



SPR 2015 POSTGRADUATE COURSE

Cardiac Imaging

Provided for further study

Visualize the Future



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?

- A. Agenesis of the corpus callosum & TGA (heart looping)
- B. Chiari III malformation/cephalocele & vascular ring
- C. focal cortical dysplasia & coarctation of the aorta
- D. holoprosencephaly & pulmonic valve stenosis
- E. agenesis of the corpus callosum & AVSD

Visualize the Future



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 embryology. In weeks 2-3 the earliest stages of cardiac development begins with formation of the heart 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

Visualize the Future



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?

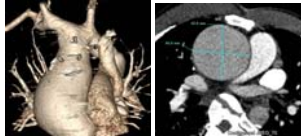
- Miller SP, et al. Abnormal brain development in newborns with congenital heart disease. N Engl J Med. 2007;357:1928-38.

Visualize the Future





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

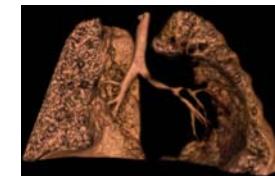


The diameter of the ascending aorta measured 6.4 cm in this patient with a history of bicuspid aortic valve. The AHA recommendation for this patient is:

- Nishimura RA, et al. 2014 AHA/ACC guideline for the management of patients with valvular heart disease: executive summary: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol. 2014;63:2438-88.



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



Visualize the Future



What is the abnormality depicted in this image?

- Noel CV, et al. Incidence of tracheobronchial anomalies found with hypoplastic left heart syndrome. *Congenit Heart Dis.* 2014;9:294-9.
- Kazim R, et al. Tracheobronchial anomalies in children with congenital cardiac disease. *J Cardiothorac Vasc Anesth.* 1998;12:553-5.



Visualize the Future



You are shown an image of the aortic root from a cardiac MRI of a 25-year-old woman with Marfan syndrome.

Which of the following statements is correct?

- Patients with Marfan syndrome and a normal aortic root diameter have a 15% risk of aortic dissection or other serious cardiac complication during pregnancy
- In pregnant women with Marfan syndrome, an aortic root diameter greater than 30 mm is a risk factor for dissection
- In patients with Marfan syndrome, pre-pregnancy surgery is recommended when the ascending aorta is ≥ 35 mm
- In pregnant women with Marfan syndrome, an increase in aortic root diameter during pregnancy is a risk factor for dissection.
- Following elective aortic root replacement, patients with Marfan syndrome have no risk for aortic dissection



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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 40 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 ≥ 45 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



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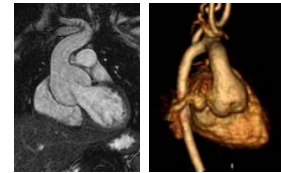
Which of the following statements is correct?

- European Society of Gynecology (ESG), et al. ESC guidelines on the management of cardiovascular diseases during pregnancy: the task force on the management of cardiovascular diseases during pregnancy of the European Society of Cardiology (ESC). Eur Heart J. 2011;32:3147–3197.
- Khairy P, et al. Pregnancy outcomes in women with congenital heart disease. Circulation. 2006;113:517-24.
- Pacini L, et al. Maternal complication of pregnancy in Marfan syndrome. Int J Cardiol 2009;136:156–161.



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

- Marfan syndrome (The genetic fault in Marfan syndrome apparently impairs aortic medial elastic fibers more extensively than impairment in Congenital Heart Disease)
- Truncus arteriosus (The risk of rupture and dissection is exceedingly rare, Root Dilation in Patients with Truncus Arteriosus [Carlo WF, McKenzie ED, Slesnick TC. Congenit Heart Dis.](#) 2011 May-Jun;6(3):228-33)
- Bicuspid aortopathy (Longitudinal outcome studies have shown a much lower acute aortic event rate for similar degrees of aortic root dilation)
- Tetralogy of Fallot and its variants (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?

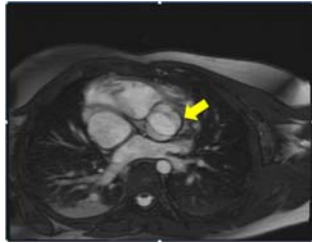
- Option A is **CORRECT**. The genetic fault in Marfan syndrome apparently impairs aortic medial elastic fibers more extensively than impairment in congenital heart disease
- Option B is **NOT** correct. The risk of rupture and dissection is exceedingly rare
- Option C is **NOT** correct. Longitudinal 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?

- Francois K. Aortopathy associated with congenital heart disease: A current literature review. Ann Pediatr Cardiol. 2015;8:25-36.
- Zanjani KS, et al. Aortic dilatation and aortopathy in congenital heart diseases. J Cardiol. 2013;61:16-21.
- Niwa K. Aortic root dilatation in tetralogy of Fallot long-term after repair--histology of the aorta in tetralogy of Fallot: evidence of intrinsic aortopathy. Int J Cardiol. 2005;103:117-9.
- Girdauskas E, et al. Bicuspid aortic valve and associated aortopathy: an update. Semin Thorac Cardiovasc Surg. 2013;25:310-6.
- Hardikar AA, et al. Surgical thresholds for bicuspid aortic valve associated aortopathy. JACC Cardiovasc Imaging. 2013;6:1311-20.
- Knirsch W, et al. Aortic aneurysm rupture in infantile Marfan's syndrome: Pediatr Cardiol. 2001;22:156-9.





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?

- Allen HD, et al., editors. Moss & Adams: Heart Disease in Infants, Children and Adolescents: Including the Fetus and Young Adult, 8th edition. Philadelphia:Lippincott Williams & Wilkins; 2012
- Allen BD, et al. Thoracic aorta 3D hemodynamics in pediatric and young adult patients with bicuspid aortic valve. J Magn Reson Imaging. 2015 Jan 22.
- Merritt BA, et al. Association between leaflet fusion pattern and thoracic aorta morphology in patients with bicuspid aortic valve. J Magn Reson Imaging. 2014;40:294-300.



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.



Visualize the Future



Coronal reformat chest MR Angiography shows total cavopulmonary Fontan connection. Which of the following is true regarding the Fontan procedure?

- Allen HD, et al., editors. Moss & Adams: Heart Disease in Infants, Children and Adolescents: Including the Fetus and Young Adult, 8th edition. Philadelphia:Lippincott Williams & Wilkins; 2012
- Navarro-Aguilar V, et al. Fontan procedure: imaging of normal post-surgical anatomy and the spectrum of cardiac and extracardiac complications. Clin Radiol. 2015;70:295-303
- Krieger E, et al. Heart failure treatment in adults with congenital heart disease: where do we stand in 2014. Heart 2014;100:1329-1334
- Sandler KL, et al. Optimizing CT angiography in patients with Fontan physiology: single-center experience of dual-site power injection. Clin Radiol. 2014;69:e562-7.



Visualize the Future



How frequent is hepatic fibrosis in patients with Fontan operation?

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



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How frequent is hepatic fibrosis in patients with Fontan operation?

- 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.



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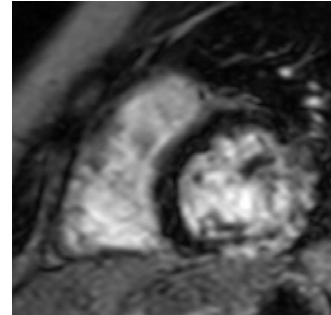


How frequent is hepatic fibrosis in patients with Fontan operation?

- Bulut OP, et al. Magnetic resonance imaging identifies unsuspected liver abnormalities in patients after the Fontan procedure. *J Pediatr.* 2013;163:201-6.
- Kiesewetter CH, et al. Hepatic changes in the failing Fontan circulation. *Heart.* 2007;93:579-84.



Visualize the Future



You are shown a short-axis image from a cardiac MRI of a 12 year-old male with Duchenne muscular dystrophy. The image most likely shows which of the following abnormalities?

- A. Pericarditis
- B. Myocardial infarction
- C. Myocardial fibrosis
- D. Improper inversion time
- E. Right ventricular dysfunction



Visualize the Future



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.



Visualize the Future



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

- Cummings KW, et al. A pattern-based approach to assessment of delayed enhancement in nonischemic cardiomyopathy at MR imaging. *RadioGraphics.* 2009;29 :89-103.
- Viallon M, et al. Head-to-head comparison of eight late gadolinium-enhanced cardiac MR (LGE CMR) sequences at 1.5 tesla: from bench to bedside. *Journal of magnetic resonance imaging. J Magn Reson Imaging.* 2011;34:1374-1387.
- Verhaert D, et al. Cardiac involvement in patients with muscular dystrophies: magnetic resonance imaging phenotype and genotypic considerations. *Circ Cardiovasc Imaging.* 2011;4:67-76.



Visualize the Future



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.



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 **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.



Visualize the Future



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

- Williams IA, et al. Fetal cerebrovascular resistance and neonatal EEG predict 18-month neurodevelopmental outcome in infants with congenital heart disease. *Ultrasound Obstet Gynecol.* 2012;40:304–309.
- Kaltman JR, et al. Impact of congenital heart disease on cerebrovascular blood flow dynamics in the fetus. *Ultrasound Obstet Gynecol.* 2005;25:32–36.

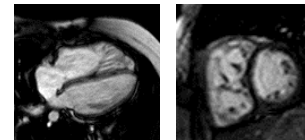


Visualize the Future



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

	Left ventricle	Right ventricle
End diastolic volume (ml)	113	191
End systolic volume (ml)	49	94
Stroke volume (ml)	64	98
Ejection fraction (%)	57	51



RV regurgitant fraction = 35%
EDV RV:LV = 1.7
RV EDVI = 134 ml/m²

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 end diastolic volume index 134 ml/m²
- E. End diastolic volume RV:LV = 1.7



Visualize the Future



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 fraction 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.



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?

- Geva T. Repaired tetralogy of Fallot: the roles of cardiovascular magnetic resonance in evaluating pathophysiology and for pulmonary valve replacement decision support. *J Cardiovasc Magn Reson.* 2011;13:9.



Pre requisite for dynamic pulmonary imaging is?

- ECG gating
- Intubation
- Wide detector CT scanner
- Helical CT scanner
- Breath held in expiration



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?

- Greenberg SB. Dynamic pulmonary CT of children. *AJR Am J Roentgenol.* 2012;199:435-40.
- Greenberg SB, et al. Dynamic pulmonary computed tomography angiography: a new standard for evaluation of combined airway and vascular abnormalities in infants. *Int J Cardiovasc Imaging.* 2014;30:407-14.



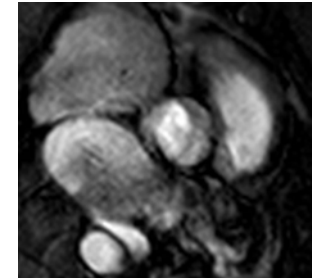
Visualize the Future



You are shown an image of the aortic root from a cardiac MRI of a 22-year-old woman.

Which of the following statements is correct?

- A. Women with bicuspid aortic valve do not have a risk of aortic dilatation and dissection.
- B. In women of childbearing age acquired aortic stenosis is more common than congenital aortic stenosis.
- C. With asymptomatic mild or moderate AS, pregnancy is well tolerated
- D. All women with a bicuspid aortic valve need not undergo imaging of the ascending aorta before pregnancy.
- E. Pre-pregnancy surgery should be considered when the aortic diameter is >40 mm



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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.
- Option E is **NOT** correct. In patients with bicuspid aortic valve and an aortic root >50 mm, pre-pregnancy surgery should be considered.



Visualize the Future



Which of the following statements is correct?

- European Society of Gynecology (ESG), et al. ESC guidelines on the management of cardiovascular diseases during pregnancy: the task force on the management of cardiovascular diseases during pregnancy of the European Society of Cardiology (ESC). *Eur Heart J.* 2011;32:3147–3197.
- Khairy P, et al. Pregnancy outcomes in women with congenital heart disease. *Circulation.* 2006;113:517-24.



Visualize the Future



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



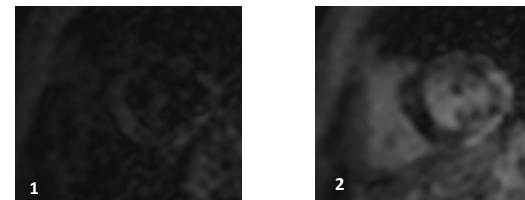
In patients with bicuspid aortopathy and no additional risk factors, surgery for the ascending aorta is recommended when the aorta measures which of the following diameters?

- 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?

- Francois K. Aortopathy associated with congenital heart disease: A current literature review. *Ann Pediatr Cardiol.* 2015;8:25-36.
- Zanjani KS, et al. Aortic dilatation and aortopathy in congenital heart diseases. *J Cardiol.* 2013;61:16-21.
- Girdauskas E, et al. Bicuspid aortic valve and associated aortopathy: an update. *Semin Thorac Cardiovasc Surg.* 2013;25:310-6.
- Hardikar AA, et al. Surgical thresholds for bicuspid aortic valve associated aortopathy. *JACC Cardiovasc Imaging.* 2013;6:1311-20.



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.



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?

- Cummings KW, et al. A pattern-based approach to assessment of delayed enhancement in nonischemic cardiomyopathy at MR imaging. *RadioGraphics*. 2009;29 :89-103.
- Viallon M, et al. Head-to-head comparison of eight late gadolinium-enhanced cardiac MR (LGE CMR) sequences at 1.5 tesla: from bench to bedside. *Journal of magnetic resonance imaging. J Magn Reson Imaging*. 2011;34:1374-1387.
- Verhaert D, et al. Cardiac involvement in patients with muscular dystrophies: magnetic resonance imaging phenotype and genotypic considerations. *Circ Cardiovasc Imaging*. 2011;4:67-76.



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

- Bicuspid aortic valve stenosis
- Ebstein malformation of tricuspid valve
- Hypoplastic left heart syndrome**
- Tetralogy of Fallot with pulmonary atresia
- 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
 - Atrioventricular septal defect
 - Congestive heart failure





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

- Miller TA, et al. Abnormal abdominal aorta hemodynamics are associated with necrotizing enterocolitis in infants with hypoplastic left heart syndrome. *Pediatr Cardiol.* 2014;35:616-21.

