

What does a cardiac surgeon want to know from you?

**An interpretation guide for imaging prior to
congenital cardiac surgery.**

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No financial disclosures.



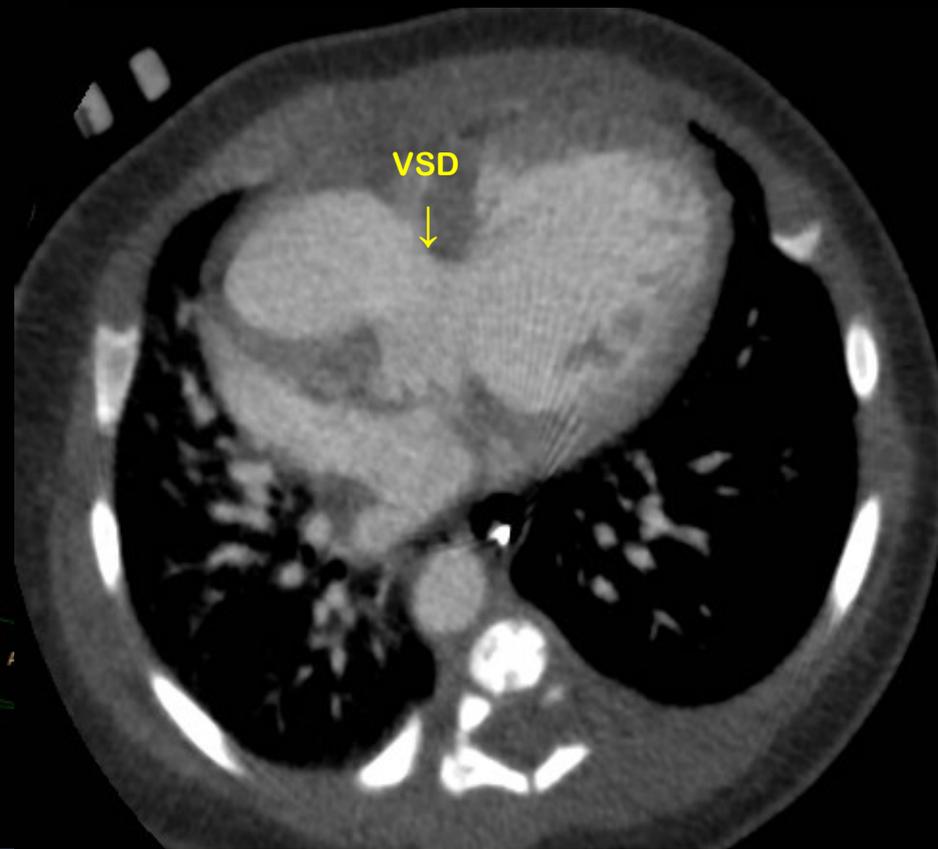
Objectives

- ✦ Describe 4 congenital heart diseases (CHD) and the type of cardiac surgery required for repair.
- ✦ Review the necessary parameters and measurements for the imaging report.
- ✦ Define the concurrent abnormalities to assess for, as well as imaging pitfalls to beware of.
- ✦ Address the key imaging questions asked by cardiac surgeons prior to repair.

Transposition of the Great Vessels (TGA)

- ✦ **D-TGA:** Aorta is connected to and originates above the right ventricle (RV), and pulmonary artery (PA) is connected to the left ventricle (LV).
 - ✦ 2nd most common cause of cyanosis from CHD.
- ✦ **L-TGA:** congenitally corrected form of TGA in which the great vessels AND the ventricles are switched, so normal blood flow is maintained.

D-Transposition of the Great Vessels



Aorta arises from RV

PA arises from LV

Surgical repair of D-TGA

- ✦ **Anatomic Correction/Arterial Switch: Jatene or Rastelli**
 - ✦ **Jatene: switch great arteries, transfer coronary arteries, repair VSD.**
 - ✦ **Rastelli: used in setting of pulmonic stenosis and ventricular septal defect (VSD)**
 - ✦ **RV to PA baffle (to bypass pulmonic stenosis) and LV to aorta baffle (via VSD)**
- ✦ **Physiologic Correction/Atrial Switch: Mustard or Senning, now antiquated procedures.**
 - ✦ **An intra-atrial baffle redirects oxygenated pulmonary vein (PV) blood to RV and deoxygenated PA blood to LV.**
 - ✦ **Disadvantage: requires RV to pump systemic blood; long term complications.**

D-TGA Concurrent Abnormalities

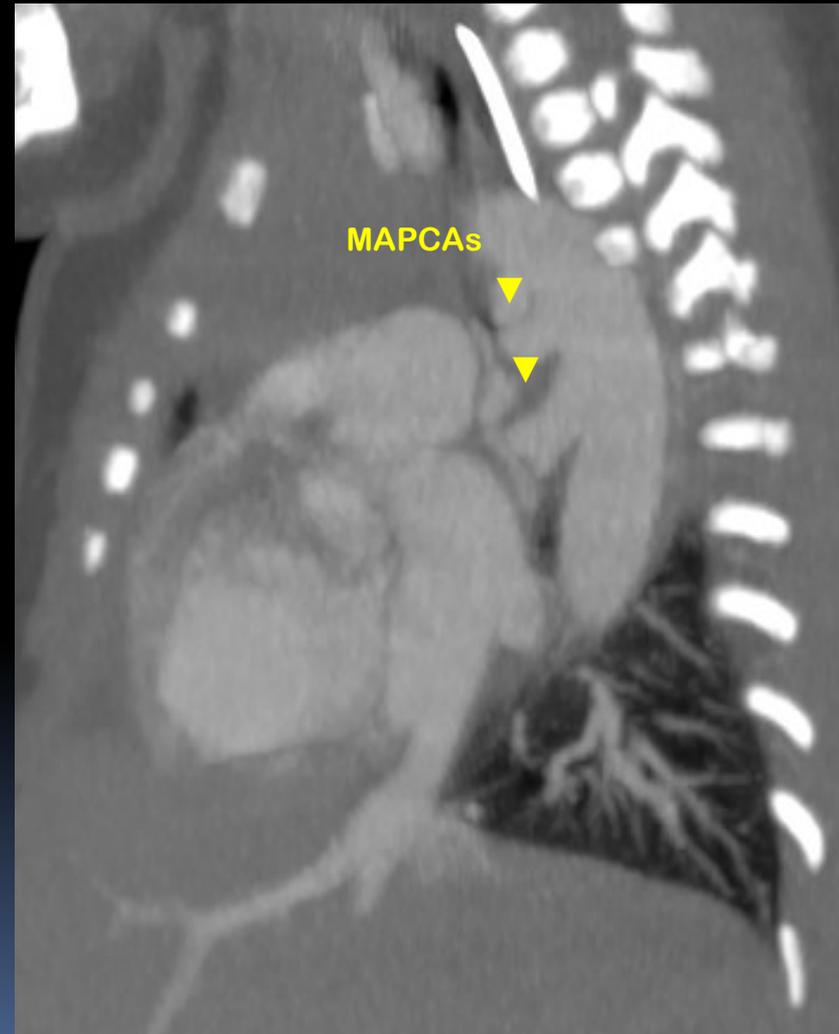
- ★ Systemic and pulmonary circulations are completely separate, so mixing/shunting must occur to sustain life.
- ★ Assess for intracardiac or extracardiac shunts as well as amount of mixing.
 - ★ VSD is present in up to 45% of infants with D-TGA.
- ★ If VSD is not present, then mixing **MUST** occur by patent ductus arteriosus (PDA), patent foramen ovale (PFO), or secundum atrial septal defect (ASD).
PFO alone is usually inadequate for mixing of blood, so assess patency of PDA.

D-TGA Concurrent Abnormalities

- ✦ 25% of patients have some form of pulmonic stenosis, so must assess patency of pulmonic valve.
 - ✦ Most often occurs in subpulmonic region.
 - ✦ If no VSD present, then “dynamic only” pulmonic stenosis may occur during systole from anterior motion of the mitral valve leaflet.
- ✦ Since aorta is malpositioned, coronary arteries arise from unprotected location.
 - ✦ Important to assess and describe their location relative to sternum, since coronary arteries are at increased risk of damage during median sternotomy due to their unprotected anterior location.

D-TGA Concurrent Abnormalities

- ✦ If the degree of shunting is small, then Major Aorto-Pulmonary Collateral Arteries (MAPCAs) develop in response.
- ✦ Long term, MAPCAs decrease blood flow through PAs, which prevents adequate growth and can prohibit cardiac surgery repair.
- ✦ Image entire thoracic aorta to identify all MAPCAs.
- ✦ Both surgical and interventional ligation techniques are employed.



D-TGA

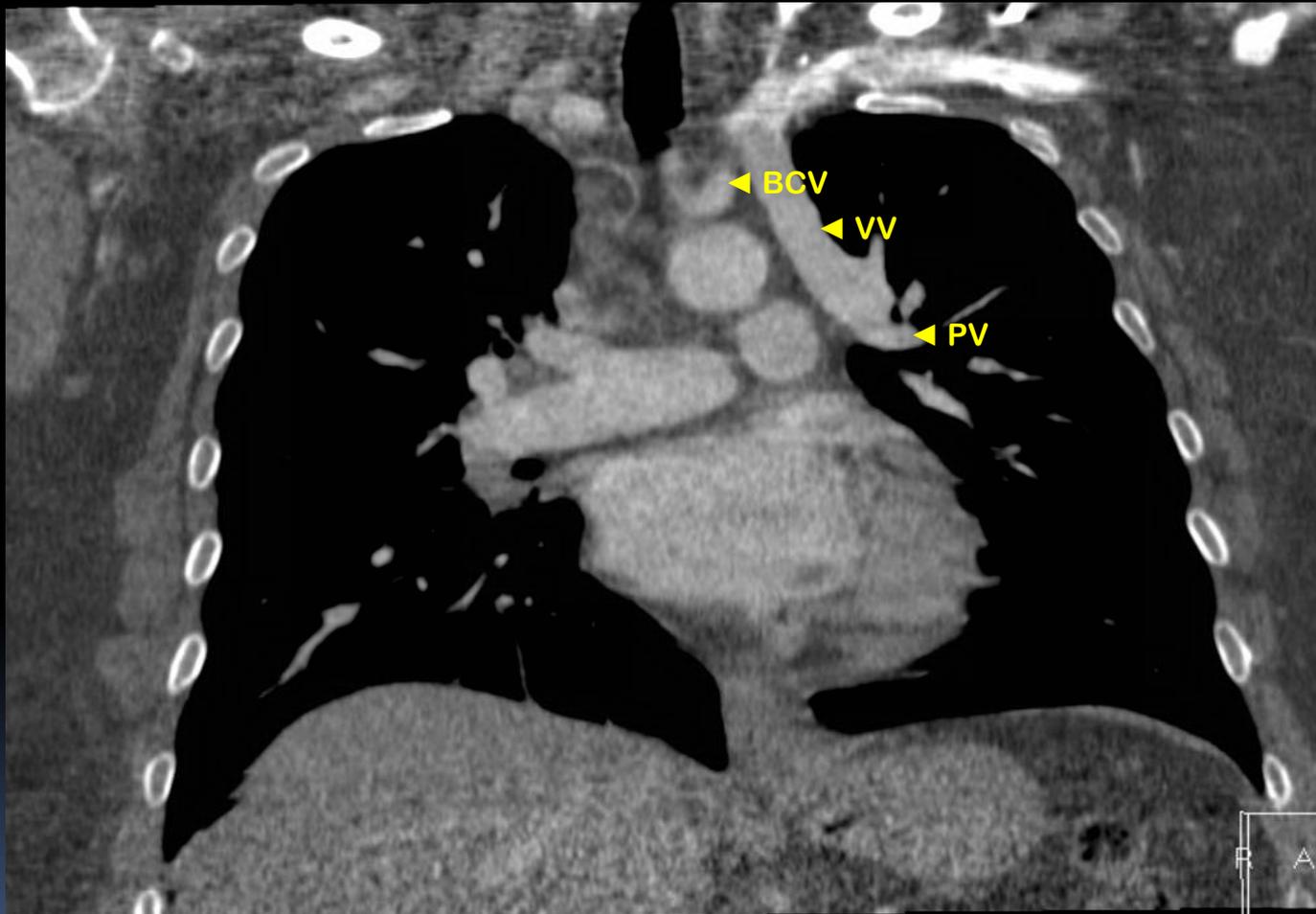
Key Points

- ✦ Describe size and location of great vessels.
- ✦ Describe where coronary arteries come off and their relationship to the sternum.
- ✦ Assess for shunts (VSD, PDA, PFO, and ASD) and size of shunt.
- ✦ Evaluate for pulmonic stenosis.
- ✦ Image entire thoracic aorta for presence of MAPCAs and describe size and location.

(Partial and Total) Anomalous Pulmonary Venous Return (PAPVR and TAPVR)

- ✦ A left-to-right shunt with anomalous connection of at least one (partial) or all (total) pulmonary veins (PVs) into a systemic vein.
- ✦ PAPVR:
 - ✦ Anomalous insertion of 1 or more PVs, usually onto the nearest systemic vein or directly into the RA.
 - ✦ Right PVs usually insert onto the vena cava, the azygous vein, or onto a hepatic vein (the latter occurs with the right inferior PV).
 - ✦ Left PVs usually insert onto the left brachiocephalic vein (via an anomalous vertical vein), the hemiazygous vein, or onto the coronary sinus.

Partial Anomalous Pulmonary Venous Return (PAPVR)



Left upper lobe pulmonary vein (PV) inserts onto the left brachiocephalic vein (BCV) via an anomalous vertical vein (VV)

Partial Anomalous Pulmonary Venous Return (PAPVR)

- ✦ Assess size and orientation of both the anomalous and normal pulmonary veins.
- ✦ Always evaluate supra- and infra-cardiac branch vessels as potential recipients of the anomalous vein.
- ✦ Evaluate degree of shunting (>50% is typically symptomatic).

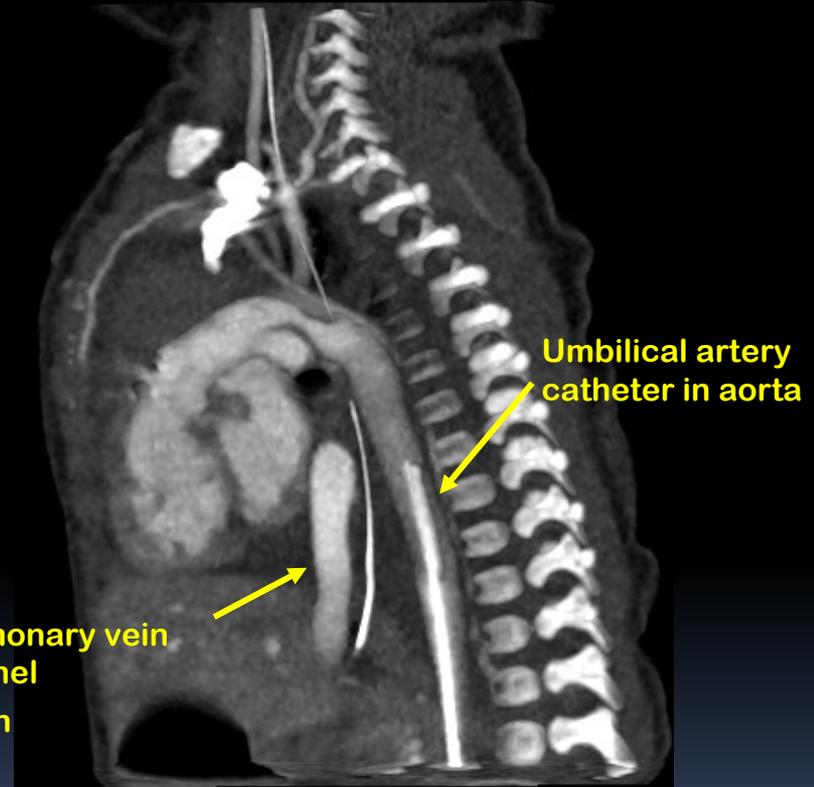
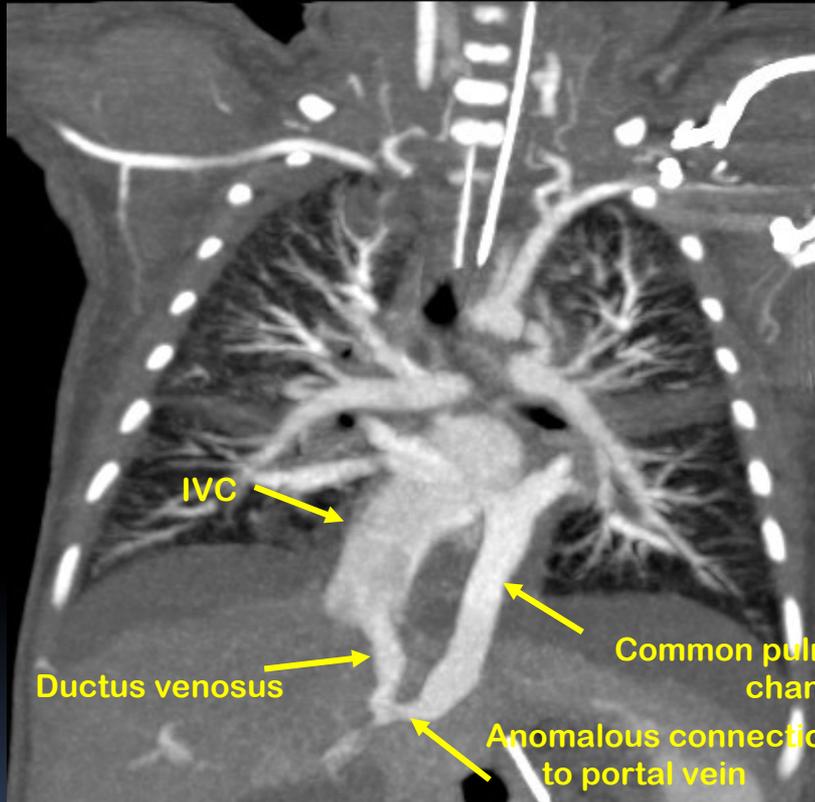
Concurrent Abnormalities in PAPVR

- ✦ Intracardiac shunt (such as ASD) can be present.
 - ✦ If anomalous PV connection is to the SVC or RA, there is a 90% chance of ASD. If anomalous vein connects to IVC, there is only a 15% chance of ASD.
- ✦ 10% of infants with ASD have a pulmonary venous anomaly. Therefore, always assess for anomalous PVs in infants with ASD.

Total Anomalous Pulmonary Venous Return (TAPVR)

- ✦ The 4 pulmonary veins often merge into a common channel prior to insertion. Total anomalous connection occurs in 1 of 3 ways:
 - ✦ Supracardiac: channel inserts on either the superior vena cava (SVC), the left brachiocephalic vein, or the azygous/hemizygous vein.
 - ✦ Intracardiac: channel or separate PVs insert onto the coronary sinus or into the right atrium (RA).
 - ✦ Look for bilateral SVCs, polysplenia, or asplenia.
 - ✦ Infracardiac: channel passes below the diaphragm to insert onto the portal vein or ductus venous tributaries. Always causes PV obstruction/back up.
- ✦ Must have a conduit for shunting of blood to the left heart to sustain life.

Total Anomalous Pulmonary Venous Return (TAPVR)



Infracardiac TAPVR

via a common PV channel draining into the portal vein

Total Anomalous Pulmonary Venous Return

- ✦ Must have a conduit for shunting of blood to the left heart to sustain life, most often an ASD or a PFO.
- ✦ Obtain a chest and abdomen CT (from mid-neck to the porta hepatis) to evaluate supra- and infra-cardiac branch vessels as potential recipients of anomalous veins.
- ✦ Contrast bolus should be timed to achieve optimal vascular enhancement. For infants, hand inject 2 mL/kg and scan immediately.
- ✦ Evaluate degree of pulmonary vein obstruction (especially for infracardiac insertions since obstruction is always present).
- ✦ All patients must be surgically repaired (urgently if there is high grade PV obstruction).

APVR

Key Points

- ✦ Determine size, location, and insertion of all 4 pulmonary veins.
- ✦ Scan from mid-neck to porta hepatis to evaluate for supra-cardiac and infra-cardiac insertions.
- ✦ If partial anomalous connection, describe location of anomalous insertion and evaluate degree of shunting. Search for concurrent presence of an ASD, which would be repaired at the time of surgery.
- ✦ If total anomalous connection, describe location of anomalous insertion, evaluate for presence of a common channel, and determine presence/absence of PV obstruction.

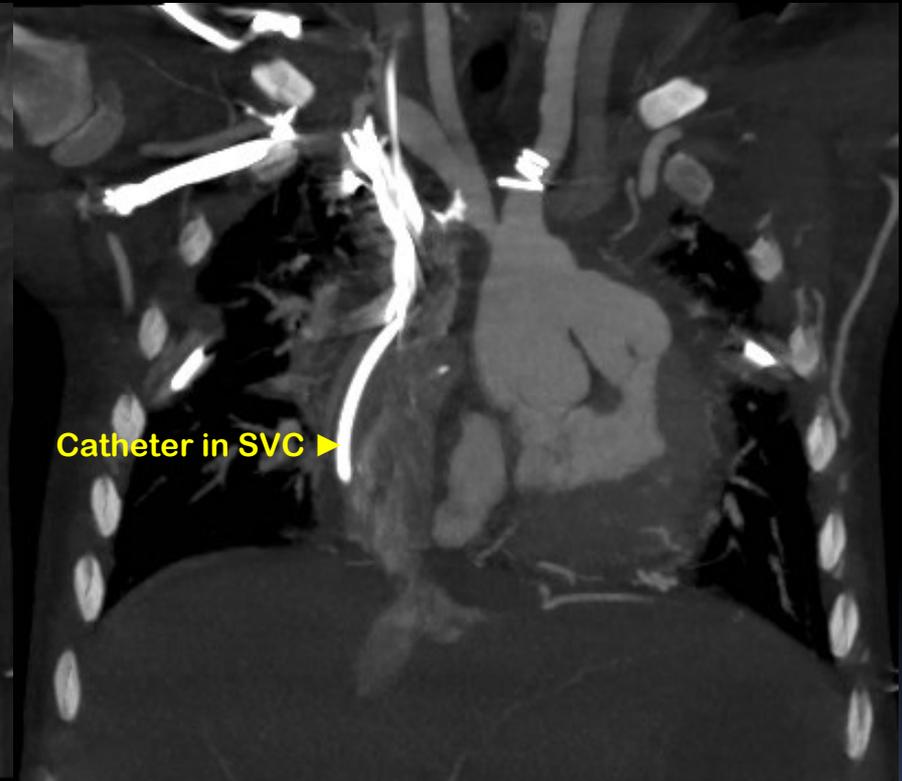
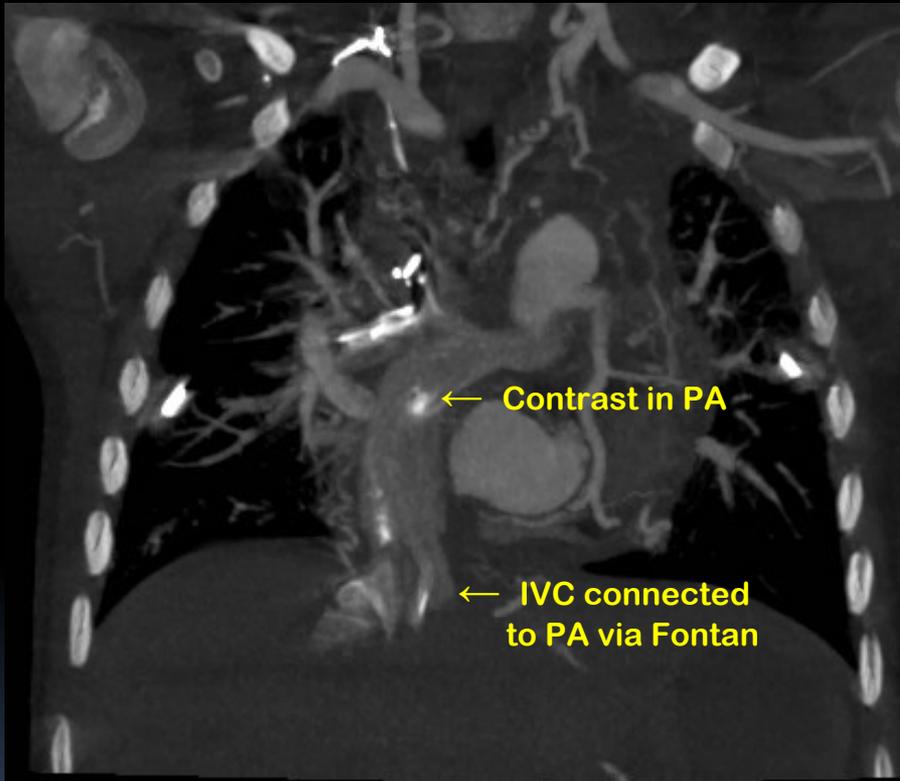
Post-Fontan Surgery Patients

- ✦ Not a CHD per se, but this represents a large group of patients with CHD that are frequently imaged prior to definitive treatment with heart transplant.
- ✦ Important to beware of complications seen on imaging in post-Fontan patients that may expedite listing for transplant.
- ✦ Many CHDs lead to palliation with the Fontan procedure:
 - ✦ Hypoplastic left heart syndrome
 - ✦ Double inlet ventricle
 - ✦ Tricuspid atresia
 - ✦ Pulmonary atresia with intact ventricular septum
 - ✦ Ebstein anomaly
 - ✦ Heterotaxy
 - ✦ Straddling atrioventricular (AV) valve
 - ✦ Crossed AV connections

Fontan Surgery

- ✦ Performed in patients with either an anatomic or functional single ventricle, or those unable to undergo biventricular repair. Usually performed as the last stage of a three-part Norwood procedure.
- ✦ Stage 1: RV redirected to supply systemic circulation, atrial septum excised to redirect pulmonary venous return to RA, Blalock-Taussig (BT) shunt created to connect right subclavian artery to right PA.
- ✦ Stage 2: BT shunt taken down and replaced with Glenn shunt in which blood from SVC is routed into the PA for oxygenation via a surgical conduit.
- ✦ Stage 3: Once patient can no longer oxygenate well, the modified Fontan is performed in which blood from the IVC is routed into the PA for oxygenation via a surgical conduit.

Fontan Surgery



Fontan connecting the IVC to the PA.

Contrast enters the SVC and is rapidly seen in the PA via Glenn shunt.

Normal mixing of contrast-opacified blood from Glenn shunt and non-opacified blood from Fontan, can be confused for thrombus.

Post-Fontan Considerations

- ✦ Important to adequately visualize patency of all anastomoses.
- ✦ To determine patency of the SVC and IVC conduits, contrast may need to be administered in the upper extremity, the lower extremity, or both.
 - ✦ Consider individualized contrast injection depending on clinical concern.
- ✦ Hypo-attenuating regions of blood in the RA, RV, and PAs may represent either thrombus or mixing of blood
 - ✦ A multiphase study may clarify whether thrombus is present.

Post-Fontan Considerations

- ✦ All Fontan patients may eventually require cardiac transplantation due to failing RV.
- ✦ Assess carefully for signs of failing Fontan:
 - ✦ Right ventricular dilatation
 - ✦ Right ventricular failure
 - ✦ Right atrial or left atrial enlargement (to suggest overload of RV)
 - ✦ Pulmonary enlargement/hypertension from systemic venous blood diversion to PAs
 - ✦ Large thrombus in Fontan circulation
 - ✦ Cirrhosis/ascites from right heart failure

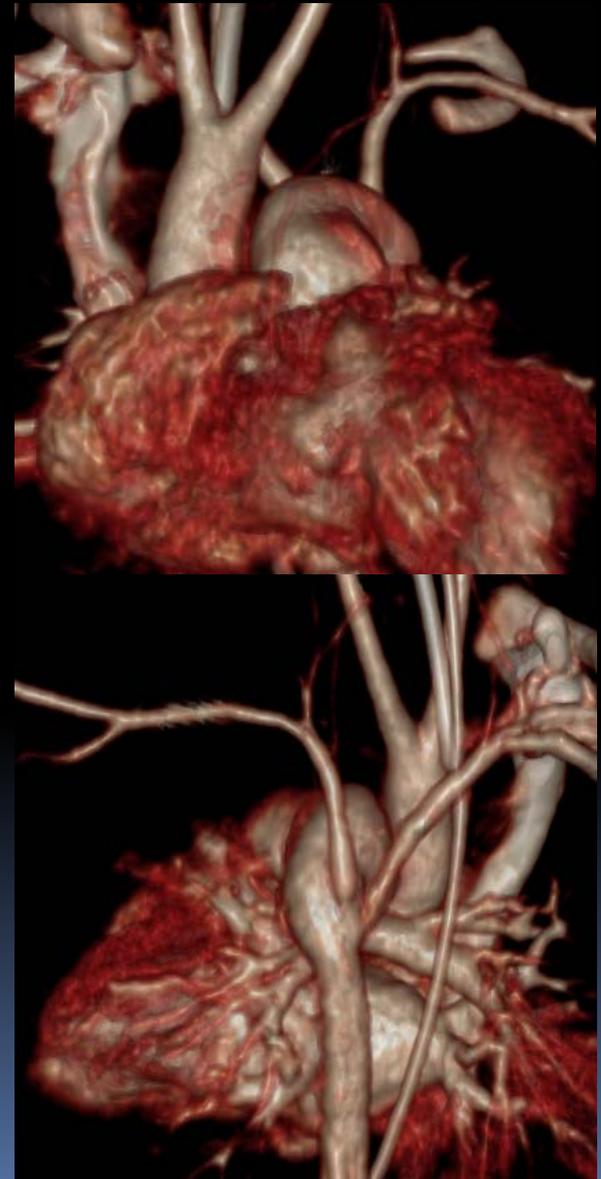
Post-Fontan Surgery

Key Points

- ✦ Evaluate patency of all anastomoses which may require individualized contrast injection to visualize SVC and IVC conduit blood flow.
- ✦ If there is concern for thrombus, multiphase study can be useful for distinguishing thrombus from admixed blood.
- ✦ Assess for signs of failing Fontan such as RV dilatation or failure, pulmonary hypertension, poor flow through SVC/IVC conduits, and evidence of hepatic overload/failure.
- ✦ Post-Fontan patients may eventually require heart transplant. Imaging findings of failing Fontan may precede clinical symptoms.

Aortic Arch Anomalies

- ✦ Various anomalies to include right aortic arch, double aortic arch, aortic coarctation, and aortic arch interruption.
- ✦ Always assess entire aortic arch as normal left aortic arch may have variant branch anatomy.
- ✦ If arch anomalies are present, describe the origin of all supra-aortic branch vessels.



Aortic Arch Anomalies

- ✦ Right aortic arch with mirror-image branching: assess for concurrent CHD.
 - ✦ 25% of Tetralogy of Fallot patients and 25-50% of Truncus Arteriosus patients have right aortic arch with mirror image branching.
- ✦ Double aortic arch: right and left arches wrap around the trachea and join posterior to the esophagus.
 - ✦ Most common type has a larger right arch and hypoplastic left arch.
- ✦ Surgical repair is indicated, especially if symptomatic.

Aortic Arch Anomalies

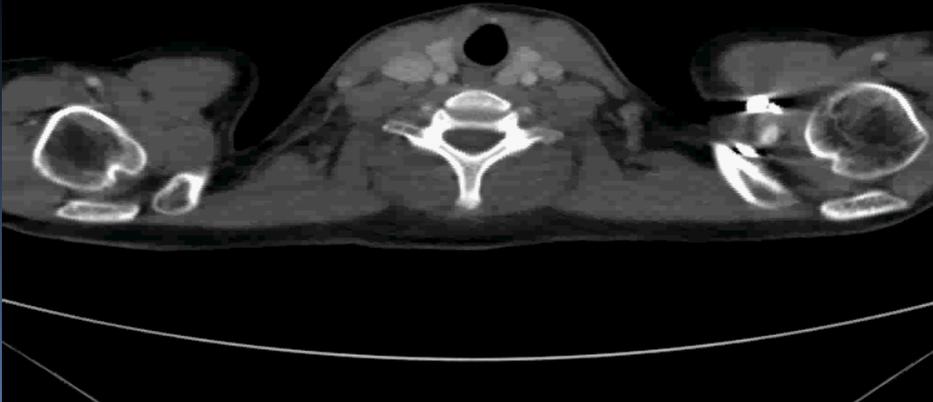
- ✦ Aortic coarctation: focal narrowing of the aorta in the region of the ductus arteriosus.
- ✦ Important to describe location of coarctation relative to ductus as pre-ductal form is most symptomatic.
- ✦ 25-50% of patients have bicuspid aortic valve so evaluate number of leaflets and for presence of stenosis.
- ✦ Evaluate for shunts given association with PDA and VSD.
- ✦ Surgical repair is indicated for children due to:
 - ✦ increased survival with earlier age of repair.
 - ✦ 75% mortality by age 46 with untreated coarctation.
- ✦ Balloon angioplasty is used in teens/adults.



Right aortic arch



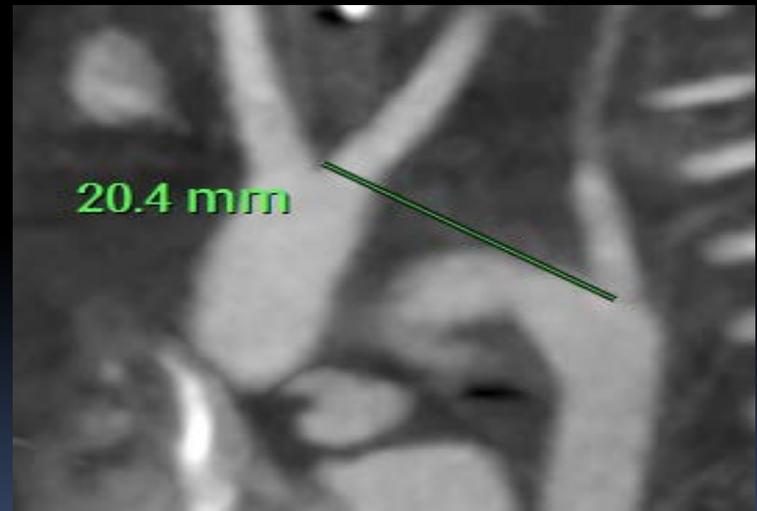
Aortic coarctation



Double aortic arch

Aortic Arch Anomalies

- ✦ Aortic arch interruption: complete discontinuity between the proximal ascending aorta and the distal descending aorta.
- ✦ Accompanied by PDA (to provide systemic blood flow) and VSD.
- ✦ Distinguish between true arch interruption versus arch hypoplasia.
- ✦ Describe site of disruption, distance between arches (for surgical planning), and which branches arise off of which arch.



2 cm gap between ascending and descending arch with left subclavian artery arising from descending arch

Aortic Arch Anomalies

Key Points

- ✦ If arch anomalies are present, describe location of all aortic branch vessels and location of anomaly relative to branch vessels.
- ✦ Evaluate for concurrent CHD, especially with right aortic arch with mirror branching.
- ✦ Assess for bicuspid aortic valve and aortic valve stenosis if coarctation is present. Describe location of coarctation relative to ductus arteriosus.
- ✦ Differentiate arch interruption from hypoplasia, measure distance between arches, and evaluate for VSD and PDA.

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