The neural tube and the cardiac tubes form at approximately the 3rd week of gestation and then fold or loop, respectively, during the 4th week of gestation. What are the paired brain and cardiac lesions which correlate with these weeks of gestation?

- Option A is NOT correct. The corpus callosum forms in week 7 of gestation while heart looping occurs in week 5 of gestation.
- Option B is CORRECT. Milestones of cardiac and brain development occur at key times in the human embryo. In weeks 2-3 the earliest stages of cardiac development begins with formation of the heat tubes and the aortic arches which coincides with formation and closure of the neural tube. Hence, Chiari III malformation/encephalocele and vascular ring are malformations which may be found together if errors in both organ systems occur at the same time.
- Option C is NOT correct. Focal cortical dysplasia is thought to originate in week 7 of gestation defects leading to coarctation of the aorta occur in weeks 2-3 of gestation.
- Option D is NOT correct. Holoprosencephaly and pulmonary valve stenosis both occur during weeks 5-7 of gestation.
- Option E is NOT correct. The corpus callosum forms in week 7 while an endocardial cushion defect originates in week 5.

The patient is a 25 year old with a history of bicuspid aortic valve. The diameter of the ascending aorta measured 6.4 cm.

The best American Heart Association recommendation for this patient is?

A. Annual cardiology follow up
B. Annual echocardiography
C. Annual CTA
D. Annual MRI
E. Ascending aorta replacement

The diameter of the ascending aorta measured 6.4 cm in this patient with a history of bicuspid aortic valve. The AHA recommendation for this patient is:

• Option A is NOT correct because cardiology follow up is appropriate when the ascending aorta is smaller than 4 cm.
• Options B, C and D are NOT correct. Imaging follow up is appropriate when the ascending aorta is 4.0 cm in diameter and annual follow up appropriate if the diameter measures 4.5 cm
• Option E is CORRECT. The AHA guideline suggests ascending aorta replacement if the diameter of the ascending aorta measures greater than 5 cm

The diameter of the ascending aorta measured 6.4 cm in this patient with a history of bicuspid aortic valve.


TGA with hypoplastic aorta

What is the abnormality?

A. Tracheal stenosis
B. Right main and lobar bronchial narrowing
C. Left main and lobar bronchial narrowing
D. Bilateral left sided bronchus
E. Tracheal bronchus (Pig bronchus)
What is the abnormality depicted in this image?

- Option A is NOT correct. There is no tracheal abnormality.
- Option B is NOT correct. The right bronchus is normal.
- Option C is CORRECT. There is narrowing of the left main and lobar bronchi.
- Option D is NOT correct. There is no airway branching anomaly.
- Option E is NOT correct. There is no airway branching anomaly.

What is the abnormality depicted in this image?


Which of the following statements is correct?

- In pregnant women with Marfan syndrome, an aortic root diameter > 40 mm and an increase in aortic root diameter during pregnancy are risk factors for dissection.
- Option A is NOT correct. Patients with Marfan syndrome and a normal aortic root diameter have a 1% risk of aortic dissection or other serious cardiac complication during pregnancy.
- Option B is NOT correct. In pregnant women with Marfan syndrome, an aortic root diameter greater than 30 mm is a risk factor for dissection.
- Option C is NOT correct. In patients with Marfan syndrome, pre-pregnancy surgery is recommended when the ascending aorta is ≥ 35 mm.
- Option D is CORRECT. In pregnant women with Marfan syndrome, an increase in aortic root diameter during pregnancy is a risk factor for dissection.
- Option E is NOT correct. Following elective aortic root replacement, patients remain at risk for dissection in the residual aorta.
Risk of aortic aneurysm and aortic dissection/rupture is most common in which of these conditions?

- Option A is **CORRECT**. The genetic fault in Marfan syndrome apparently impairs aortic medial elastic fibers more extensively than impairment in congenital heart disease.
- Option B is **NOT** correct. The risk of rupture and dissection is exceedingly rare.
- Option C is **NOT** correct. Longitudinal outcome studies have shown a much lower acute aortic event rate for similar degrees of aortic root dilation.
- Option D is **NOT** correct. Risk of dissection and rupture seem to be less common in TET patients.

Risk of aortic aneurysm and aortic dissection/rupture is most common in which of these conditions?

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58th Annual Meeting & Postgraduate Course

You are shown a MR image of the aortic valve. The abnormality shown (see arrow):

A. Is always associated with cyanotic congenital defect.
B. Is not associated with aortic root dilation.
C. Is the most common congenital malformation occurring in approximately 10% of the general population.
D. CT/MR imaging is more adequate to evaluate the proximal aorta from the valve annulus to the takeoff of the great vessels.
E. Echocardiography assessment of the aortic root is adequate for this aortic valve abnormality.

Which is true regarding the abnormality highlighted by the arrow?

- Option A is NOT correct. While bicuspid aortic valve can be associated with complex congenital heart disease it is the most common congenital malformation occurring in up to 2% of the general population.
- Option B is NOT correct. It is always associated with aortic dilation extending from aortic annulus to the takeoff of the great vessels.
- Option C is NOT correct. It occurs in up to 2% of the general population.
- Option D is CORRECT. CT/MR adequately assess the entire aorta.
- Option E is NOT correct. Echocardiography, while adequate for evaluation of the aortic root, may not provide adequate imaging of the entire ascending aorta.

Which is true regarding the abnormality highlighted by the arrow?


Coronal reformat chest MR Angiography shows total cavopulmonary Fontan connection. Fontan operation:

- A. Is a complete repair with no risk for heart failure.
- B. Arrhythmias are not a complication.
- C. It is only performed in patients with tricuspid atresia.
- D. Routine CT angiography of the chest in Fontan patients produces diagnostic imaging for pulmonary thromboembolic disease.
- E. MRI allows non-invasive functional and anatomical assessment before and after surgery, and early detection of cardiac and extracardiac complications.
Coronal reformat chest MR Angiography shows total cavopulmonary Fontan connection. Which of the following is true regarding the Fontan procedure?

- Option A is NOT correct. Fontan operation is a palliative procedure with high risk of heart failure from either ventricular dysfunction or chronic venous congestion.
- Option B is NOT correct. Arrhythmias are significant complication. Patients with Fontan palliation often require implantation of pacemaker.
- Option C is NOT correct. It was initially performed in patients with tricuspid atresia. Currently performed as well in patients with single ventricle physiology such as hypoplastic left heart syndrome, pulmonary atresia with hypoplastic RV double inlet left ventricle, and heterotaxy syndrome.
- Option D is NOT correct. Routine CT angiography of the chest in Fontan patients produces suboptimal imaging for pulmonary thromboembolic disease. Single injection often results in suboptimal studies that either cannot be interpreted or are interpreted incorrectly. Dual simultaneous injection through an upper and lower extremity is preferred to achieve optimal opacification of the pulmonary arteries.
- Option E is CORRECT: MRI allows non-invasive functional and anatomical assessment before and after surgery, and early detection of cardiac and extracardiac complications. The limitation of MRI are patients with pacemakers.

How frequent is hepatic fibrosis in patients with Fontan operation?

A. 20%
B. 40%
C. 60%
D. 80%
E. 100%

- In the recent study by Bulut et al, 23/23 MRI scans demonstrated morphologic liver changes of fibrosis with varying degrees of reticular contrast enhancement compatible with fibrosis and congestion.
How frequent is hepatic fibrosis in patients with Fontan operation?


Which of the following abnormalities is present in this 12-year-old male with Duchenne muscular dystrophy?

- Option A is NOT correct. Pericardial inflammation can be associated with myocarditis, which could result in the late gadolinium enhancement seen on the image. However, the given history is not suggestive of this diagnosis. It should also be noted that some studies have suggested an inflammatory component (myocarditis) to the myocardial damage seen in muscular dystrophy patients.
- Option B is NOT correct. In an adult being worked up for chest pain, this could represent scar due to myocardial infarction. However, in a 12-year-old with Duchenne muscular dystrophy, infarction related to coronary artery occlusion is highly unlikely.
- Option C is CORRECT. The image is from a late gadolinium enhancement sequence from a cardiac MRI. The myocardium is properly nulled and the blood pool is bright. The arrow points to an area of bright signal (contrast retention) in the mid inferior, inferolateral and anterolateral wall of the left ventricular myocardium, consistent with late gadolinium enhancement due to fibrosis.
- Option D is NOT correct. The inversion time is adequate, with a nulled myocardium and bright blood pool.
- Option E is NOT correct. The abnormality is in the left ventricular myocardium. Additionally, a single static image is not adequate to diagnose right ventricular dysfunction.

Which of the following abnormalities is present in this 12-year-old male with Duchenne muscular dystrophy?

In what manner has brain perfusion been shown to be altered in patients with congenital heart disease?

A. Cerebral blood flow is normal in fetuses with left sided obstructive lesions.
B. Fetuses with right sided obstructive lesions have reduced cerebral blood flow.
C. Cerebral blood flow patterns and flow dynamics are not influenced by duration of deep hypothermic circulatory arrest or bypass.
D. Neurodevelopmental outcome may be worse in HLHS than in other forms of LSOL because cerebrovascular flow is more severely reduced.
E. Cerebral blood flow is most often well preserved in fetuses with congenital heart disease because the placenta helps to regulate blood pressure.

What criterion for pulmonary valve replacement is present in this patient?

A. Left ventricle ejection fraction 57%
B. Right ventricle ejection fraction 51%
C. Right ventricle regurgitant fraction 35%
D. Right ventricle and diastolic volume index 134 ml/m2
E. End diastolic volume RV/LV = 1.7

14 year old with history of tetralogy of Fallot repair. Cardiac MRI ventricle volumes and calculations are summarized below.

<table>
<thead>
<tr>
<th>Metric</th>
<th>Left ventricle</th>
<th>Right ventricle</th>
</tr>
</thead>
<tbody>
<tr>
<td>End diastolic volume (ml)</td>
<td>113</td>
<td>191</td>
</tr>
<tr>
<td>End systolic volume (ml)</td>
<td>49</td>
<td>94</td>
</tr>
<tr>
<td>Stroke volume (ml)</td>
<td>64</td>
<td>98</td>
</tr>
<tr>
<td>Ejection fraction (%)</td>
<td>57</td>
<td>51</td>
</tr>
<tr>
<td>RV regurgitant fraction (%)</td>
<td>35</td>
<td></td>
</tr>
<tr>
<td>EDV RV/LV</td>
<td>1.7</td>
<td></td>
</tr>
<tr>
<td>RV EDVI</td>
<td>134 ml/m2</td>
<td></td>
</tr>
</tbody>
</table>

[Future] Visualize the Future

In what manner has brain perfusion been shown to be altered in patients with congenital heart disease?

- Option A is NOT correct. Fetuses with HLHS – an obstructive left sided lesion – have abnormal cerebral blood flow dynamics. There are two mechanisms by which cerebral perfusion is reduced: intracardiac mixing of oxygenated and deoxygenated blood leads to decreased O2 delivery to the brain, and cerebral perfusion occurs retrograde via the ductus arteriosus which limits volume and pressure of blood flow.
- Option B is NOT correct. Intracardiac mixing of oxygenated and deoxygenated blood leads to decreased O2 delivery to the brain. Since venous return is directed away from the right side of the heart (the right side is obstructed) a larger volume of blood flow is directed to the left and therefore to the brain as well.
- Option C is NOT correct. Duration of deep hypothermic circulatory arrest is typically longer in patients with HLHS patients who have been shown to have worse neurodevelopmental outcome compared to other congenital heart patients who undergo surgical repair with shorter arrest times.
- Option D is CORRECT. Fetuses with HLHS have decreased MCA-PI compared to normal fetuses. It is because of abnormalities in cerebral blood flow in the fetus with HLHS and alterations of cerebral blood flow during deep hypothermic circulatory arrest that patients with HLHS have poor neurodevelopmental outcomes.
- Option E is NOT correct. The relative resistance in the cerebral versus umbilical arterial vasculature may be inappropriate in fetuses with CHD.
Cardiac MRI ventricle volumes and calculations are summarized below. What criterion for pulmonary valve replacement is present in this patient with a prior tetralogy of Fallot repair?

- Options A and B are NOT correct. The left and right ventricle ejection fractions are normal. Left ventricle ejection fraction less than 55% and right ventricle ejection from less than 47% are criteria for pulmonary valve replacement.
- Option C is CORRECT. A regurgitant fraction greater than 25% is a criterion for pulmonary valve replacement, but additional criteria must also be present.
- Option D is NOT correct. Right ventricle end diastolic volume index can be a criterion for pulmonary valve replacement. Although no consensus for how large the RV EDVI needs to be, all publications suggest that the RV EDVI limit is 150 ml/m² or greater.
- Option E is NOT correct. The end diastolic volume right ventricle to left ventricle ratio can be used as a measure of right ventricle enlargement, but the criteria calls for the ratio to be 2.0 or greater.

Which of the following is a prerequisite for dynamic pulmonary imaging?

- Option A is NOT correct. Dynamic pulmonary CT is not an ECG gated study.
- Option B is NOT correct. Dynamic pulmonary CT can be performed in intubated or self ventilated patients. Intubation is not a prerequisite.
- Option C is CORRECT. Dynamic pulmonary CT requires wide detector scanners.
- Option D is NOT correct. Dynamic pulmonary CT cannot be performed on a helical scanner.
- Option E is NOT correct. Scanning is performed during one entire respiratory cycle.

Which of the following is a prerequisite for dynamic pulmonary imaging?


Which of the following statements is correct?

- In pregnant women with asymptomatic mild to moderate aortic stenosis, pregnancy is well tolerated. Also patients with severe AS may sustain pregnancy well, as long as they remain asymptomatic during exercise testing and have a normal BP response during exercise.
- Option A is NOT correct. Women with bicuspid aortic valve have a risk of aortic dilatation and dissection. Approximately 50% of the patients with a bicuspid aortic valve and aortic stenosis have dilatation of the ascending aorta. Dissection does occur, although less frequently than in Marfan patients.
- Option B is NOT correct. In women of childbearing age the main cause of aortic stenosis is congenital bicuspid aortic valve.
- Option C is CORRECT. In pregnant women with asymptomatic mild to moderate aortic stenosis, pregnancy is well tolerated.
- Option D is NOT correct. All women with a bicuspid aortic valve should undergo imaging of the ascending aorta before pregnancy. In patients with bicuspid aortic valve and an aortic root >50 mm, pre-pregnancy surgery should be considered.

Which of the following is a prerequisite for dynamic pulmonary imaging?

In patients with bicuspid aortopathy and no additional risk factors, surgery for the ascending aorta is recommended when the aorta measures which of the following diameters?

A. > 45 mm
B. > 50 mm
C. > 55 mm
D. > 60 mm
E. > 40 mm

• Option C is CORRECT. Surgery to the ascending aorta in BAV patients is mandated
  – for an ascending aorta of >50 mm, when the patient has a family history of dissection, or the size progression is >5 mm/year
  – for an ascending aorta of >55 mm, when there are no risk factors
  – for an ascending aorta of >45 mm, when surgery for the valve is obligatory; replacement of the aortic sinuses needs to be individualized

In patients with bicuspid aortopathy and no additional risk factors, surgery for the ascending aorta is recommended when the aorta measures which of the following diameters?


You are shown short-axis inversion time scout images from a cardiac MRI of a 12 year-old male with Duchenne muscular dystrophy. Compared to the image on the right, the image on the left displays which of the following?

A. Pericarditis
B. Myocardial infarction
C. Myocardial fibrosis
D. Improper inversion time
E. Right ventricular dysfunction
Two short-axis inversion time scout images from a 12-year-old male with Duchenne muscular dystrophy are shown. Compared to the image on the right (1), the image on the left (2) displays which of the following?

- Option A is NOT correct. The image on the right does show subepicardial enhancement, which could be myocarditis associated with pericarditis. However, the image on the left does not clearly display any pericardial abnormality.
- Option B is NOT correct. While the image on the right does show late enhancement in the lateral and free wall, the image on the left does not clearly display this abnormality.
- Option C is NOT correct. While the image on the right does show late enhancement in the lateral and free wall, consistent with myocardial fibrosis in this patient with Duchenne muscular dystrophy. The image on the left does not clearly display this abnormality.
- Option D is CORRECT. The image on the left displays low signal in both the myocardium and blood pool (200 ms inversion time). The image on the right was obtained at a proper inversion time and shows adequate nulling of the myocardium (arrow) with bright signal in the blood pool (arrowhead). Also note the late enhancement of the lateral and inferior wall on the right image, not clearly seen on the left image.
- Option E is NOT correct. No right ventricular abnormality is displayed.

Which of the following conditions is most commonly associated with necrotizing enterocolitis?

A. Bicuspid aortic valve stenosis  
B. Ebstein malformation of tricuspid valve  
C. **Hypoplastic left heart syndrome**  
D. Tetralogy of Fallot with pulmonary atresia  
E. Ventricular septal defect

Which of the following conditions is most commonly associated with necrotizing enterocolitis?

- Options A, B, D, and E are NOT correct. The risk of necrotizing enterocolitis in association with congenital heart disease is highest in patients with hypoplastic left heart syndrome.
- Option C is CORRECT. CHD is a risk factor when necrotizing enterocolitis occurs in term infants.  
  - Systemic-to-pulmonary shunt (Norwood)  
  - Patent ductus arteriosus (Hybrid)  
  - Aortopulmonary window  
  - Truncus arteriosus  
  - Atroventricular septal defect  
  - Congestive heart failure
Which of the following conditions is most commonly associated with necrotizing enterocolitis?