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Episode 1

What are key imaging features to identify the right atrium in a congenital cardiac malformation?

Potentially the best imaging feature to identify the right atrium is identification of the atrium into which the coronary sinus drains. Additional features are identification of the atrium from which the suprahepatic inferior vena cava drains. The superior vena cava insertion is more variable and less reliable to identify the true right atrium.

What are key imaging features to identify the left atrium in a congenital cardiac malformation?

The left atrium is best identified by searching for the atrium which has the finger-like atrial appendage. Pulmonary vein insertion is less reliable to identify the true left atrium.

What are key imaging features to identify the right ventricle in a congenital cardiac malformation?

Key features that can help identify the right ventricle include presence of the moderator band, identification of a more apical attachment of the atrioventricular valve (insertion of the AV valve is typically more apical in the right ventricle compared to the left ventricle), presence of the infundibulum, and a trabeculated appearance of the septum compared to a smoother septum on the left ventricular side.

What are key imaging features of Tetralogy of Fallot?

Tetralogy of Fallot is the result of failure of the right ventricular outflow track to fuse with the interventricular septum. On radiography look for decreased pulmonary blood flow and a boot-shaped heart with an upturned cardiac apex and possible right aortic arch. The four classic features of Tetralogy of Fallot are:

1. Right ventricular hypertrophy
2. Ventricular septal defect
3. Pulmonary stenosis
4. Overriding aorta

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What is the prognosis in terms of survival of individuals with Tetralogy of Fallot following appropriate surgical repair?

Prognosis is excellent. Following surgical repair, there is at least 90% survival at 35 years following surgery. Note that degree of right ventricular outflow tract stenosis guides timing of repair and degree of clinical symptoms and symptom onset. Severe stenosis requires earlier surgical repair.

What is the most common type of atrial septal defect: ostium primum or ostium secundum?

Ostium secundum accounts for around 60% of atrial septal defects compared to 35% for ostium primum. Note that the final 5% is from sinus venosus atrial septal defects.

Classic associations to remember for board exams:

Ostium primum: associated with endocardial cushion defects, common in Trisomy 21 (Downs)

Ostium secundum: Holt-Oram syndrome which is autosomal dominant with congenital heart defects like ASD and VSD and coarctation of the aorta along with upper limb anomalies like radial and thumb aplasia and hypoplasia of the clavicle.

Sinus venosus ASD: partially anomalous pulmonary venous return

What are the components of the endocardial cushion?

The endocardial cushion results from formation of the lower atrial septum, ventricular septum, and the septal leaflets of the tricuspid and mitral valves.

What is the difference between a partial, transitional, and complete endocardial cushion defect?

A partial endocardial cushion defect has a canal passing through either the mitral or tricuspid valves but not both and may be asymptomatic. A transitional endocardial cushion defect has a canal through both the mitral and tricuspid valves and the atrial and/or ventricular septum. A complete endocardial cushion defect has a large septal defect and may have either a common or separate mitral valve and tricuspid valve. A complete endocardial cushion defect will have symptoms of congestive heart failure and a large left to right shunt and mitral regurgitation. Note that all of these will have an ostium primum atrial septal defect.

Nearly half of all individuals with an endocardial cushion defect will have Down's syndrome. On imaging expect diffuse cardiac enlargement, a gooseneck deformity of the left ventricular outflow tract on angiography, and right greater than left increased pulmonary vascularity. A hypersegmented sternum and 11 ribs would be a manifestation of associated Trisomy 21.

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What are common imaging findings suggestive of an atrial septal defect?

Atrial septal defects (without other associated cardiac anomalies) would manifest with enlargement of the right atrium and right ventricle with normal appearing left atrium and left ventricle. Associated asymmetric right pulmonary artery enlargement may be seen.

What is cor triatriatum?

As the name suggests—triatriatum—this is an entity in which there are essentially 3 atria due to to a congenital anomaly in which a membrane divides the left atrium into two with anterior and posterior chambers. Clinical symptoms are often pulmonary hypertension in children and this can be fatal. This appears similar to mitral stenosis on a chest radiograph with a normal-sized heart and pulmonary edema. There may be an associated atrial septal defect and anomalous pulmonary venous drainage. Early surgical intervention often leads to best outcomes.

What is the most common cyanotic congenital heart disease?

Tetralogy of Fallot

What is the so called pentalogy of Fallot?

The 4 findings characteristic of tetralogy of Fallot (ventricular septal defect, right ventricular outflow tract obstruction, overriding aorta and right ventricular hypertrophy) with additional finding of atrial septal defect or patent ductus arteriosus.

True or false? Tetralogy of Fallot is associated with a right-sided aortic arch in greater than 50% of cases.

False. A right-sided aortic arch is seen in approximately 25% of cases of Tetralogy of Fallot.

What are some of the most common causes of right ventricular outflow tract obstruction in cases of tetralogy of Fallot?

With tetralogy of Fallot, right ventricular outflow tract obstruction can result from valvular anomalies such as a hypoplastic pulmonary valve annulus or bicuspid pulmonary valve. Other contributors to right ventricular outflow tract obstruction can include infudibular stenosis and pulmonary artery hypoplasia.

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Tetralogy of Fallot, along with many other congenital cardiac anomalies, have VACTERL associations. What are the principle components of VACTERL?

V: vertebral anomalies

A: anorectal anomalies such as anal atresia

C: cardiac anomalies and cleft lip

TE: tracheoesophageal atresia and/or esophageal atresia

R: renal anomalies and/or radial ray anomalies

L: limb anomalies such as polydactyly

What congenital cardiac anomalies are commonly associated with congenital rubella infection?

Congenital rubella infection is associated with both ventricular septal defects as well as tetralogy of Fallot. Additional features of congenital rubella infection can include deafness, intrauterine growth restriction, mental impairment and microcephaly. Note that rubella is one of the TORCH infections (Toxoplasomsosis, Other (syphilis, varicella zoster, parvovirus B19), Rubella, Cytomegalovirus, Herpes simplex virus) and primary infection by the mother is much higher risk for the developing infant than viral reinfection or reactivation.

Which trisomies are classically associated with congenital cardiac anomalies?
Notable trisomies associated with congenital cardiac anomalies include trisomy 13, trisomy 18, and trisomy 21. Note that nearly all cases of trisomy 13 and 18 will have a congenital cardiac anomaly and roughly half of trisomy 21 will have an associated congenital cardiac anomaly.

Name some cardiovascular anomalies associated with Turner syndrome.

Turner syndrome is notably associated with a bicuspid aortic valve and aortic coarctation with a notable risk of aortic dissection.

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Episode 2

What are the 5 T's of cyanotic congenital heart disease?

A mnemonic to help you remember common causes of cyanotic congenital heart diseases as the 5 T's mnemonic, as follows:

Tetralogy of Fallot
Truncus arteriosus
Transposition of the great arteries
Total anomalous pulmonary venous return
Tricuspid valve anomalies

Cyanotic congenital heart disease can be divided based on conditions with increased versus decreased pulmonary vascularity. What are some cyanotic congenital heart conditions that present with increased pulmonary vascularity?

Cyanotic congenital heart disease entities that present with increased pulmonary vascularity include a large ventricular septal defect, several types of total anomalous pulmonary venous return, and several types of truncus arteriosus.

What are some congenital heart disease entities that present with decreased pulmonary vascularity?

Cyanotic congenital heart disease entities that present with decreased pulmonary vascularity include tetralogy of Fallot, Ebstein anomaly with coexisting atrial septal defect, and hypoplastic right heart syndrome.

What are classic features of an Ebstein anomaly?

Ebstein anomaly results from abnormal tricuspid valve development with tricuspid regurgitation that often presents with hydrops fetalis. Classic imaging features include marked right-sided heart enlargement with a very enlarged right atrium. A buzzword for the appearance of the heart is the "box-shaped" heart on a chest radiograph. I remembered this as the "reverse Nirvana" lesion when studying for boards given one the band's hit songs "Heart Shaped Box" being the reverse of the "Box-Shaped Heart". On cross-sectional imaging expect apical displacement of the septal and posterior leaflets of the tricuspid valve. You can use the mitral valve attachment as a surrogate for the expected normal position of the tricuspid valve which, in Ebstein anomaly, will be apically displaced in relation to the mitral valve. Note an association with maternal lithium use—and "Lithium" was also a hit Nirvana song—and association with trisomies 13 and 21 and Turner syndrome. Ebstein anomaly may coexist with other congenital cardiac anomalies.

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What are key features of partial anomalous pulmonary venous return (PAPVR)?

The underlying abnormality of PAPVR is anomalous connections of some, but not all, pulmonary veins with the systemic circulation instead of the expected drainage of all pulmonary veins to the left atrium. Four common subtypes exist: supra-cardiac, cardiac, infracardiac and mixed. Supracardiac PAPVR has an association with persistent left superior vena cava. Cardiac PAPVR commonly sees pulmonary veins drain to the right atrium and/or coronary sinus. Infracardiac PAPVR can have drainage of pulmonary vein(s) to the portal vein, inferior vena cava or hepatic veins. PAPVR has a strong associated with atrial septal defects in nearly half of cases.

What are common features of Scimitar syndrome?

Scimitar syndrome is a form of PAPVR and is also known as hypogenetic lung syndrome. Scimitar syndrome involves an anomalous pulmonary vein (PAPVR) that drains a hypoplastic lung, most commonly connecting to the inferior vena cava, but also can be seen with anomalous drainage to the portal vein or right atrium. Adults with Scimitar syndrome are prone to repeated pulmonary infections and this may be a clinical presenting symptom provided to you on a multiple choice question stem. If presenting as an infant, heart failure is likely which may be an indication of coexisting congenital heart disease. If the right to left shunt is significant pulmonary hypertension can develop (the Eisenmenger phenomenon).

What is total anomalous pulmonary venous return (TAPVR)?

TAPVR results when all pulmonary veins have abnormal drainage versus PAPVR where only some of the pulmonary veins drain anonymously. TAPVR is a cyanotic congenital heart entity (one of the 5 T's). In TAPVR, all of the pulmonary veins connect to a structure that will eventually drain into the right atrium and death will ensue unless there is a coexisting right to left shunt via either a large patent foramen ovale or atrial septal defect. As in PAPVR, there is supracardiac, cardiac, infracardiac, and mixed patterns of TAPVR. Supracardiac is most common. On imaging look for right heart enlargement due to the increased right heart blood flow with normal left atrial size.

What is the classic descriptor on chest radiograph of supracardiac TAPVR?

The snowman appearance on a frontal chest x-ray is characteristic of supracardiac TAPVR. The snowman appearance results from a dilated vertical vein, superior vena cava, and brachiocephalic vein on top—all of which form the snowman's head, with the enlarged right atrium forming the body.

True or false? TAPVR is associated with asplenia?

True. TAPVR is associated with heterotaxy and asplenia.

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Episode 3

Patent ductus arteriosus involves a patent connection between which structures?

Patent ductus arteriosus involves patency of the duct between the aorta and the pulmonary artery/pulmonary arterial system.

Within what time from does the ductus arteriosus normally close?

Approximately 24 to 48 hours after birth the ductus arteriosus will functionally close. Anatomic full closure can take about a month.

Why can a patent ductus arteriosus potentially be lifesaving with certain congenital cardiac anomalies?

In certain congenital cardiac anomalies, maintaining persistence of the ductus arteriosus can be key for survival or well-being of the infant as this allows blood to flow between the aorta and pulmonary arterial system. In cases of tetralogy of Fallot, pulmonary atresia, hypoplastic left heart, and Eisenmenger syndrome, this aberrant blood flow pathway through the patent ductus arteriosus can be beneficial.

What types of treatments can be considered to close a patent ductus arteriosus?

Treatments include endovascular coiling/closure, surgical clipping or ligation, or medical therapy.

What medical therapy exists to help keep the ductus arteriosus open and what medical therapy exists to help close the ductus arteriosus?

To help keep the ductus arteriosus open, prostaglandin E1 can be used.

To help close the ductus arteriosus, indomethacin can be used.

Note on board exams, associations to keep in mind with patent ductus arteriosus include prematurity, cyanotic heart disease, and maternal rubella.

With patent ductus arteriosus, imaging appearance can vary depending on the direction of blood flow through the ductus and the presence of co-existing cardiac abnormalities. However, in an uncomplicated case, imaging may show cardiomegaly with left heart enlargement, AP window obscuration, and pulmonary edema.

For atrial septal defect, is the ostium primum or secundum type most common?

Secundum atrial septal defects are most common. With atrial septal defects, secundum is #1, NOT #2 as the name could suggest.

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True or false: Secundum atrial septal defects often do not close on their own.

False. Secundum atrial septal defects often do close on their own. Primum atrial septal defects are less likely to close on their own and can also be difficult to close with device closure as they are often close to atrioventricular valvular tissue.

Note some key associations with atrial septal defects include:

Ostium primum atrial septal defect: Down's syndrome

Sinus venosus atrial septal defect: partial anomalous pulmonary venous return

The egg on a string appearance on a frontal chest radiograph is classic for which entity?

Transposition of the great arteries (classic for D-TGA). Top cause of cyanosis in early neonatal life, often related to closure of the patent ductus arteriosus and/or patent foramen ovale which close after birth and help provide blood flow between the pulmonary and aortic circulation. Risk factors include maternal diabetes. D-TGA requires surgery to correct. L-TGA often doesn't require surgery but is "congenitally corrected".

On board exams, if you are shown a frontal radiograph of an infant that has cyanosis and a right aortic arch, what are the top two congenital cardiac anomalies you should first consider?

Truncus arteriosus and tetralogy of Fallot. If there is increased pulmonary vascularity you would suspect truncus arteriosus. If decreased pulmonary vascularity you would expect tetralogy of Fallot.

We already covered the 5 T's of cyanotic congenital heart disease on the prior episode. What are the most common differential considerations for non-cyanotic congenital heart disease?

Congenital heart disease without cyanosis should most commonly make you consider atrial and ventricular septal defects, patent ductus arteriosus, post-ductal aortic coarctation, and partial anomalous pulmonary venous return.

True or false: Adrenal insufficiency can cause small heart size?

True. Adrenal insufficiency (Addison's) is a top differential consideration for a heart that is too small. Additional considerations can include malnourishment/cachexia of various causes and constrictive pericarditis.

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Congestive heart failure in a newborn should make you think more about right-sided or left-sided cardiac obstruction?

Functional left heart cardiac obstruction is associated with newborn congestive heart failure. Think of entities like pre-ductal aortic coarctation, mitral stenosis, aortic stenosis, hypoplastic left heart, cor triatriatum, and infracardiac total anomalous pulmonary venous return. Note that with total anomalous pulmonary venous return, having a large patent foramen ovale or atrial septal defect aids in survival. Supracardiac total anomalous pulmonary venous return is most common and gives the classic snowman appearance on a frontal chest radiograph. Infracardiac pulmonary venous return is less common, but still shows up a lot on board exams and will show pulmonary edema in a new born.

True or false: Total anomalous pulmonary venous return is associated with asplenia?

True. In fact, nearly all cases of asplenia will have co-existing total anomalous pulmonary venous return. Note that asplenia is essentially bilateral right-sidedness. The majority of these individuals will also have an endocardial cushion defect. Also remember the association between Downs syndrome and endocardial cushion defects.

True or false: Truncus arteriosus essentially always has an associated ventricular septal defect.

True. Truncus arteriosus is a cyanotic congenital anomaly wherein a common trunk provides both the systemic and pulmonary circulation instead of a separate pulmonary artery and proximal thoracic aorta. Truncus arteriosus is classically taught to always have an associated ventricular septal defect. Other high-yield associations include DiGeorge (Catch 22) syndrome aka 22q11.2 deletion syndrome, CHARGE syndrome, and a right aortic arch.

CATCH 22

C=cardiac anomalies

A-abnormal facies

T=thymic aplasia

C=cleft palate

H=hypocalcemia/hypoparathyroidism.

22=22q11.2 deletion

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CHARGE: From CHD7 mutation

C=coloboma (eye formation anomaly)

H=heart defects

A=atresia choanae

R=retarded growth and development

G=genital hypoplasia

E=ear anomalies with possible deafness

True or false: Turner syndrome is associated with post-ductal aortic coarctation.

False. Turner's syndrome is associated with pre-ductal aka infantile aortic coarctation. Adult-type coarctation is post-ductal. In pre-ductal coarctation the patent ductus arteriosus become an important conduit for blood to flow from the pulmonary circulation into the more distal aorta.

Note that aortic coarctation in general is strongly associated with a bicuspid aortic valve and berry aneurysms among other anomalies. Look for rib notching of the 4th to 8th ribs, but not 1st and 2nd ribs as they have separate blood supply from the costocervical trunk as well as the "figure 3 sign" on a frontal chest radiograph from the aortic anomaly.

Note that Turner syndrome is also highly tested on board exams. Other Turner associations include horseshoe kidney, gonadal dysgenesis, hypothyroidism due to thyroid antibodies, antenatal cystic hygroma on ultrasound, etc.

Note:

Content I haven't covered on these episodes includes surgical repair/correction for many of these congenital defects. Studying the surgical repair options is best with visual diagrams and you can find plenty of info on this in existing board prep books and online. You should be comfortable with the options and imaging appearances of surgical repair of transposition of the great arteries and tetralogy of Fallot, among other congenital cardiac anomalies.