Introduction to Imaging of Congenital Heart Disease

Chest radiography with MR and CT Correlation

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The Segmental Approach to Congenital Heart Disease

Provides accurate anatomical and physiological diagnosis of CHD to allow more appropriate and better selection of therapeutic options: medical, surgical, or both
Steps in the Segmental Approach to Congenital Heart Disease

- Anatomic type of each of the 3 major segments. For example \{S,D,D\}, \{I,L,L\}, \{S,D,L\} etc.
- How each segment is connected to the adjacent segment (CAVC, DILV, TGA etc.)
- Associated anomalies within each segment, or between them (TAPVR, ASD, VSD, cleft mitral valve etc.)
- How the segmental combinations and connections, with or without the associated malformations, function
Chest Radiography

Morphology of CHD cannot be determined by chest radiographs.

But it is one of the primary means of determining physiology at birth.
Physiology

• Too much Pressure (Afterload)
  – Right sided obstruction (cyanosis)
  – Left sided obstruction

• Too much Volume (Preload)
  – Left to right shunts
  – Valvular incompetence

• Intermixing (cyanosis)

• Poor contractility (bad myocardium)
Radiographic approach to CHD

- **Pulmonary venous hypertension**
  - Left sided obstruction
  - Poor contractility
- **Increased pulmonary blood flow (PBF):**
  - Left to right shunts
  - Intermixing states with unobstructed PBF
- **Decreased pulmonary blood flow**
  - Intermixing states and/or obstructed PBF
- **Normal pulmonary blood flow**
  - Mild right sided obstruction, mild forms of CHD
Radiographic Appearance Helps to Determine Initial Palliation

- **Pulmonary venous hypertension:**
  - Correct left sided obstructive lesions

- **Increased pulmonary blood flow:**
  - Pulmonary artery banding or close defect
  - Fix left sided obstruction (ie. Coarct/VSD)

- **Decreased pulmonary blood flow:**
  - Augment PBF with a shunt

- **Normal pulmonary blood flow:**
  - No immediate palliation
Can you summarize CHD treatment in one word?
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Plumbing
Congenital Heart Disease Treatment

Two Ventricle Repair

• One step palliation

Single Ventricle Repair

• Staged palliation
  Initial palliation
  Final palliation
Chest Radiograph - Cardiomegaly with Pulmonary Venous Congestion and Edema

Left sided obstructive lesions such as coarctation

Rx = Fix the left sided obstruction

Chest radiograph of a newborn with severe coarctation of the aorta showing moderate cardiomegaly with pulmonary vascular congestion and diffuse edema.
Lesions causing pulmonary overcirculation such as multiple VSD’s

Rx = Patch repair or device closure or PA Banding
Multiple VSD’s. Frontal and lateral chest radiographs showing cardiomegaly with left atrial enlargement and increased pulmonary blood flow. Since pulmonary artery banding can cause distortion and abnormal growth of the pulmonary arteries, pulmonary over circulation from large simple left to right shunts is currently treated more definitively with patch repair or device closure if possible. If a patient has multiple shunt lesions such as several muscular ventricular septal defects (VSD’s) associated with severe respiratory symptoms from pulmonary over circulation, pulmonary artery banding may be used to decrease pulmonary blood flow to give time for some of the VSD’s to spontaneously close.
Chest Radiograph - Decreased Pulmonary Blood Flow

TOF, Pulmonary Atresia

Modified BTS
Tetralogy of Fallot with pulmonary valve atresia. Chest radiograph of a newborn with cardiomegaly, upturned cardiac apex, concave MPA segment, right aortic arch and severely diminished pulmonary blood flow to both lungs. Oblique MIP image from an MRA shows a modified Blalock-Taussig shunt (BTS) connecting the proximal left subclavian artery to the left pulmonary artery. When there is significant obstruction to pulmonary blood flow, surgeons now prefer to do a full repair in the neonate if the pulmonary arteries are confluent and normal in size. Initial palliation with a modified BTS is performed when the pulmonary arteries are very small and/or the lungs are supported by systemic aortopulmonary collaterals. Complete repair is then performed when the child is older and the pulmonary arteries have grown larger from the increased flow through the BTS.
Single Ventricle - Intermixing States with Obstructed Systemic Blood Flow

Situs Inversus, DORV, Coarctation

“2 step palliation” - Fix the obstructive lesion (coarctation) then do single ventricle palliation with the Fontan operation
Coronal oblique MIP images (a and b) from an MRA show situs inversus with a left-sided liver and levocardia. Both the aorta and MPA arise from a right ventricle consistent with double outlet right ventricle (DORV). Sagittal oblique MIP image shows a markedly hypoplastic transverse arch (arrow) and a large PDA supporting the descending aorta. The coarctation increases systemic arterial pressure, resulting in increased pulmonary blood flow through the MPA. Therefore, the initial palliation is to relieve the obstruction on the systemic side by fixing the coarctation. If a 2 ventricle repair can not be performed, then the patient will undergo a single ventricle palliation to a Fontan.
Outline

- Left sided obstructive lesions
  - Mitral and aortic valve
- Left to right shunts
- Cyanotic congenital heart disease
  - Admixture lesions
  - Right sided obstructive lesions
  - Tricuspid and pulmonary valves
Left Sided Obstructive Lesions

- Mitral Stenosis
- Cor triatriatum
- Supramitral ring
- Valvular aortic stenosis
- Sub and supravalvular aortic stenosis
- Coarctation
- Hypoplastic left heart syndrome
  - Fontan Palliation
Chest radiograph of a teenager with cardiomegaly, down-turned cardiac apex indicating left ventricular enlargement and severe left atrial enlargement due to combined mitral stenosis and regurgitation. Notice the dilated left atrial appendage on the frontal view and the enlarged LA causing compression of the bronchus intermedius on the lateral view.
Chest radiograph of a young child showing cardiomegaly with left atrial enlargement and pulmonary vascular congestion due to congenital mitral stenosis.
Normal Mitral Valve – 4C
Normal Mitral Valve

Diastole

Systole

3C

Short Axis
Previous slide titled **Normal Mitral Valve:**

Still frame SSFP images in 3 chamber (3C) and short axis (SA) planes showing the appearance of the normal mitral valve open in diastole and closed in systole. The diastolic short axis image from the base of the heart shows the 2 leaflet mitral valve has a disc shape and the 3 leaflet tricuspid valve has an oval shape when they have opened (long arrows). At the mid ventricular level during systole, the papillary muscles of the mitral valve are contracted and thickened (small arrows) which prevents the leaflets from prolapsing back into the left atrium as the left ventricle contracts.
35 year old with history of coarctation repair and VSD closure in infancy
Congenital mitral stenosis from a parachute mitral valve. Long axis views of the heart (a = 4 chamber, b = 3 chamber) showing a stenotic mitral valve with thickened leaflets (arrows). The short axis image (c) shows that the stenotic mitral valve (arrow) has a small rounded shape when it opens during diastole because both valve leaflets are attached via chordae tendinae to one papillary muscle.
2 year old boy with failure to thrive

Chest radiograph shows cardiomegaly with pulmonary vascular congestion and interstitial edema. An unusual circular structure on the frontal view is in the expected location of the LA. This patient has cor triatriatum and this circular structure is the obstructed proximal chamber within the left atrium.
Cor triatriatum occurs during embryologic development when the common pulmonary vein draining all the pulmonary veins from the developing lungs does not become fully incorporated into the left atrium. A persistent membrane or web divides the left atrium into 2 separate chambers which causes a variable degree of obstruction to pulmonary venous drainage. The confluence of the pulmonary veins proximal to the obstructing membrane dilates, becoming the “third” atrium.
Two chamber (a) and 4 chamber (b and c) SSFP images from a cardiac MRI show an obstructing membrane proximal to the mitral valve that divides the dilated left atrium into 2 chambers (arrows). Note the small opening within the membrane on the second 4C image (c).
Supra Mitral Stenosis

Supramitral ring

Cor triatriatum
Shone’s Syndrome

Complex of four obstructive anomalies of the left heart:

- Supra-mitral ring
- Parachute mitral valve (stenosis)
- Subaortic stenosis
- Coarctation of the aorta
Aortic Stenosis
Normal Aortic Valve

Short Axis

Diastole

Systole

Long Axis

Diastole

Systole
Bicuspid Aortic Valve

Short Axis

Long Axis

Systole

Diastole
Previous slide titled Bicuspid Aortic Valve:

Long and short axis images of the aortic valve with fusion of the intercoronary commissure. The valve leaflets are thickened and in systole the open valve has an oval shaped orifice and turbulent flow extends into the ascending aorta. In diastole, the LV is dilated due to severe aortic regurgitation with the regurgitant jet arising from failure of coaptation of the left non commissure (arrows).

Sinus = the aortic root has 3 sinuses (outpouchings of the wall creating a cloverleaf)
Cusp = leaflet – the normal aortic valve has 3
Commissure = coaptation point between the aortic valve leaflets – there are 3
Valvular Aortic Stenosis
Subaortic Stenosis with Coarctation

Subaortic stenosis or narrowing of the LV outflow tract (LVOT) during embryologic development can result in decreased flow through the ascending aorta and transverse arch that can lead to arch hypoplasia and / or coarctation. This follows the accepted principle “No flow = No grow.”
Subvalvular Aortic Stenosis

6 year old s/p repair of complete CAVC, now with subaortic stenosis

3 chamber views show a linear band of tissue (arrow in a) crossing the LVOT inferior to the aortic valve. The band of tissue obstructs the LVOT resulting in systolic turbulent flow (arrow in b) that extends into the ascending aorta consistent with subaortic stenosis.
Supravalvular Aortic Stenosis
Williams Syndrome

- Elfin Facies
- Hypercalcemia
- Supravalvar Aortic Stenosis
- PA & other vessel stenoses (renal arteries)

SSFP white blood (a), black blood (b) and anterior volume rendered image from an MRA (c) show supravalvular aortic stenosis with the characteristic narrowing of the sinotubular junction creating an “hourglass” configuration.
Cardiomegaly with Pulmonary Venous Hypertension - Edema

Differential Diagnosis:
- Left sided obstruction
- Large Lt to Rt Shunts
- Valvular regurgitation
- Cardiomyopathy
- Conduction abnormality
- Tumor

Critical Aortic Stenosis
Coarctation

- Ductal theory - obstruction of aortic isthmus resulting from constriction of anomalous ductal tissue invading wall of aorta
- Hemodynamic theory – “No flow no grow” associated with diffuse hypoplasia of the aortic arch
- Associations: bicuspid aortic valve, VSD, Shone and Turner syndromes

Focal Diffuse
Coarctation

- “3” Sign
- Rib notching
- LV enlargement
- Dilated ascending aorta (if associated aortic valve stenosis)
Chest radiograph findings in patients with simple coarctation. Frontal chest radiograph (a) shows the “3” sign (arrow) and notching of the inferior aspect of the upper ribs. Note the ascending aortic contour is also prominent due to post-stenotic dilatation from a stenotic bicuspid aortic valve. A second patient with coarctation has even more dramatic rib notching on the chest radiograph (b) due to dilated tortuous intercostal arteries (MRA MIP images c and d). The enlarged intercostal arteries have retrograde flow to the descending aorta distal to the coarctation to support the systemic circulation of the lower body.
Coarctation “3 Sign”
Newborn with CHF and Hypotension  Asymptomatic 19 year old with Hypertension

Newborn and young adult with coarctation. Newborns tend to present more acutely with hypotension and CHF due to more severe, long segment coarctation. Older patients present asymptptomatically with hypertension and tend to have focal coarctation at the aortic isthmus. Focal coarctation progresses slowly over time which allows collateral circulation to develop to support the circulation to the lower body.
Coarctation – MRI assessment

- Extent of narrowing
- Collateral vessels
- Aortic root and ascending aorta dimensions (BAV)
- Left ventricular size and function
- Descending aorta flow and gradient measurement
MR imaging of coarctation is performed pre-operatively in older patients when echocardiography windows do not permit full evaluation of the extent of narrowing and collateral circulation. Sagittal oblique MIP MRA image (a) and volume rendered posterior image (b) show numerous collaterals extending to the descending aorta (white arrows). The hemodynamic effects of the narrowing can be assessed by measuring flow velocity ($V$) at the level of obstruction (c. sagittal oblique phase velocity image) that can then be used to calculate a gradient using the modified Bernoulli equation: 

$$\text{Gradient (mmHg)} = 4V^2$$

A gradient measuring above 20 mm Hg warrants treatment with either balloon dilation or surgery to relieve the obstruction.
Surgical Repair of Coarctation

“End to End”

“End to Side”
Surgical Repair of Coarctation
There have been several approaches to the surgical repair of coarctation. The “end to end” approach involves resection of the narrowed segment, including all ductal tissue, and creating a direct anastomosis between the proximal and distal aortic segments. The “end to side” approach involves elongating the anastomosis especially when there is long segment narrowing that involves the transverse arch.

The patch and subclavian flap repairs widen the narrowed segment of the aorta and there may not be complete resection of ductal tissue with increased risk of re-coarctation. Also both the patch and subclavian flap repairs have a significant incidence of aneurysm development at the repair site.
Coarctation – Post-op Assessment

- Restenosis
- Collaterals
- Aneurysm
- Transverse arch hypoplasia
- Aortic stenosis/ regurgitation

MRI following balloon dilatation and stenting or surgical repair of coarctation. A large pseudoaneurysm from an uncontrolled intimal tear is a potential complication of balloon dilatation and stenting (a). Patients with severe transverse arch hypoplasia (b) may require a jump graft between the ascending and descending aorta (arrow).
HLHS results from a variable degree of left ventricular and aortic hypoplasia due to abnormal development of the mitral and aortic valves that ranges from stenosis to atresia. The most severe LV and aortic hypoplasia typically occurs when both the mitral and aortic valves are atretic. This diagram of HLHS shows a hypoplastic LV and aorta due to aortic valvular atresia. The dilated RV supports the systemic circulation to the aorta via a PDA. Both an ASD and a PDA are needed to get oxygenated blood to the systemic circulation.
Hypoplastic Left Heart Syndrome

S/P Balloon Atrial Septostomy
Previous slide titled Hypoplastic Left Heart Syndrome:

Initial chest radiograph in a baby with HLHS (a) shows a small heart and dilation of the left atrial appendage (arrow) due to a restrictive or small ASD which is obstructing the pulmonary venous return to the right atrium and ventricle. Following balloon atrial septostomy (b), an ASD has been created which decompresses the left atrium (the LA appendage is no longer dilated on the chest radiograph) and the heart is larger because there is now more pulmonary venous return to the right ventricle to support the systemic circulation.
The Norwood operation for HLHS assigns the RV as the systemic ventricle. The MPA is divided and the pulmonary valve is used as the neo aortic valve. The aortic arch is reconstructed off the pulmonary valve and a small BT shunt is placed between the right subclavian or innominate artery and the branch pulmonary arteries to support the pulmonary circulation. A recent modification of the procedure uses a small conduit between the RV and the PA’s in place of the BT shunt called the Sano modification. Lastly, the atrial septum is excised to allow unimpeded pulmonary venous return to enter to the right atrium and right ventricle.
HLHS s/p
Norwood
Types of CHD requiring Fontan

For palliation (functionally univentricular hearts with anatomy precluding 2 ventricle repair)

- Tricuspid atresia
- Unbalanced atrioventricular canal defect
- Hypoplastic left heart syndrome
- Double inlet or outlet ventricle
- Heterotaxy
- Pulmonary atresia with intact ventricular septum
- Ebstein’s anomaly
- Straddling atrioventricular valve
- Crossed atrioventricular connections
Fontan Operation

- Functional single ventricle
- Staged surgical palliation
- Separation of systemic and pulmonary venous return
- “Good” ventricle becomes the systemic pump
- Systemic venous return is routed to the pulmonary arteries without a ventricular pump
- Variety of surgical techniques
Single Ventricle

Pulmonary Obstruction
- MBTS or No Intervention
  - Stage 2
    - (4-6 mo)
  - Stage 3
    - (>18 mo)

Systemic Obstruction
- Stage 1 Norwood/Sano
  - Bidirectional Glenn
    - Stage 2
      - (4-6 mo)
  - Fontan
    - Stage 3
      - (>18 mo)

No Pulmonary Obstruction
- PA Band
The Fontan operation has undergone a number of revisions over the years. The classic Fontan operation, first performed in patients with tricuspid atresia, involved dividing the pulmonary arteries and connecting the SVC to the RPA and connecting the right atrial appendage to the LPA. Note that all of the blood from the lower body, including the hepatic venous blood, returns primarily to the left lung. This operation is no longer performed because the lack of hepatic venous blood entering into the right lung can result in the development of arteriovenous malformations (AVM’s) in the right lung. A vasoactive agent within the blood is thought to be degraded by liver enzymes and filtered out of the blood while passing through the liver before getting to the lungs, that prevents the development of AVM’s. This process is similar to the hepatopulmonary syndrome which develops in patients with cirrhosis of the liver due to portal to systemic shunts.

The modified Fontan involves a direct anastomosis of the right atrial appendage to the MPA and is often complicated by right atrial dilation, arrhythmias and thrombus formation. The Fontan operation was then revised to the currently used approaches of a fenestrated lateral tunnel baffle or an extracardiac conduit in order to create laminar flow through the Fontan pathway to the pulmonary arteries.
Tricuspid Atresia s/p Lateral Tunnel Fontan

Passive PBF requiring systemic venous pressure to be greater than pulmonary venous pressure
Status post Fontan for Tricuspid Atresia – PICC placed in RPA
Previous slide titled Status post Fontan for Tricuspid Atresia – PICC placed in RPA:

The requisition says “Check PICC placement”. Frontal radiograph (a) of the chest shows opacity in the right lower lobe. The PICC has been advanced via the right subclavian vein and extends down the right side of the mediastinum entering into the right hilum (arrow). Knowing that the patient has had the Fontan operation is important for understanding the course of the catheter that extends from the SVC into the right pulmonary artery. Coronal oblique MIP images from an MRA (b and c) show the Fontan pathway with the SVC and IVC anastomosis to the RPA (arrow).
Pulmonary Embolism

- Increased risk in CHD patients
- 17% of Fontan patients can have silent PE *(Varma et al. *JACC* 2003)*
- Chronic PE can result in elevated PVR and Fontan failure
Imaging of Intracardiac Thrombus

Intracardiac thrombus can be visualized on SSFP images (a) and using the myocardial delayed enhancement sequence (b) with a prolonged TI time. This patient has thrombus within the right atrium following a direct right atrial to pulmonary artery anastomosis “old style” modified Fontan.
Left Sided Obstructive Lesions

- Mitral Stenosis
- Cor triatriatum
- Supramitral ring
- Valvular aortic stenosis
- Sub or supravalvular aortic stenosis
- Coarctation
- Hypoplastic left heart syndrome
  - Fontan Palliation
Thank You For Your Attention !!!