Congenital Aortic Anomalies

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16 mo with chronic cough
16 mo with chronic cough
Question 1

Which vessel configuration is **LEAST** likely based on the preceding images?

1. Right aortic arch with an aberrant left subclavian artery
2. Pulmonary sling
3. Double aortic arch
4. Left arch with an aberrant right subclavian artery
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Double Aortic Arch
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During embryonic vascular development, which aortic/branchial arch contributes to the normal left-sided aortic arch?

1. II arch
2. III arch
3. IV arch
4. VI arch
During embryonic vascular development, which aortic/branchial arch contributes to the normal left-sided aortic arch?

1. II arch
2. III arch
3. IV arch
4. VI arch

Answer 2
Aortic Arch Development

Third arches: carotid arteries
Fourth arches: aortic arch (AA) on the left side, BCA on right
Sixth arches: right & left PA’s and the ductus arteriosus (DA)
Seventh intersegmentals: subclavian arteries
First, second, and fifth arches involute.

Based on Edwards’ hypothetical embryonic double aortic arch model
Anatomy:

• double arches encircle the esophagus and trachea (most common vascular ring)
• various forms, most commonly right arch is dominant with a more superior apex and descending aorta on the left

Epidemiology:

• vascular rings comprise 1% of surgically managed cardiovascular malformations (about half of those are double arches)
• no gender or race predilection
Clinical presentation:

- respiratory difficulties like stridor but also including apparent life-threatening events
- dysphagia, vomiting, choking, usually >2 yrs of age

Associations:

- usually without other cardiovascular anomalies
- 20% have chromosome band 22q11 deletion
Double Aortic Arch

Imaging:

- Chest Radiographs - may show indentation of trachea or indeterminate arch sidedness
- Esophagram - bilateral and posterior indentation of esophagus
- CT - anatomy, determine dominant arch for surgical planning (contralateral thoracotomy), evaluate airway compression
- MRI - anatomy, as CT
Right Aortic Arch with Aberrant Left Subclavian Artery
Right arch w/ ab LSC

interruption of the dorsal segment of the left arch between the LCCA and LSCA with regression of the right ductus arteriosus
Right Aortic Arch w/ ab LSC

- left ligamentum arteriosum can form a vascular ring

- Kommerell’s diverticulum - remnant of left dorsal aortic root

- can be associated with a midline descending aorta

- Two major regions of potential airway compression
  1. level of arch and ab subclavian artery
  2. level of carina or main bronchi
9 yo with headaches
Aortic Coarctation

Focal
Long Segment w/ Collaterals
Which of the following is a characteristic of aortic coarctation?

1. Reverse figure-of-three sign on frontal chest radiograph
2. Associated with berry aneurysms of the circle of Willis
3. First and second rib notching secondary to collateral vessels
4. Right ventricular hypertrophy
Which of the following is a characteristic of aortic coarctation?

1. Reverse figure-of-three sign on frontal chest radiograph

2. Associated with berry aneurysms of the circle of Willis

3. First and second rib notching secondary to collateral vessels supplied by costocervical trunk

4. Right ventricular hypertrophy
Aortic Coarctation

Pathophysiology:

- defect in vessel media, posterior infolding which may be circumferential
- located near the ductus arteriosus
- can have tubular hypoplasia of arch
- post-stenotic dilation
- collateral vessels (intercostal and internal mammary)

Epidemiology:

- 5-10% of all congenital cardiac lesions
- untreated - < 20% survive to age 50 yrs
- males > females up to 2:1
Aortic Coarctation

Clinical presentation (depends on severity):

• infancy - CHF, acidosis, poor perfusion of lower body

• post infancy - asymptomatic, murmur, htn, HA, leg cramps, cold feet

Associations:

• Turner’s syndrome (XO)
• Bicuspid Aortic Valve
• VSD
• PDA
• Berry aneurysms of the circle of Willis
Aortic Coarctation

Imaging:

• Chest Radiographs - “3” sign, rib notching, edema, CM
• Esophagram - reverse “3” sign
• CT - anatomy (site, severity, morphology, collaterals)
• MRI
  anatomy
  gradient (peak velocities)
  collateral flow using phase contrast techniques
  evaluate for bicuspid AV, VSD, or LVH
6 yo with dysphagia
6 yo with dysphagia
Cervical Arch with Aneurysm

- perhaps formation of aortic arch from third rather than fourth arch or lack of caudal migration of 4th arch

- rare anomaly but with 20% complicated by aneurysm

- often asymptomatic, can have vascular ring-like symptoms or supraclavicular pulsatile mass
6 day old with abnormal echo
Question 4

Which type of interrupted aortic arch is demonstrated?

1. Type A
2. Type B
3. Type C
4. Type D
Which type of interrupted aortic arch is demonstrated?

1. Type A
2. Type B
3. Type C
4. Type D

Answer 4
Interrupted Aortic Arch

33-42% Type A
53-65% Type B
1-4% Type C

Classification by Celoria and Patton
Interrupted Aortic Arch

Anatomy:
- complete discontinuity between ascending and descending aorta
- simple or complex
  - simple - VSD and PDA
  - complex - truncus, TGA, double outlet RV

Epidemiology:
- 1% of congenital heart disease
- 25% have DiGeorge syndrome
Interrupted Aortic Arch

Clinical presentation:

• heart failure (particularly with closure of ductus arteriosus), cyanosis, poor peripheral pulses

Imaging:

• Chest Radiographs - CM with increased pulm vascularity and edema in critically ill newborn midline trachea and inconspicuous aortic knob

• CT - anatomy, determine type

MRI - anatomy, CT
17 yo with scoliosis
Question 6

Which is often associated with Marfan syndrome?

1. Coagulopathy
2. Arterial tortuosity
3. Mitral valve prolapse
4. Coronary artery aneurysm
Which is often associated with Marfan syndrome?

1. Coagulopathy

2. Arterial tortuosity - Loeys-Dietz feature

3. Mitral valve prolapse

4. Coronary artery aneurysm - Kawasaki’s feature
Marfan Syndrome

• **Definition:** Inherited connective tissue disorder with an autosomal dominant transmission

• **Incidence:** 1 per 5000

• 25% represent new mutations

• **Fibrillin protein dysfunction which prevents proper microfibril formation.**

• **Premature medial degeneration with risk of aortic dilation and subsequent dissection or rupture**
Marfan Syndrome

Imaging:

screening and surveillance with MRA
evaluate for progressive aortic enlargement and complications
References:


