Anatomical spectrum of coarctation

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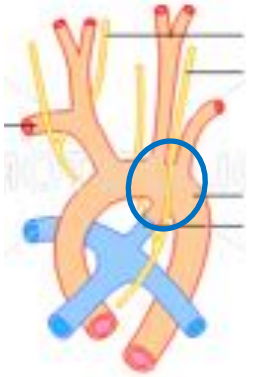
Coarctation of the aorta
Definition

International Society for Nomenclature of Paediatric and Congenital Heart Disease (ISNPCHD)

- "A congenital cardiovascular malformation of the aorta in which there is a discrete luminal narrowing of the aorta (below the lower limit of normal adjusted for body size).
- Usually just distal to the left subclavian artery, at the junction of the arterial duct and the aortic arch,
- But also anywhere in the thoracic or abdominal aorta.
- Aortic coarctation can be associated with varying degrees of proximal and distal aortic hypoplasia."



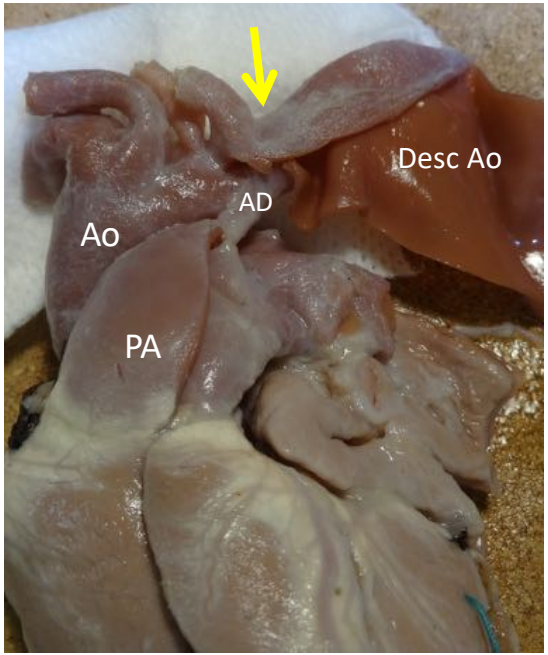
Nomenclature for congenital and paediatric cardiac disease: The IPCCC and the ICD-11.
Franklin RCG et al. Cardiol Young 2017;27:1872-1938.

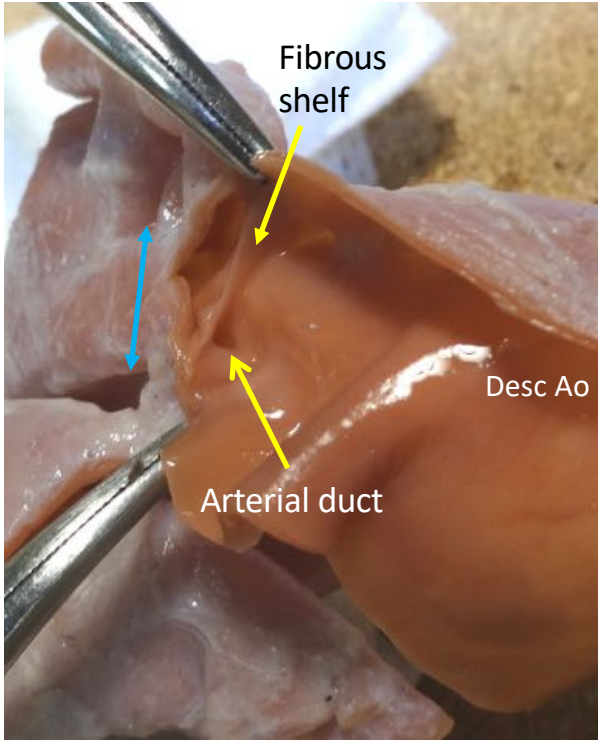
Coarctation of the aorta
Two major anatomic forms

- « Infantile type » = hypoplastic Ao arch
  - Infants, antenatal
  - Hypoplastic (tubular) aortic arch
  - The descending aorta is supplied by the arterial duct (ductal dependency)
  - Secondary to LVOT obstruction (flow lesion)
- « Adult type » = isolated type of CoA
  - Also frequent in infants !!
  - Discrete isthmic narrowing
  - The descending aorta is supplied by the ascending aorta
  - Isolated or minor intracardiac anomalies (BAV)

Coarctation : isolated type

Coarctation : isolated type



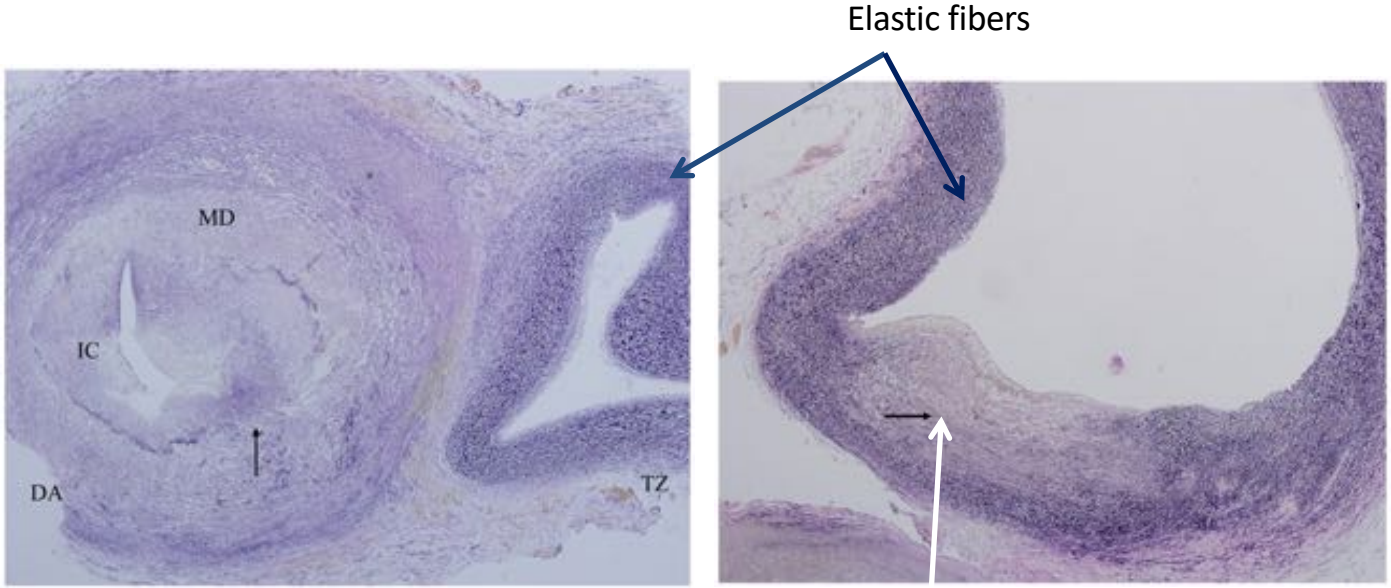


Coarctation : the role of the ductal tissue

- Ductus : smooth muscle cells
- Aorta : elastic fibers but no SMC
- 15 infants with CoA:
  - Abnormal extension of ductal tissue (>50% of the total circumference) in the CoA segment in 11/15
  - Enhanced apoptosis with cystic medial necrosis : risk of aneurysm formation

Kim JE et al. Int J Cardiol 2010;145:177-8

Coarctation : the role of the ductal tissue



Ductus arteriosus

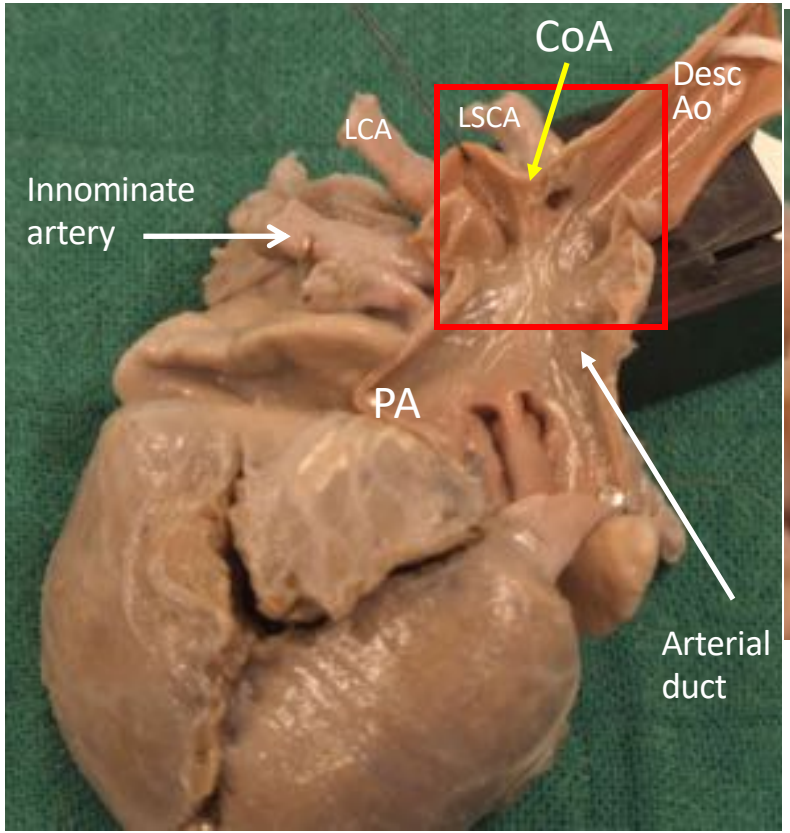
Aorta

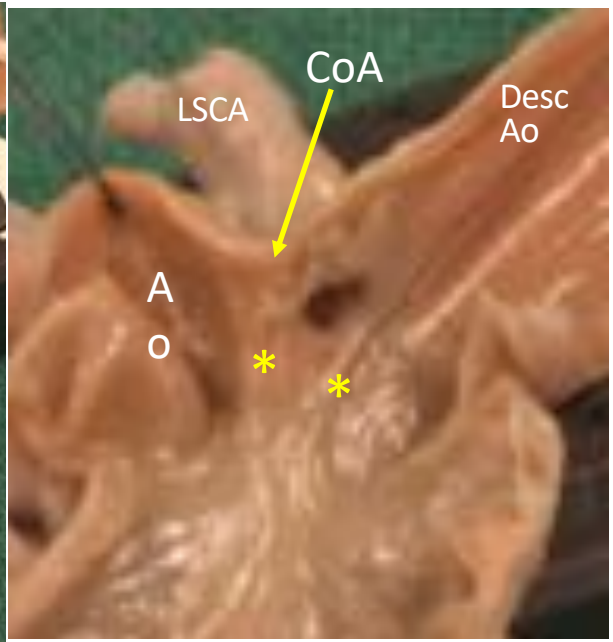
Ductal tissue in the aortic media :
« transition zone »

« Considerable amount of ductal tissue at coarctation site and transition zone »

Kim JE et al. Int J Cardiol 2010;145:177-82

Coarctation and ductal tissue



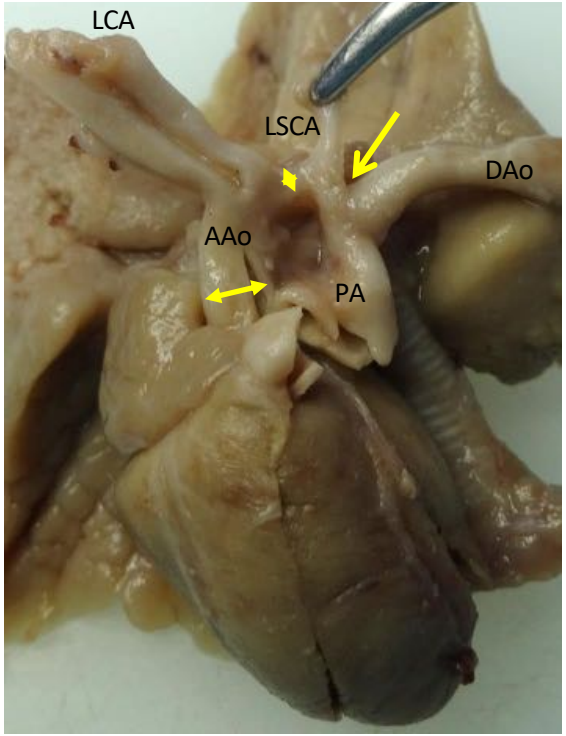


* Fibrous shelf

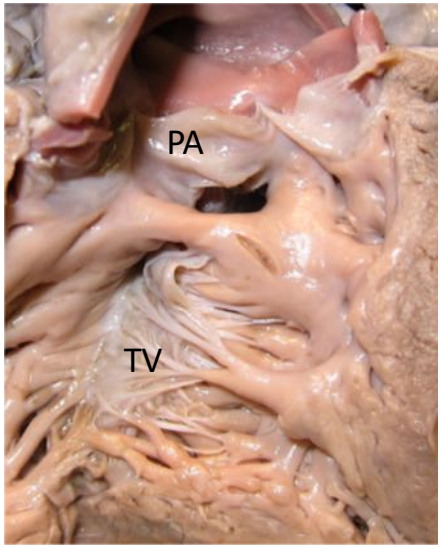
Coarctation with hypoplasia of the aortic arch:
Infantile type

Coarctation of the aorta
Tubular hypoplasia of aortic arch

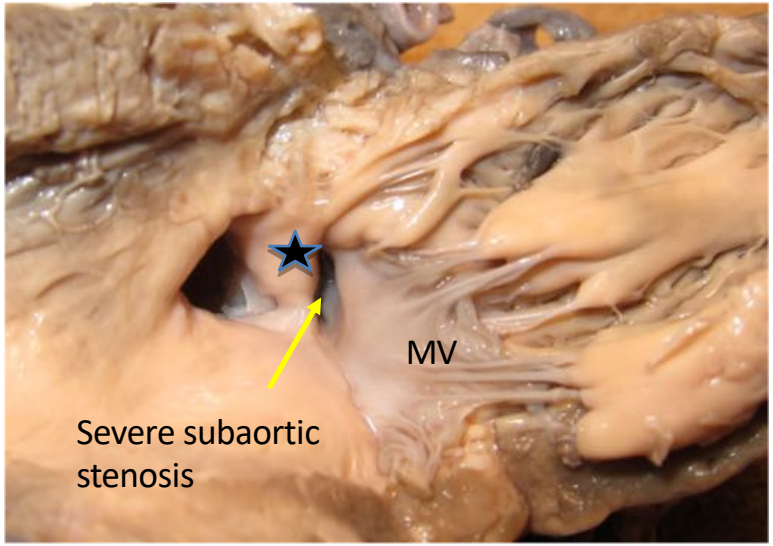
- Always secondary to reduced flow through the aorta in the fetal life
- Two main etiologies
  - Outlet VSD with posterior malalignment of the outlet septum
  - Multilevel left ventricular outflow tract obstruction
- Complex anomalies (DORV, DOLV, DILV…) : subaortic obstruction



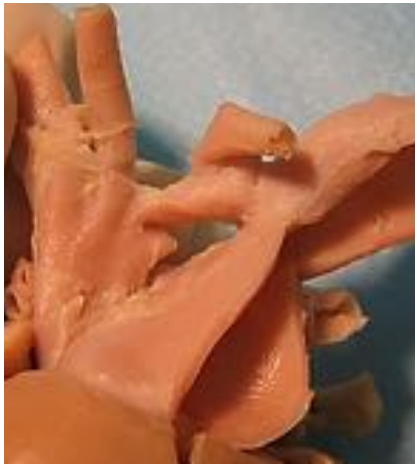
Neonatal coarctation with outlet VSD, posterior malalignment of the outlet septum



Right ventricle

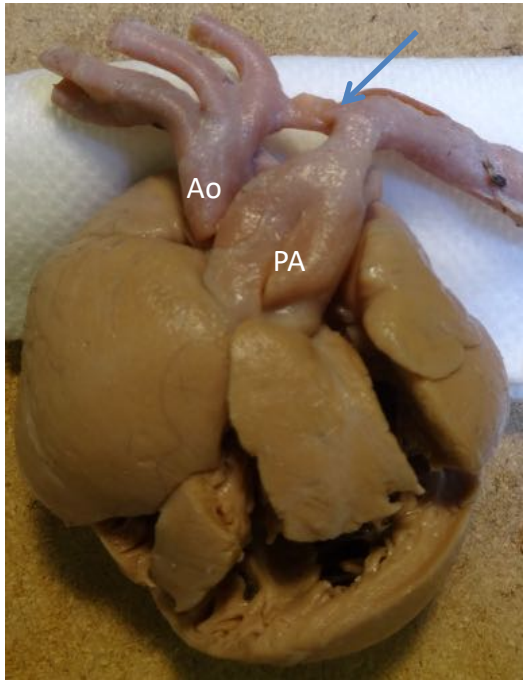


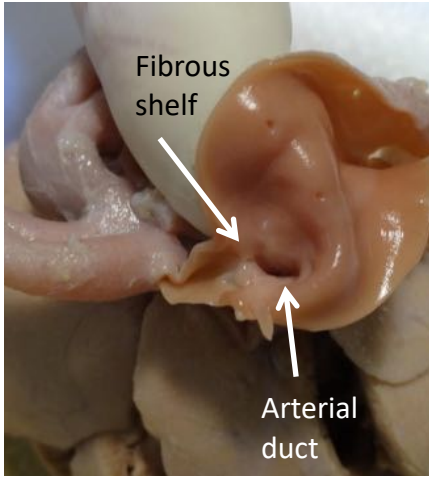
Left ventricle

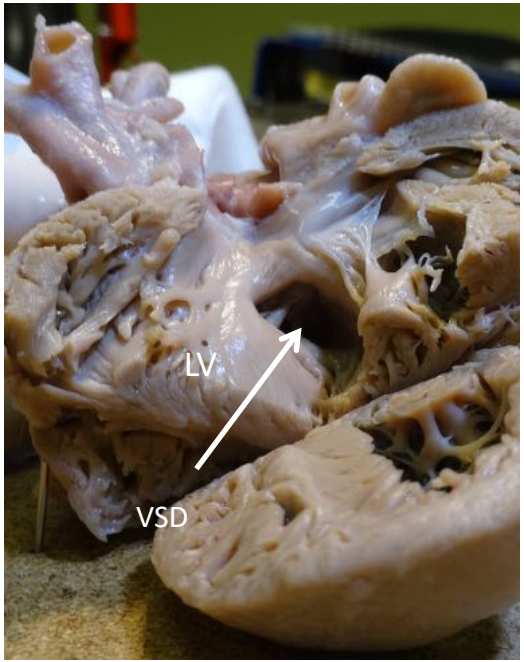


★ Outlet septum

Neonatal coarctation with VSD
Isthmic hypoplasia



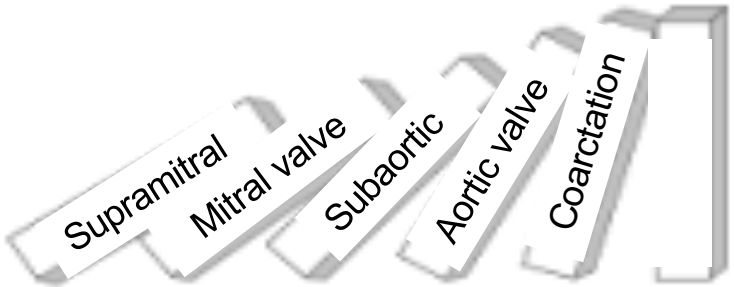




Muscular inlet VSD

Multilevel LVOT obstruction : the Shone 's complex

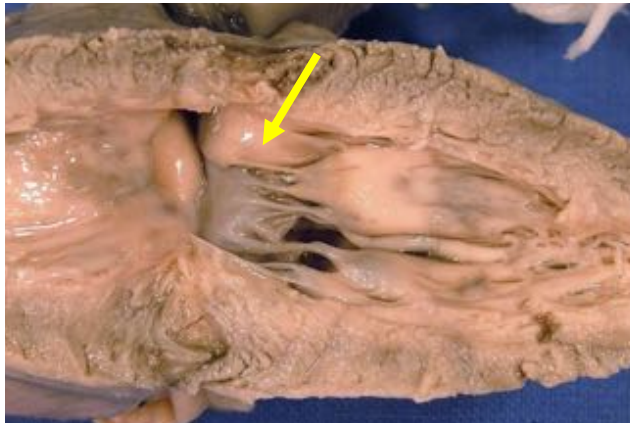
- Combination of obstructive lesions of both inlet and outlet of the LV
  - Supravalvular mitral ring
  - Parachute mitral valve
  - Subaortic stenosis
  - ***Coarctation of the aorta***
- Often incomplete : multilevel LVOT obstruction

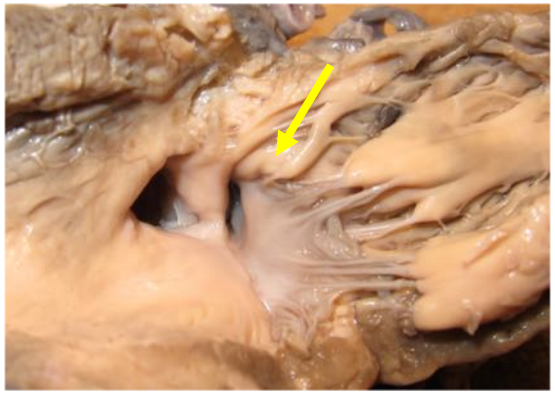


Shone and al. Am J Cardiol 1963

Subvalvular aortic stenosis : the muscle of Moulaert

- Anterolateral muscle bundle of the LV
- Exists in 40% of normal hearts
- Remnant of the ventriculo-infundibular fold
- Can participate in LVOT obstruction





Moulaert AJ et al. Am J Cardiol 1976;37:78-81.

Tubular hypoplasia of the aortic arch
The role of the ductal tissue

- The extension of the ductal tissue does not explain hypoplasia of the aortic arch
- But explains the ductal dependency and the necessity of PGE1
- Kim et al. : 11/15 infants with CoA had an hypoplastic aortic arch

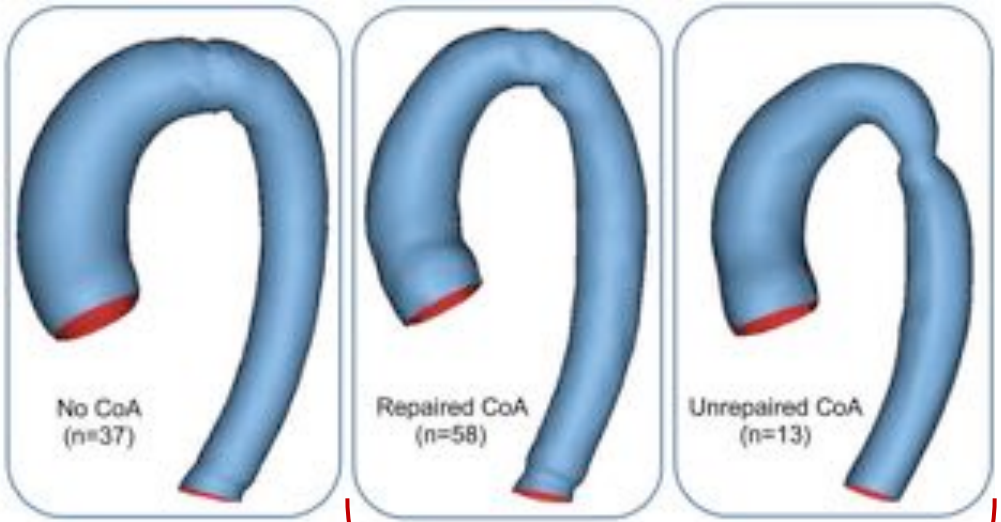
Kim JE et al. Int J Cardiol 2010;145:177-82

Coarctation of the aorta: a disease of the whole aorta

Coarctation : a disease of the whole aorta

3D MRI
108 patients
with bicuspid
aortic valve

Repaired CoA: ascending aorta less dilatated



CoA : more gothic architecture

Sophocleous F. et al Eur J Card Thorac Surg 2019

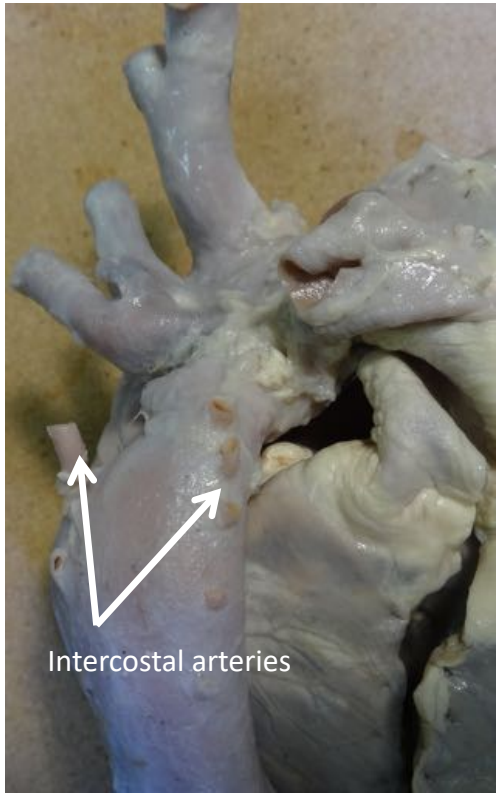
Coarctation and bicuspid aortic valve

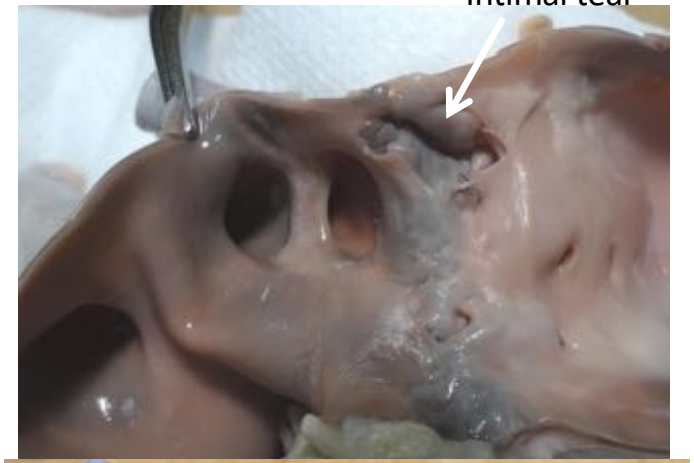
- BAV : the most frequent of all CHD (1-2% of the population)
  - 7% of patients with BAV have aortic CoA
  - But 75% of patients with CoA have BAV
  - Most often R-L BAV

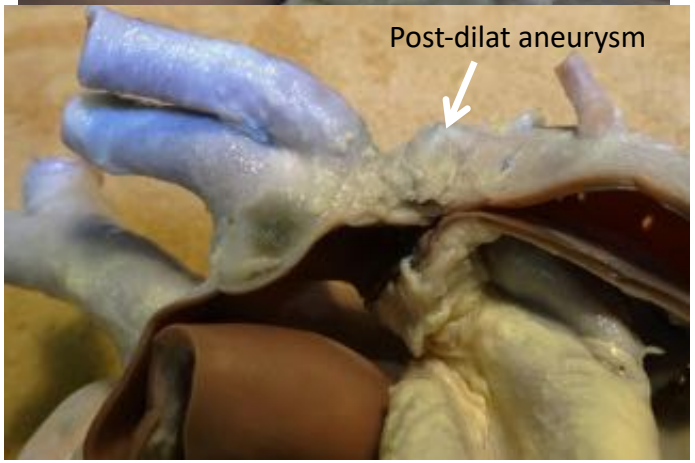
- In both anomalies : alterations of the aortic wall : media smooth muscle and intracellular matrix
  - Aortic aneurysms
  - Risk of dissection and rupture, increased if BAV+CoA

Sinning C. et al. Cardiovasc Diagn Ther 2018

Isolated CoA: balloon dilatation



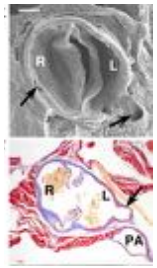




R-N vs R-L bicuspid aortic valve: different embryologic mechanisms

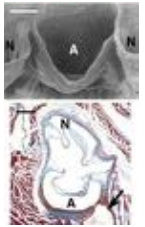
R-N BAV

- ***Abnormal*** formation of the outflow tract cushions / of the aortic valve
- The outflow tract septum is normal
- Aortic stenosis and regurgitation



R-L BAV (75%)

- ***Excessive*** fusion of the lateral endocardial OT cushions
- Anomaly of outflow tract septation
- Neural cardiac crest and second heart field
- Associated with CoA, VSD
- And with dilatation of the ascending aorta



Fernandez B. et al. J Am Coll Cardiol 2009;54:2312-8.

Coarctation : the role of cardiac neural crest

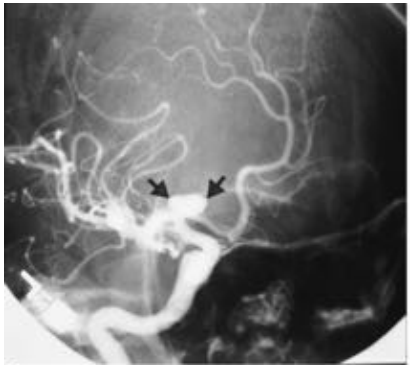
- Neural crest cells : common embryonic origin for :
  - Aortic valve
  - Media of the ascending aorta
  - Media of the intracranial arteries

Shin YW et al. PLoS One 2014
Rosenquist et al. Anat Rec 1990
Pfaltzgraff ER et al. J Mol Cell Cardiol 2014

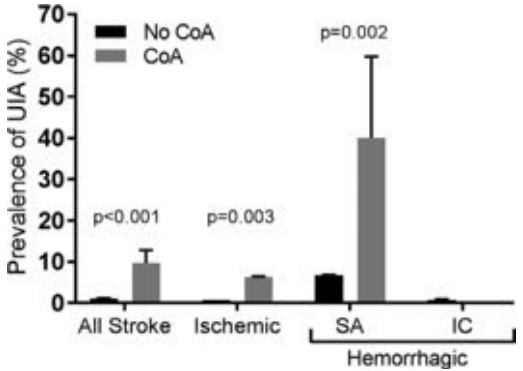
- Similar structure of cross-linked elastin and collagen
- « Neurocristopathy could be an unifying pathogenesis of BAV, coarctation, aortic root dilatation, aortic and cervical arterial dissection due to vascular fragility »
- Descending aorta : different origin (somitic mesoderm)

Coarctation and intracranial aneurysms

- Coarctation is associated with intracranial aneurysms
- National Inpatient Sample 2005-2014:
  - 4 894 582 stroke patients
  - 207 had CoA
- Patients with CoA have :
  - Ischemic stroke 15.9 yrs younger, and hemorrhagic stroke 28.5 yrs younger than the general population
  - More subarachnoid hemorrhage (11.8% vs 4.8%)
  - More unruptured intracranial aneurysms (9.7% vs 1.1%), especially in patients with hemorrhagic stroke (23.3% vs 2.5%)
- Patients with CoA must have a lifelong surveillance
- Screening for intracranial aneurysms



Perloff JK. Am J Cardiol 2010

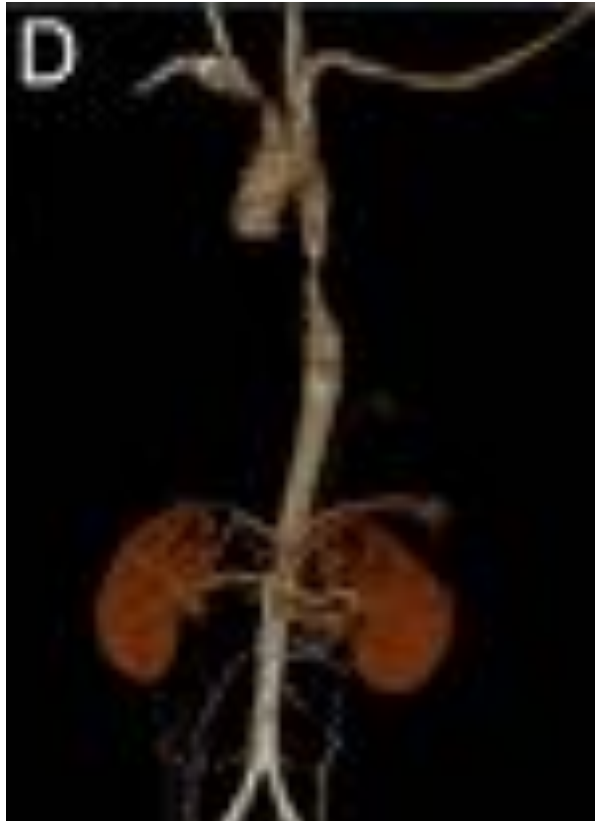


Pickard SS et al. J Am Heart Assoc 2018

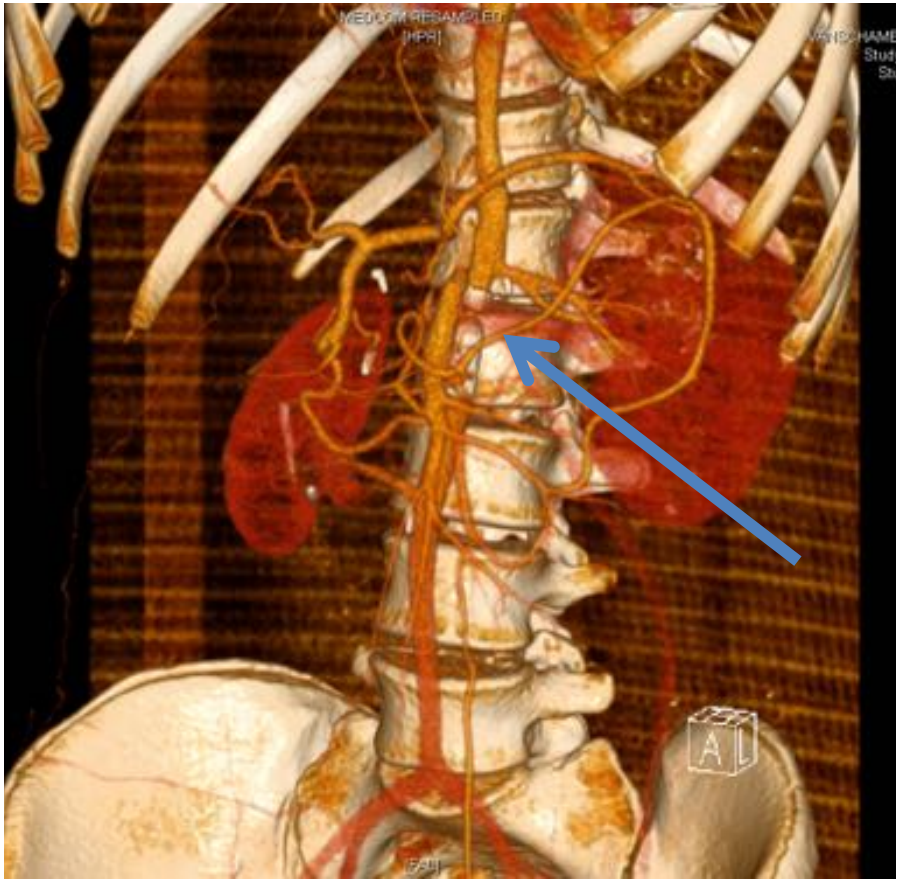
Coarctation of the aorta: anatomic variants

Atypical coarctation: congenital stenosis of the mid-thoracic aorta

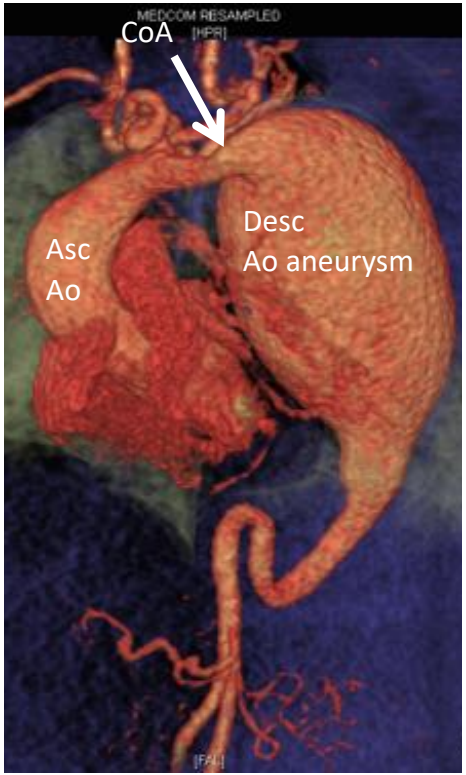
Park HK et al. JACC 2009; 53:2098

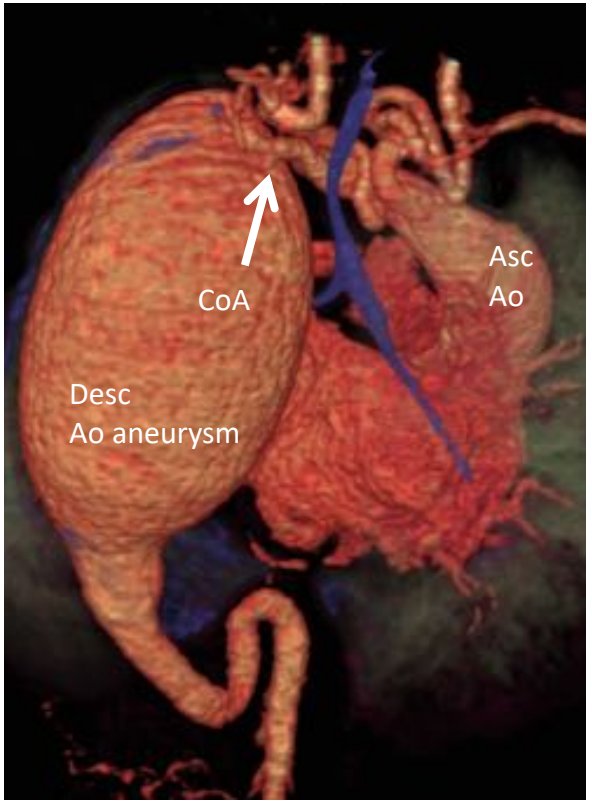


Coarctation of the abdominal aorta

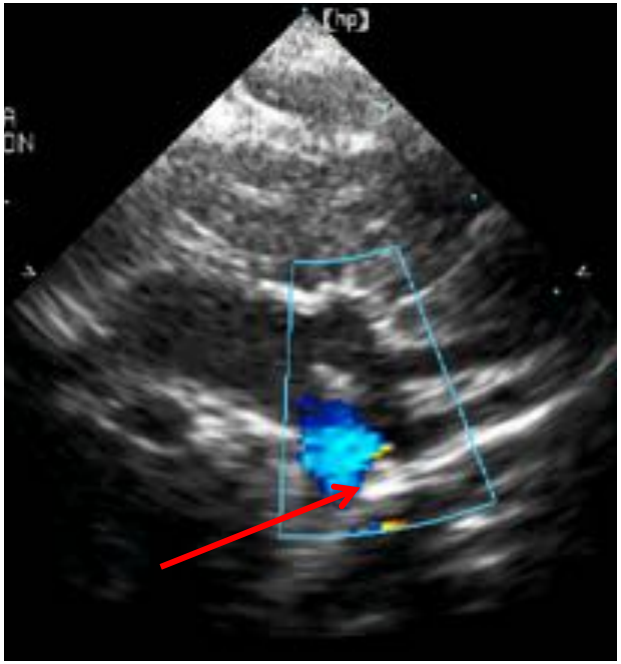


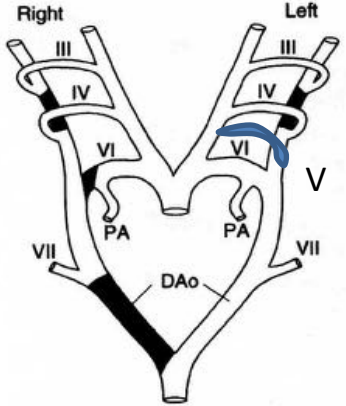
Arterial tortuosity syndrome and coarctation

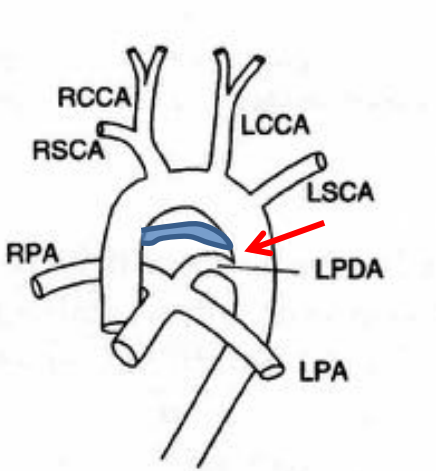




Persistent 5th arch artery





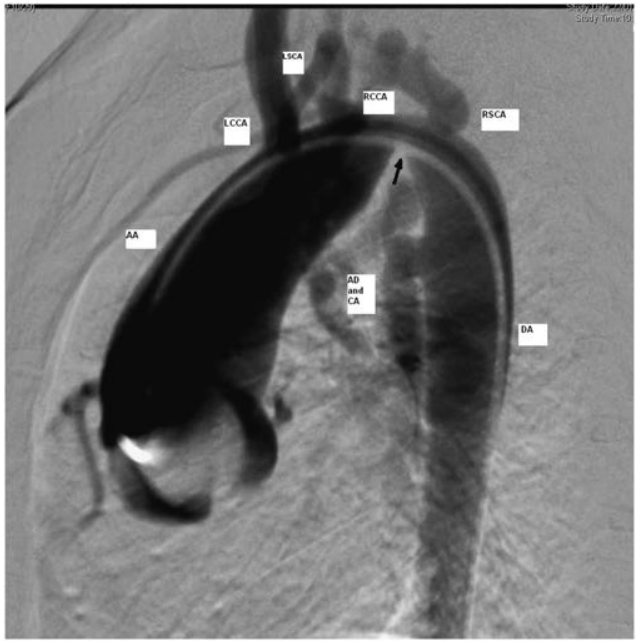


Coarctation and right-sided aortic arch

- Coarctation or interrupted aortic arch with right-sided aortic arch : always between the RCA and the RSCA

  McElhinney et al. Ann Thorac Surg 1999

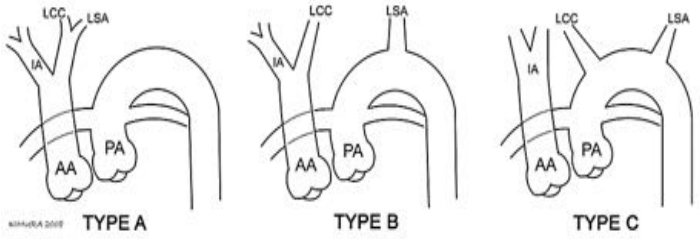
- Tetralogy of Fallot with coarctation
  - Right-sided Ao arch: between RCA and RSCA
  - Left-sided aortic arch: isthmus



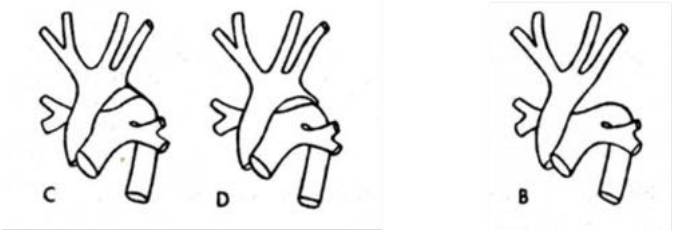
Perdreau E et al, Cardiol Young 2014

Interrupted aortic arch

- IAA type A: extreme form of isthmic coarctation
- Two anatomic types:
  - Atretic aortic arch
  - Absent aortic arch



Celoria and Patton
Am Heart J 1959



Van Mierop
Am J Cardiol 1984

Conclusion

- The infantile type, with hypoplastic aortic arch, is always the consequence of subaortic obstruction, often associated with complex intracardiac anomalies
- So-called isolated coarctation, also usually located at the isthmus, is a disease of the whole aorta including the aortic valve and the intracranial arteries.
  - Life-long surveillance
  - Screening for intracranial aneurysms