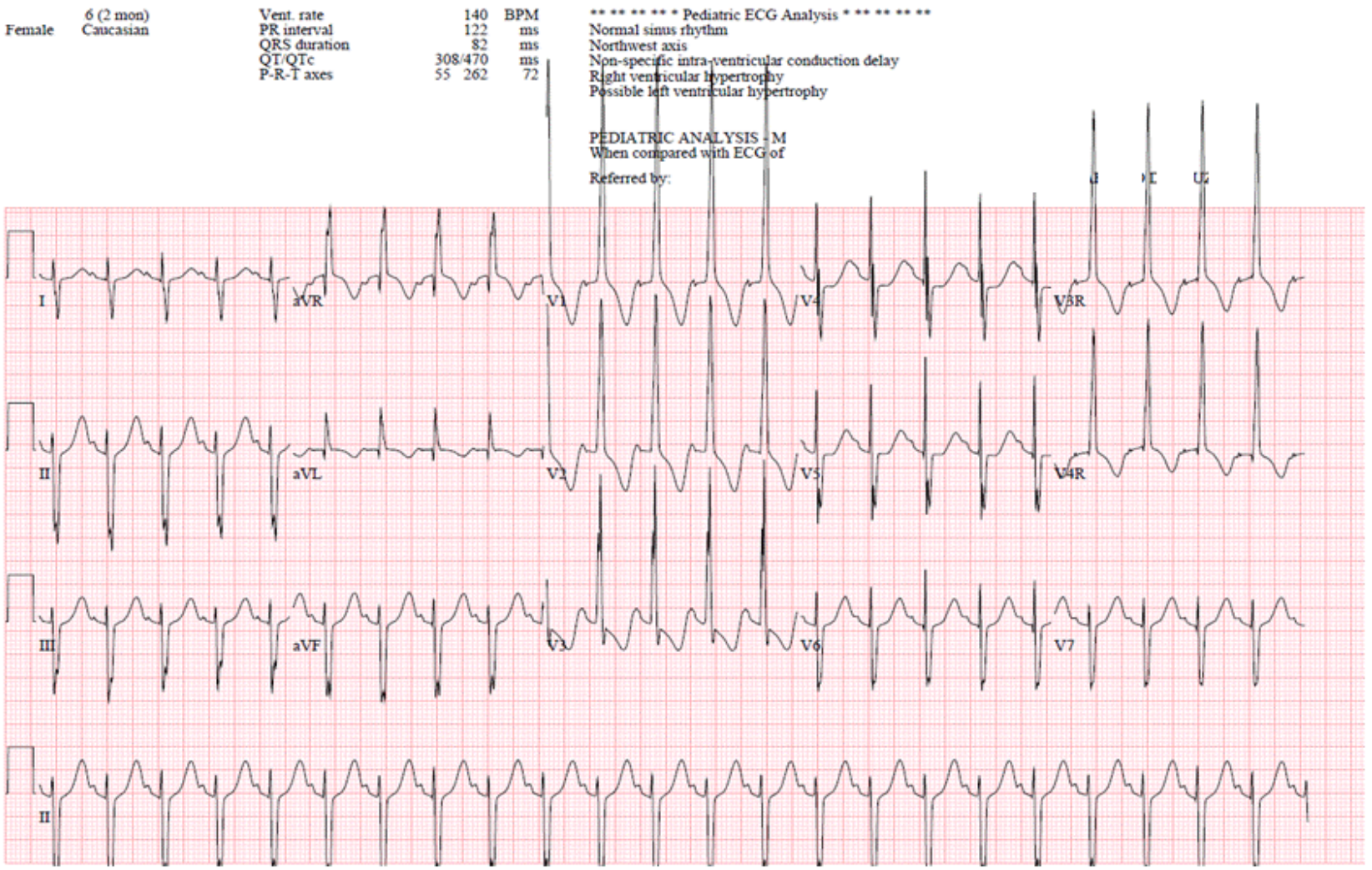


FIGURE 3. EKG OF A PATIENT WITH ATRIOVENTRICULAR SEPTAL DEFECT

This EKG shows the biventricular hypertrophy, as well as the northwest-axis deviation, with negative QRS axes in leads I and aVF.

PATENT DUCTUS ARTERIOSUS

- Typically closes functionally within 24 hours and anatomically within 3-4 weeks in most cases
- EXAM - a loud systolic or continuous murmur of blood shunting left to right during the entire cardiac cycle, loudest over the left precordium; diastolic rumble of transmitral flow into an enlarged LV; features of CHF on exam and CXR
- MANAGEMENT – ibuprofen or indomethacin, catheterization laboratory closure (as small as 700 g), surgical ligation

AORTIC VALVE STENOSIS

- Categorized as **critical** if the blood flow through the valve is insufficient, and needs additional contribution from the PDA
- EXAM - ejection systolic murmur, loudest over the aortic region, ejection click (if stenosis is moderate or mild); EKG shows left ventricular hypertrophy
- MANAGEMENT - critical AS – catheter-based vs surgical valvuloplasty in the newborn period to get off PGE
- Timing of reintervention determined by severity of the gradient across the aortic valve

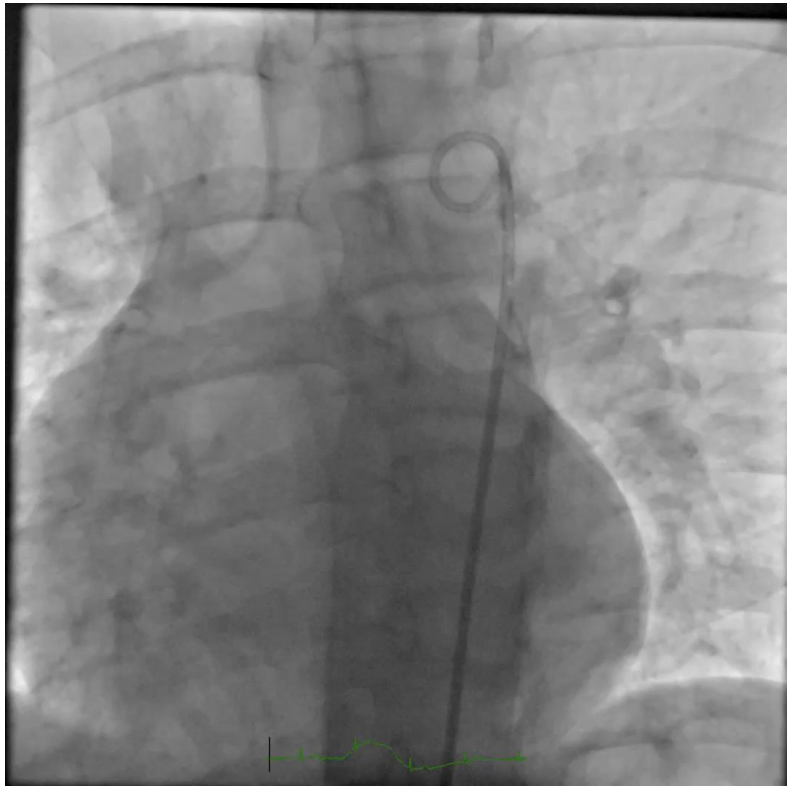
PULMONIC VALVE STENOSIS

- Categorized as **critical** if the blood flow to the lungs through the pulmonary valve is insufficient and needs additional contribution from the PDA
- EXAM - ejection systolic murmur, loudest over the pulmonic region, ejection click (if stenosis is moderate or mild); EKG – right ventricular hypertrophy
- MANAGEMENT - critical PS – catheter-based balloon valvuloplasty procedure in the newborn period to get off PGE
- Timing of reintervention determined by severity of the gradient across the pulmonary valve

COARCTATION OF AORTA

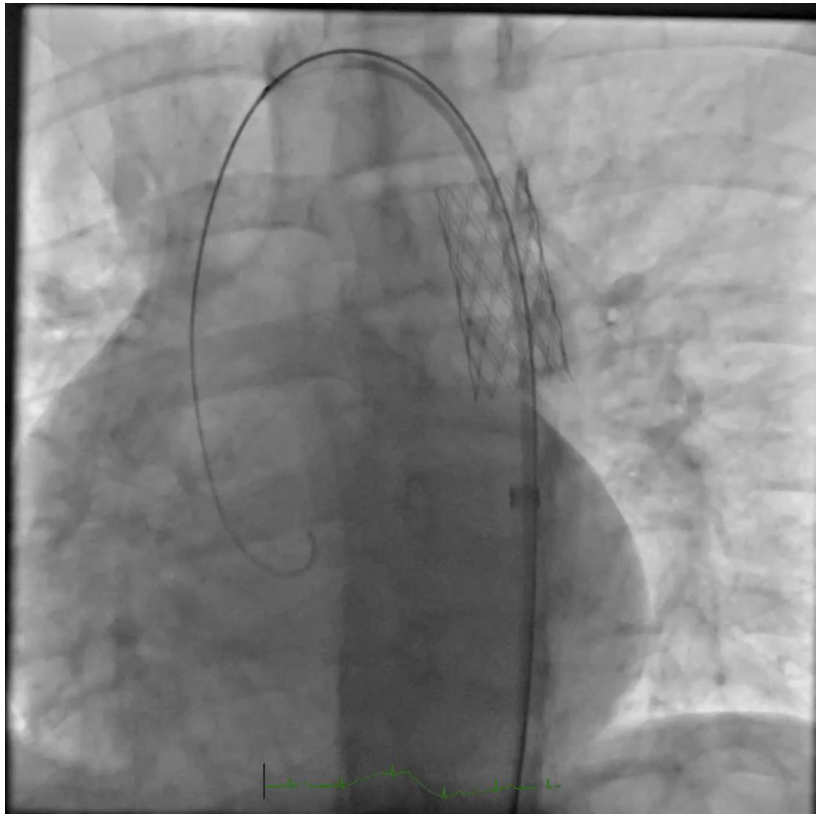
- EXAM - poor pedal or femoral pulses, shock, failed newborn screen, gradient >20 mm Hg between the upper- and lower-extremity BP; harsh systolic murmur over the back; if collaterals develop, may hear continuous murmur over the chest wall
- MANAGEMENT - surgery, balloon angioplasty, and stent placement for the coarctation in older children; close follow-up for re-coarctation and hypertension

VIDEO 1. ANGIOGRAPHY - COARCTATION OF THE AORTA



An aortogram in a teenager with coarctation of the aorta. We see narrowing of the descending aorta.

VIDEO 2. ANGIOGRAPHY - COARCTATION OF THE AORTA AFTER ANGIOPLASTY AND STENT PLACEMENT



An aortogram in the same patient after treatment with a stent in the cardiac catheterization laboratory. We see that the narrowing is abolished with placement of a stent.

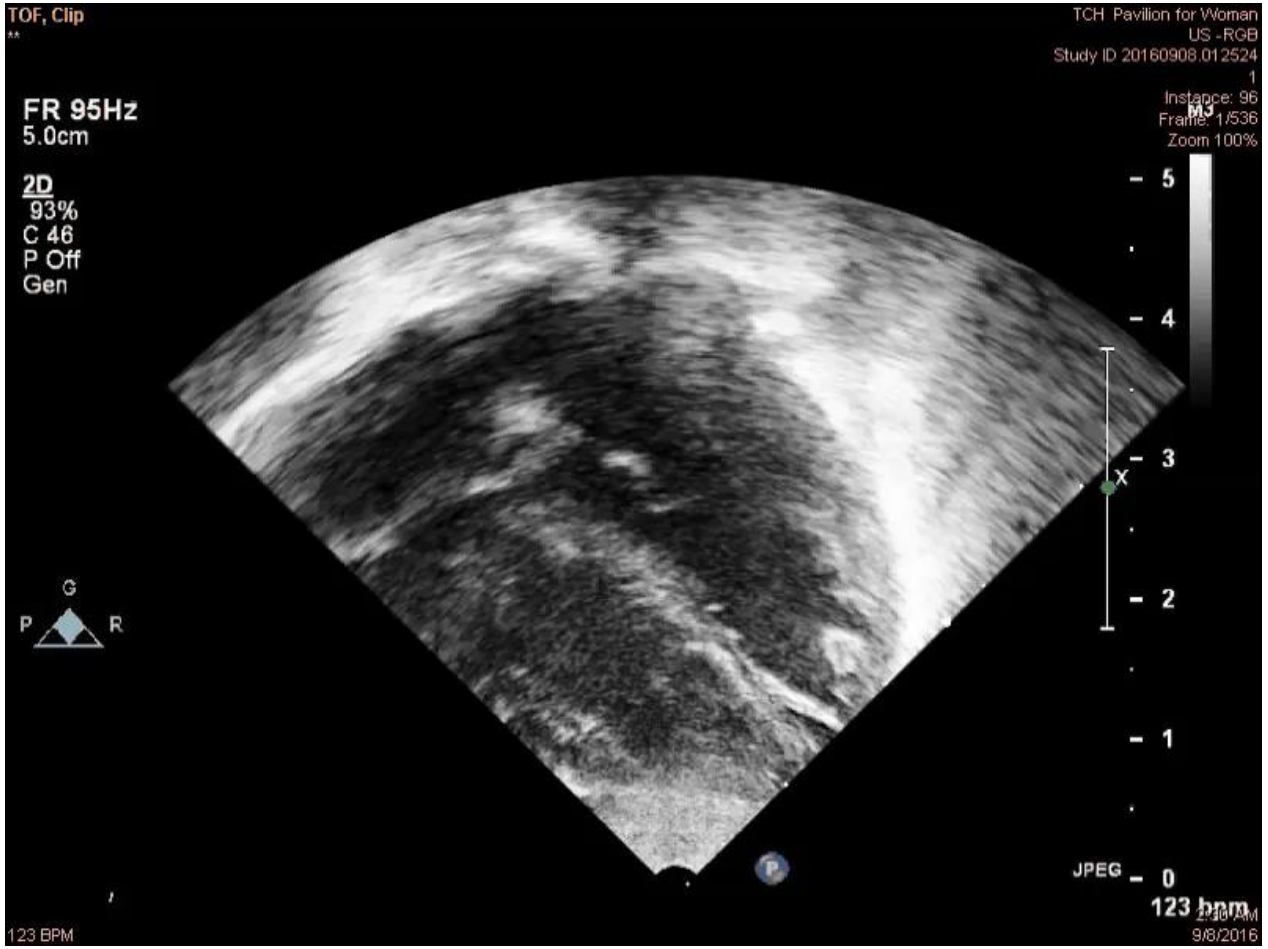
INTERRUPTION OF THE AORTA

- Discontinuation of the arch and distal continuation through a PDA past the point of interruption
  - Type A - interrupted at the isthmus distal to the left subclavian artery
  - Type B - interrupted between the origins of the left subclavian and left common carotid arteries (most common and frequently associated with chromosome 22 abnormalities)
  - Type C - interrupted between the origins of the innominate and the left common carotid arteries (most rare)
- MANAGEMENT – surgery as a neonate; close follow-up for re-coarctation

TETRALOGY OF FALLOT

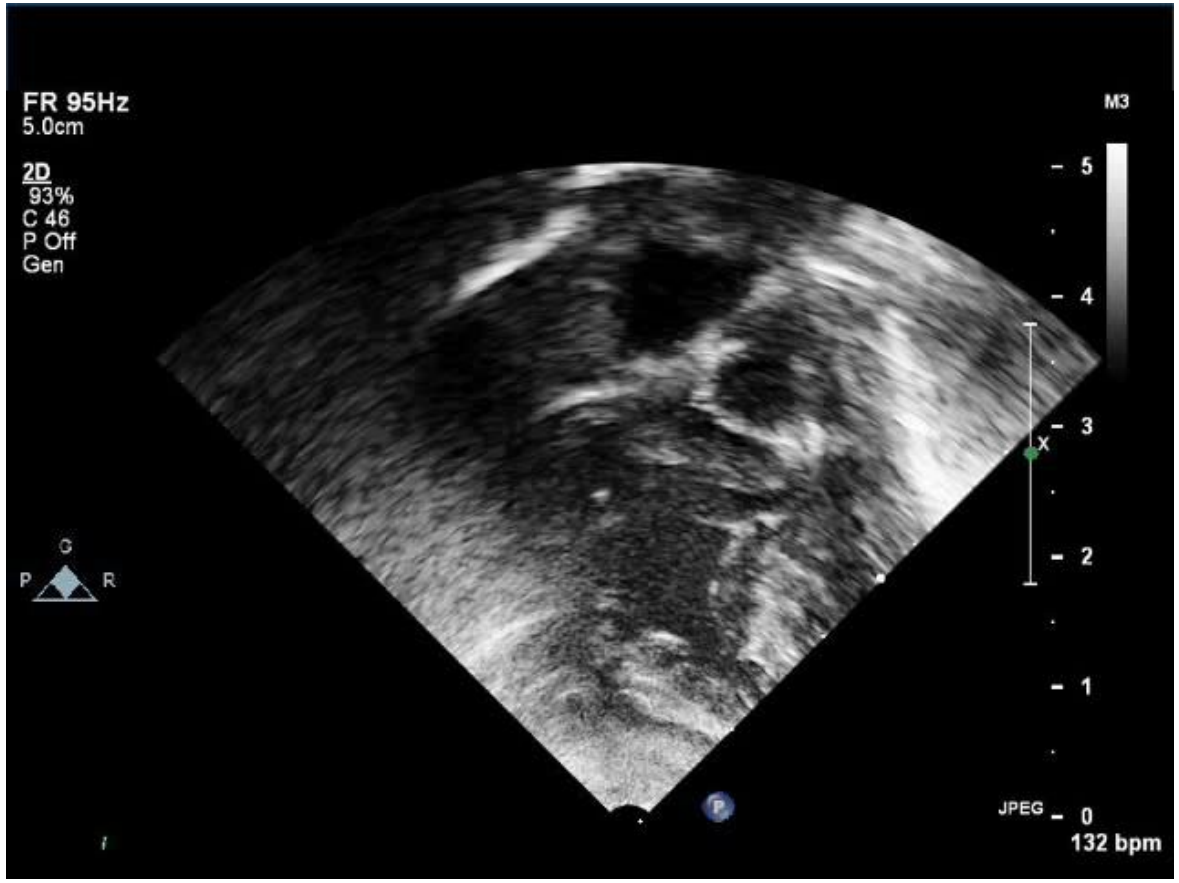
- Most common cyanotic CHD – 5% of CHD
- Anterior malalignment of the interventricular septum, leading to
    1. VSD
    2. Overriding of the VSD by the aorta
    3. Narrowing of the pulmonary outflow tract, causing RV outflow (infundibular) obstruction
    4. Consequent RV hypertrophy
- EXAM - ejection systolic murmur over the pulmonic area; single second sound; “tet spell” during agitation or fever or other concurrent illness
- MANAGEMENT – palliation with modified Blalock-Taussig-Thomas (mBTT) shunt, complete repair with VSD closure and resection of RV outflow obstruction

VIDEO 3. ECHOCARDIOGRAM - TETRALOGY OF FALLOT



Four-chamber view of the heart, with the left side of the patient on the reader's right. We see the VSD and the right ventricular hypertrophy. We can discern the aorta arising from the left ventricle overriding the VSD. As the video plays, it sweeps through the heart anteriorly. We see the pulmonary artery, which appears narrow. We also see the hypertrophied muscle bundles of the right ventricle, especially in the subvalvar region, which come very close together in systole. This creates a setup for dynamic obstruction to blood flow out of the right ventricle.

VIDEO 4. ECHOCARDIOGRAM - TETRALOGY OF FALLOT



Four-chamber view shows the doming, narrow pulmonary valve and infundibular narrowing from muscle below the pulmonary valve in the right ventricular outflow tract.

“TET SPELLS”

- Acute episode of desaturation and decrease in pulmonary blood flow
- EXAM - cyanosis, pulmonic murmur is softer (due to decreased pulmonary blood flow)
- MANAGEMENT - combination of heart rate control and vascular resistance manipulation
  - Calm the patient
  - Knee-chest position (to increase systemic vascular resistance)
  - Initiate oxygen
  - IV fluid bolus
  - IV metoprolol (to slow down the heart rate) and IV phenylephrine (to increase systemic vascular resistance)

TRANSPOSITION OF THE GREAT ARTERIES

- Second most common cyanotic CHD – most common presenting in the first week of life
- EXAM – cyanosis within 12 hrs of life; narrow mediastinal silhouette on CXR (due to the orientation of the great vessels and thymic regression); EKG normal or RV hypertrophy; usually no murmur; single S2 with a loud aortic component (due to orientation of the great vessels)
- MANAGEMENT – Surgery within the neonatal period – arterial switch procedure

TRUNCUS ARTERIOSUS

- 2%-5% of CHD
- EXAM – systolic murmur heard loudest over the left sternal border, as well as a loud S2; CHF features in the first 48 hrs of life; one-third have 22q11.2 deletion
- MANAGEMENT – temporize with diuretics, surgery as a neonate
- Surgery - Truncal outflow and truncal valve committed to the aorta and branch pulmonary arteries committed to a conduit placed from the RV to the PA; *expect normal saturation levels after repair*
- Close cardiology follow-up – may outgrow their RV to PA conduit or the truncal valve (now their aortic valve) may become regurgitant

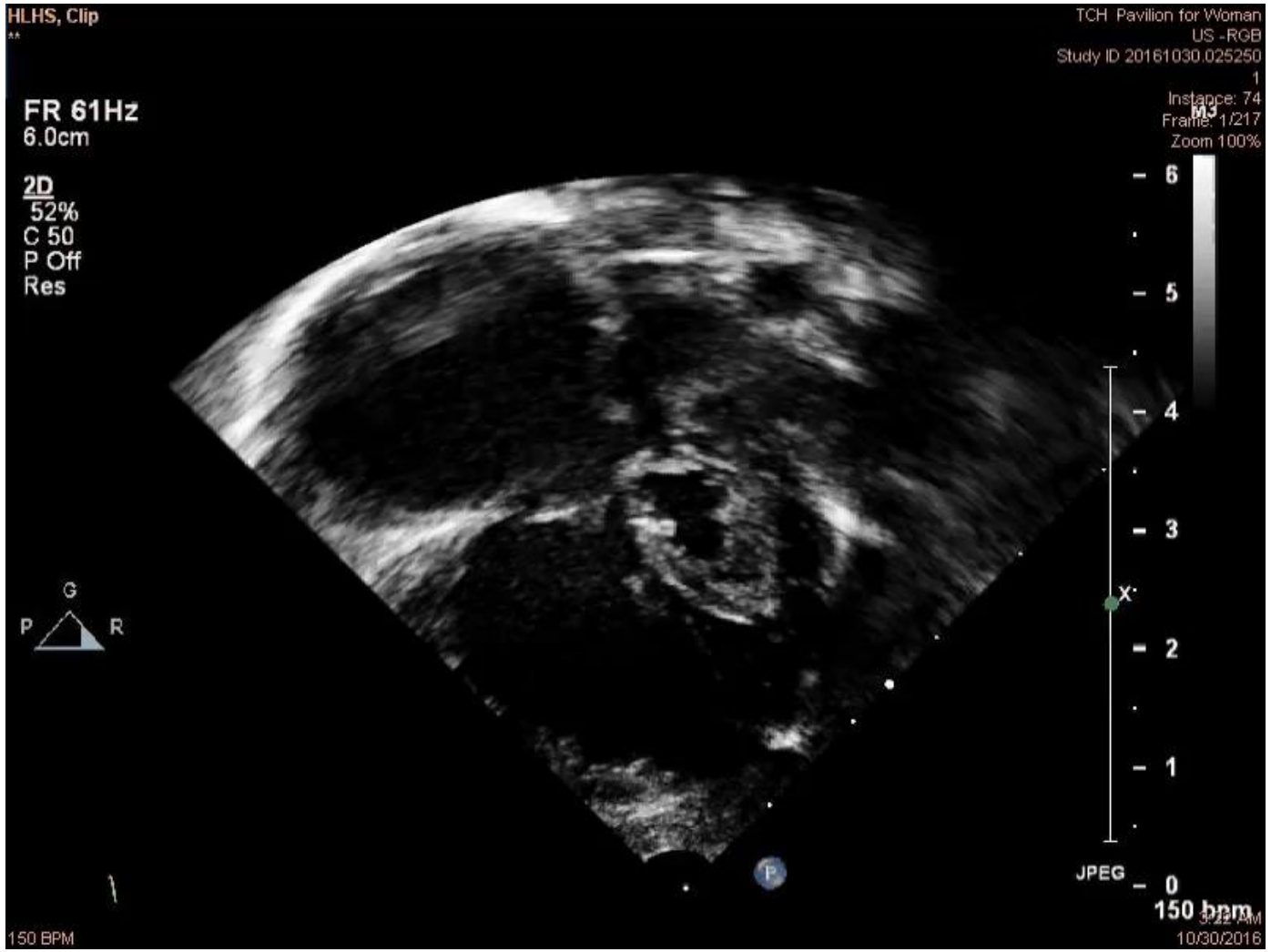
TOTAL AND PARTIAL ANOMALOUS PULMONARY VENOUS DRAINAGE (TAPVD, PAPVD)

- "Partial" when at least 1 pulmonary vein returns to the LA and "total" when none of the pulmonary veins return to the LA
- Obstructed - pulmonary venous return impeded, leading to pulmonary venous hypertension and pulmonary edema
- EXAM - respiratory distress and cyanosis within 12-24 hours of life; CXR finding of "whiteout" of the lung fields
- PAPVD (may or may not be associated with an ASD) - later in life in childhood with signs similar to that of an ASD
- MANAGEMENT – surgery – emergency for obstructed veins, elective for unobstructed veins; follow closely for recurrence of obstruction

HYPOPLASTIC LEFT HEART SYNDROME (HLHS)

- Severe stenosis or atresia of the mitral and aortic valves; hypoplastic LV and ascending aorta
- EXAM – ductal-dependent for systemic flow; signs of shock and CHF when duct starts to close; CXR with right ventriculomegaly
- Corresponding lesion with small right-sided structures – tricuspid stenosis/atresia with pulmonary stenosis or pulmonary atresia; pulmonary atresia with intact ventricular septum – ductal dependent for pulmonary blood flow
- MANAGEMENT – single-ventricle palliative pathway; watch out for features of CHF

VIDEO 5. ECHOCARDIOGRAM - HLHS



Four-chamber view of the heart, with the left side of the patient on the reader's right. We can discern the diminutive left-sided chambers, with the hypoplastic left atrium and the hypoplastic left ventricle, and compensatory dilation and hypertrophy of the right ventricle.

SINGLE-VENTRICLE PATHWAY

- Stage I - stabilize both cardiac outputs, either by surgically constructing a “neo-aorta” by using the main PA tissue to supplement the ascending aorta, creating an mBTT shunt (or RV to PA conduit) and creating an ASD
- Stage II - Glenn procedure – SVC anastamosed to the pulmonary arterial system to direct part of the deoxygenated venous blood directly to the lungs – expect saturation levels in the 80s
- Stage III – Fontan procedure – IVC anastamosed to the pulmonary arterial system – all deoxygenated blood goes to the lungs, so no mixing – normal saturation levels

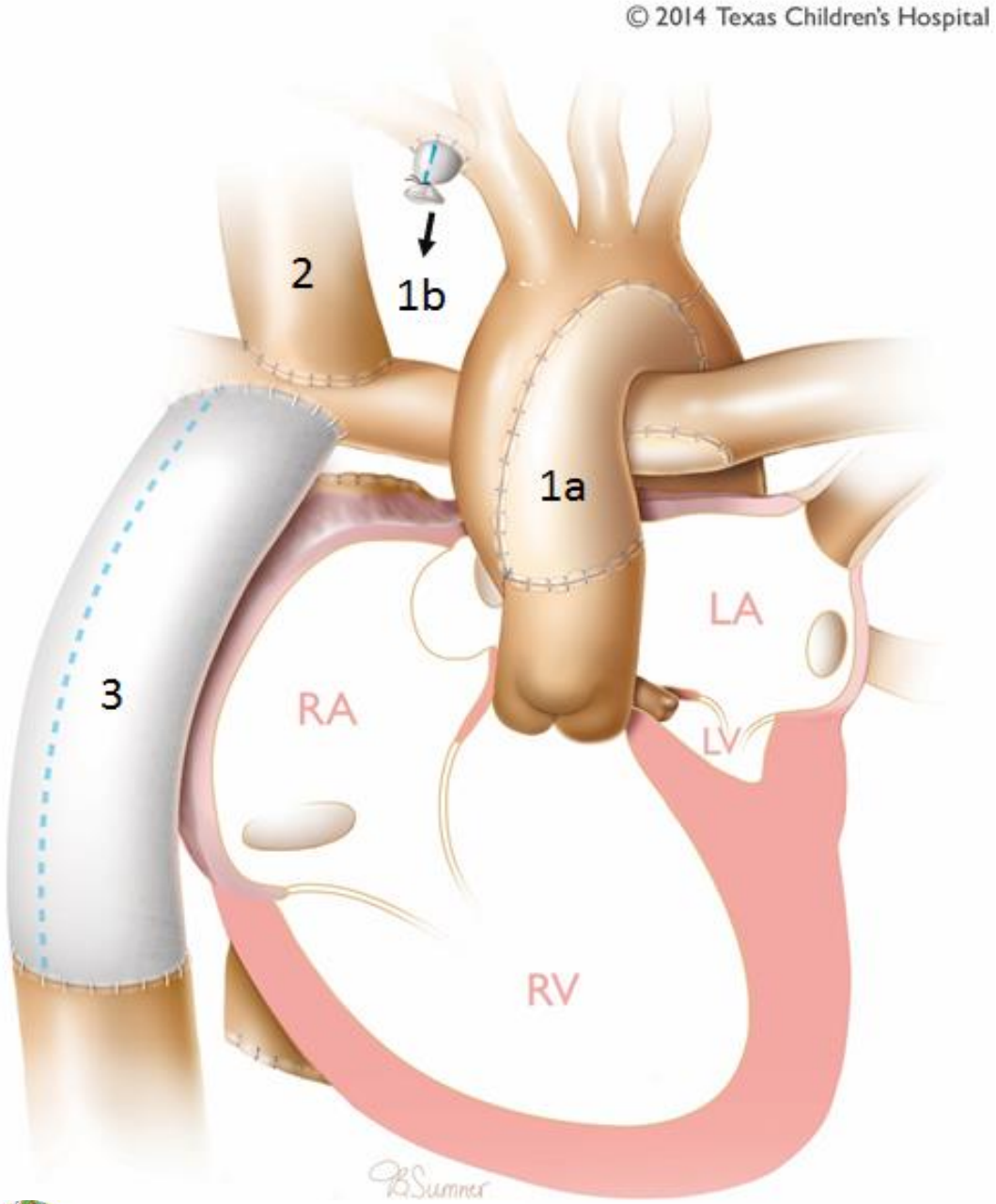


FIGURE 4. SCHEMATIC DIAGRAM SHOWING THE SHUNTS AND SITES OF ANATSAMOSES FOR THE SINGLE-VENTRICLE PALLIATION PATHWAY

1a - reconstructed the neoaorta by using the hypoplastic ascending aorta, as well as the normal main pulmonary artery, to ensure stable systemic outflow.

1b - site of the modified Blalock-Taussig-Thomas shunt to direct blood flow from the right subclavian artery into the pulmonary circulation.

2 - Glenn anastomosis between the SVC and the right pulmonary artery and mBTT shunt takedown

3 - Fontan anastomosis connects the remaining deoxygenated blood coming in through the IVC through the Fontan conduit to the pulmonary circulation.

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IMMUNIZATIONS

- No live vaccines 2 weeks before or 6 weeks after cardiopulmonary bypass (CPB)
- Most children should receive influenza vaccine
- RSV PROPHYLAXIS WITH PALIVIZUMAB - age <12 months with unrepaired cyanotic CHD or those receiving diuretic therapy for CHF in acyanotic CHD
- Children who meet the criteria for RSV prophylaxis after CPB surgery – consider a postoperative dose of 15 mg/kg of palivizumab

SUBACUTE BACTERIAL ENDOCARDITIS

- Six months after cardiac surgery or interventional catheterization procedures to repair a lesion or occlude a defect
- Residual defects around a patch or device
- Unrepaired cyanotic CHD
- Prosthetic or bioprosthetic valves or conduits implanted percutaneously or surgically
- Prior history of endocarditis or valvular heart disease in the context of heart transplantation

POSTOPERATIVE CARE AND TRANSITION

- STERNAL PRECAUTIONS – for 6-8 weeks, no “tummy time,” close supervision needed in crowded places, avoid picking up or lifting the child up by the arms
- Age-appropriate car seat
- POSTCATHETERIZATION – back to full activities within 3-5 days
- NEURODEVELOPMENTAL OUTCOMES – developmental pediatrician referral for high risk - including open heart surgery/cyanotic CHD or those with prematurity/known genetic syndrome/CPR
- TRANSITION – by a cardiologist to an adult congenital specialist