Prenatal diagnostics of aortic rings and slings

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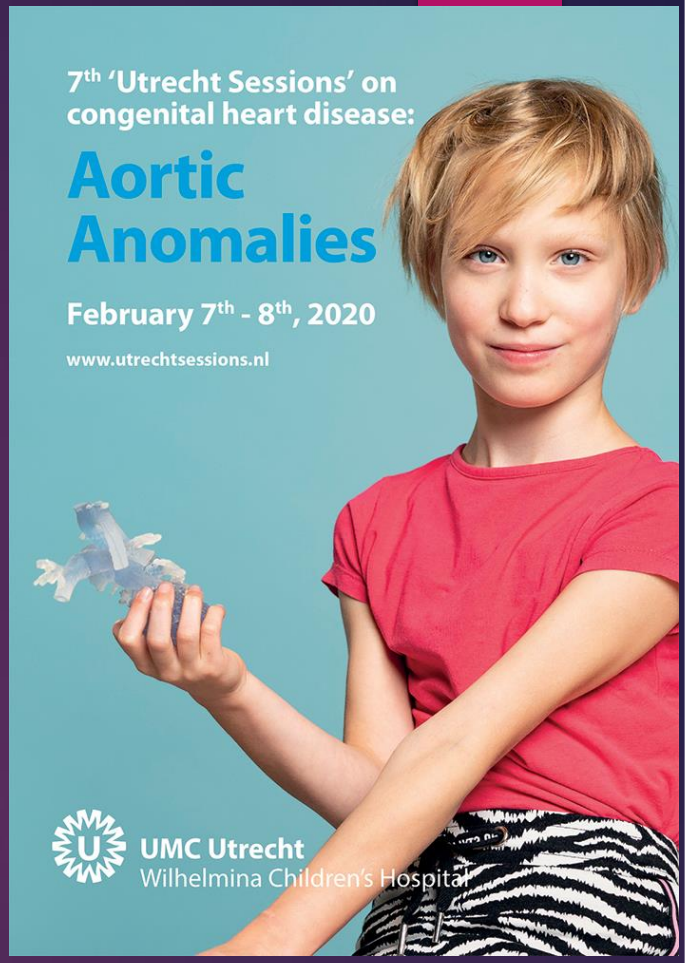
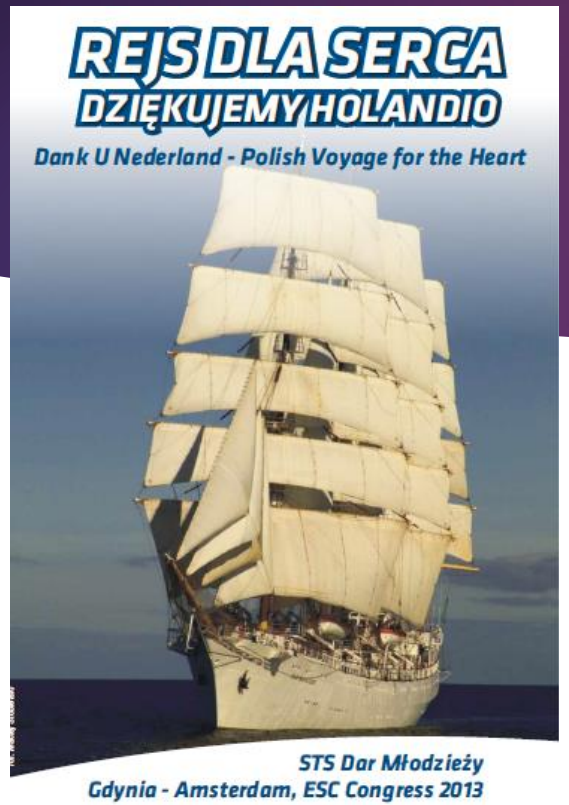


Let's start from history …



World Charity Support for Polish Children with Congenital Heart Defects

Medical Center	Years	No. of operated children and trained physicians, nurses and medical staff
Wilhelmina Kinderziekenhuis Utrecht, The Netherlands	1983 – 1990	426 children 22 physicians, nurses and medical staff

Eind februari 1984 kwamen vier Poolse kinderen – Marcin, Ania, Jacek en Paulina – in Nederland aan om in het Wilhelmina Kinderziekenhuis in Utrecht een hartoperatie te ondergaan.
Daarmee ging een uniek projekt van start.
Uniek, omdat niet alleen Marcin, Ania, Jacek en Paulina, maar hopelijk zeer veel andere Poolse hartpatiëntjes de komende jaren in Utrecht geopereerd zullen kunnen worden.
Uniek bovendien, omdat gelijktijdig met de vier kinderen een aantal Poolse artsen arriveerden die in het Wilhelmina Kinderziekenhuis zullen worden opgeleid.
Dit projekt voorziet met andere woorden niet alleen in directe hulp aan kinderen in nood, maar biedt tevens uitzicht op verbetering van de medische mogelijkheden in Polen.
'Blijvend bruikbare hulp' noemen ze dat in het Wilhelmina Kinderziekenhuis.

HONORARY DISTINCTIONS AND AWARDS FOR THE LEADERS OF THE POOLSE HARTPATIENTJES NAAR NEDERLAND PROJECT Utrecht 1983-1990

"Bene Merito" HONORARY DISTNICTIONS:

The "Bene merito" honorary distinction is conferred upon the citizens of the Republic of Poland and foreign nationals in recognition of their merits in promoting Poland abroad.

- Prof. Jan Walter Stoop
- Prof. Francois Hitchcock
- Dr Marianne Karelse - Nijsen

1984
WKZ Heart Team

1997
meeting with
prof. F. Hitchcock, Zabrze

How detection rate of certain CHD changed in Poland ?

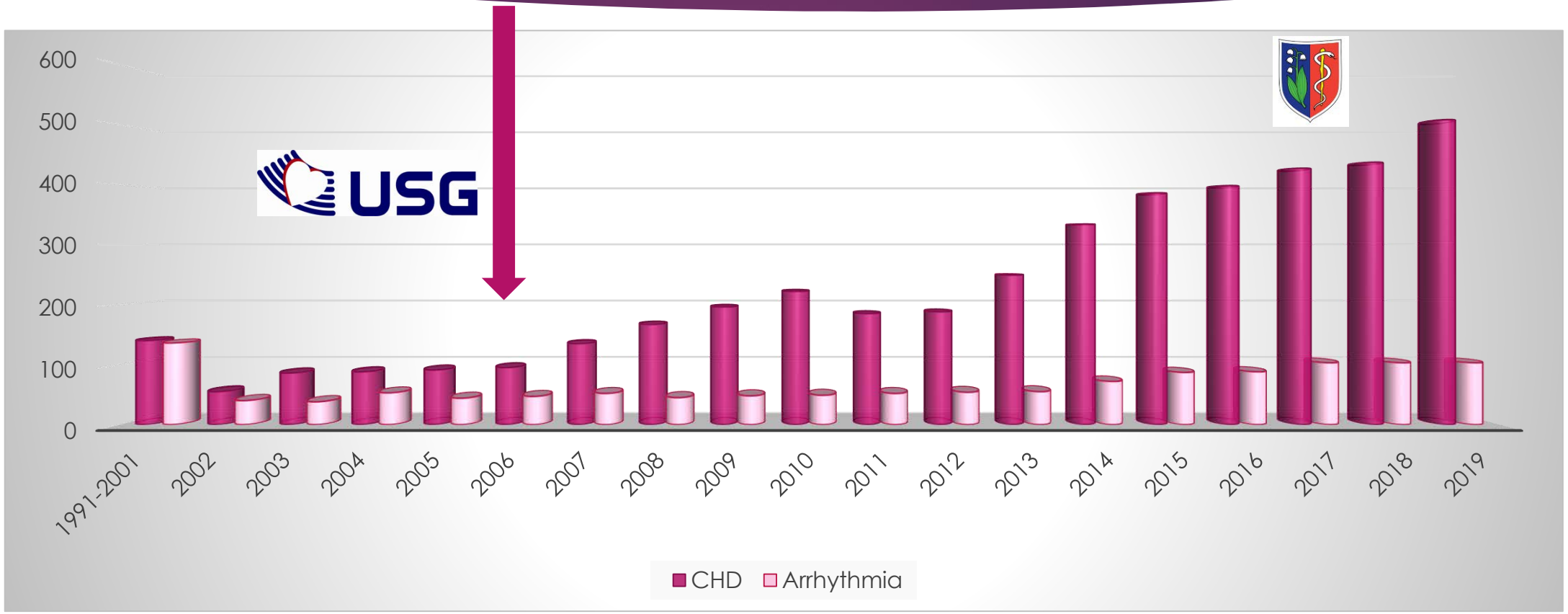
CHD	VIEW	2004	2012	2017
ALL / YEARLY		269	716	808
HLHS	4ch abn	25	86	55
AVSD	4ch N	22	53	45
TGA	4ch N	3	48	42
TOF	4ch N	10	50	46
AS	4ch dif	6	16	36
RAA	4ch N	0	0	**24**
ARSA	4ch N	0	0	**23**

Based on data www.orpkp.pl

Fetal cardiology in Warsaw

2019 **4371 ECHO**

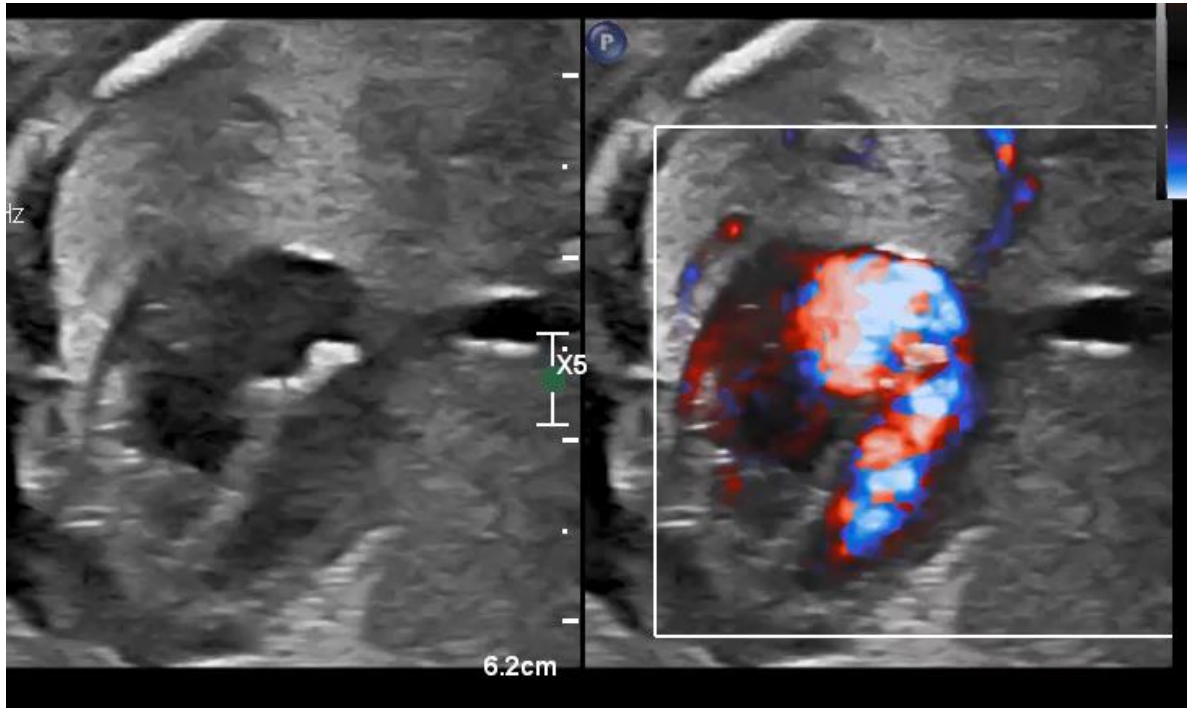
CHD	**1157**	**2390 abnormal**
arrhythmia	**173**	
ECM	**724**	



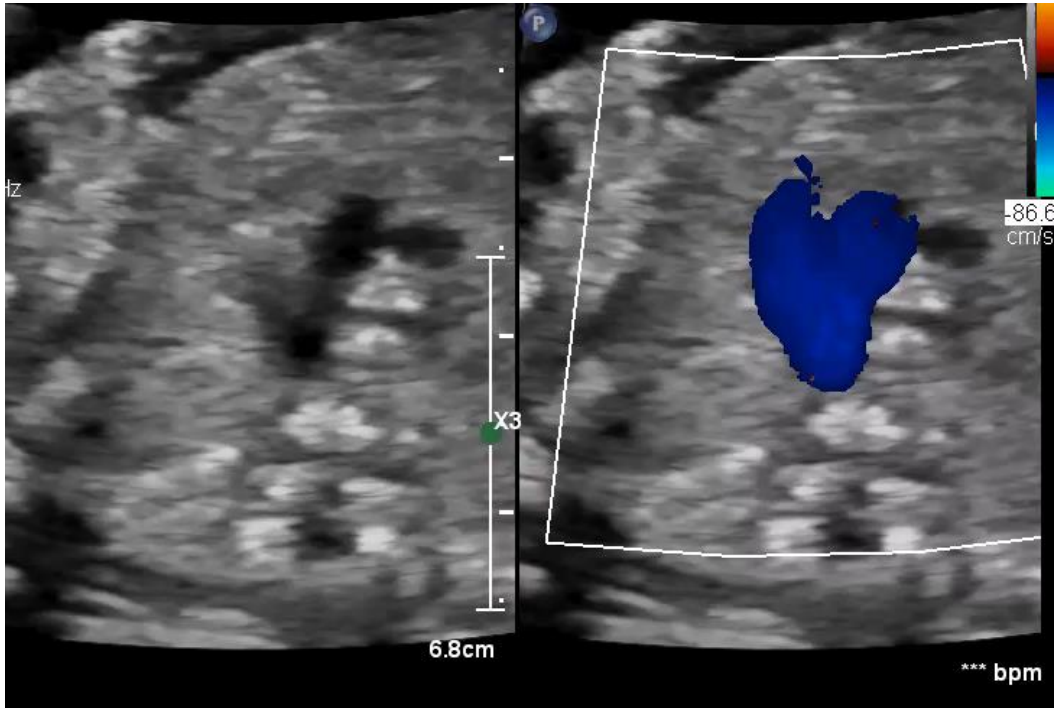
ToP - HLHS (206 between 2011 – 2018): **6%**

What obstetricians should see …

4 chamber view

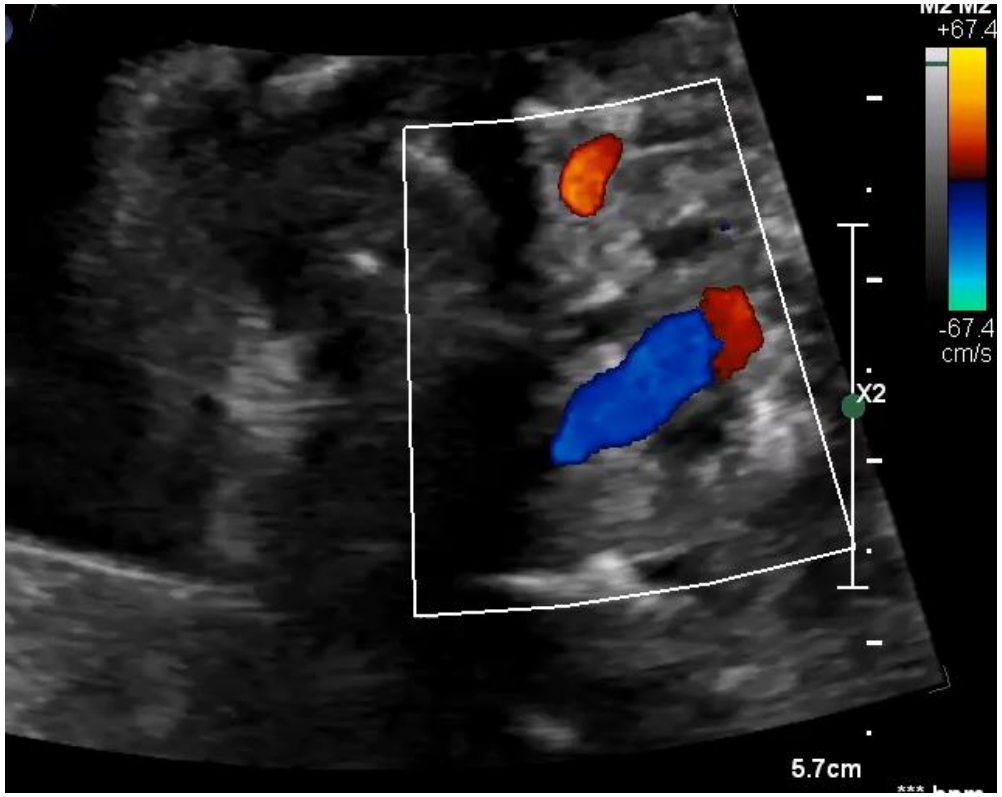


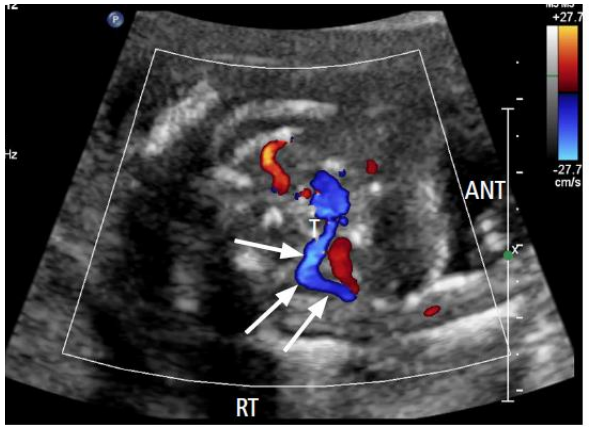
3VV-T 3 vessel – trachea view

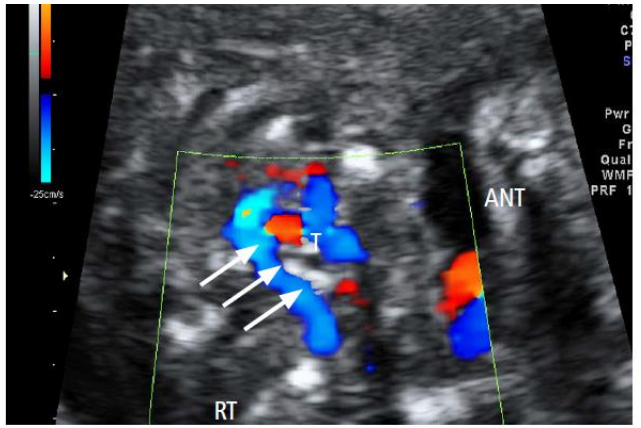


How can we improve diagnosis of the aortic rings ?

Look at coronal plane

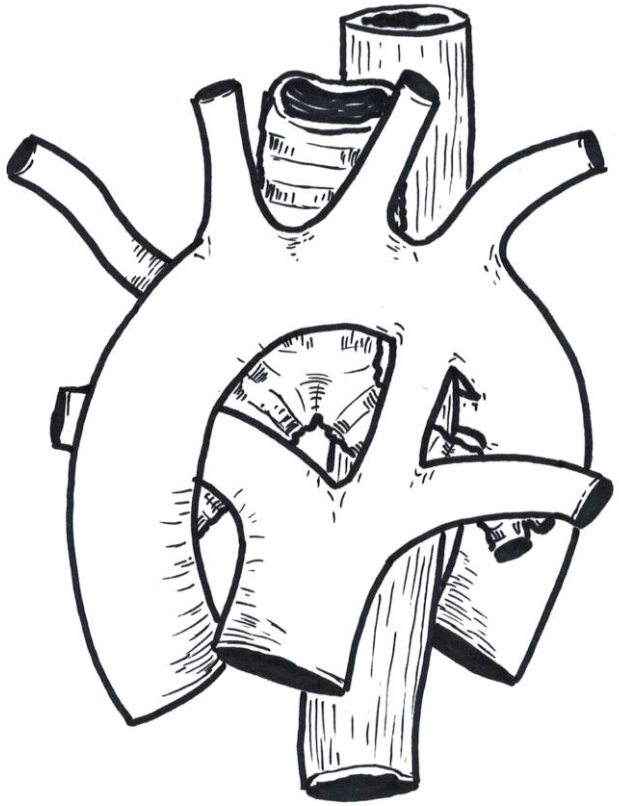


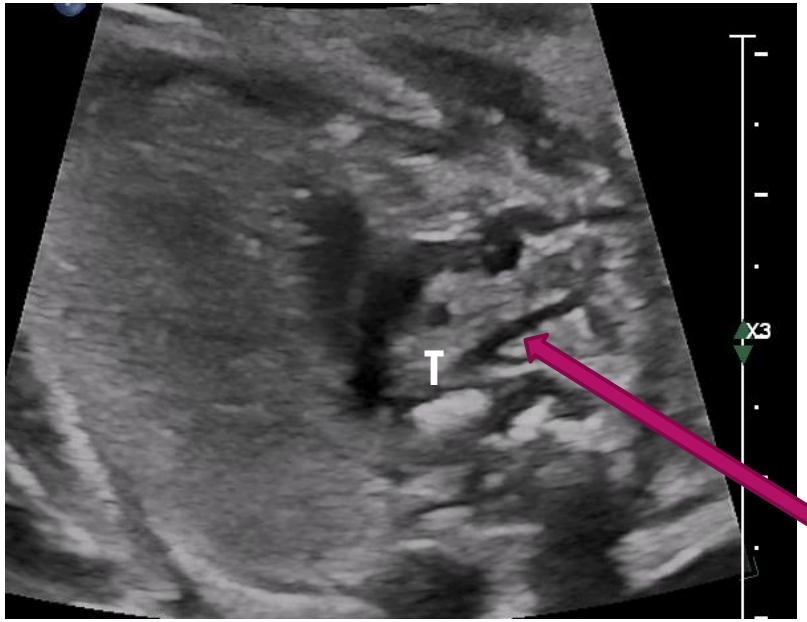




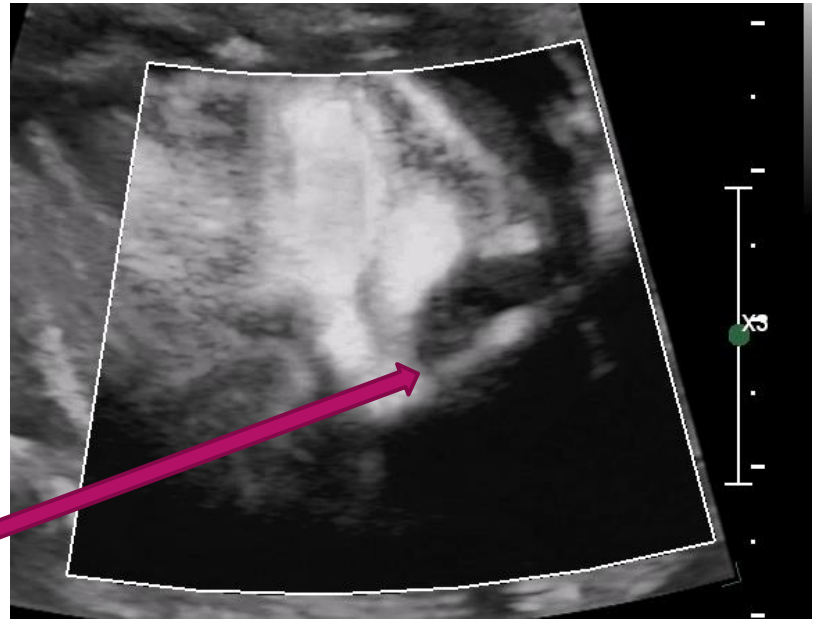
Song MJ et al., Ultrasonography, 2017;36:278-283

The most common vascular ring - ARSA

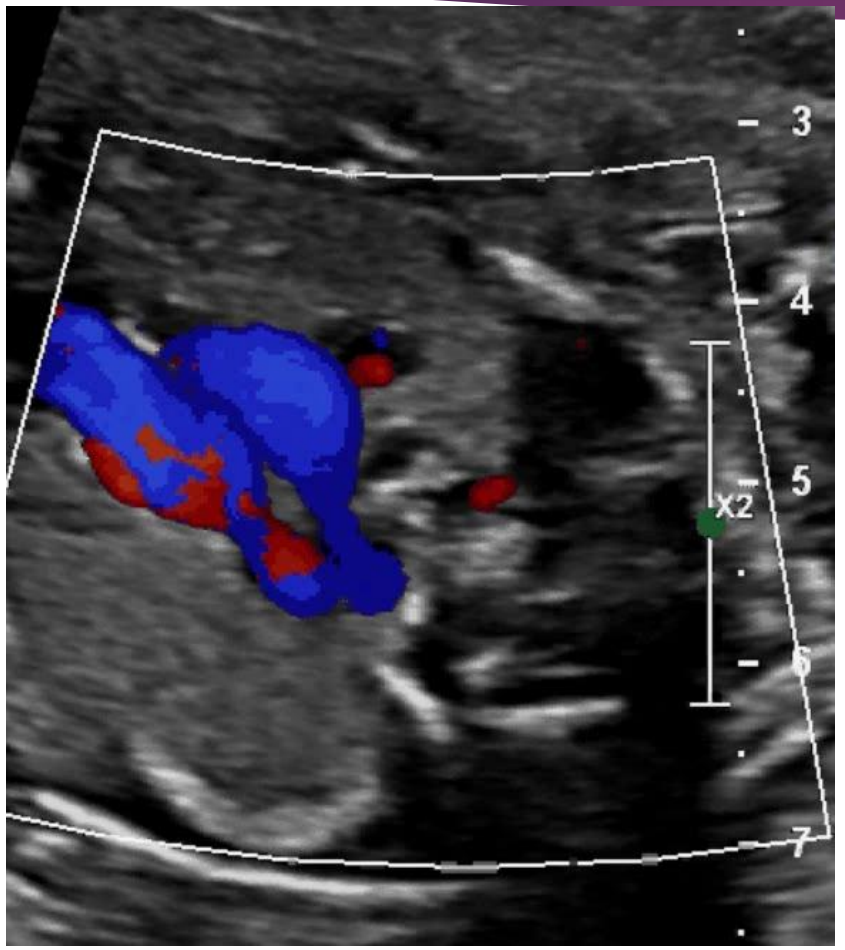


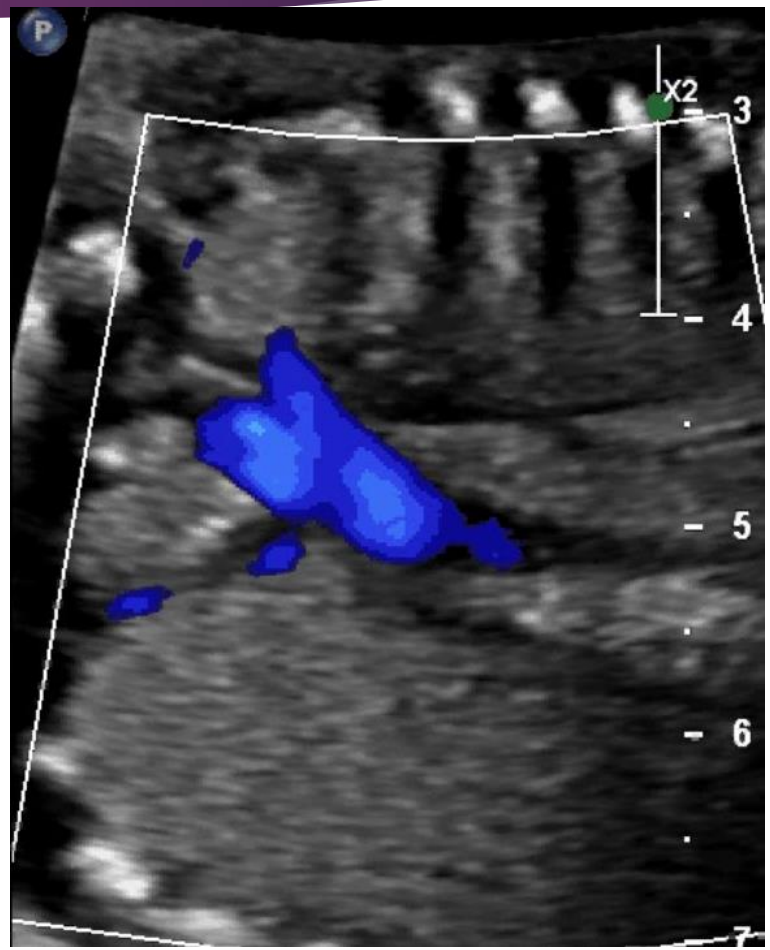


ARSA

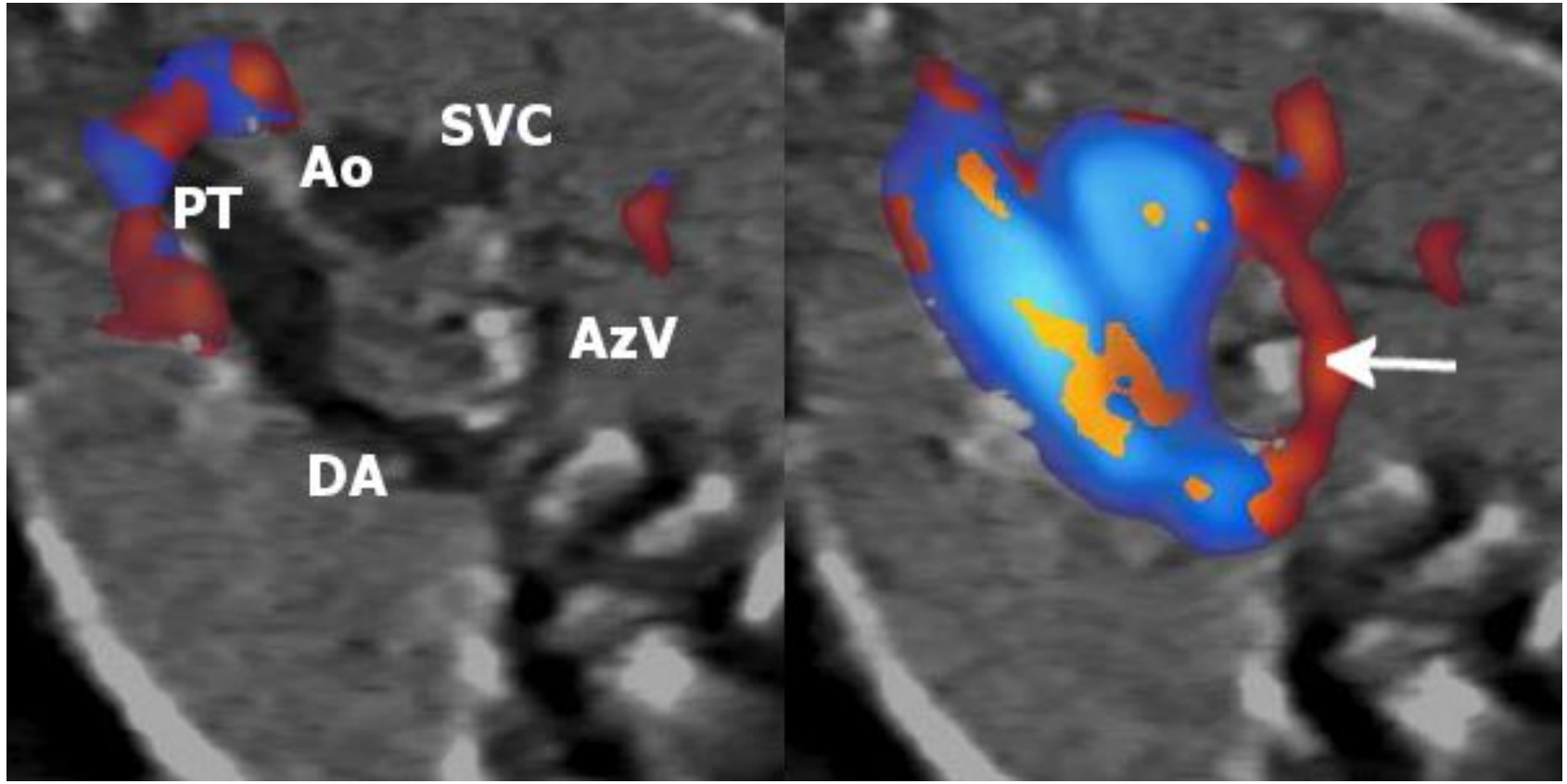


ARSA in 3VV-T and frontal view



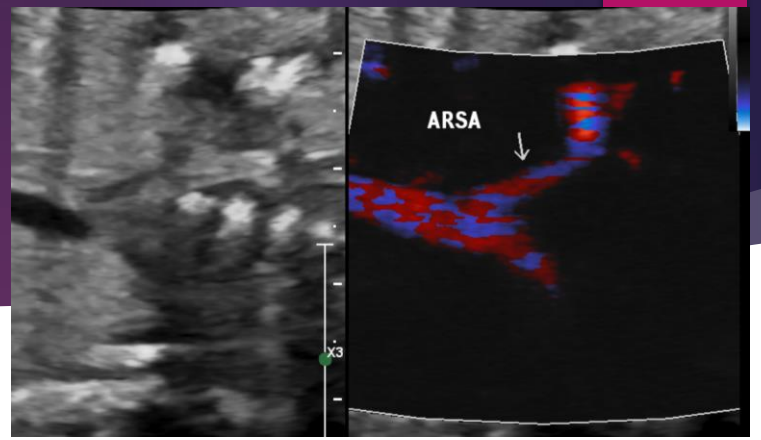


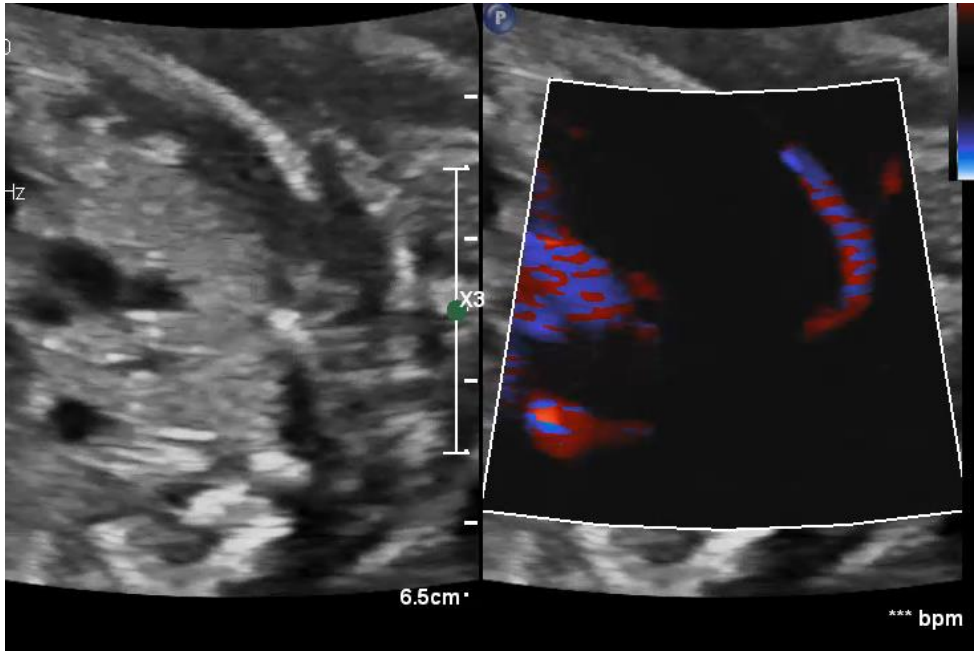
Mistakes of ARSA diagnosis is … likely



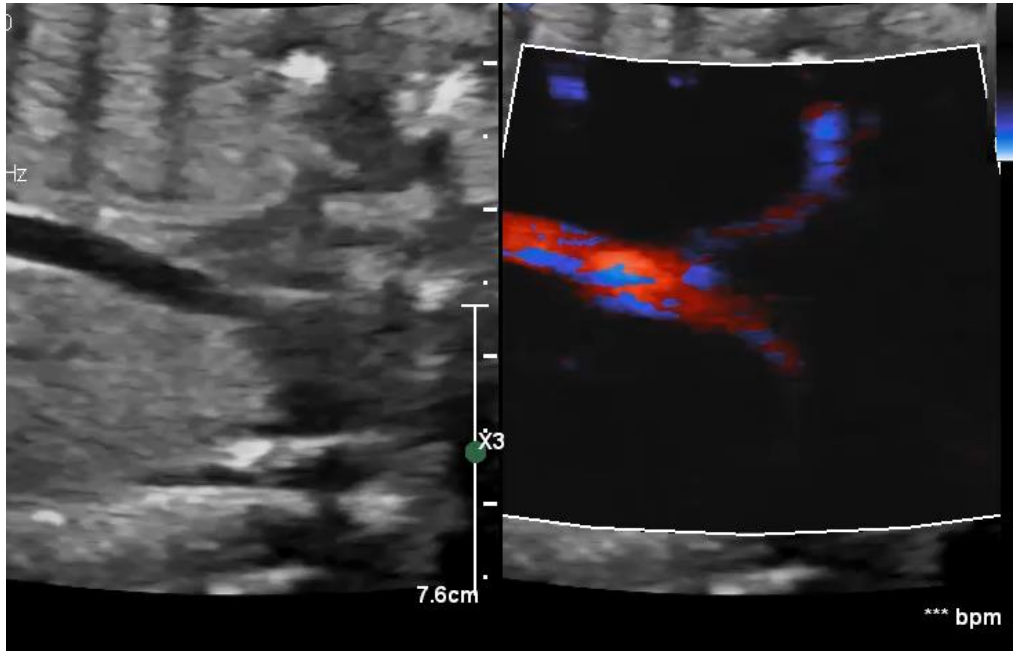
Azygos vein – not ARSA!
Be aware of the different course and character of flow!

MFI flow helps





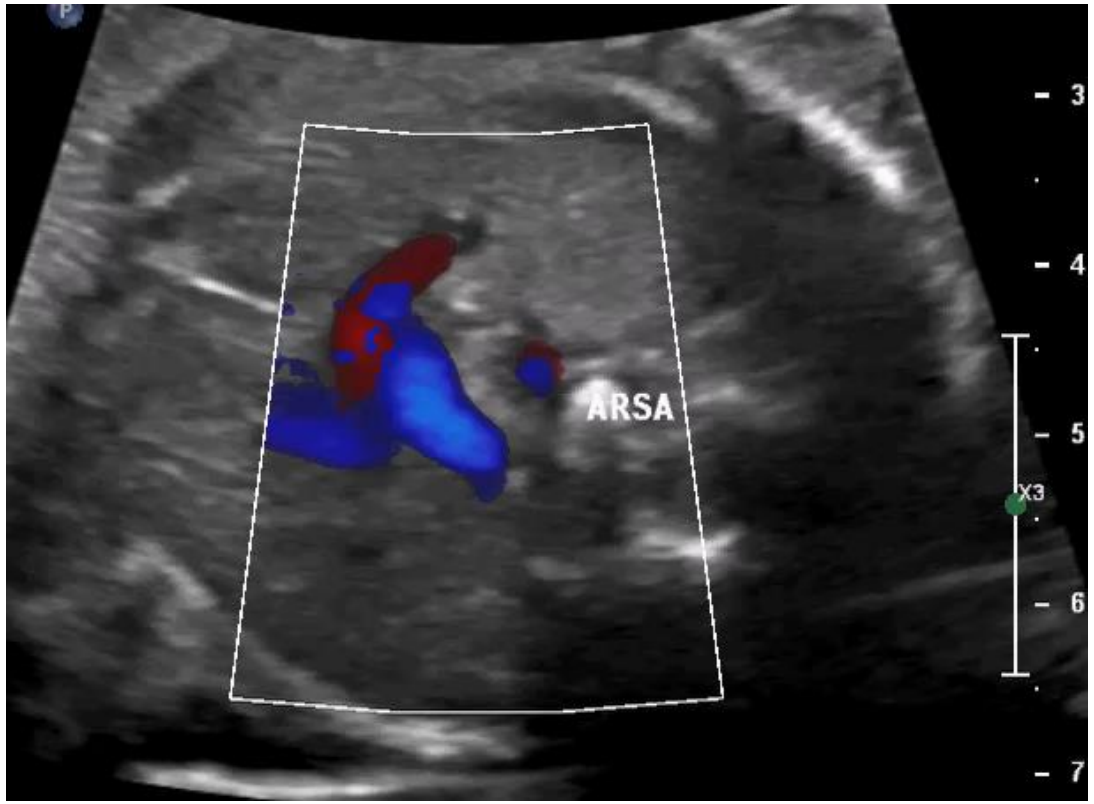
24 weeks



24 weeks

ARSA & genetic problems

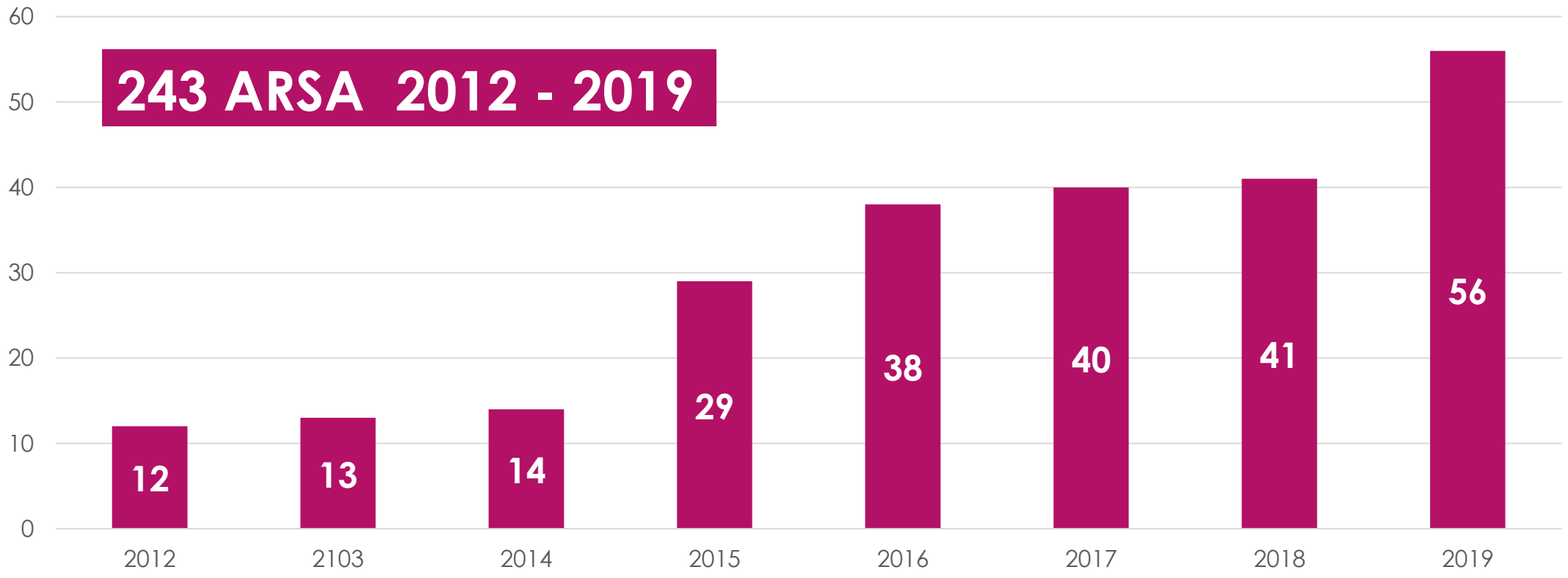
Microarray analysis has no additional value in fetal aberrant right subclavian artery: description of 268 pregnancies and systematic literature review



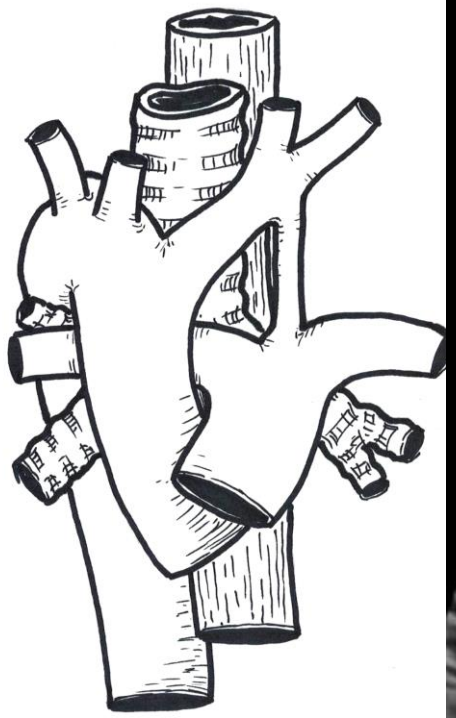
- If ARSA is an isolated finding can be a soft marker of Down syndrome not and indication for genetic test
- Incidence:
  - Isolated ARSA: T21 1:268 (0,4%)
  - Non-isolated ARSA: T21 1:22 (4,5%)
  - Isolated ARSA from literature: T21 13:579 (2,2%); none 22q11.2

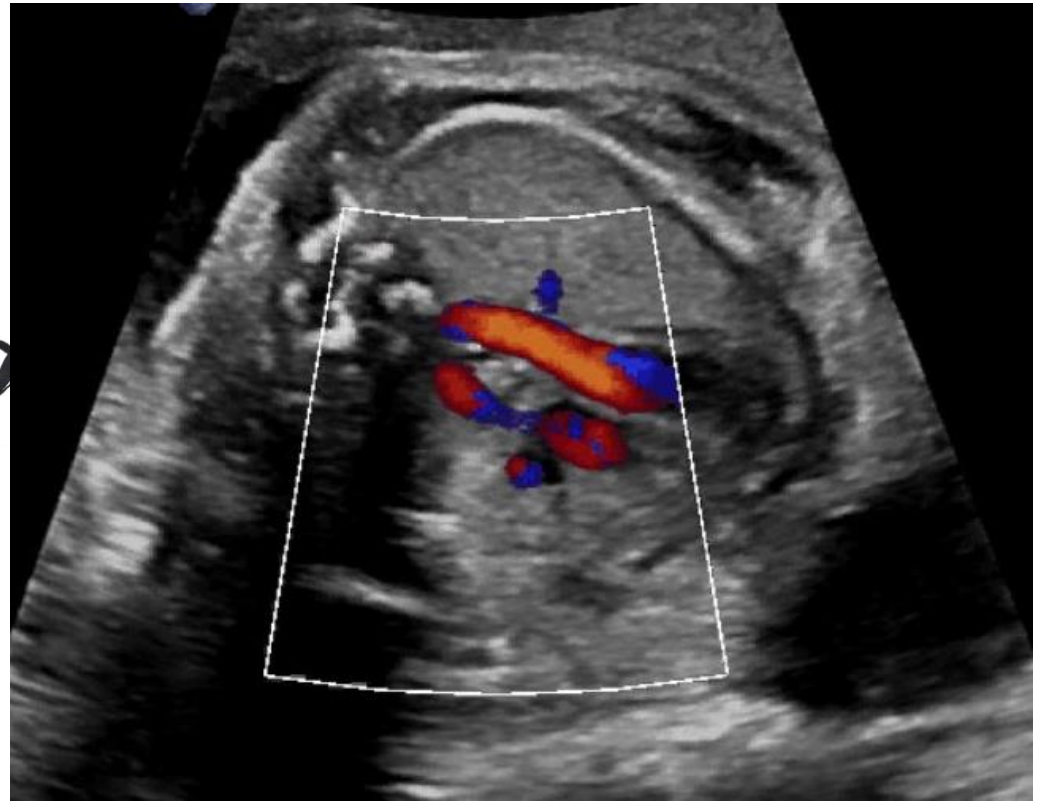
Sagi-Dain L et al. 2019;53:810-815

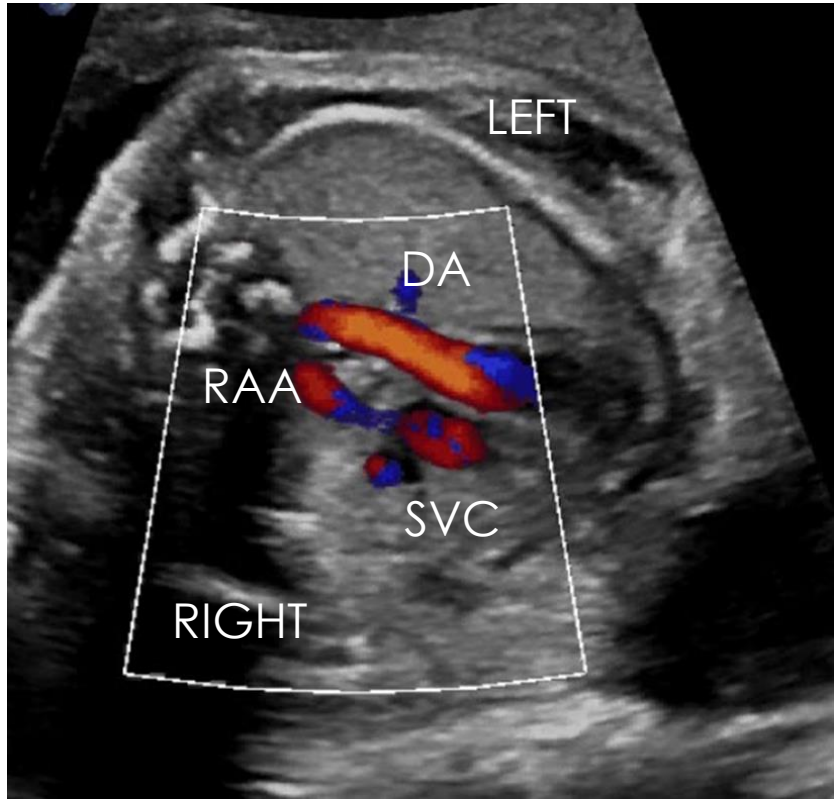
ARSA in the own material



Right aortic arch

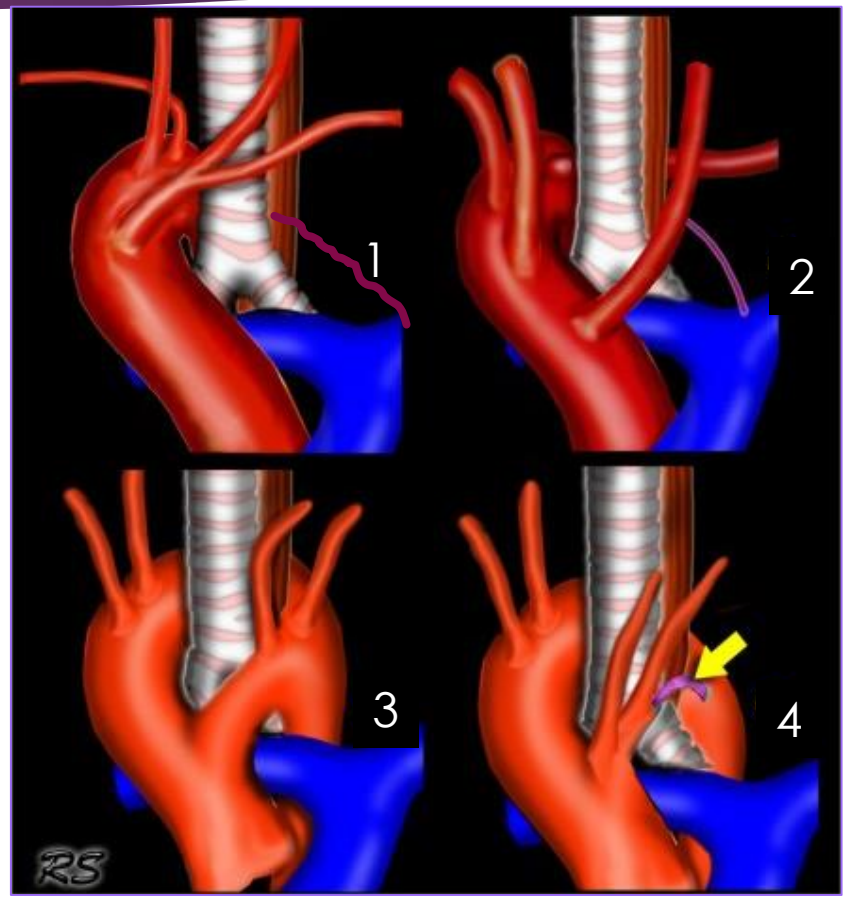






Right aortic arch

- RAA – mirror image and LDA(1)
- RAA – mirror image and RDA
- RAA & ALSA & LDA (2)
- RAA in Double Aortic Arch
  - Both arches equal in size (3)
  - Hypoplastic left arch, atretic distal part (4)



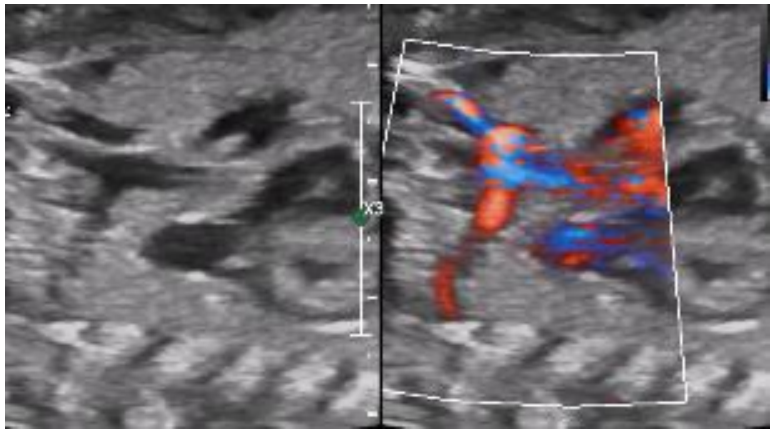
http://www.radiologyassistant.nl/en/ *Siegel MJ, Smithuis R*

RAA – step by step diagnosis

4 CH view ThAo on the right side

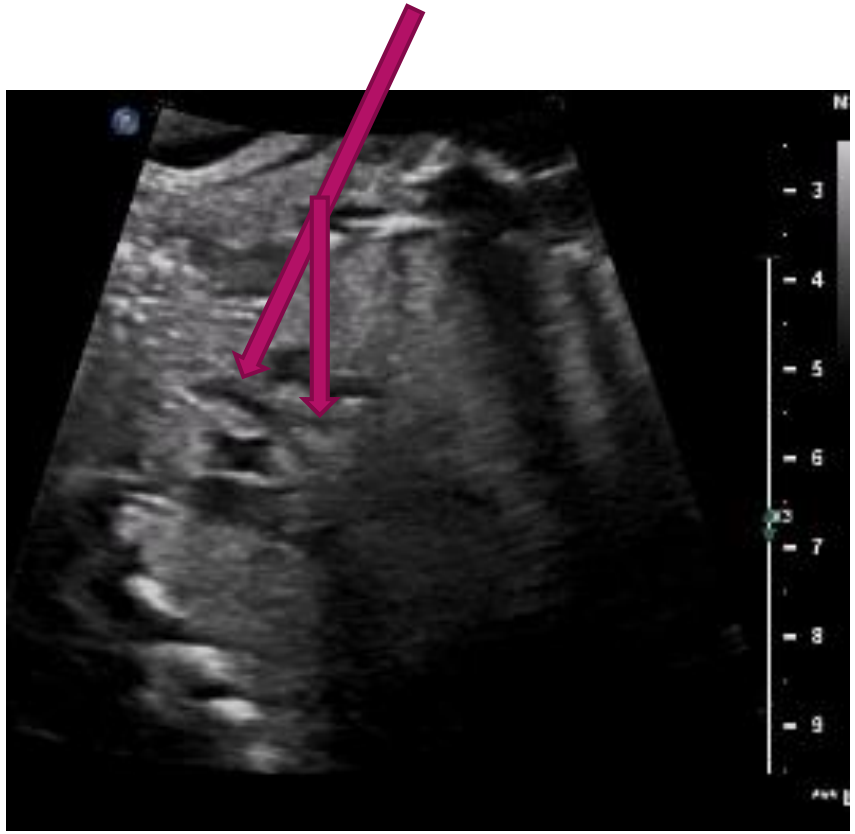
Ao & SVC on the right side

Colour Doppler necessary to evaluate arch branches, DA, PAs

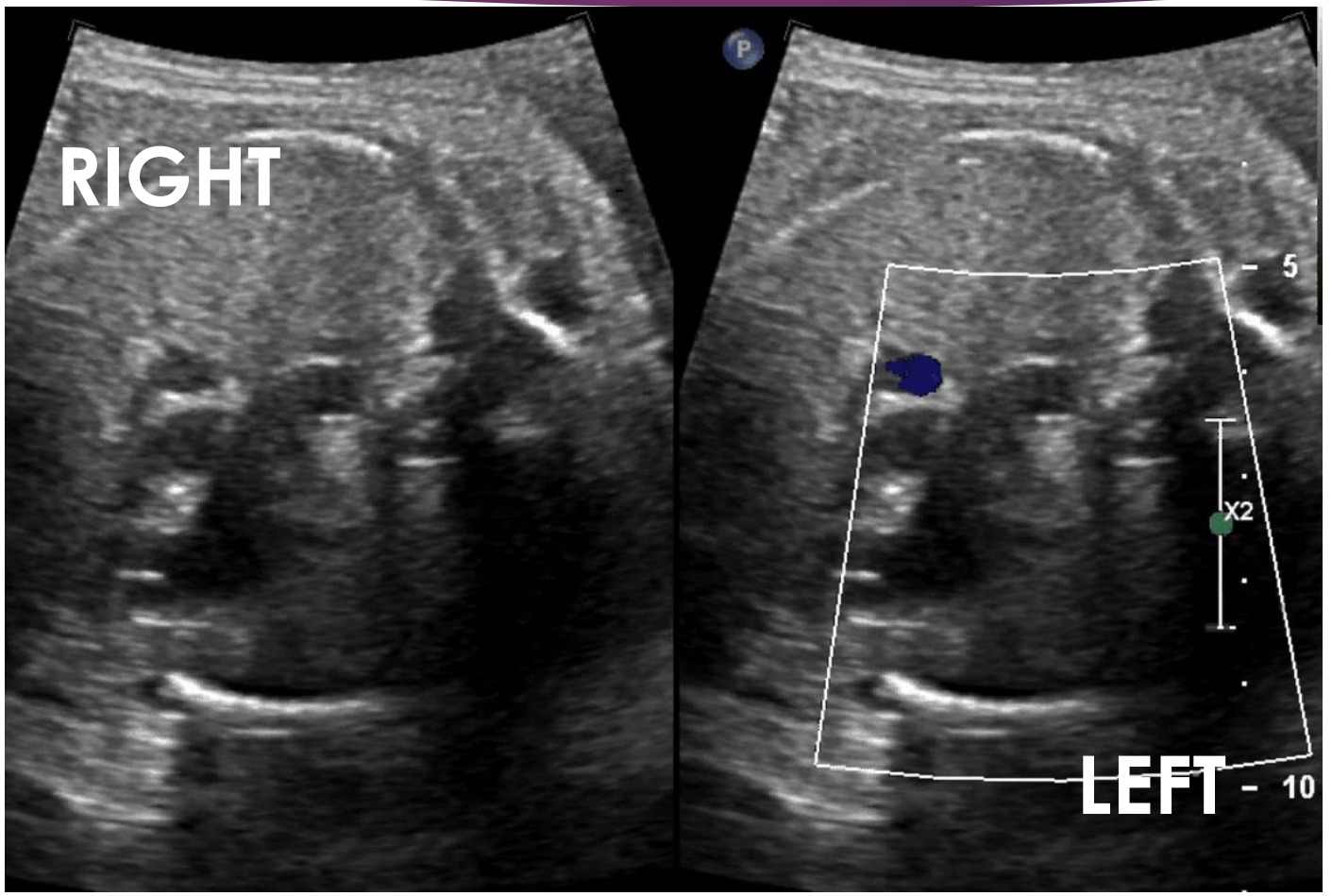


Not only vessels are seen

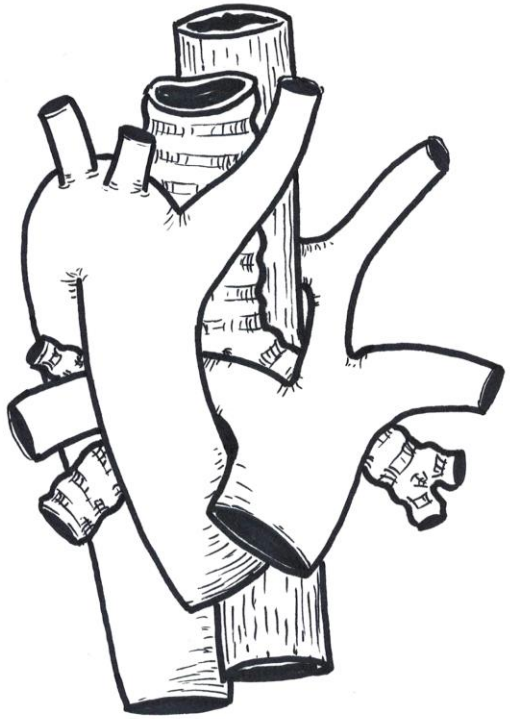
Trachea & bronchus

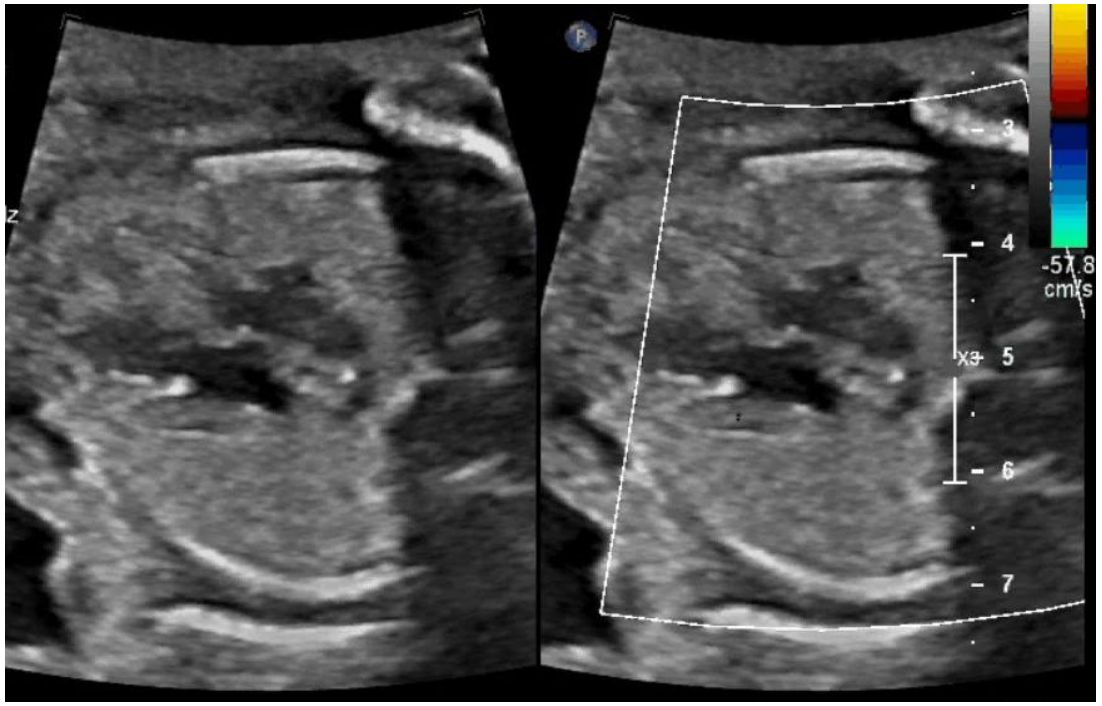


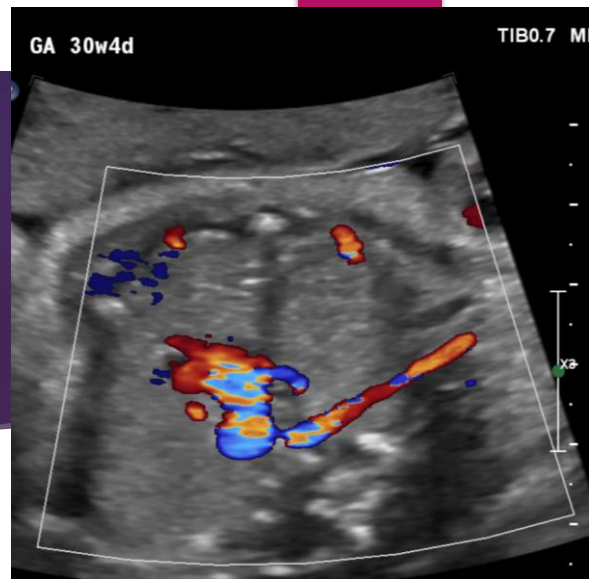
RAA & RDA



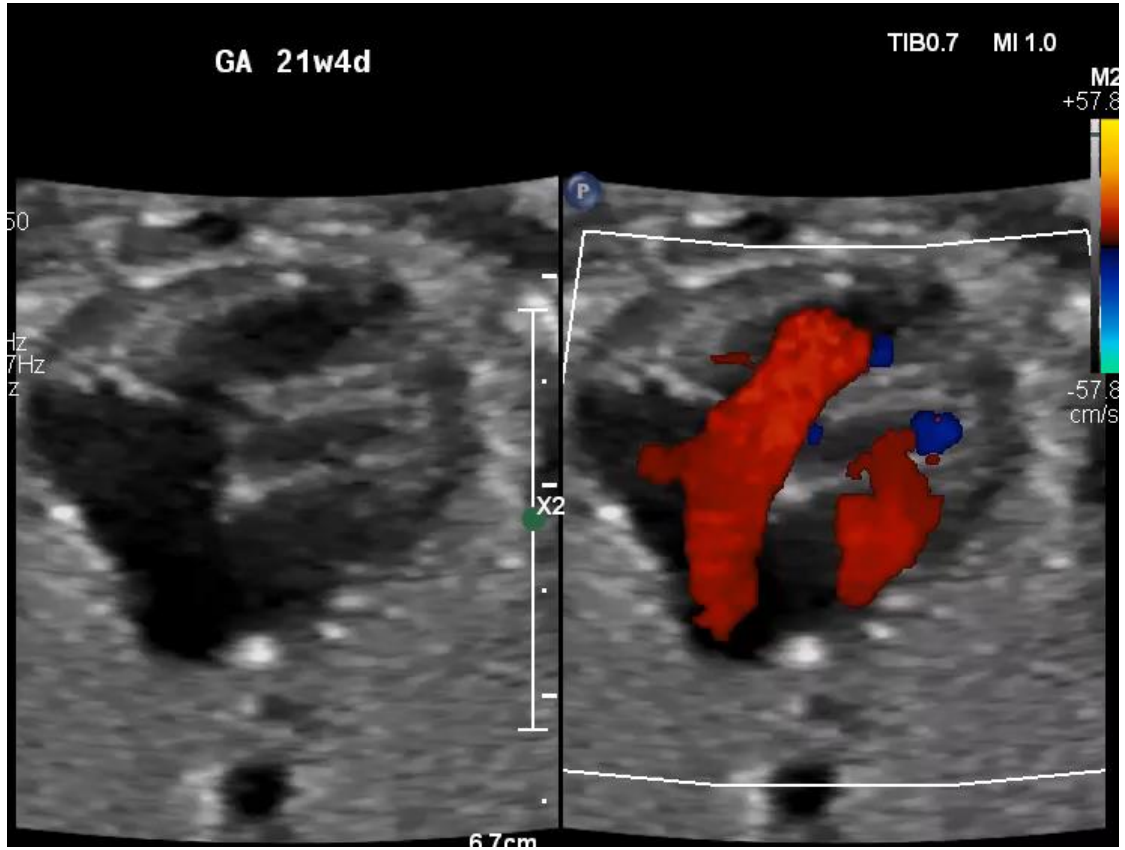
RAA & ALSA – caused vascular ring

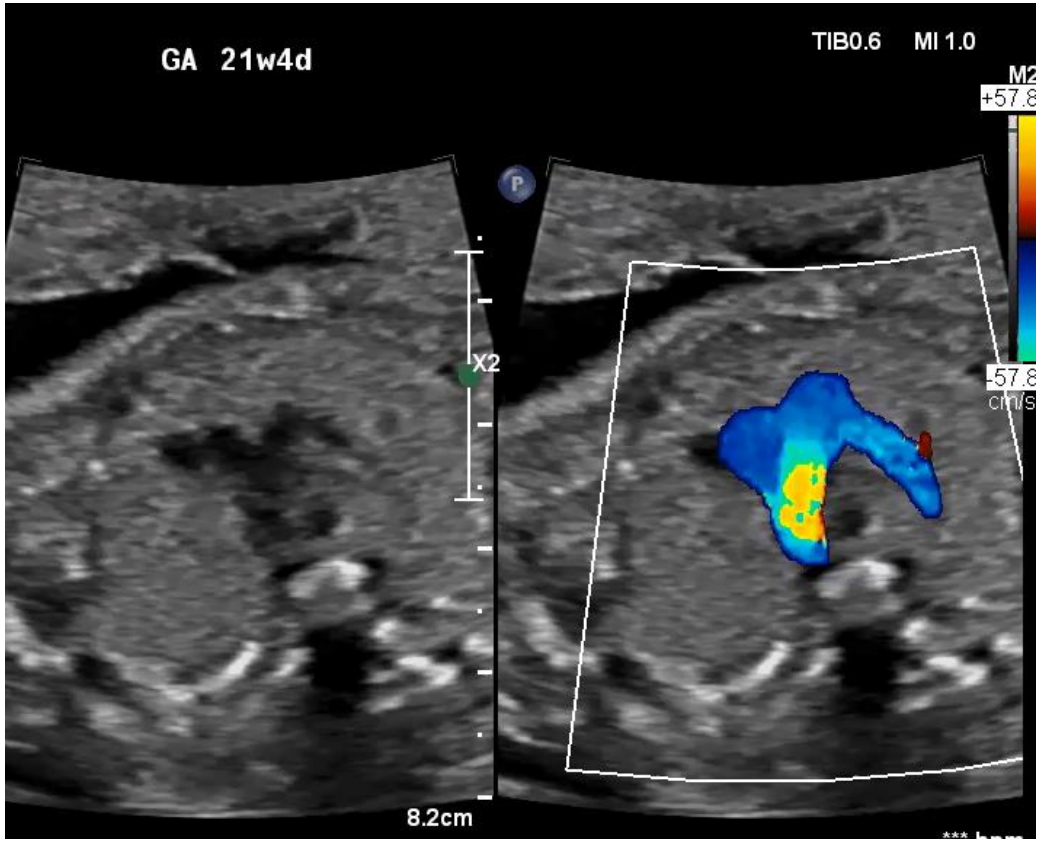




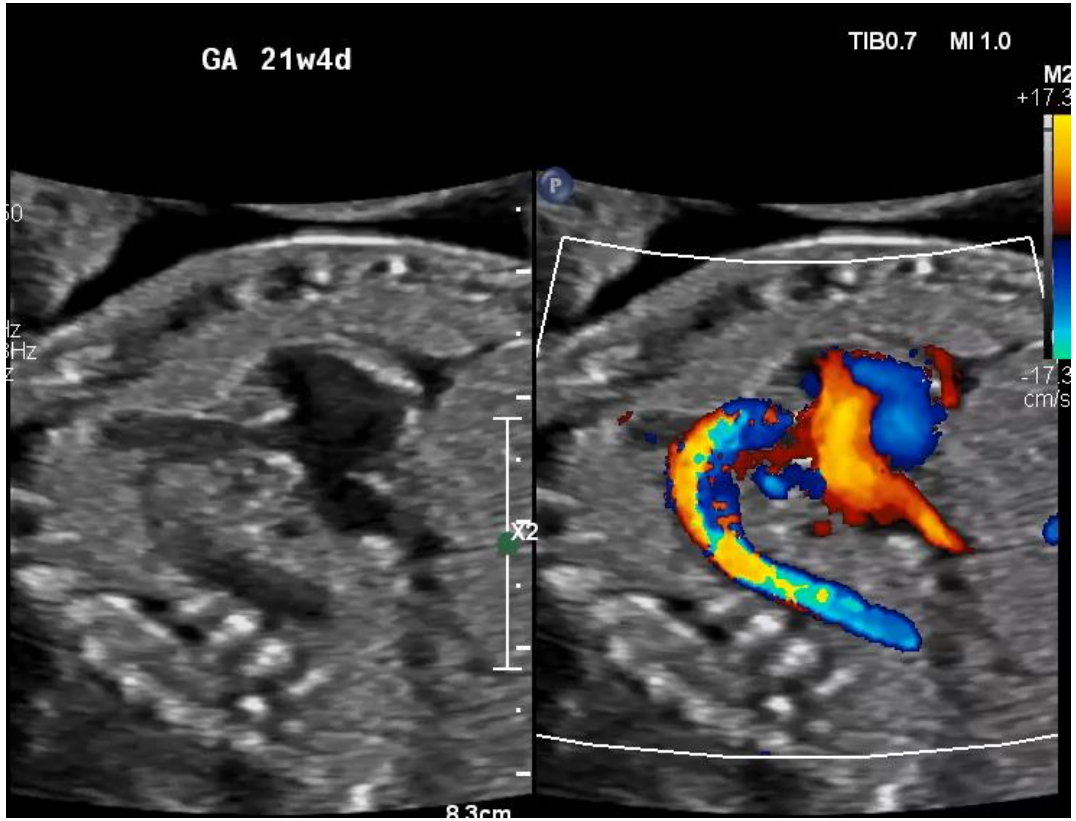


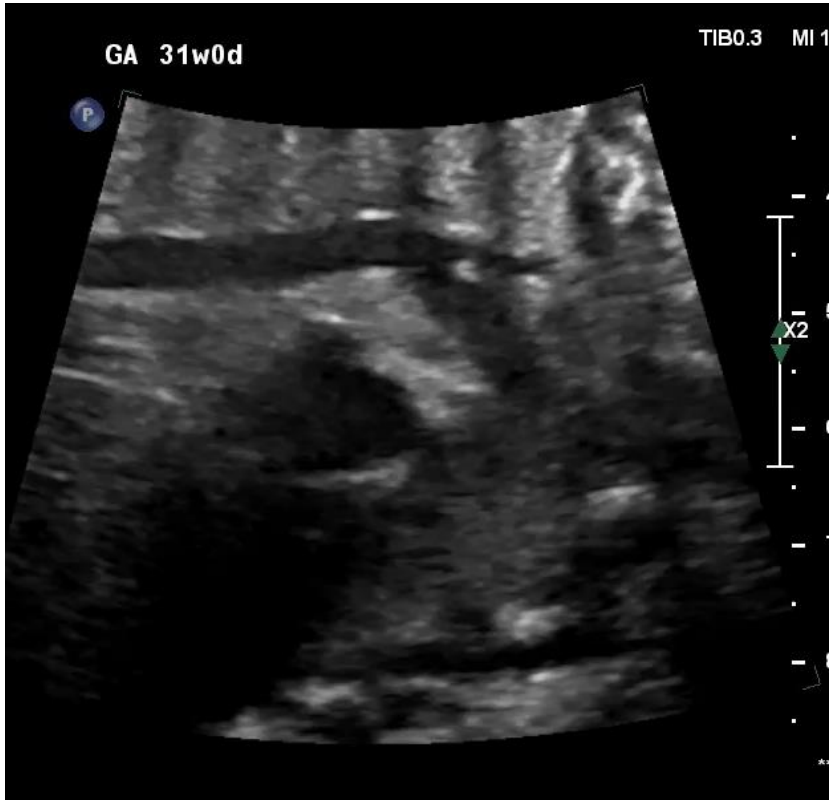
Be careful if you see RAA & RDA …





Progression of the disease …





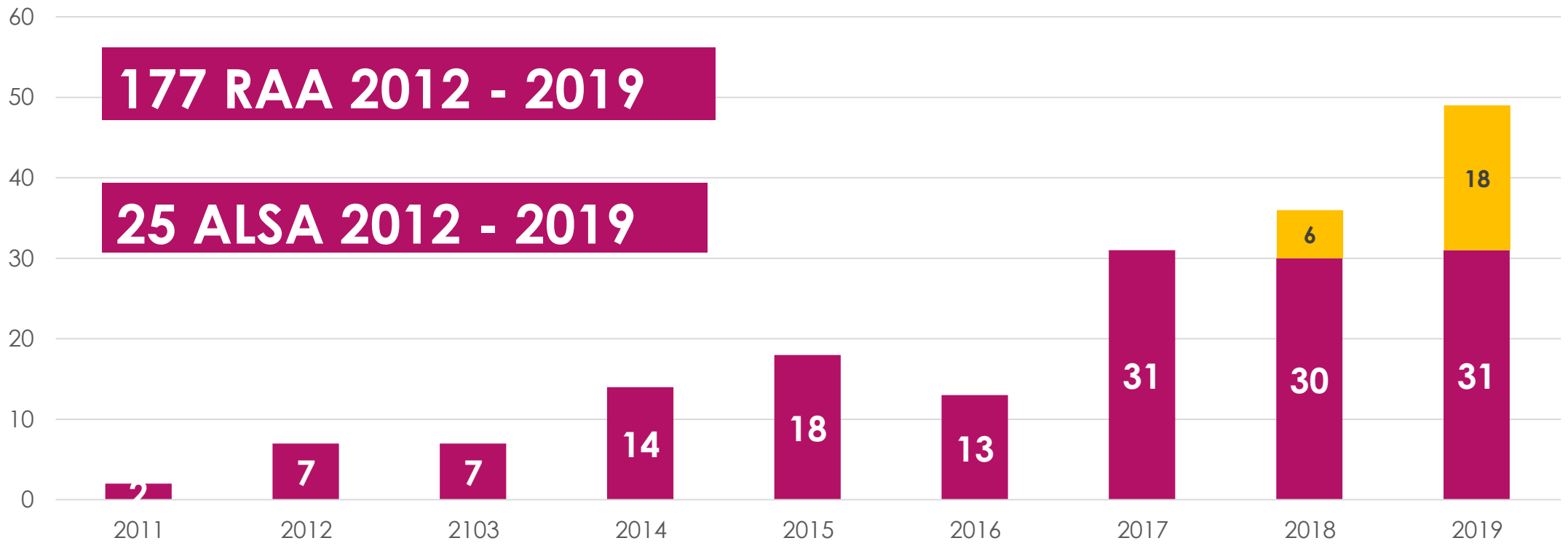
Left PA filled by left DA

Final diagnosis: DOUBLE DUCT DEPENDENT lesion

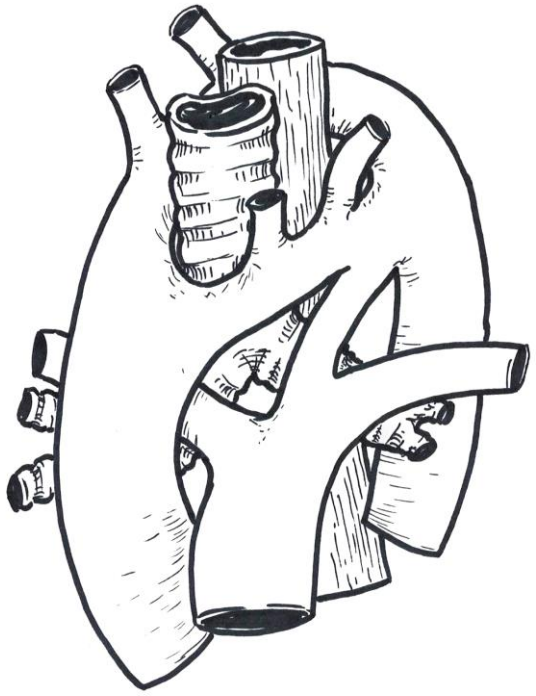
- RAA & double DA & ALSA
  - Aortic coarctation
- Left pulmonary artery from LEFT arterial duct
- VSD

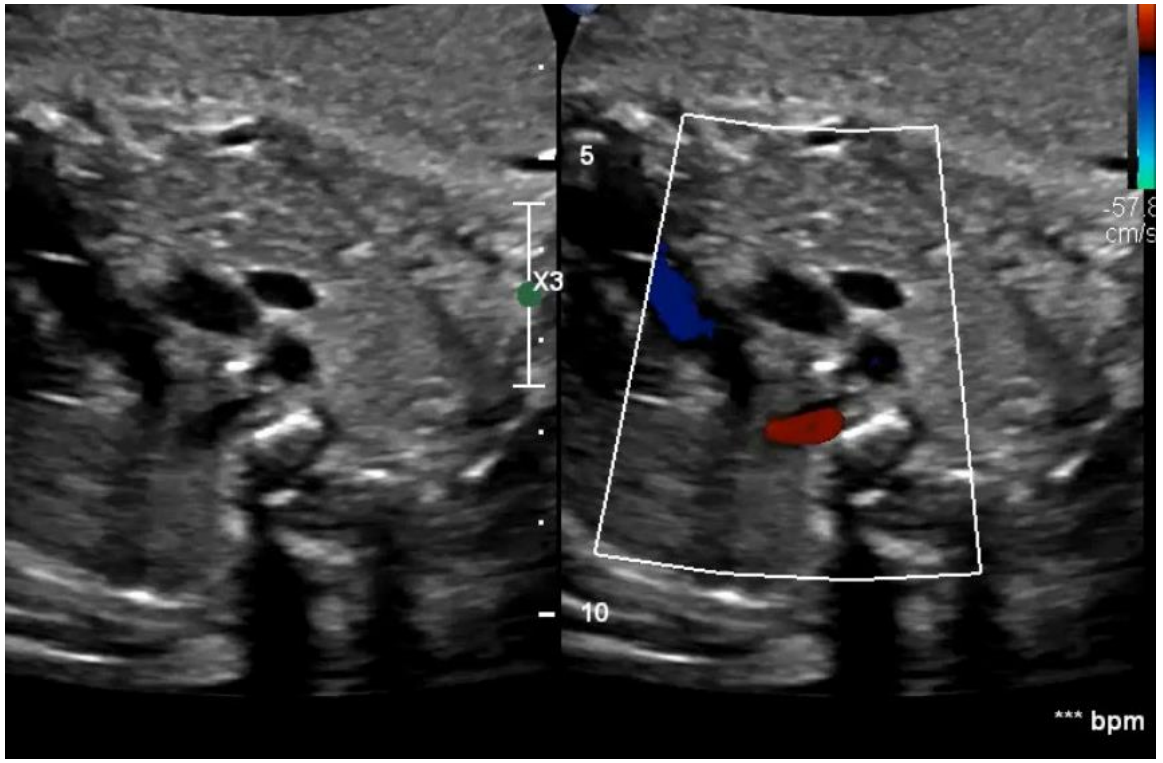
From BENIGN anomaly like abnormal position of all vessels (SVC, Ao and ductus arteriosus on the right side) to SEVERE ductal dependent lesion

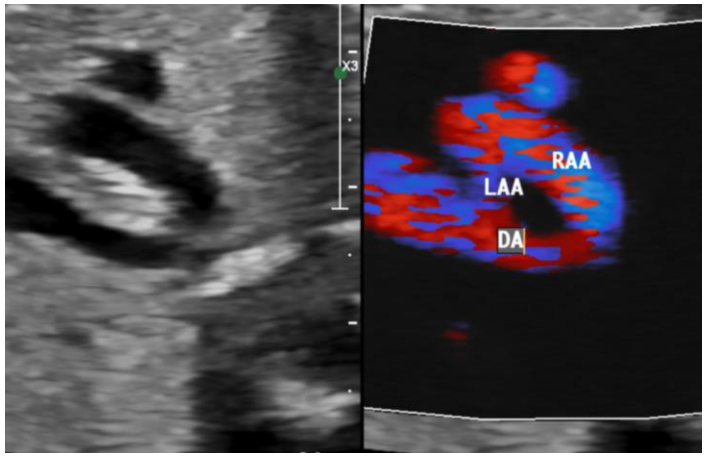
RAA in the own material



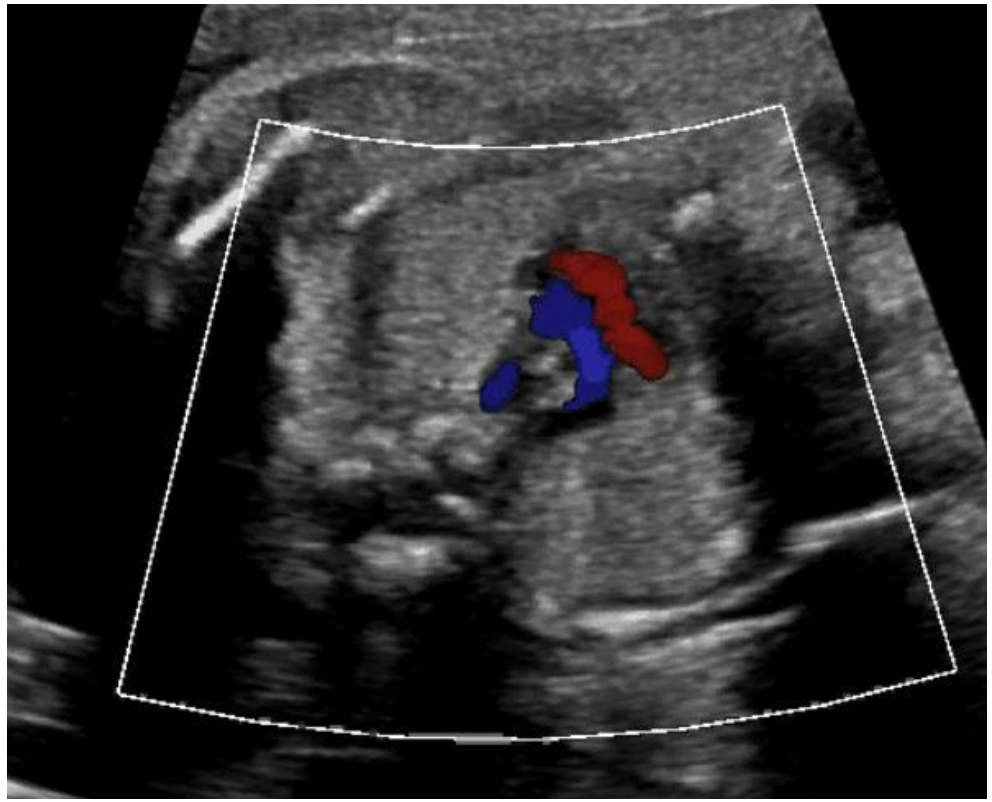
Double AORTIC ARCH

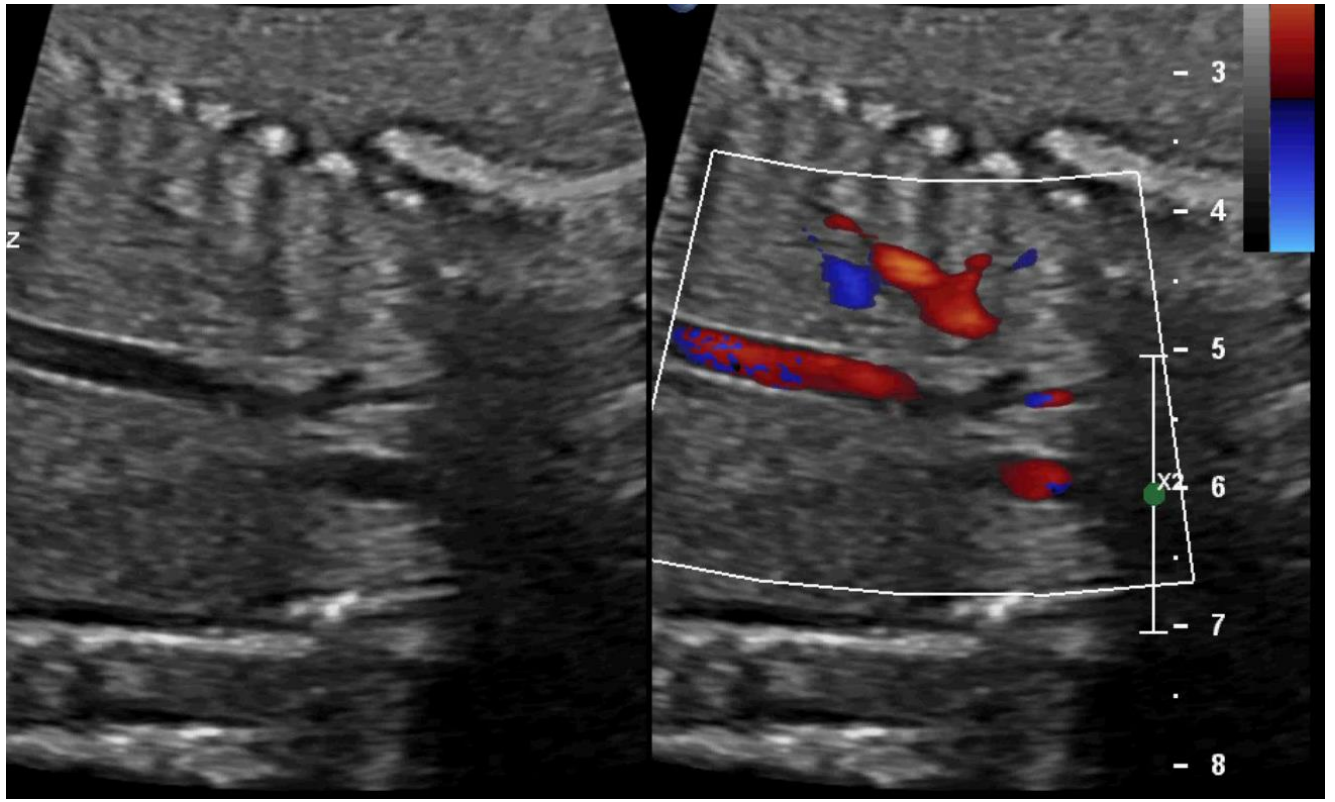




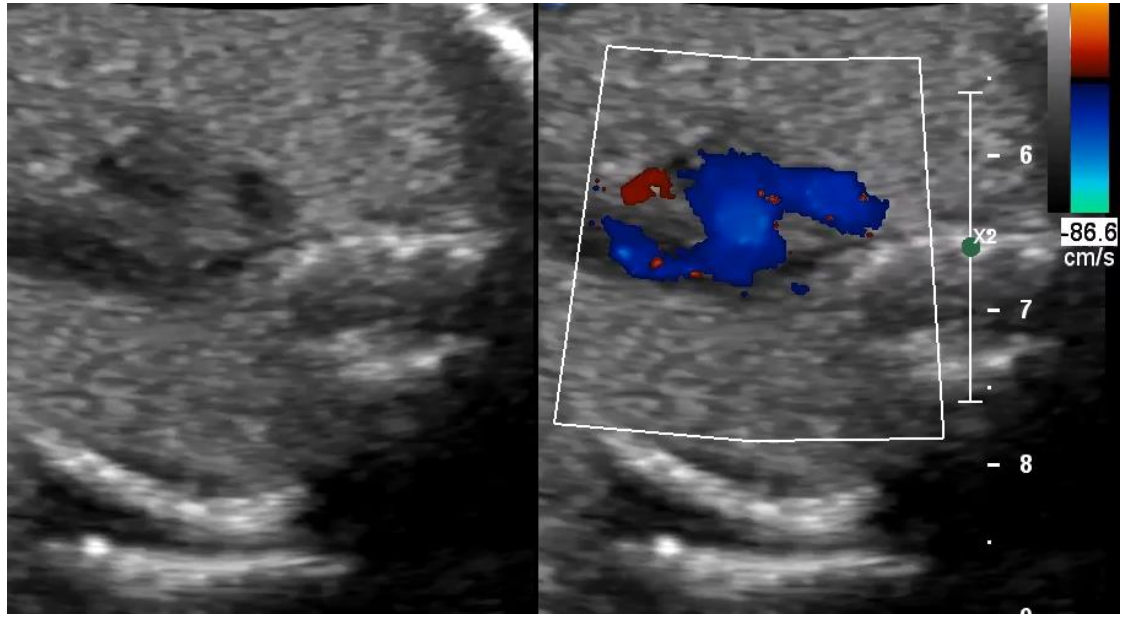


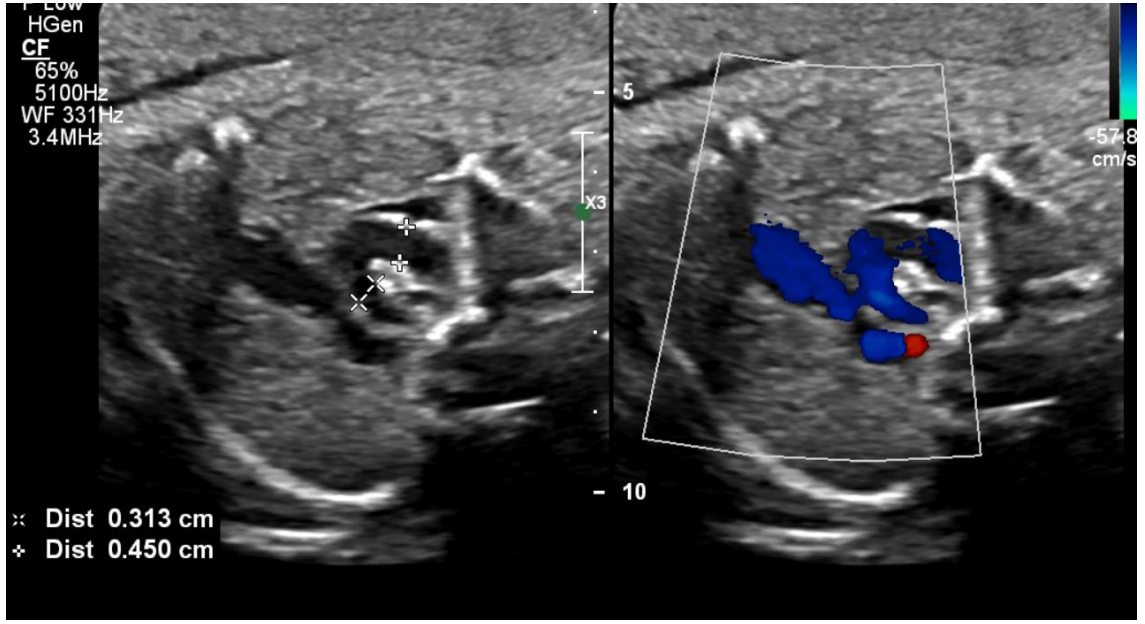
DAA in different planes



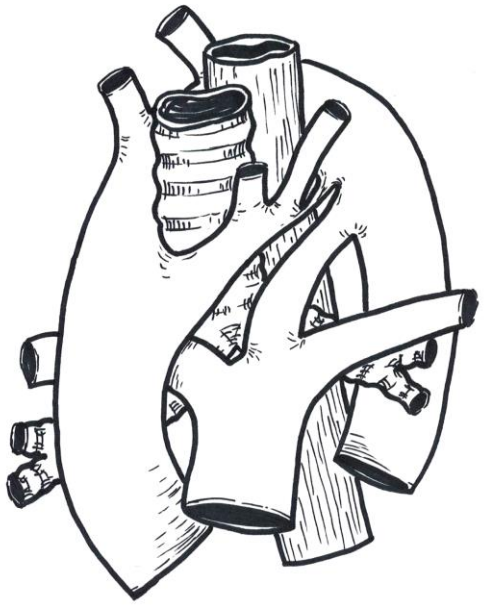


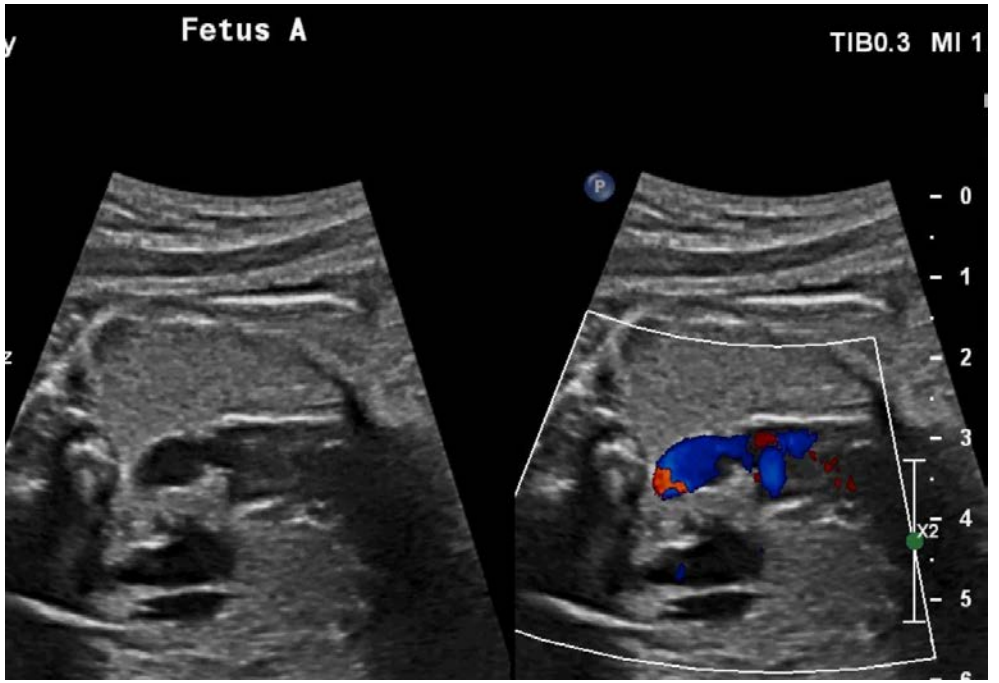
RAA both arches eqal in size



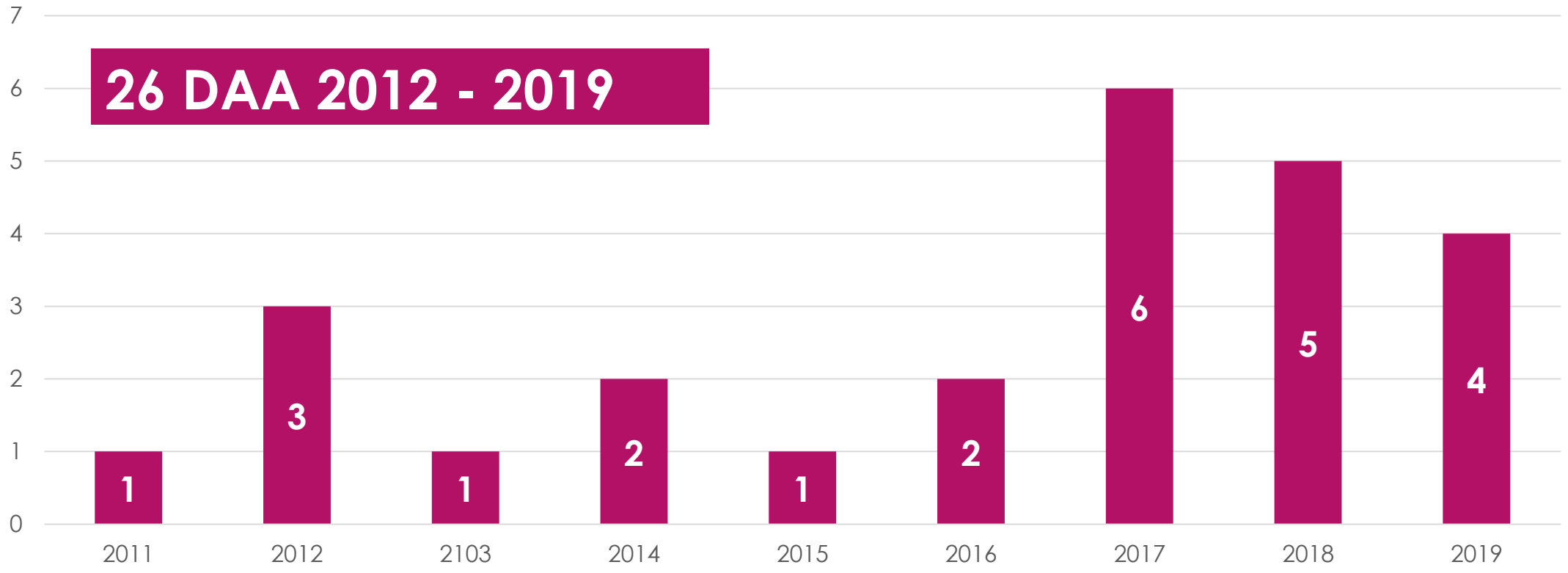


DAA with hypoplastic left arch





DAA in the own material



Other prenatal issue if vascular ring is detected

ULTRASOUND

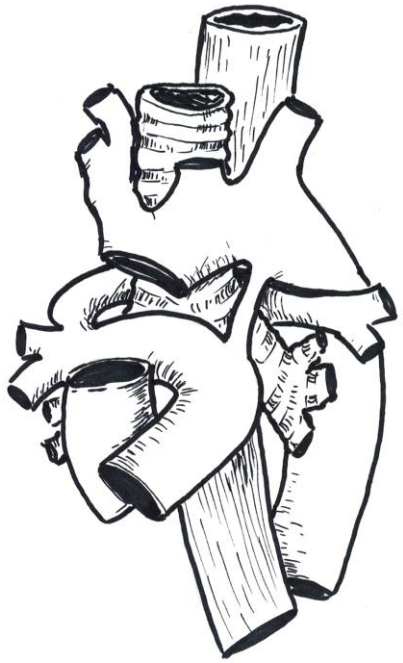
- If vascular ring is present – all vessels crossing from one side to the other behind the esophagus and traches – RISK for POLIHYDRAMNION
  - Check mediastinum in cases of „idiopathic" polyhydramnios
- Look at the THYMUS

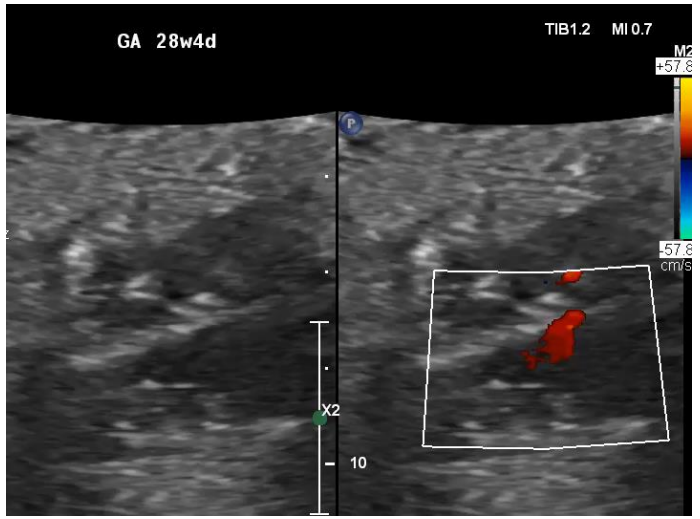
GENETICS

- Important: first trimester screening and additional ultrasound markers
- Isolated RAA
  - Chromosomal 8,2%
  - 22q112 deletion 3,7%
- Double aortic arch
  - No enough data concerning incidence of genetic problems – should be considered as in RAA ?

NO DATA ABOUT OTHER MARKERS PROVIDED

Pulmonary artery sling: lPA from rPA

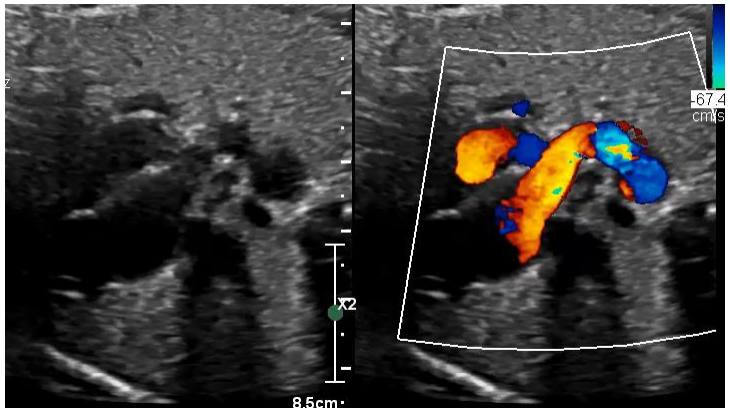


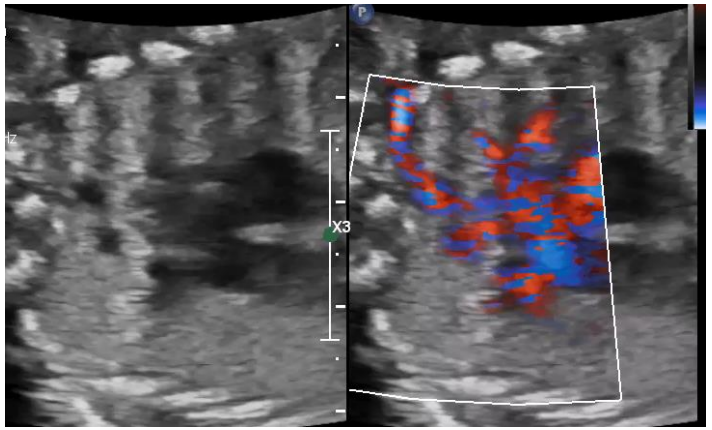


Remember

- Echocardiographic imaging of vascular rings is easier in fetuses than after birth
  - Not all abnormal vessels create vascular rings
- If any kind of abnormal position of the vessels is seen careful evaluation of anatomy is obligatory
  - All head – neck vessels
  - Pulmonary artery branches
  - Arterial ducts
- „BENIGN" pathology at screening can be severe ductal dependent lesion !!!

“If vascular ring is diagnosed prenatally – when and what kind of postnatal evaluation should be done ?”





THANK YOU

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