Aortic arch anomalies

Coarctation of the Aorta
Interrupted Aortic Arch
Echocardiography

V.Tomek, J. Marek, J. Škovránek, J. Gilík

Kardiocentrum,
University Hospital Motol,
Prague, Czech Republic

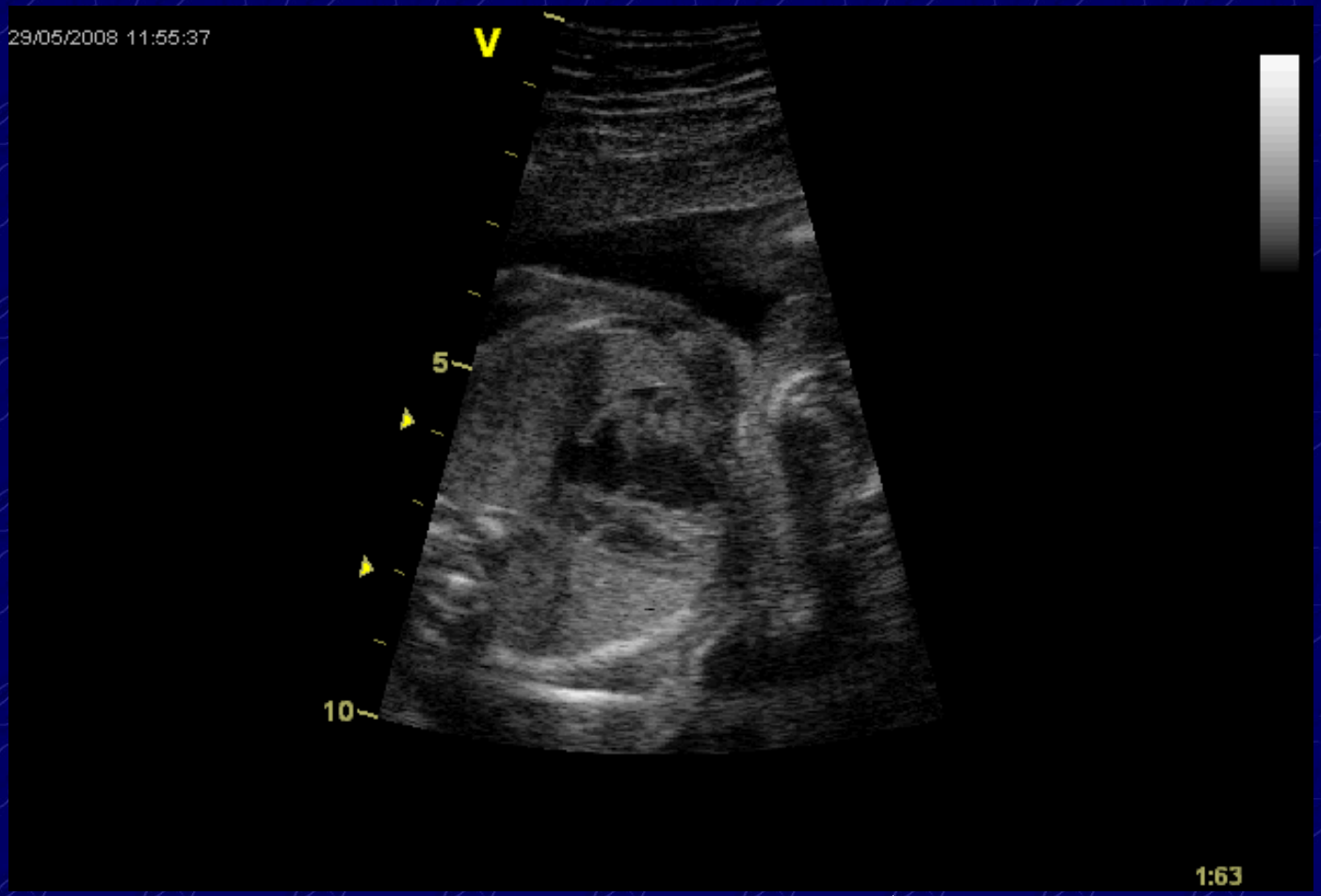
No disclosures

Coarctation of the Aorta

- occurs in 5 – 7% of CHD/ 7th form
- refers to narrowing of the AO isthmus
- circumferential shelf, more prominent along the posterior wall of AO isthmus *(juxtaductal)*
- wide anatomic spectrum, the length of COA varies from discrete to long- segment
- AOA is often elongated/hypoplastic
- >50% complex: + BAO, VSD, AS, MS, SV, TGA, DORV…

Coarctation of the Aorta

Prenatal assessment



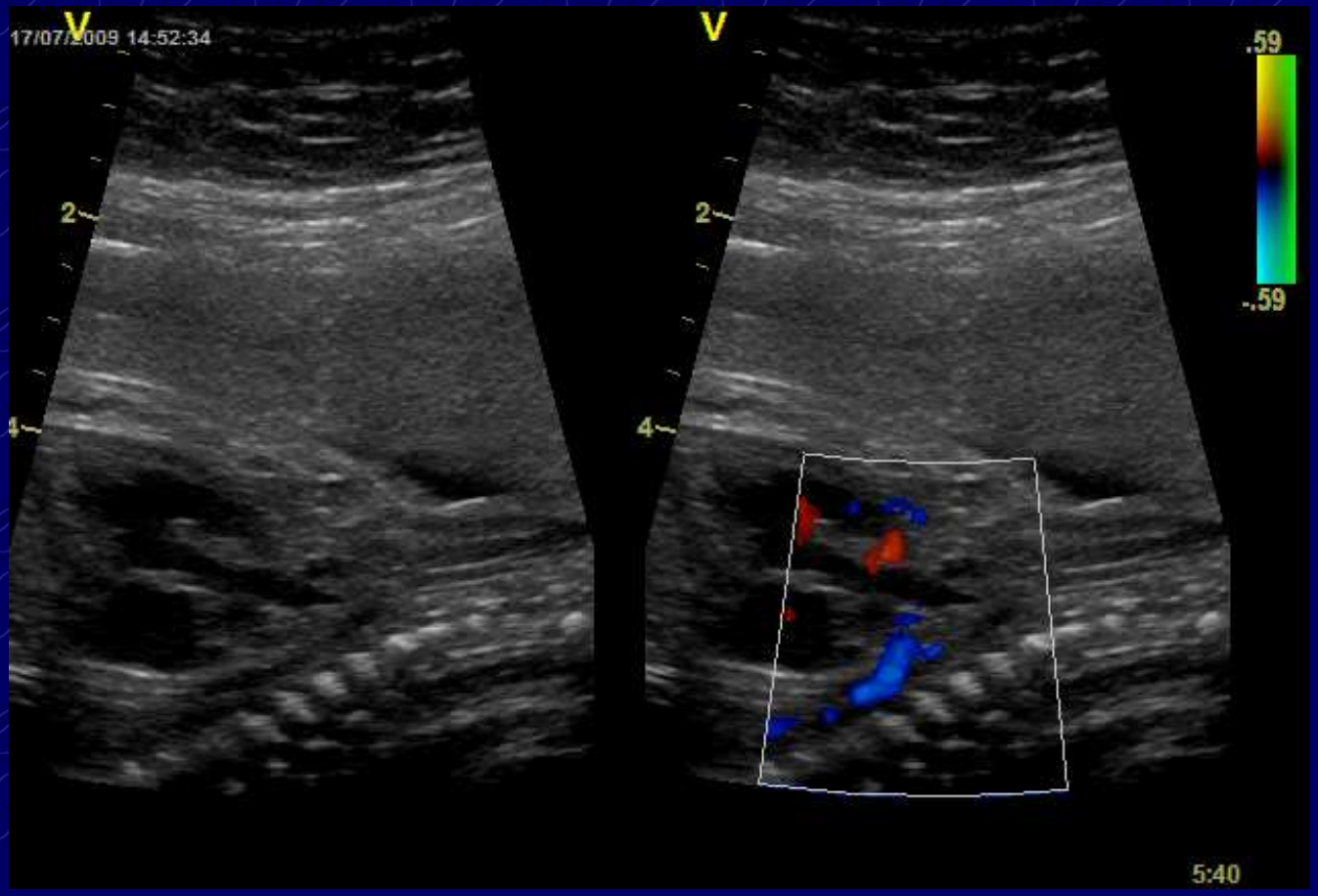
Asymmetry of ventricular size/ ↑LV afterload, ↓output

LSVC/CS – powerful indicator

Pasquini, Heart. 2005

Coarctation of the Aorta

Prenatal assessment

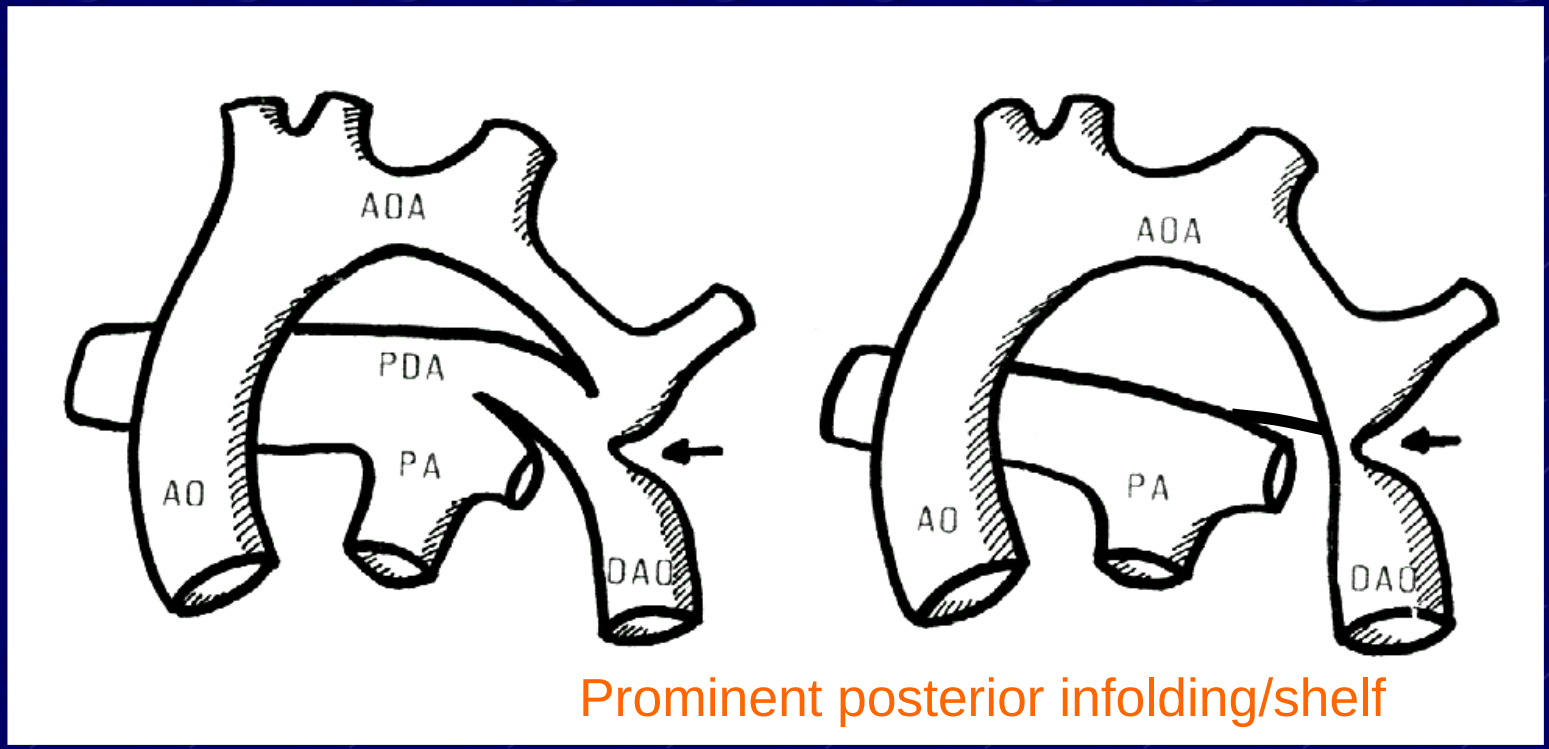


- Oblique sagittal plane
- Transverse arch hypoplasia is freq. associated

Clear depiction of the AO end of the arterial duct

Coarctation of the Aorta

Neonatal (2/3) **Adult (1/3)**



Prominent posterior infolding/shelf

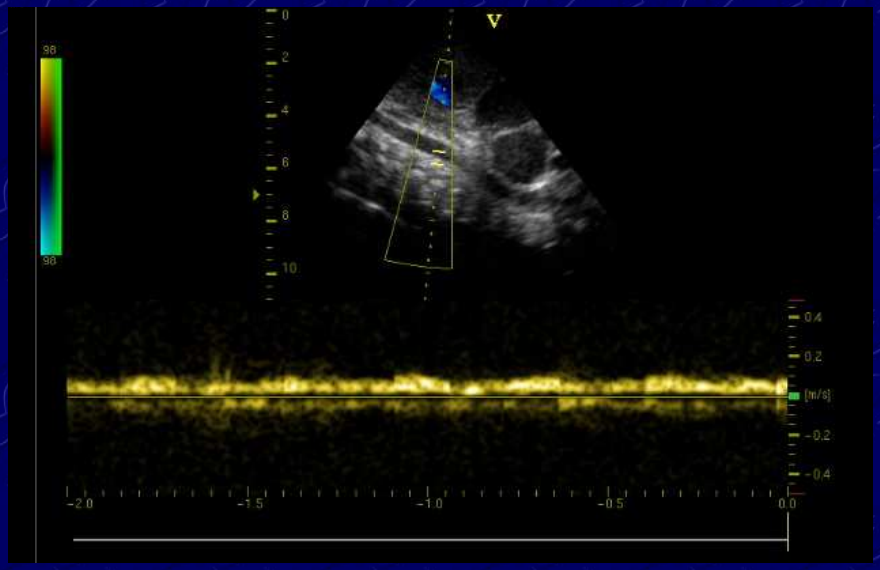
Manifests as the consequence of arterial duct closure

Coarctation of the Aorta

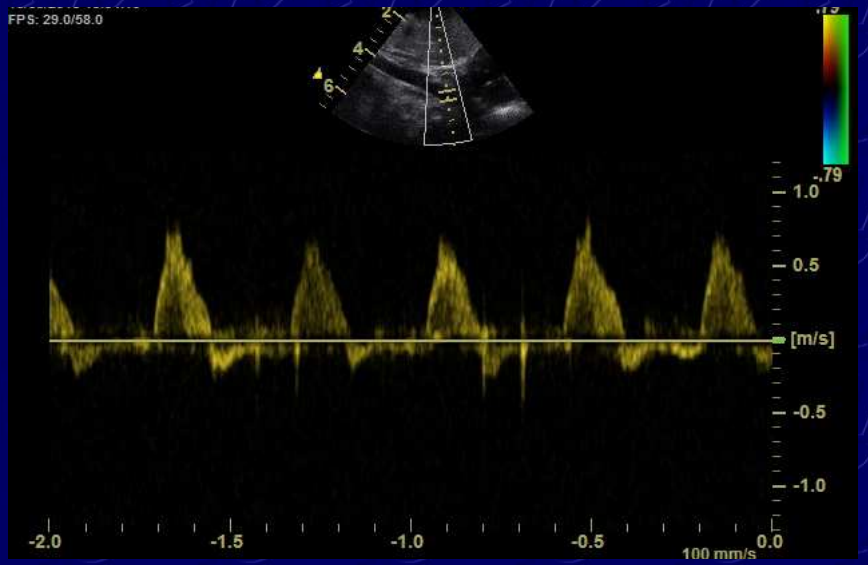
Pulsed Doppler flow pattern in abdom. AO

1. - abdominal situs determination

Critical COA



Closed PDA



Opened PDA

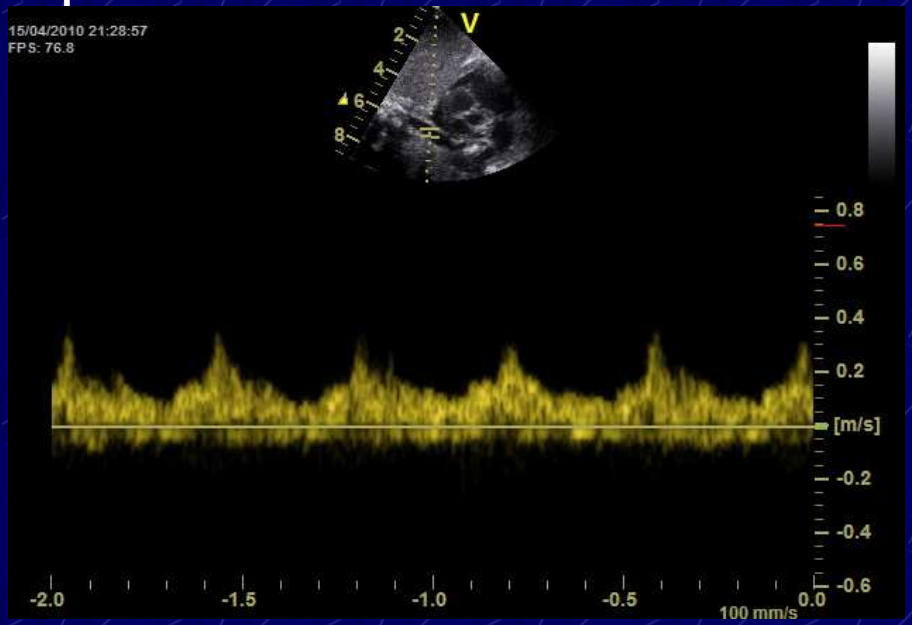
Low flow velocity
Minimal phasic variation
Indicating↓body perfusion

Slightly decreased or normal amplitude
Diastolic component is reversed or normal in case of PH

Coarctation of the Aorta

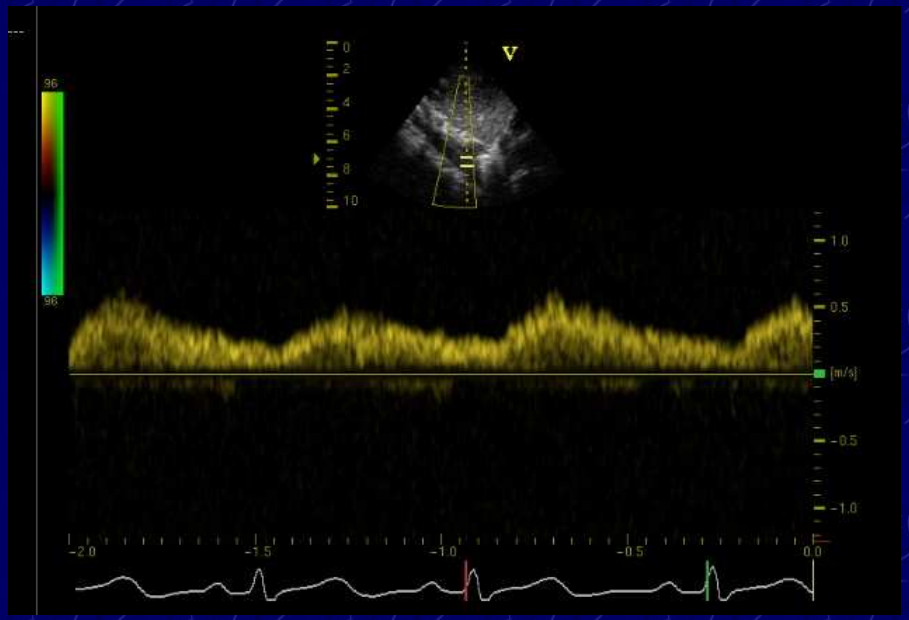
Pulsed Doppler flow pattern in abdom. AO

COA + restrictive PDA,
preserved LV function



Low syst.wave amplitude

COA – 5 years old

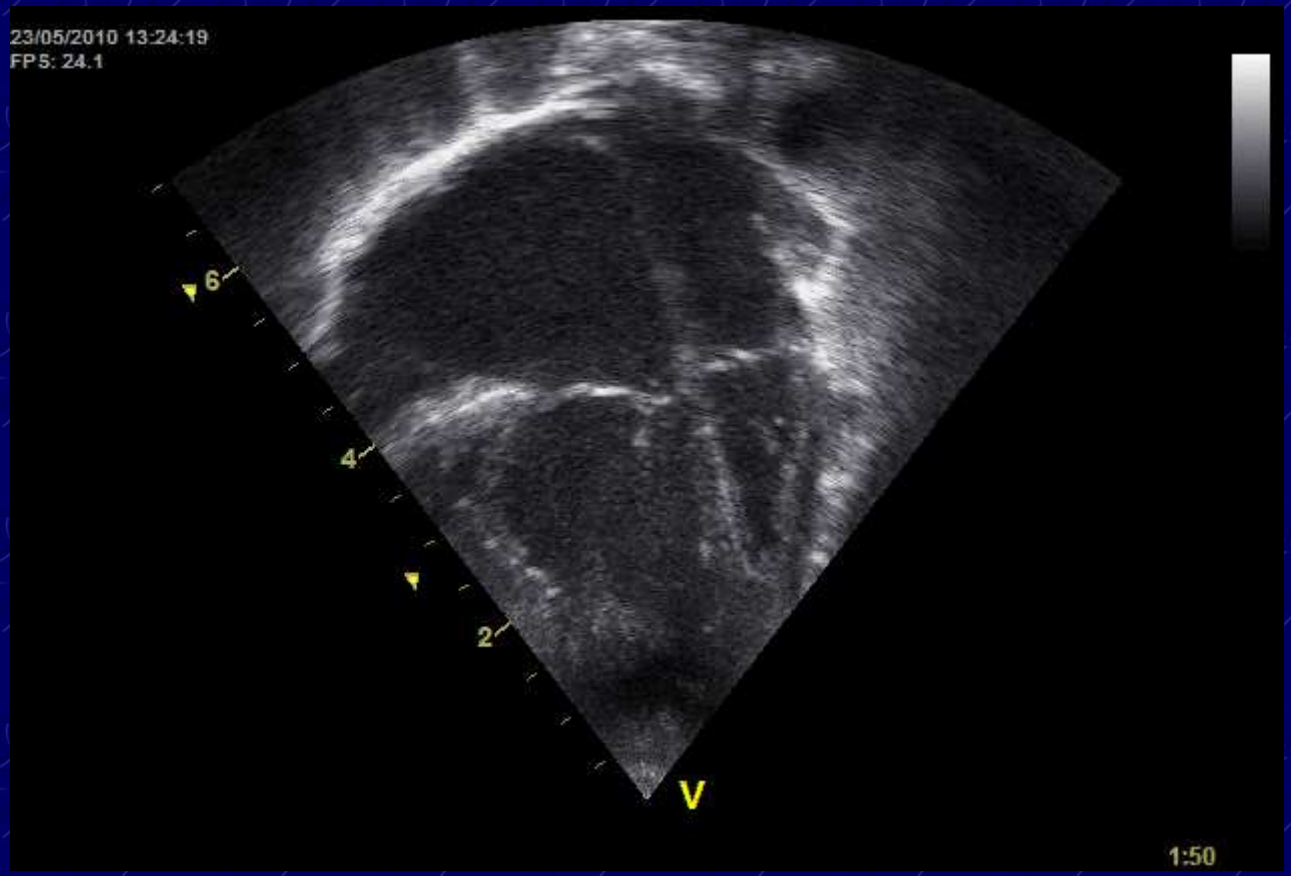


Presence of collaterals

Antegrade diast.flow (= prox.obstr.)

Coarctation of the Aorta

The function and morpholgy of LV

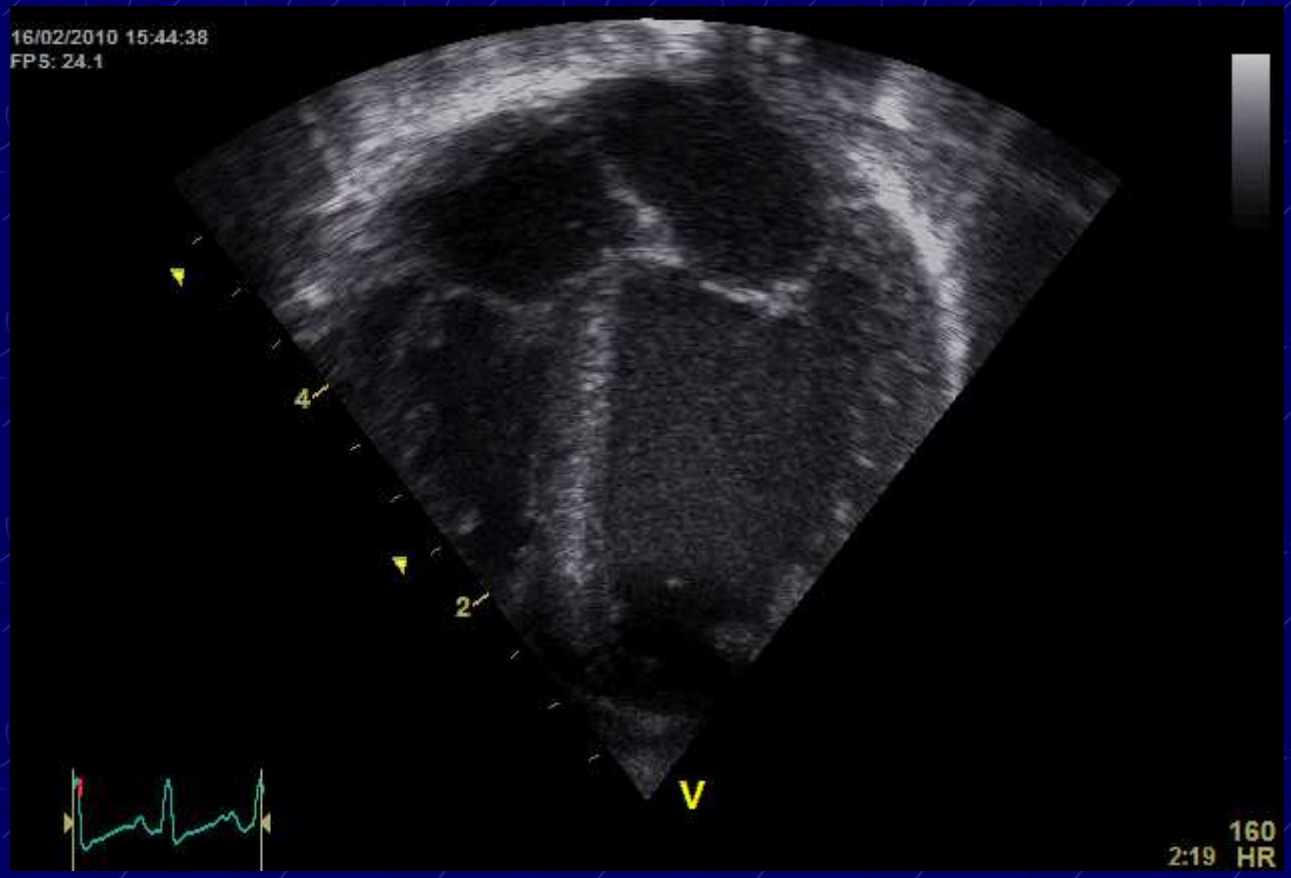


Subnormal volume, enlarged RV, TR, PH

↓ mitral and aortic size

Coarctation of the Aorta

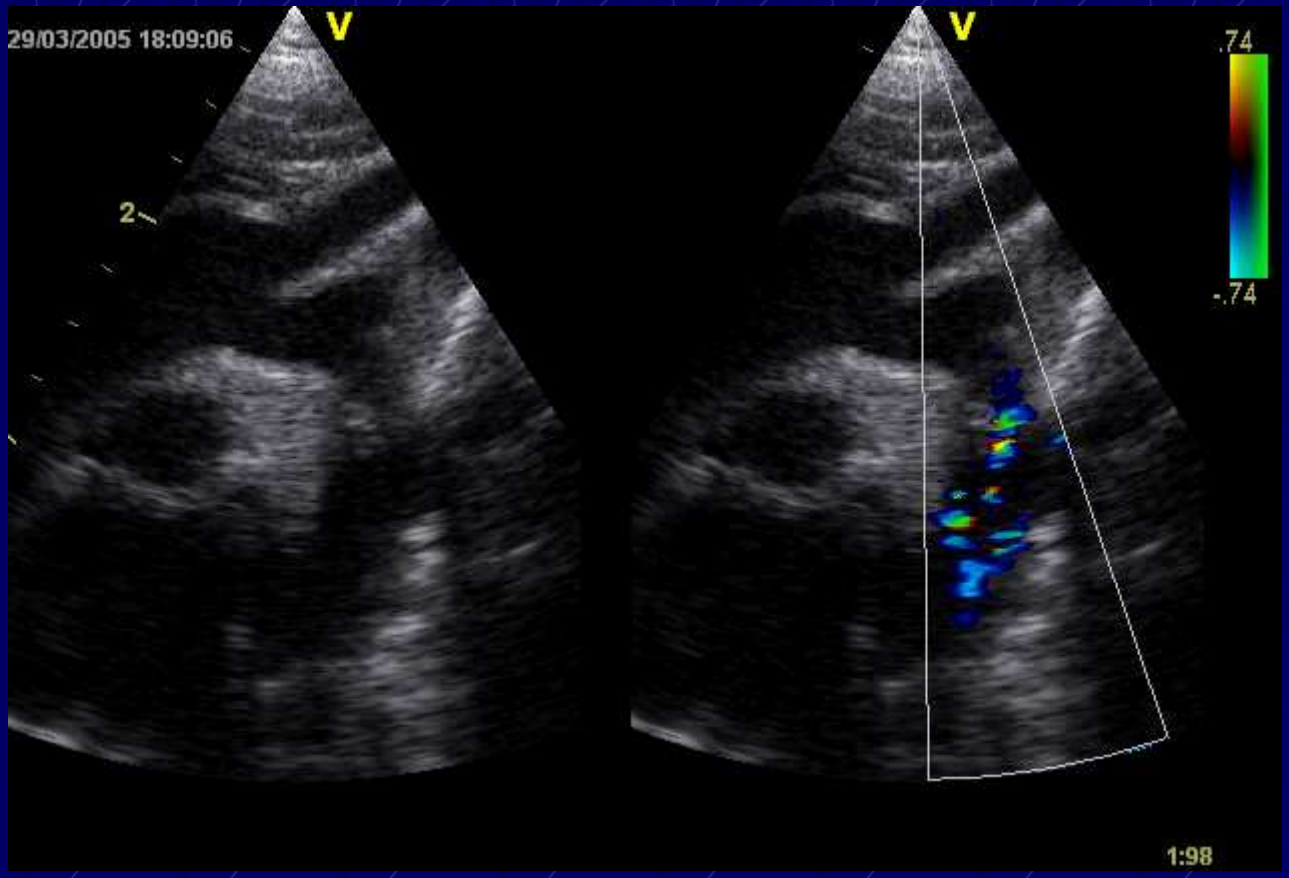
The function and morpholgy of LV



Normal/ enlarged volume, no PH, LV hypertrophy

Coarctation of the Aorta

Imaging of the AOA and isthmus



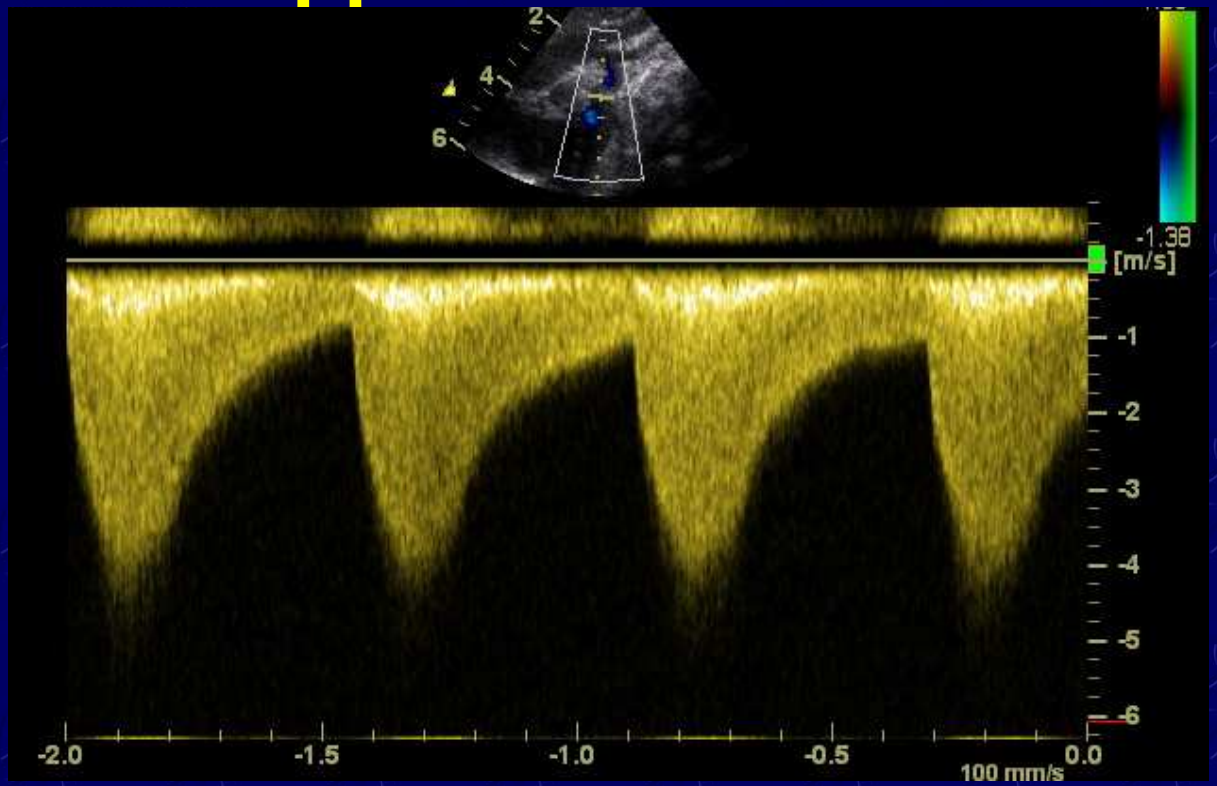
- Suprasternal notch view
- Oblique sagittal plane

Severe form, normal/ decrease LV function

Coarctation of the Aorta

Imaging of the AOA and isthmus

CW Doppler



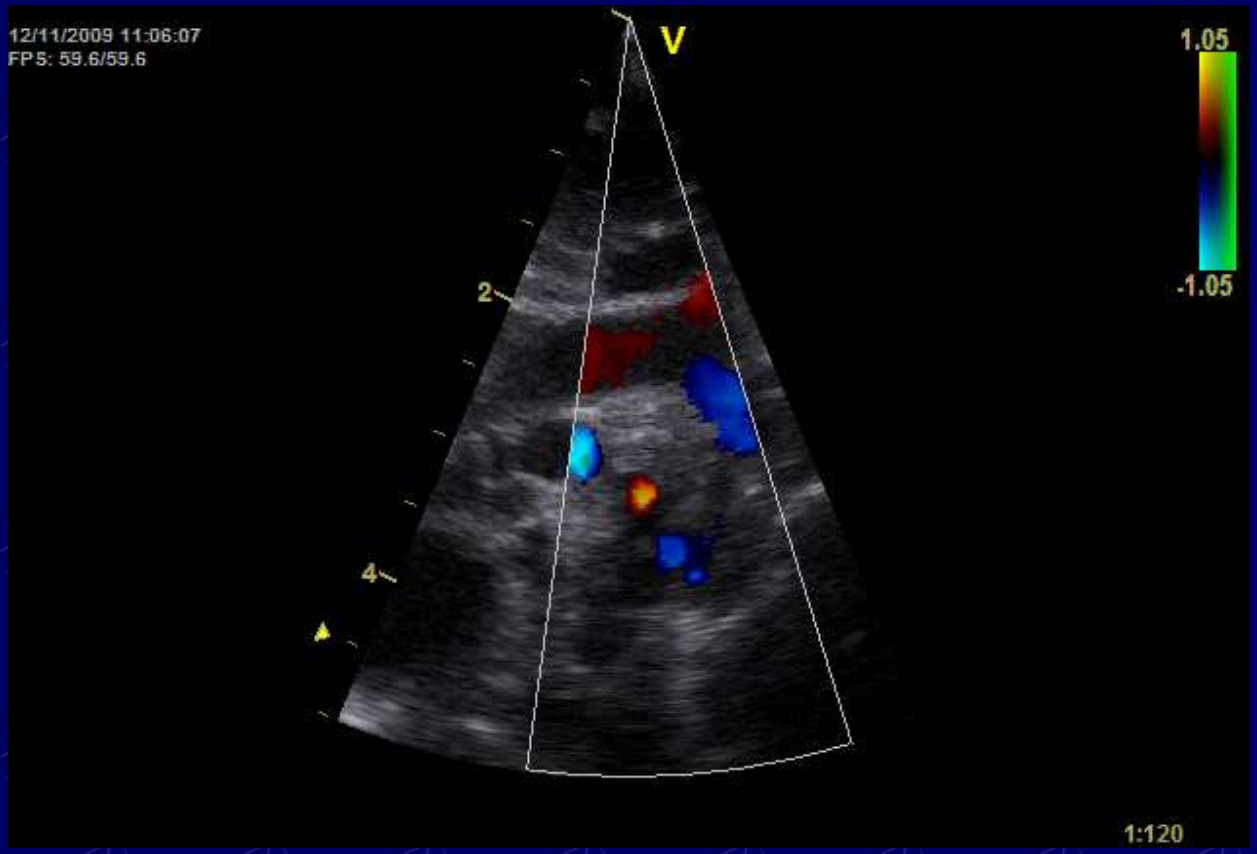
- „Serrated“ pattern, rapid accel.,early systolic peak, gradual diastolic deceleration continuous antegrade flow throughout diastole

↑ *velocity – preserved syst. function, restrictive/no PDA*

Coarctation of the Aorta

Imaging of the AOA and isthmus

Critical



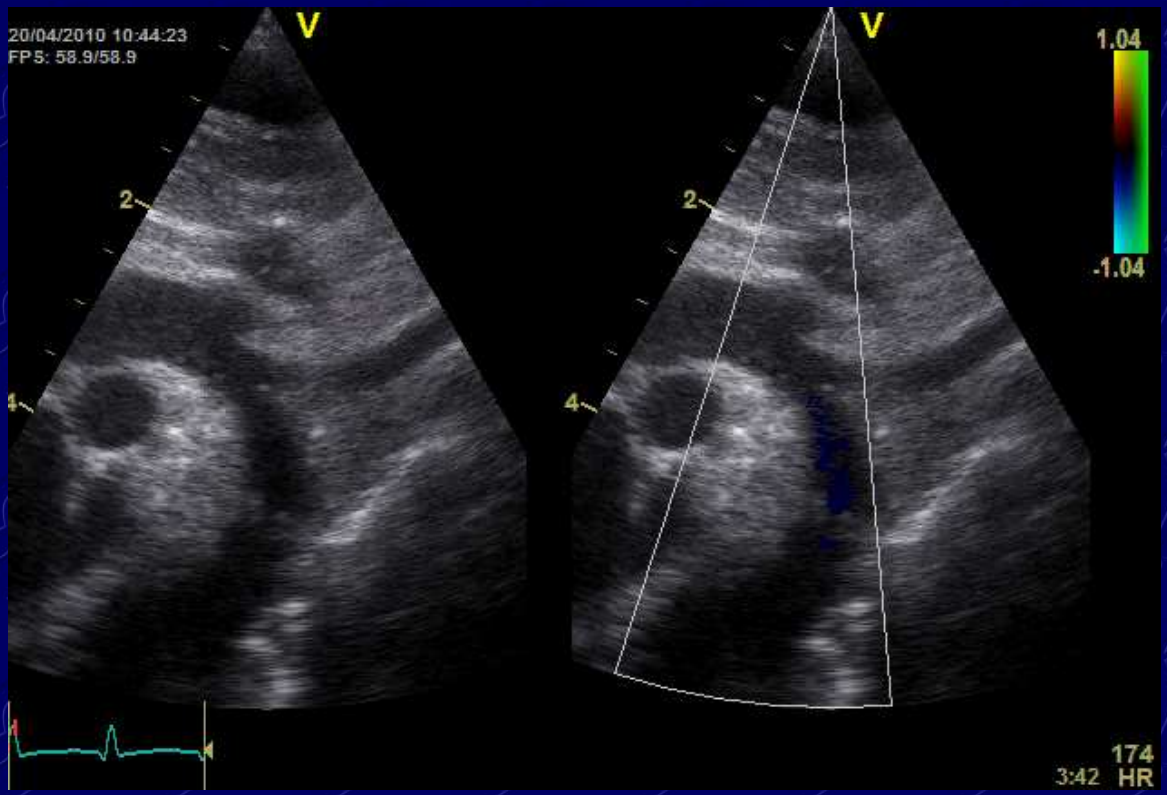
Decreased LV function: gradient 0 – 20mmHg

Low gradient in pts with large VSD and preserved LV function !!

Coarctation of the Aorta

Imaging of the AOA and isthmus

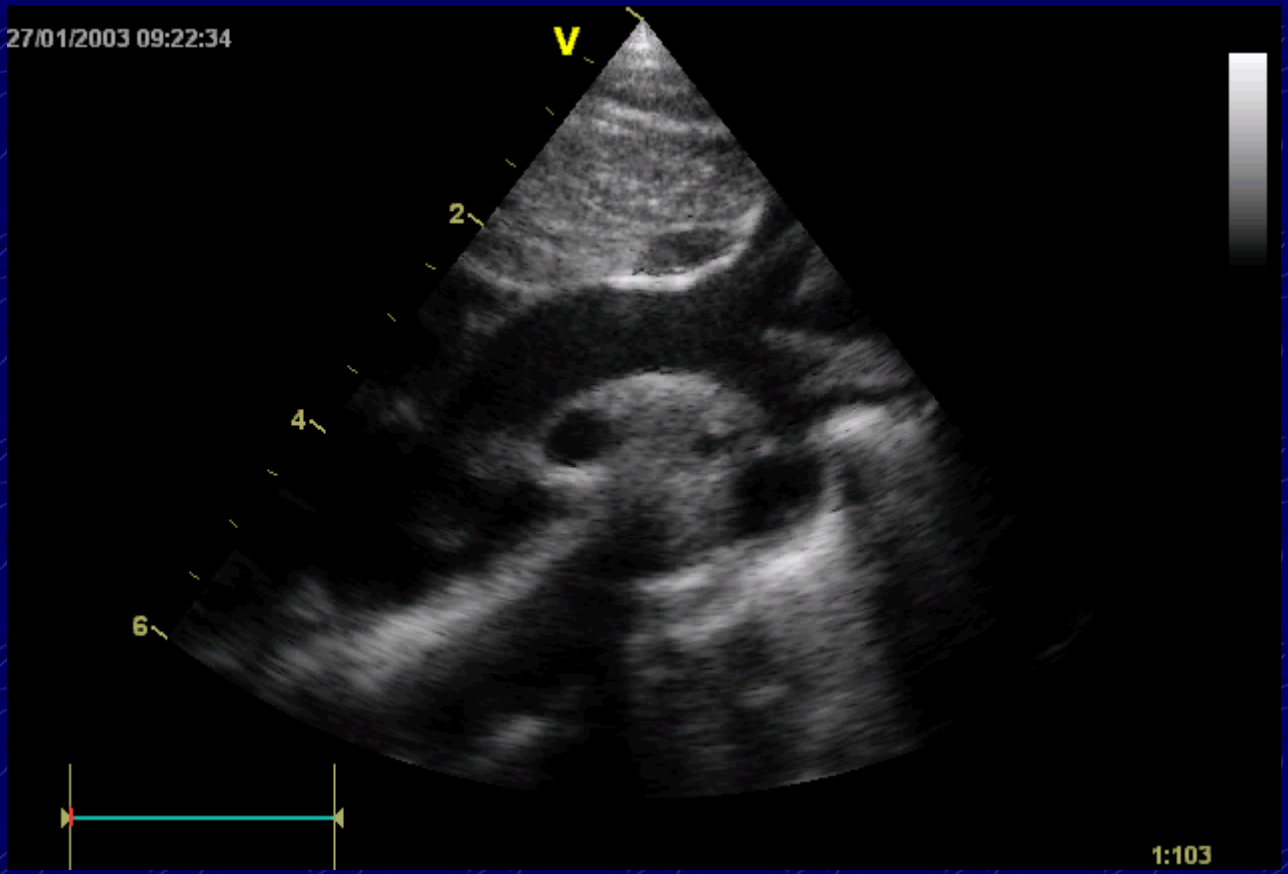
Mild or developing COA



Mild isthmic narrowing, usually no typical „ridge“

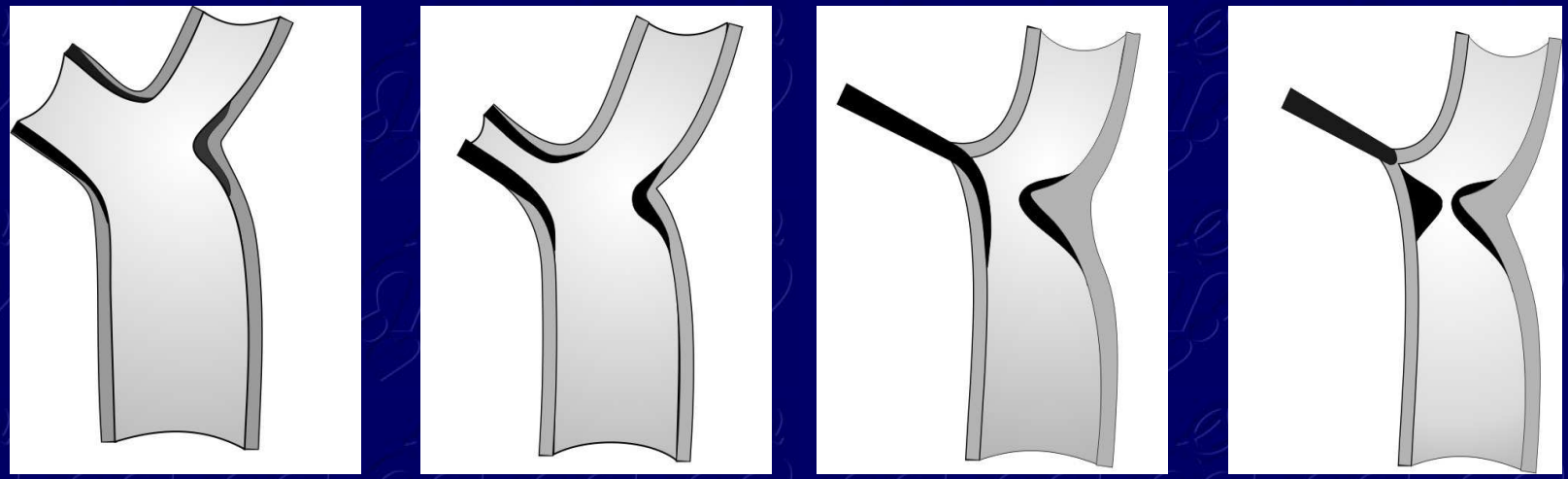
Coarctation of the Aorta

„False“ coarctation



Coarctation of the Aorta

Gradual development of COA during closure of PDA



Histology: ductal tissue circumferentially surrounding the juxtaductal portion of the AO

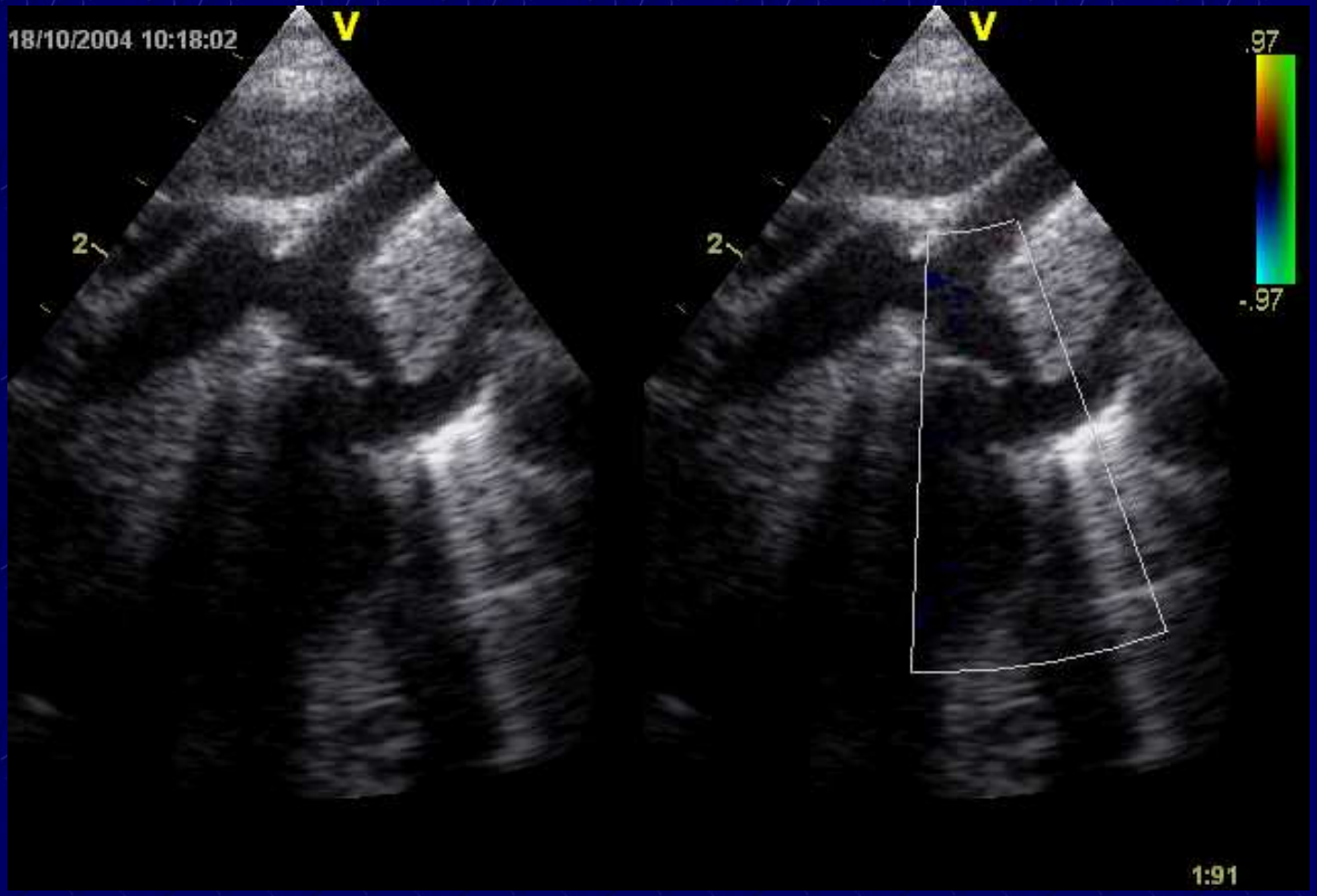
Ho, Circulation 1979

- no discrepant pulses
- no murmur
- heart is „normal“
- COA is hidden by PDA
- differential cyanosis only

Diagnosis may be difficult if PDA opened

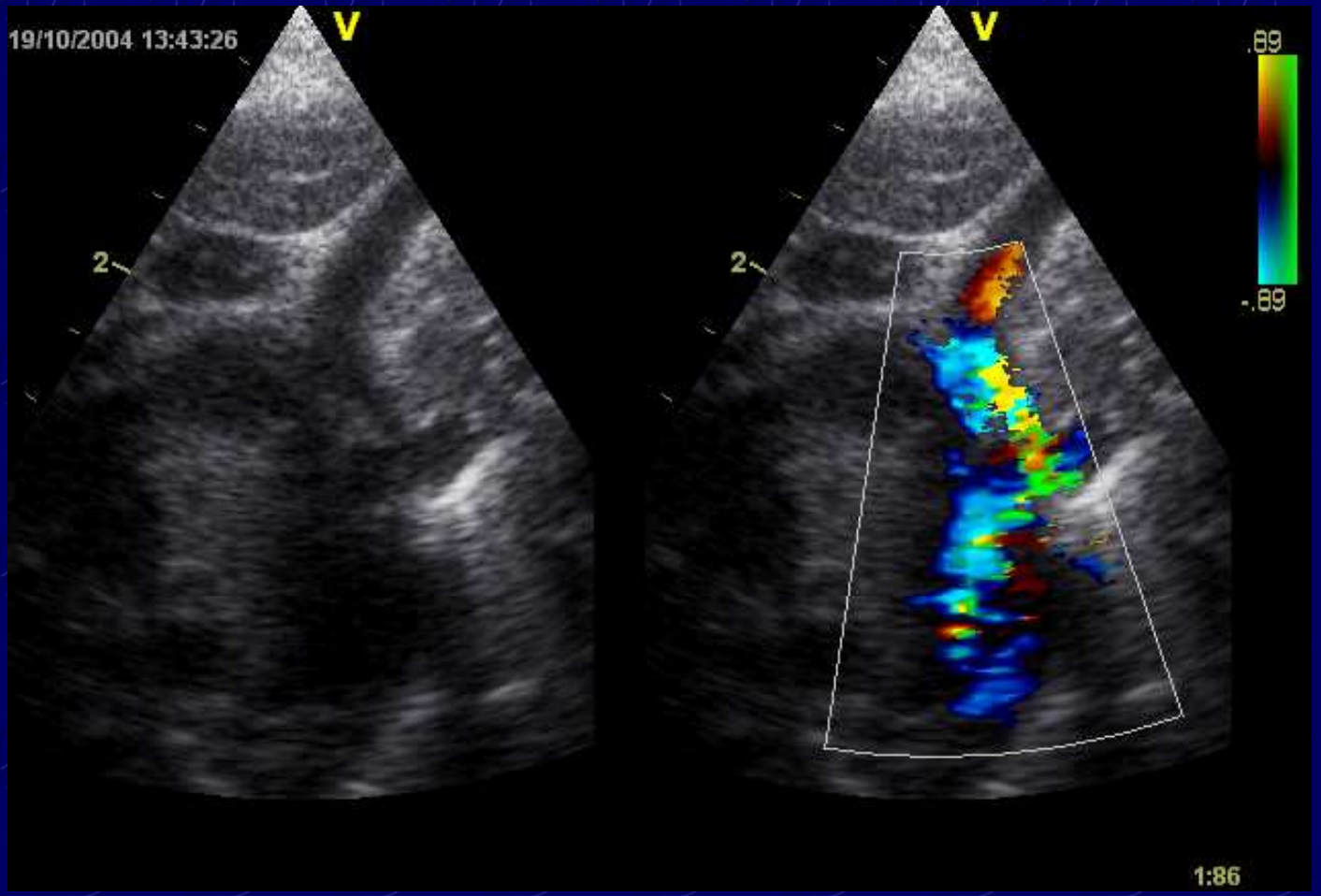
Coarctation of the Aorta

Gradual development of COA during closure of PDA



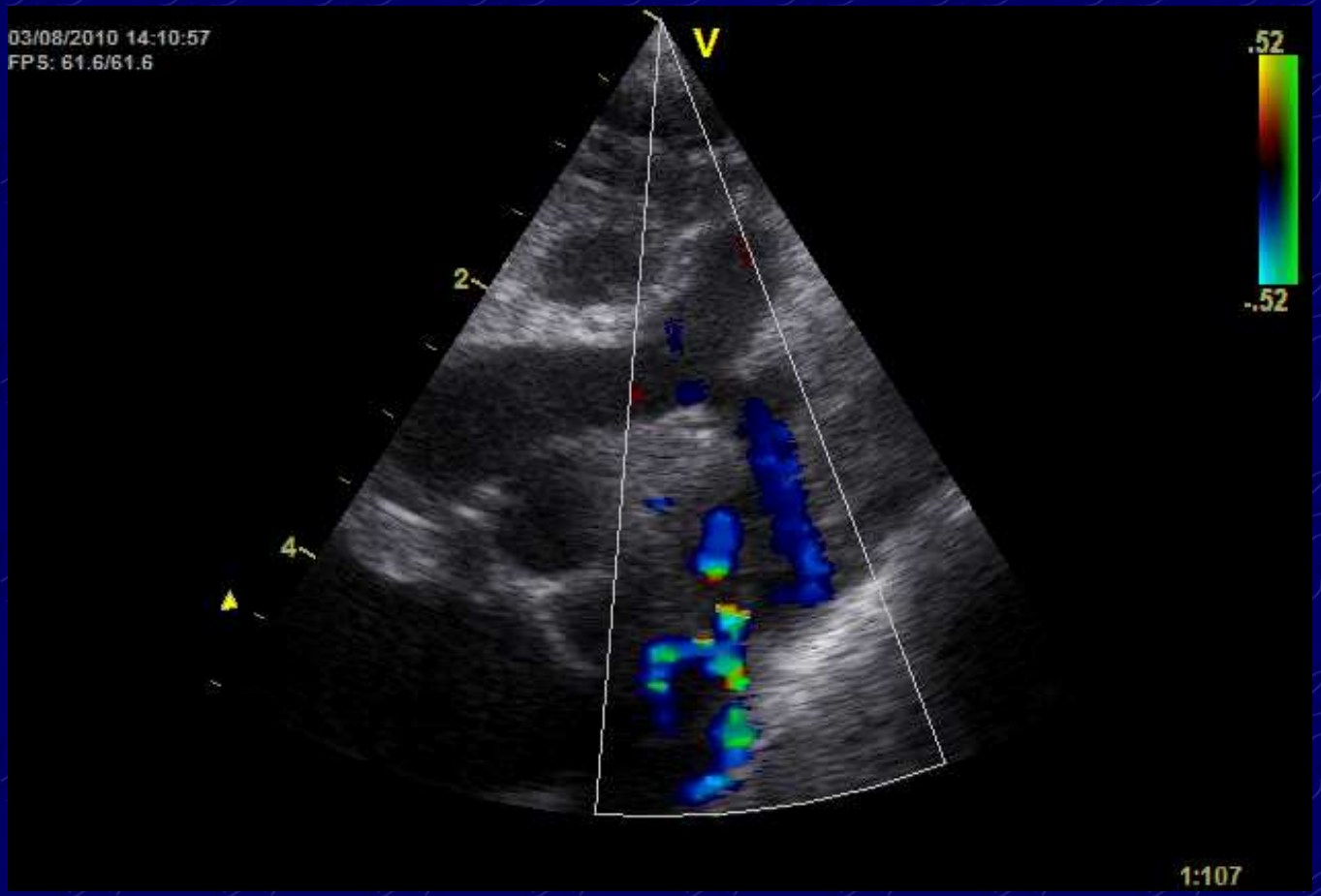
Coarctation of the Aorta

Gradual development of COA during closure of PDA



Coarctation of the Aorta

Gradual development of COA during closure of PDA



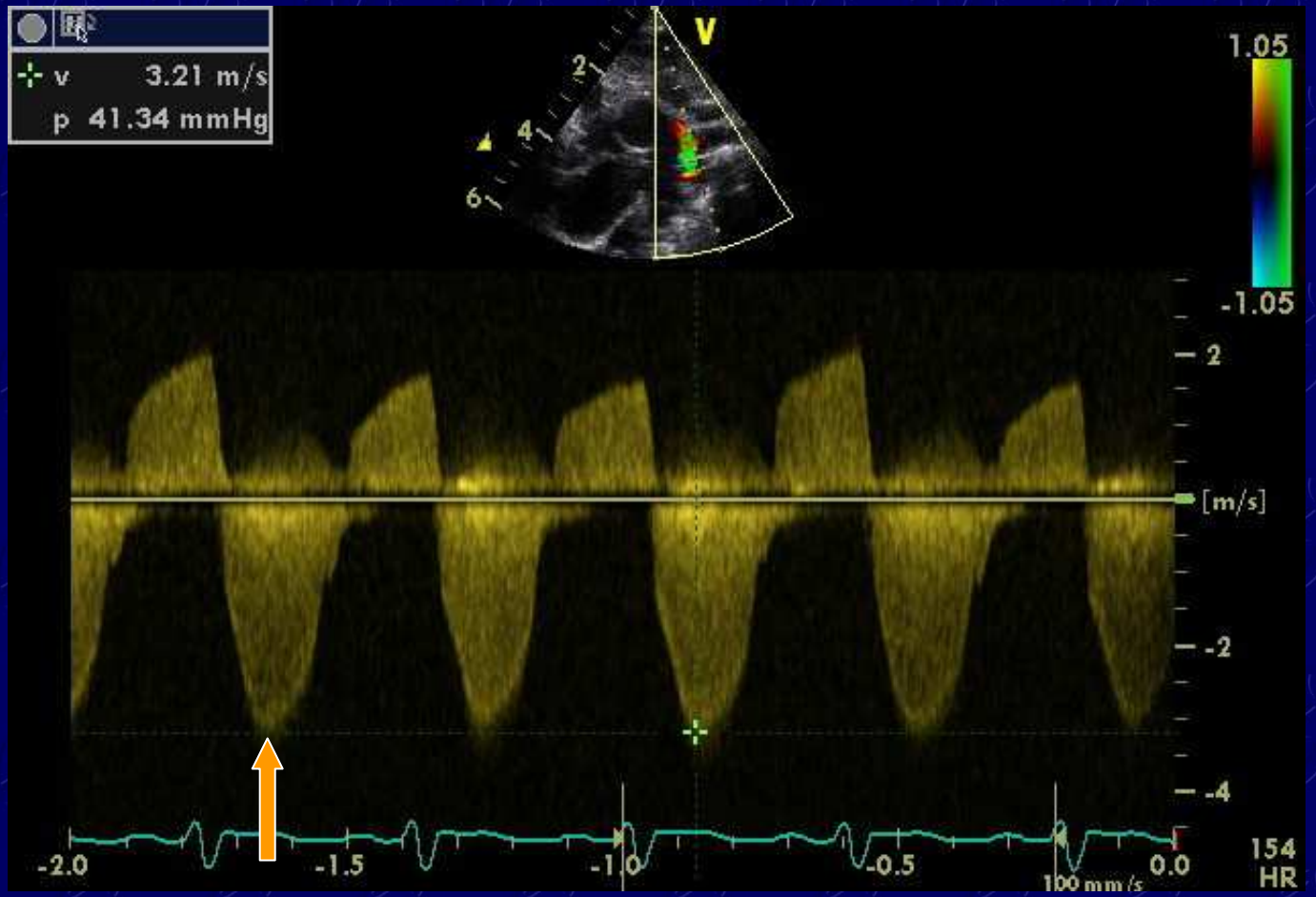
Coarctation of the Aorta

Imaging of the arterial duct

- The left infraclavicular window

Coarctation of the Aorta

Imaging of the arterial duct



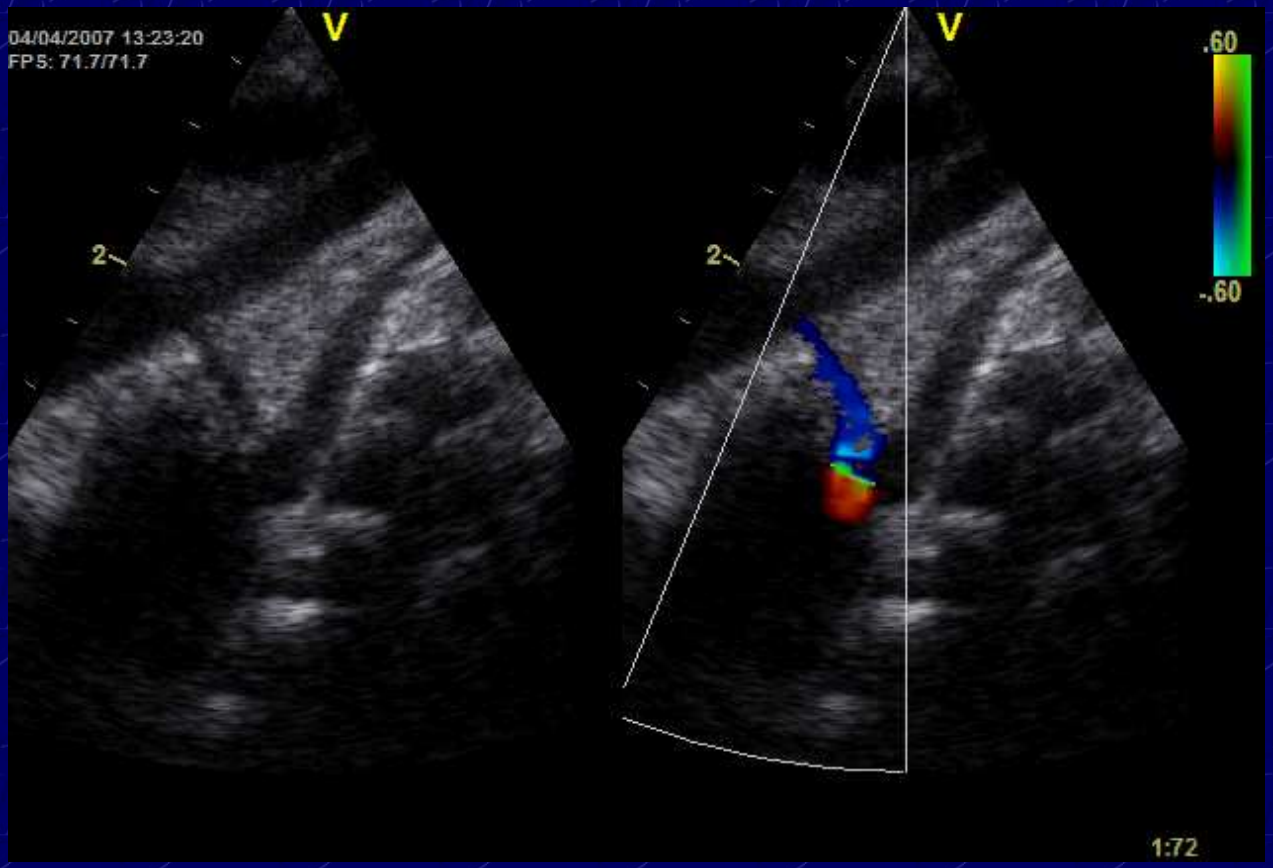
Coarctation of the Aorta

Imaging of the arterial duct

- The left infraclavicular window

Coarctation of the Aorta

Hypoplastic aortic arch



- The diameter of the narrowest segment
- 2D image measurement
- the Z-score is calculated

Hypoplastic = diameter Z-score is less than –2.0

Coarctation of the Aorta

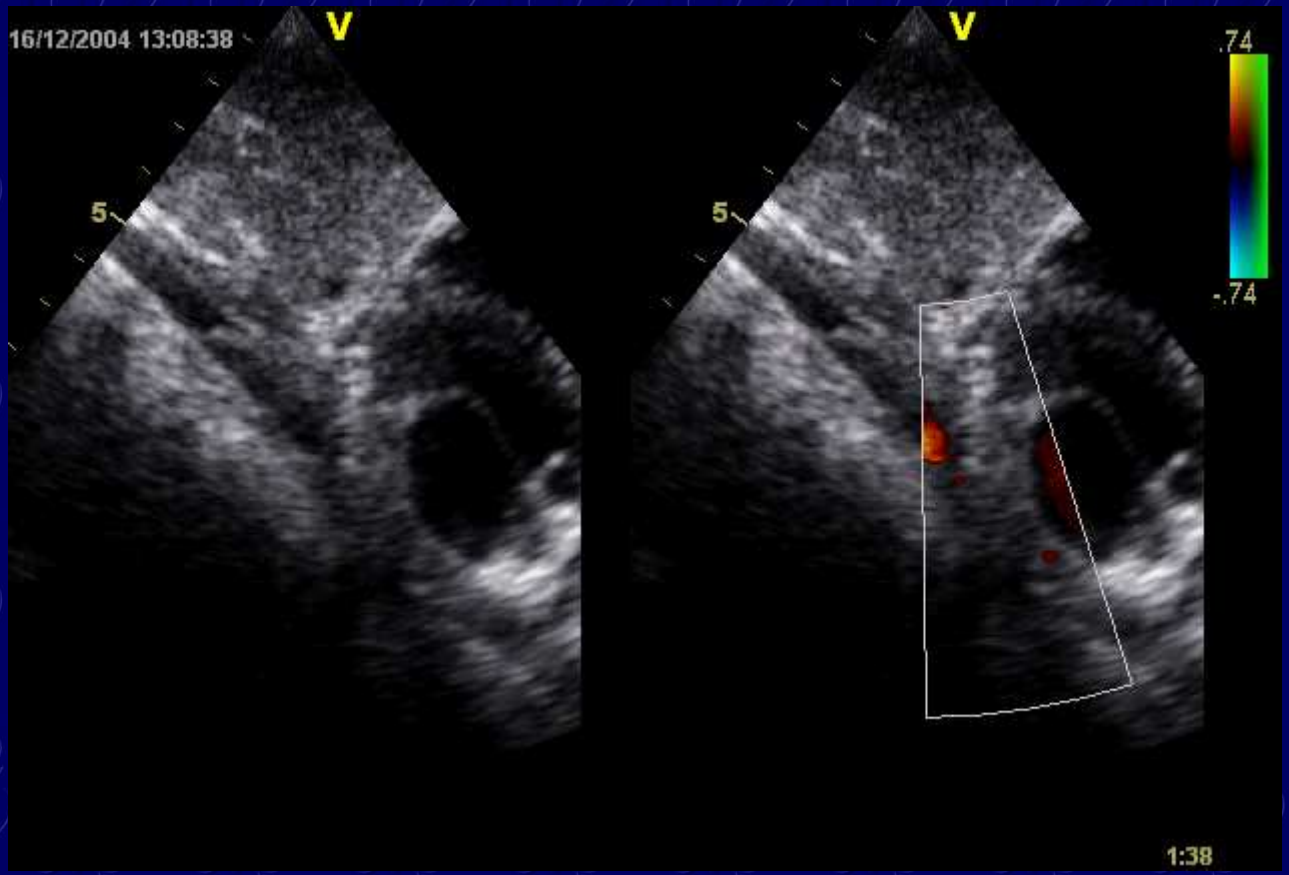
Rare forms

Coarctation of the aortic arch

Coarctation of the Aorta

Rare forms

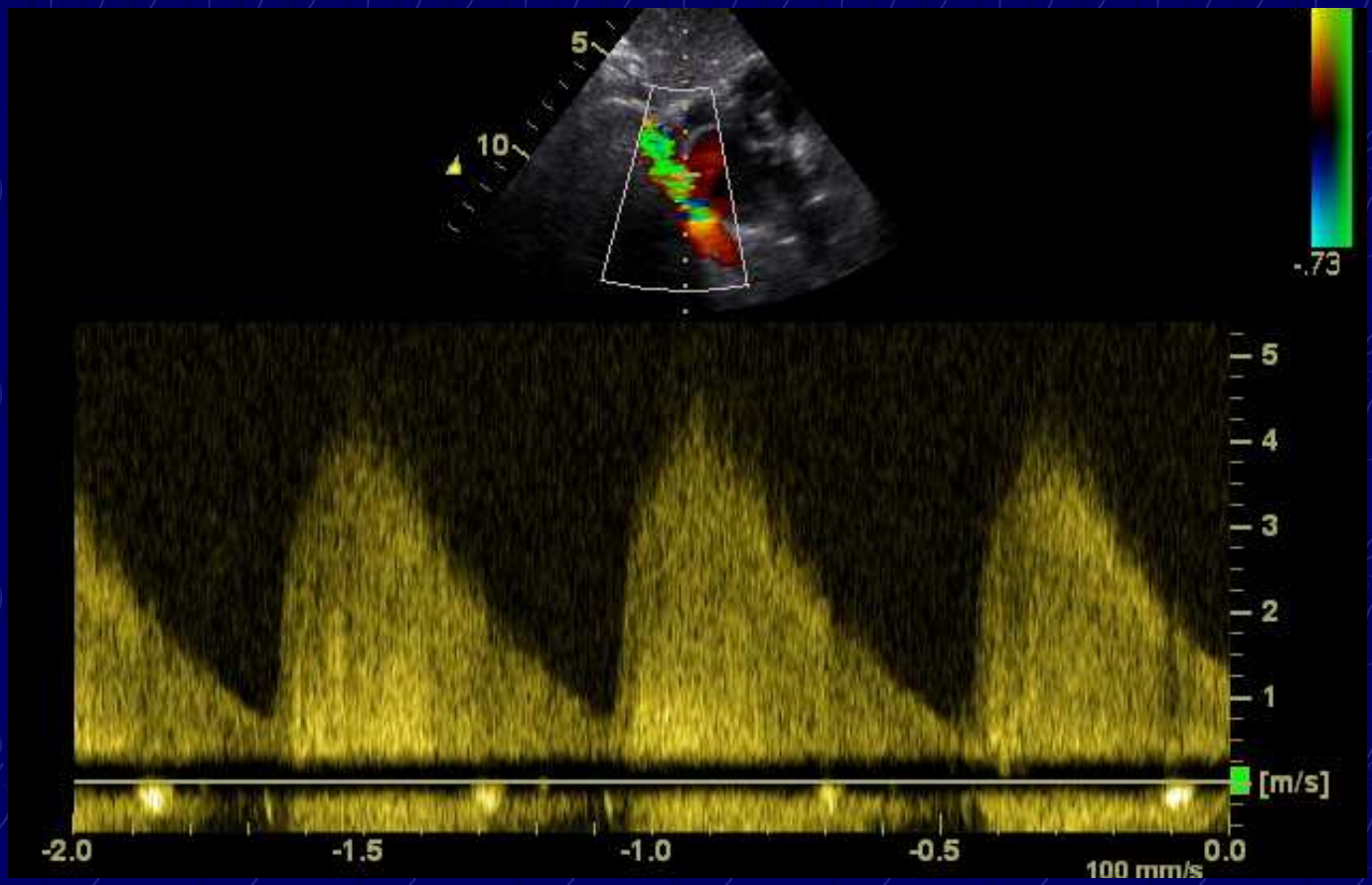
Coarctation of thoracic aorta



Coarctation of the Aorta

Rare forms

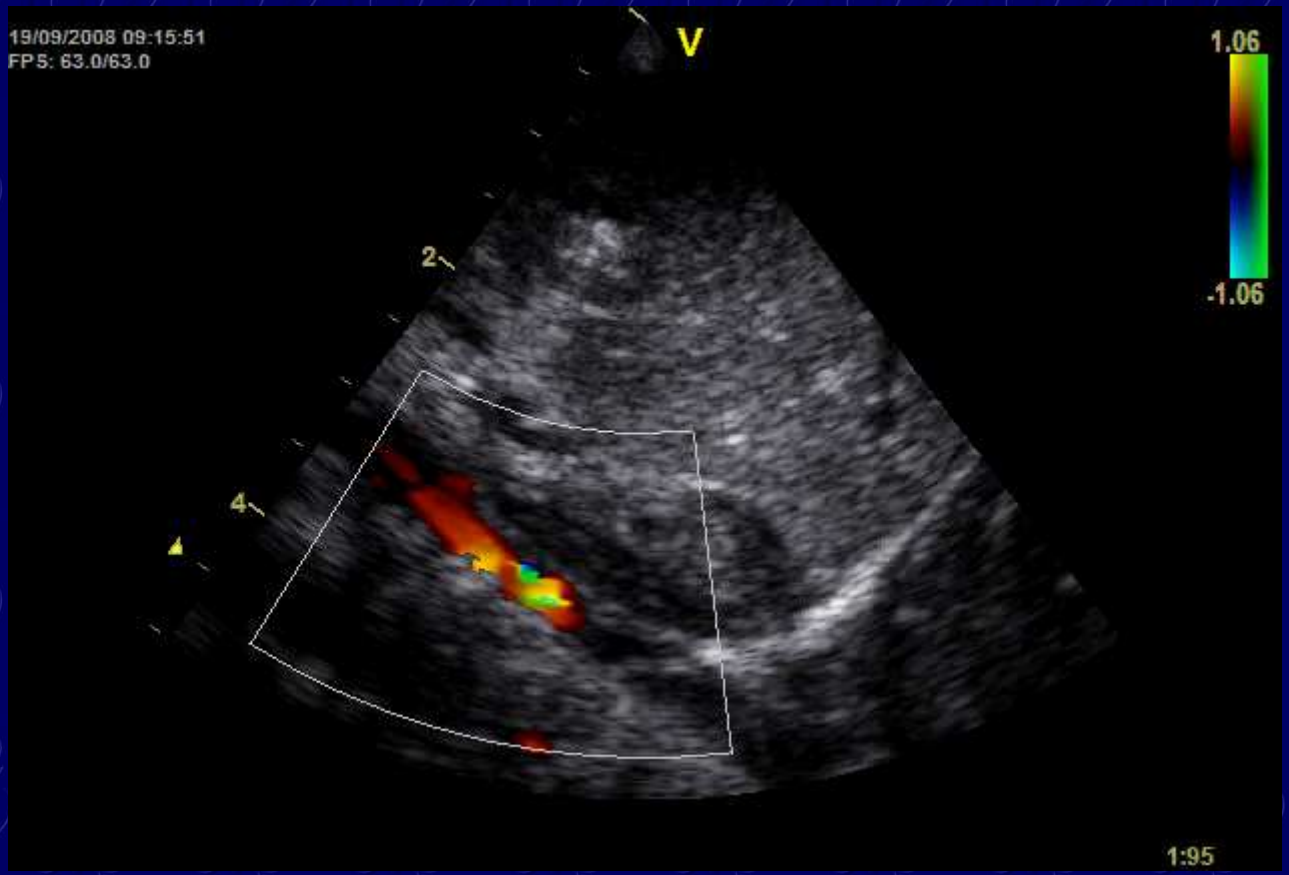
Coarctation of thoracic aorta



Coarctation of the Aorta

Rare forms

Coarctation of abdominal aorta



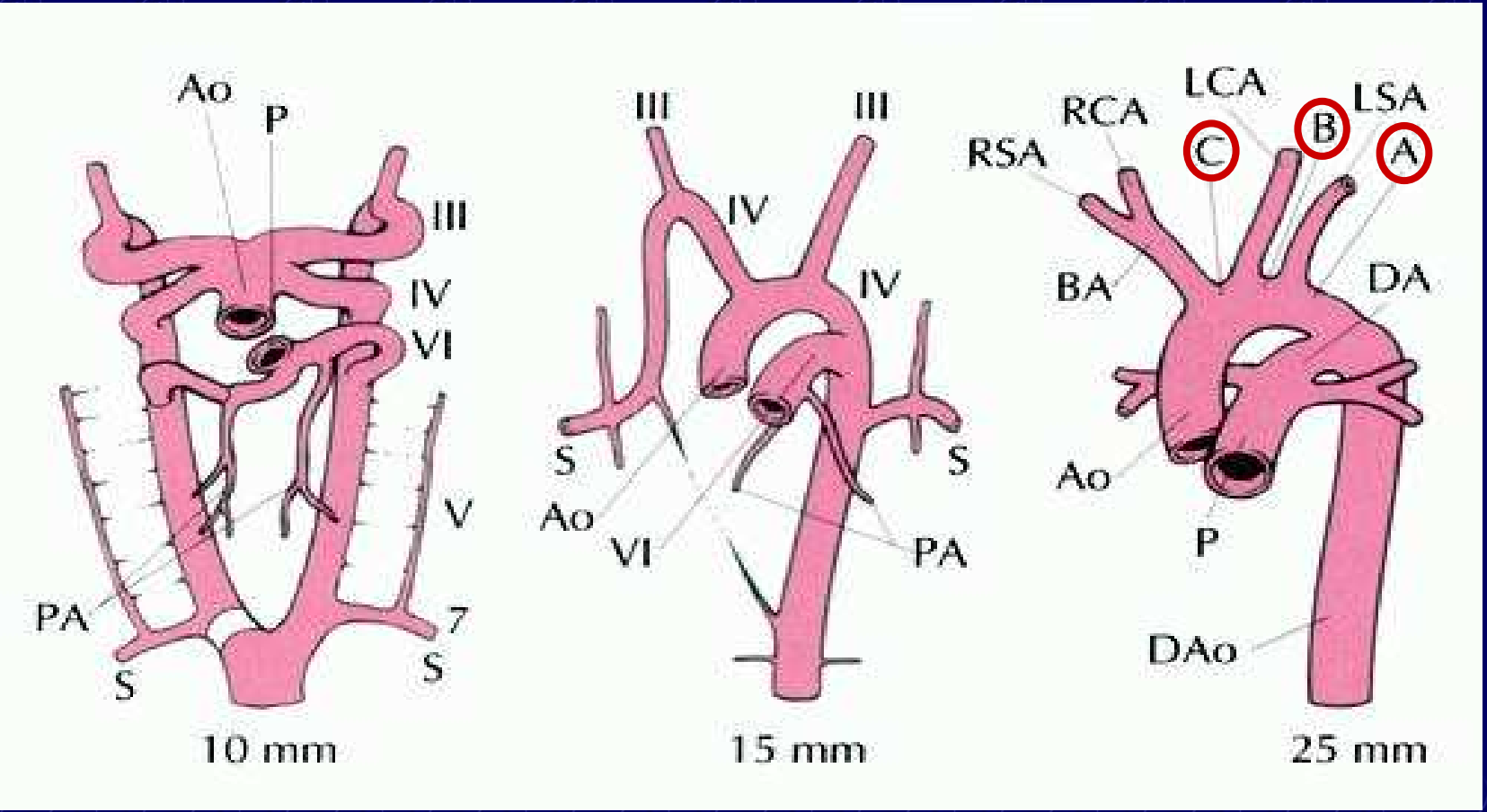
Interrupted aortic arch

- definition: discontinuity between two adjacement segment of the aortic arch

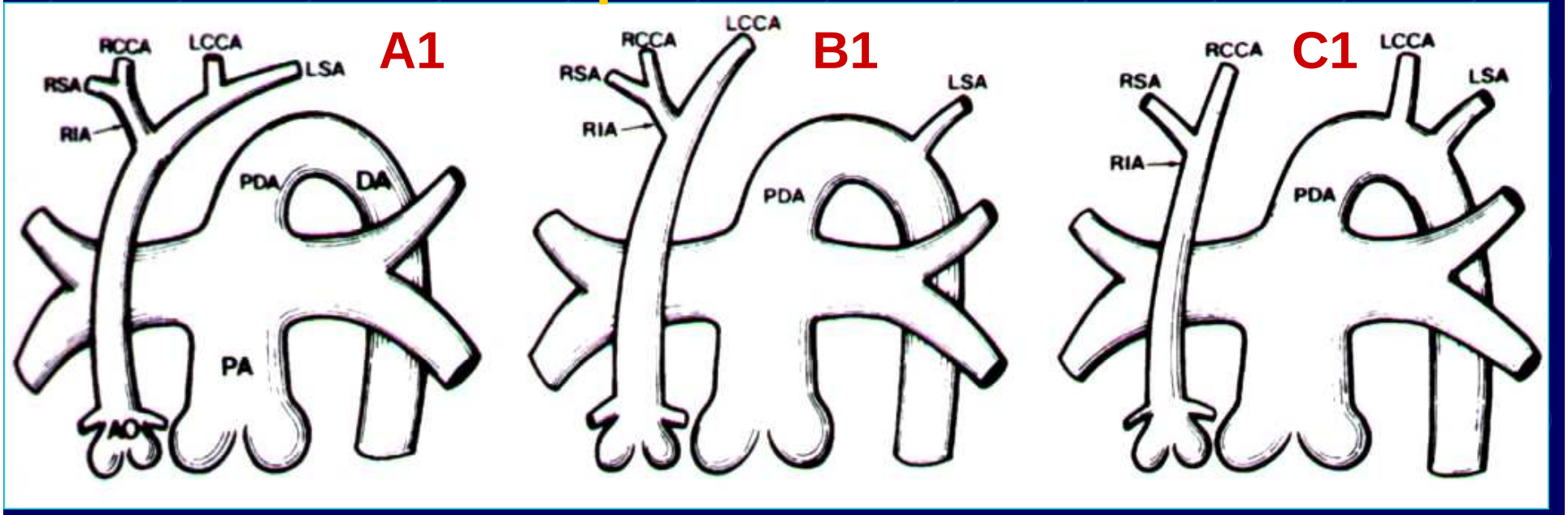
- 0.38% of all and 1.3 % of critical CHD

Interrupted aortic arch

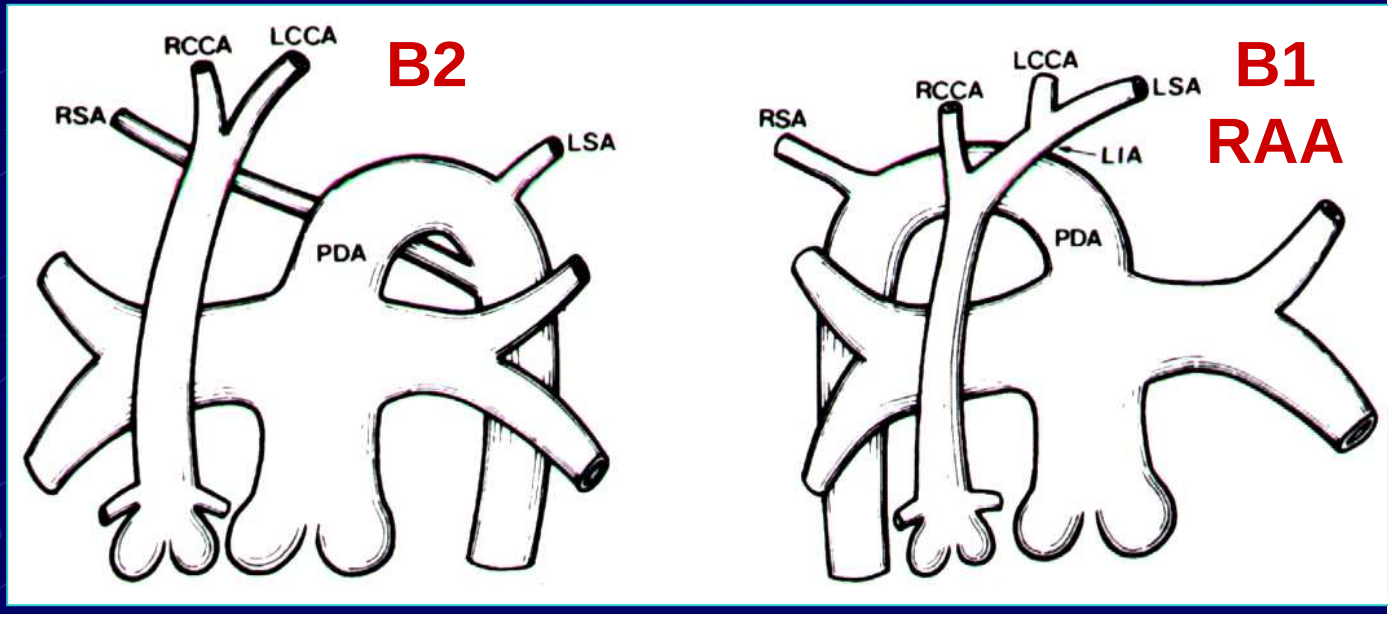
Embryonic arch diagram



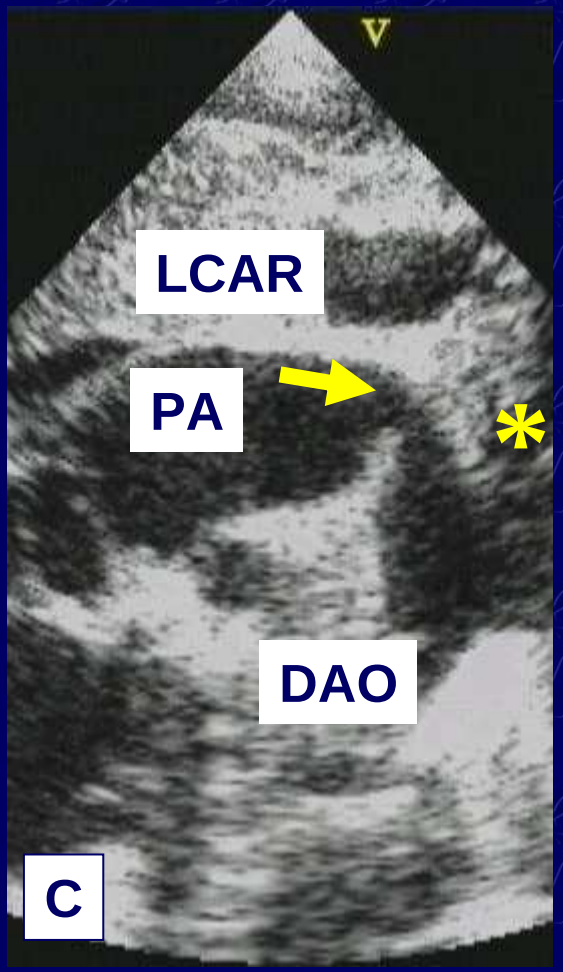
Interrupted aortic arch



Classification according to the site

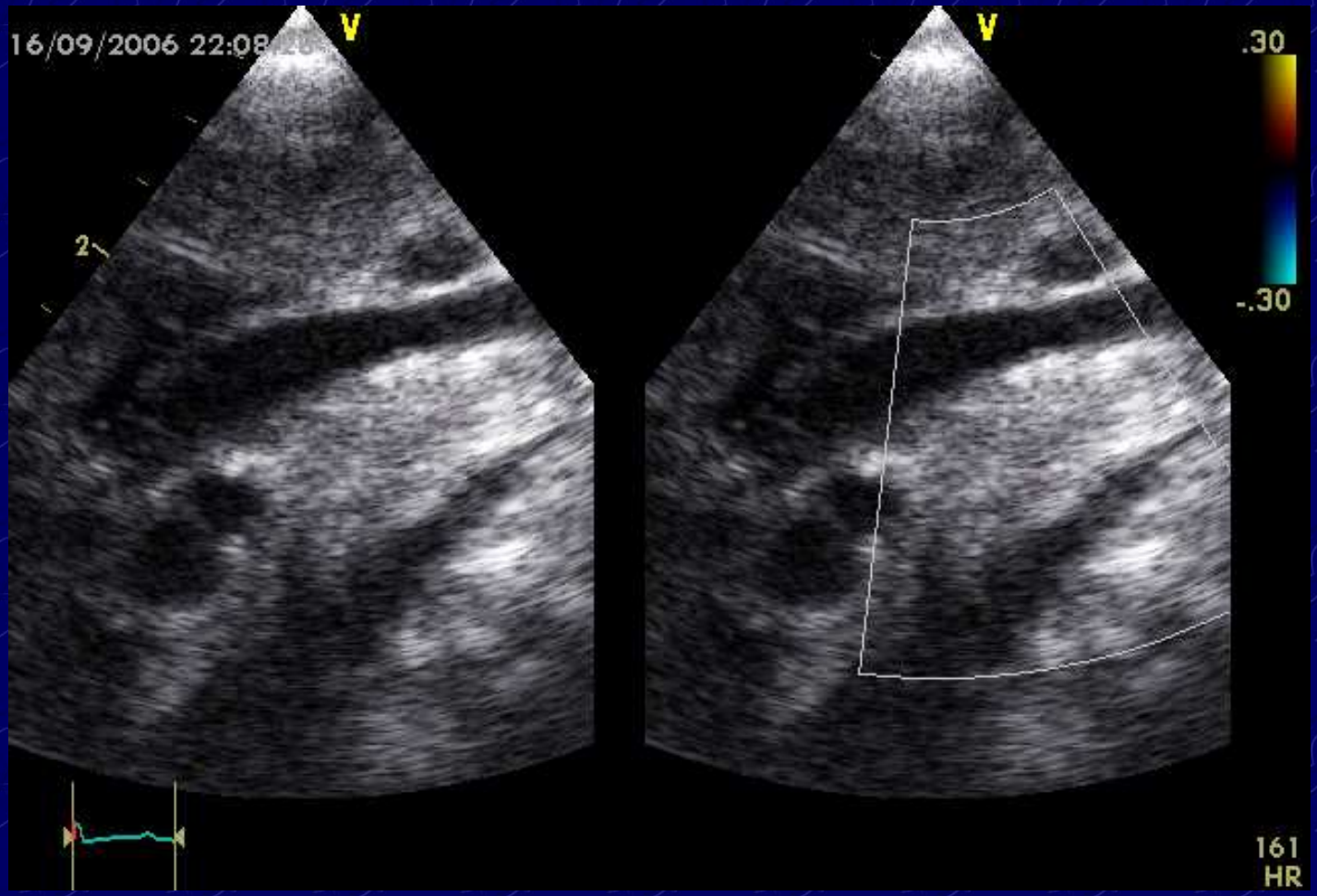


Interrupted aortic arch B1

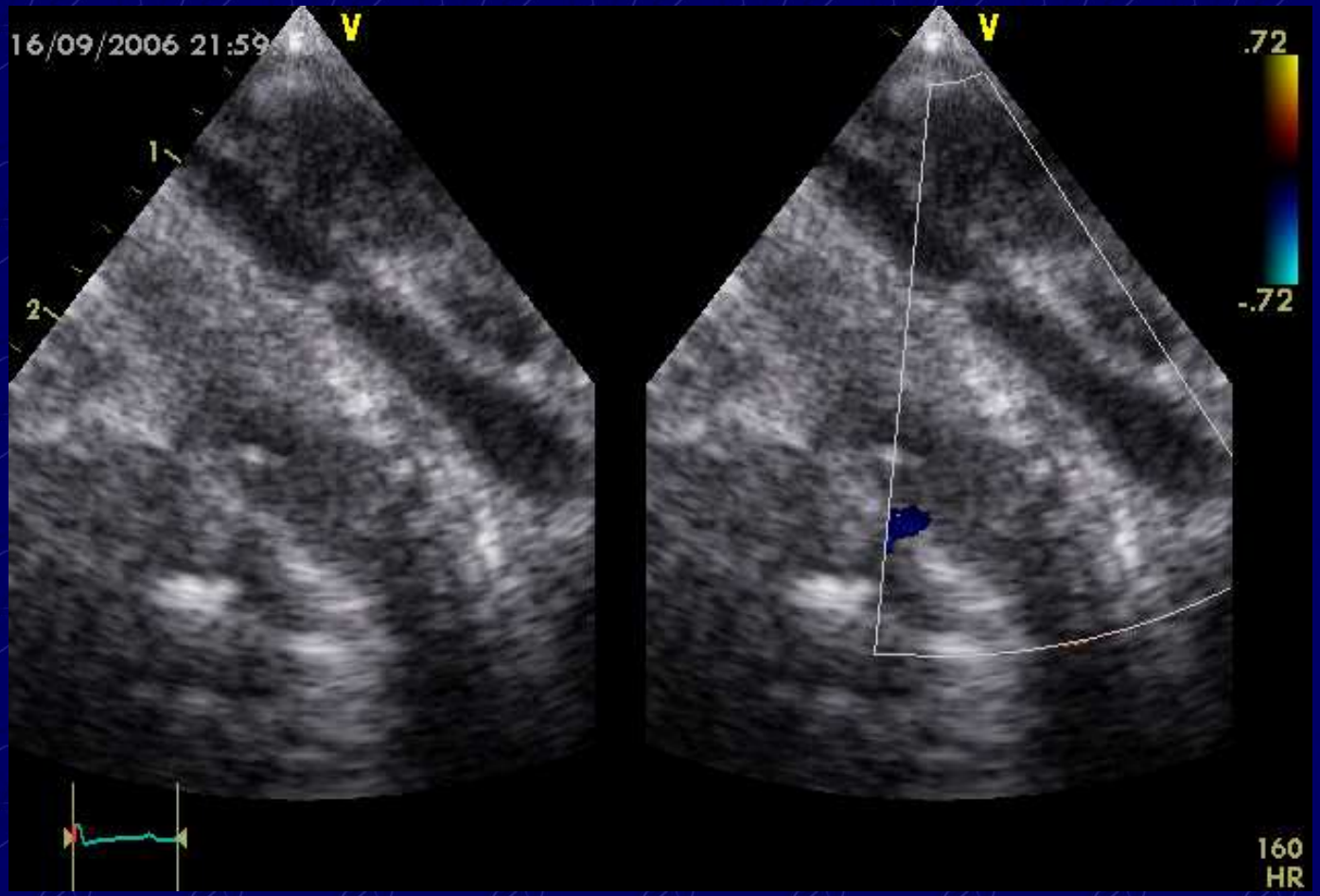


Serial views to visualize AOA anatomy
Sagital v. toward R shoulder – clockwise rotation

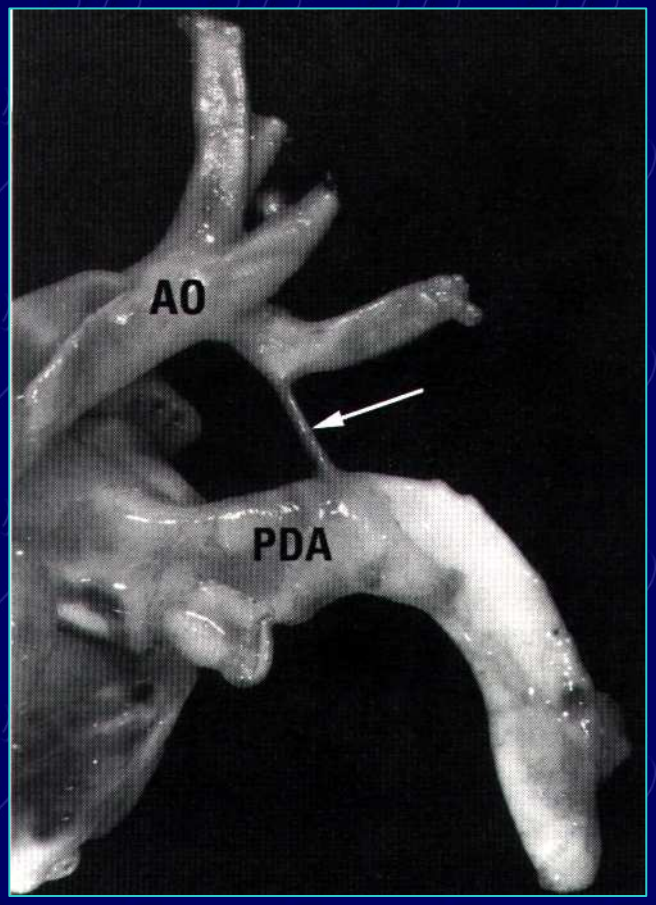
Interrupted aortic arch B2

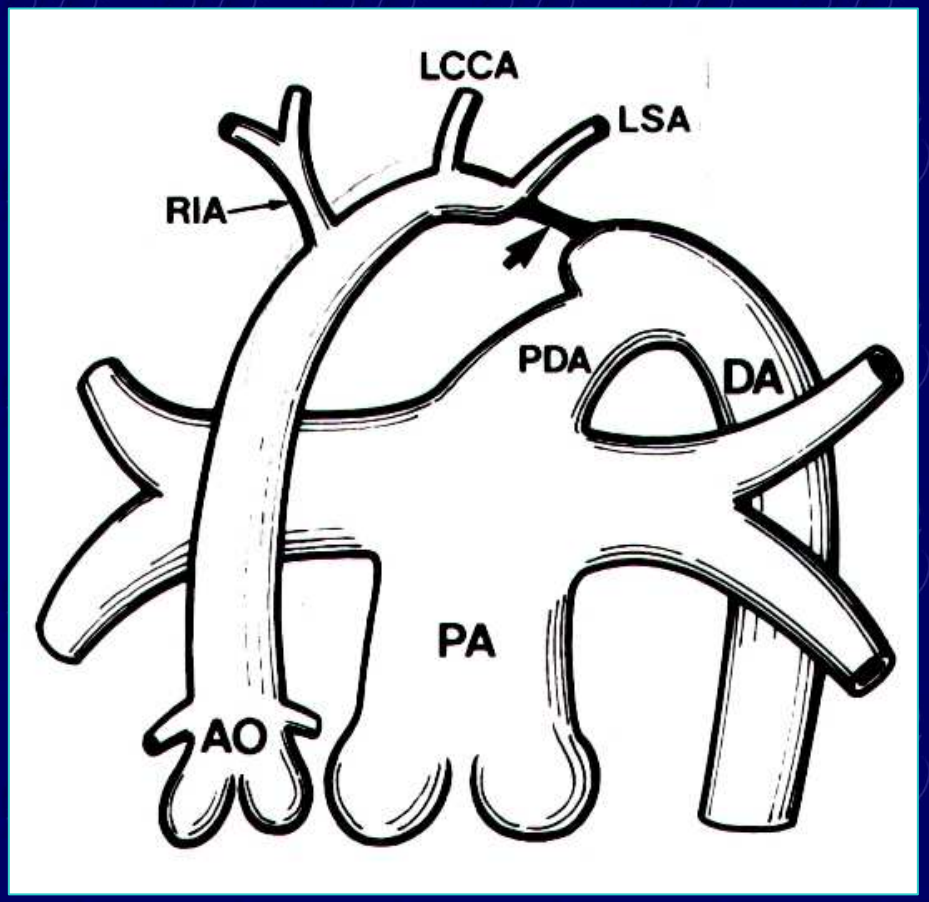


Interrupted aortic arch B2



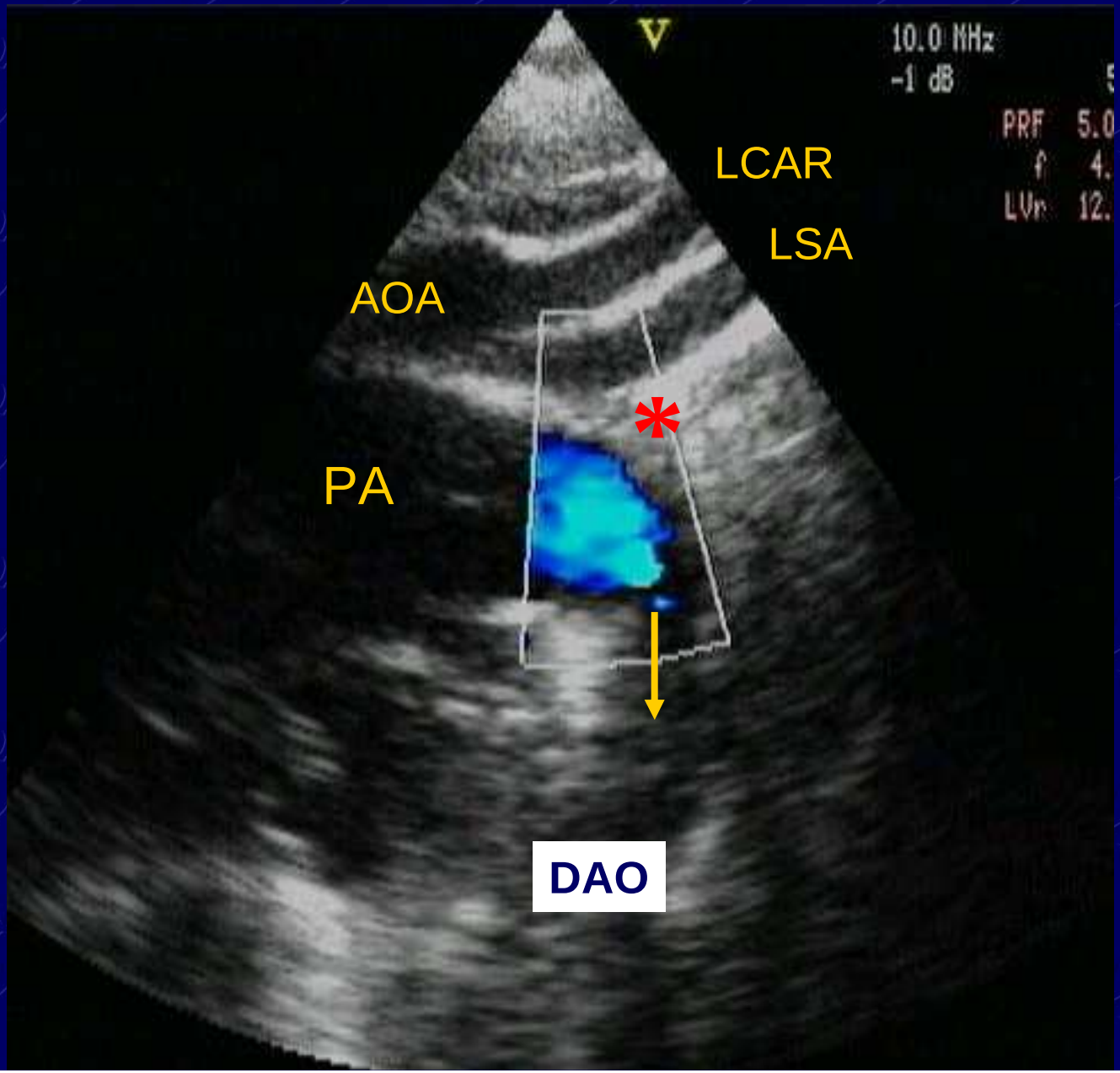
Interrupted aortic arch (A) x aortic arch atresia



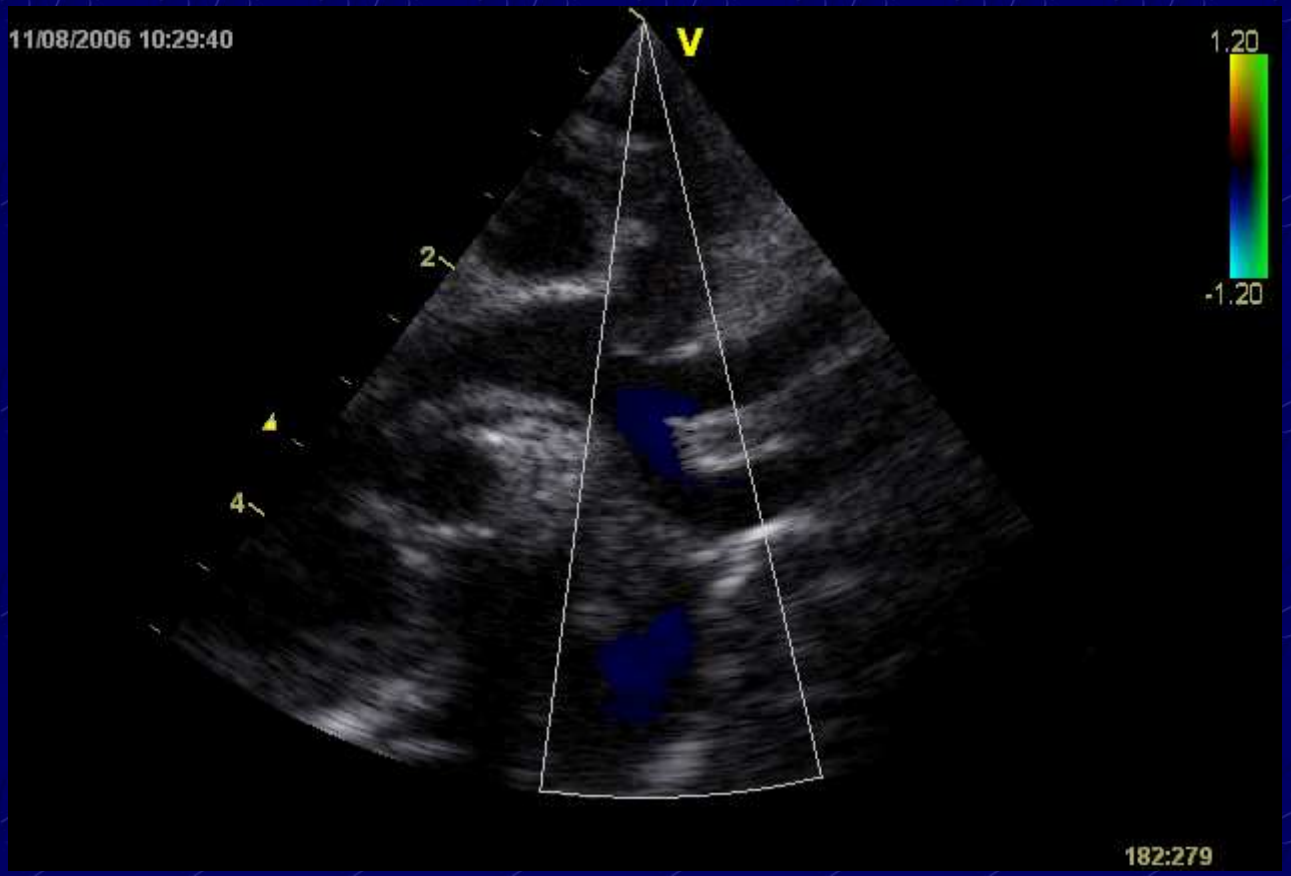


Anatomic continuity through a fibrous strand/ lumen completely obstructed

Interrupted aortic arch A1

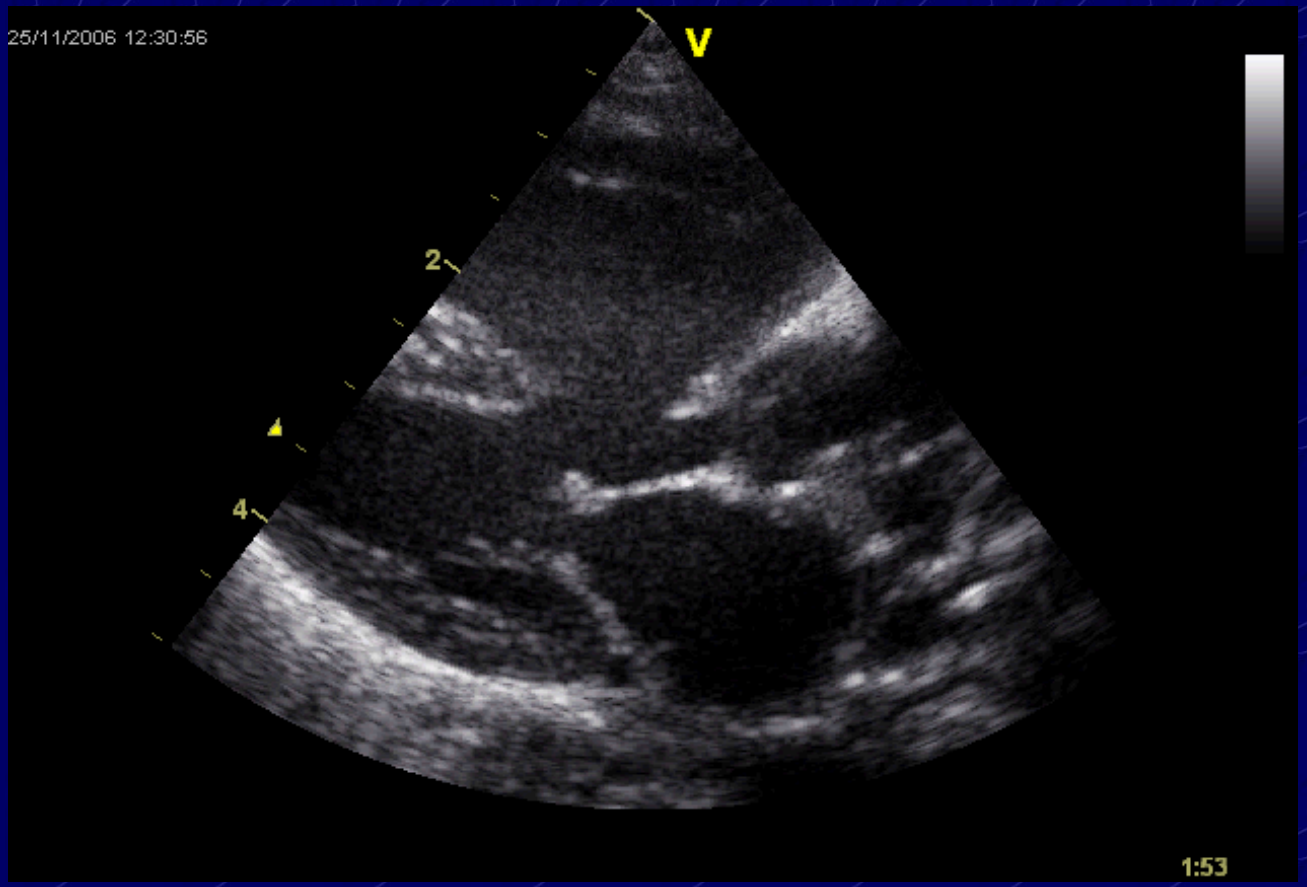


Interrupted aortic arch (A) x aortic arch atresia



Interrupted aortic arch

VSD + LVOT obstruction



LVOT area < 0.7-0.8cm²/m² develop subAO obstruction *Minich 1992, Geva 1993*

LVOT/DAO ratio < or = 1.0 recomm. subAO resection *Ge, J Am Soc Echo.1997*