Commonly Missed Congenital Heart Disease Lesions and High Yield Views to Avoid These

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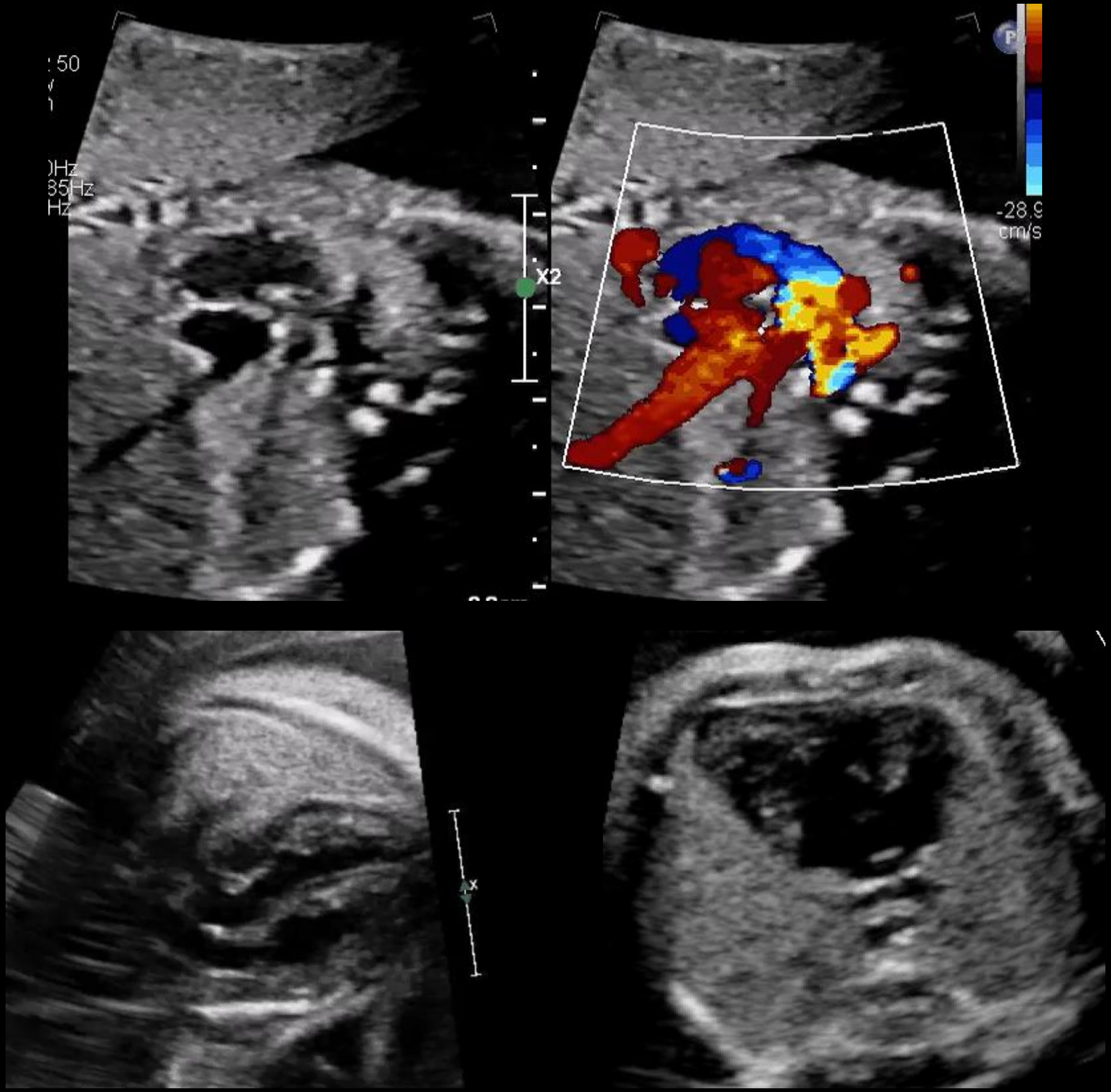

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- I have no financial relationship with any manufacturer of any commercial product and/or provider of commercial services discussed in the conference.
- I do not intend to discuss an unapproved/investigative use of a commercial product or device in my presentation

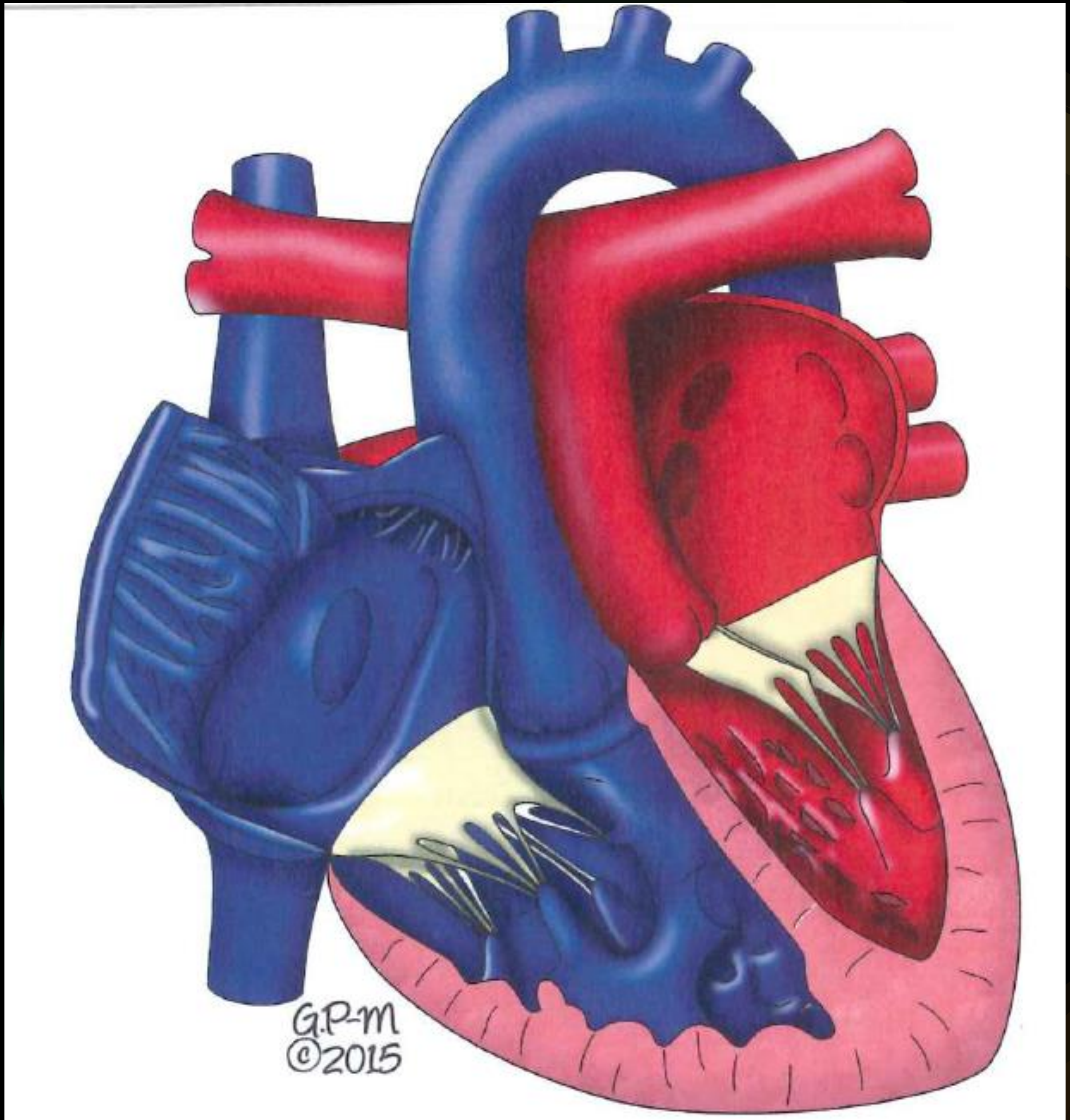
Overview

- Objectives
- Dextro-Transposition of Great Arteries (D-TGA)
- Total anomalous pulmonary venous return (TAPVR)
- Conclusion

Dextro-Transposition of the Great Arteries (D-TGA)

D-Transposition of the Great Arteries

- Most common form of cyanotic CHD presenting in the newborn
- Comprises 5-7% of all CHD
- ~3 per 10,000 live births
- 70% male (tend to be LGA and full term)
- Chromosomal abnormalities are rare
- Low prenatal detection rate



Atlas of Congenital Heart Disease Nomenclature. Ezon, Goldberg, and Kyle

Anatomy

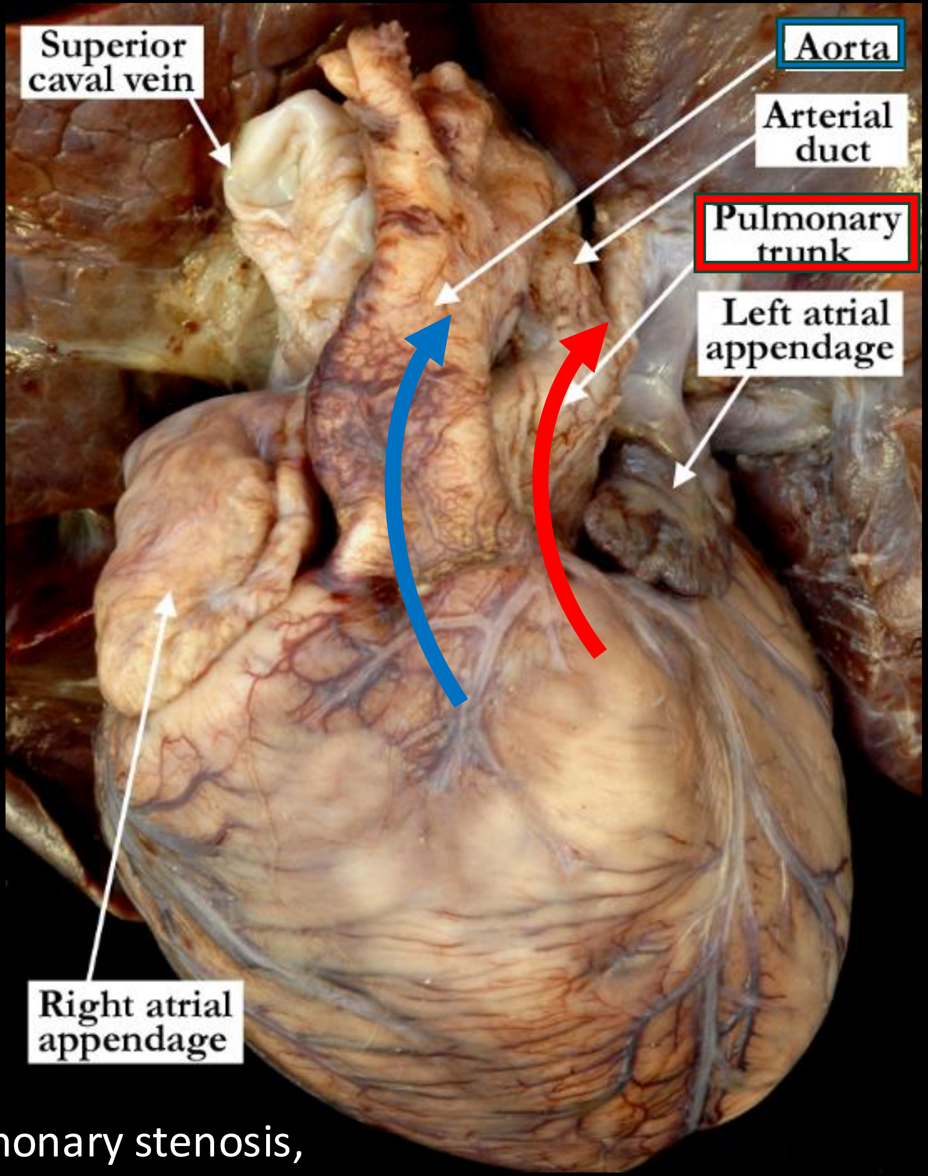
D-TGA arises when the "great arteries" have reverse origins due to embryologic failure of normal septation

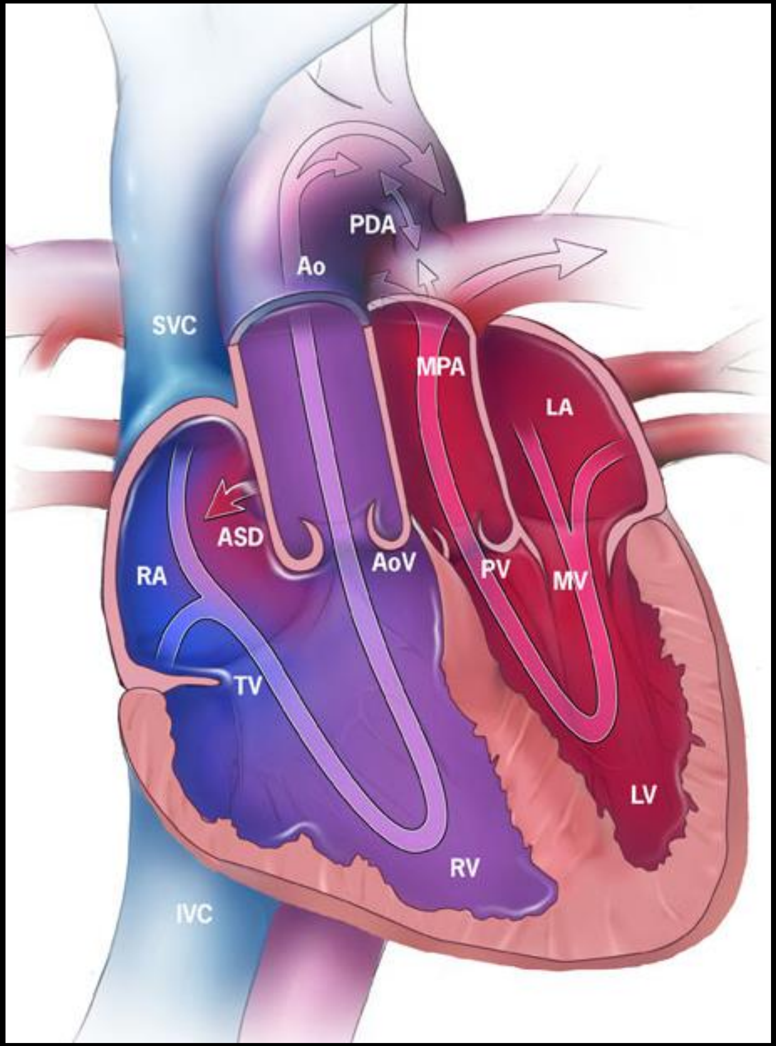
Aorta arises from the right ventricle

Pulmonary trunk arises from left ventricle

Aorta is anterior/rightward to pulmonary trunk; aorta is normally posterior to PT.

This orientation causes the great arteries to course in parallel. (Key finding by echo)





Can be associated with VSD (40%), pulmonary stenosis, coarctation of aorta, coronary anomalies

DTGA Physiology

Most common form of cyanotic CHD presenting in the newborn

Deoxygenated “blue blood” returns to right atrium via SVC/IVC and ejects from right ventricle through aorta to the body

Oxygenated “red blood” returns to left atrium from lungs via pulmonary veins and ejects from LV through pulmonary arteries back to the lungs

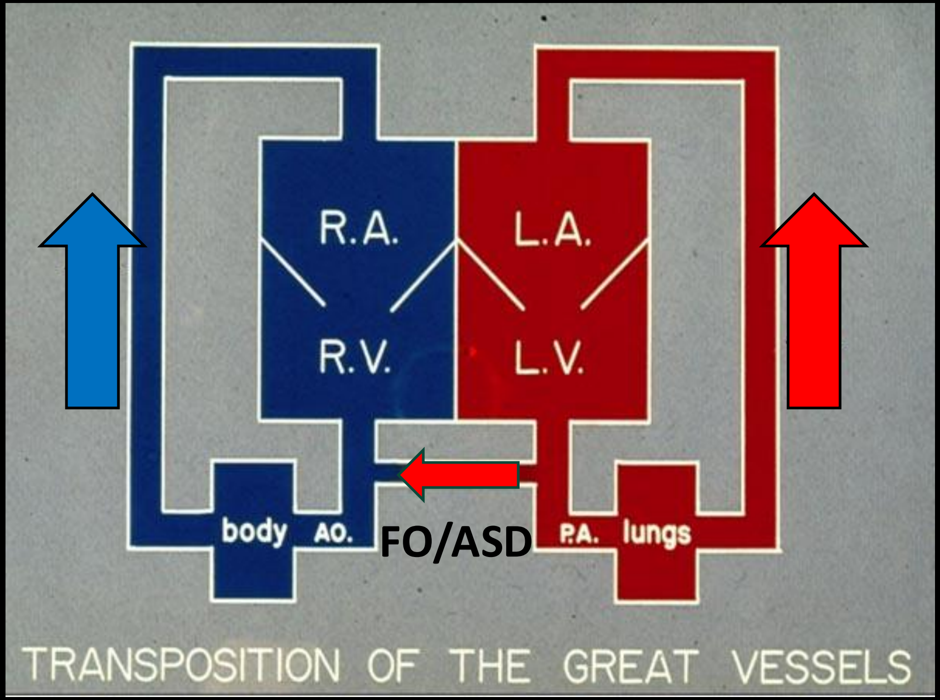
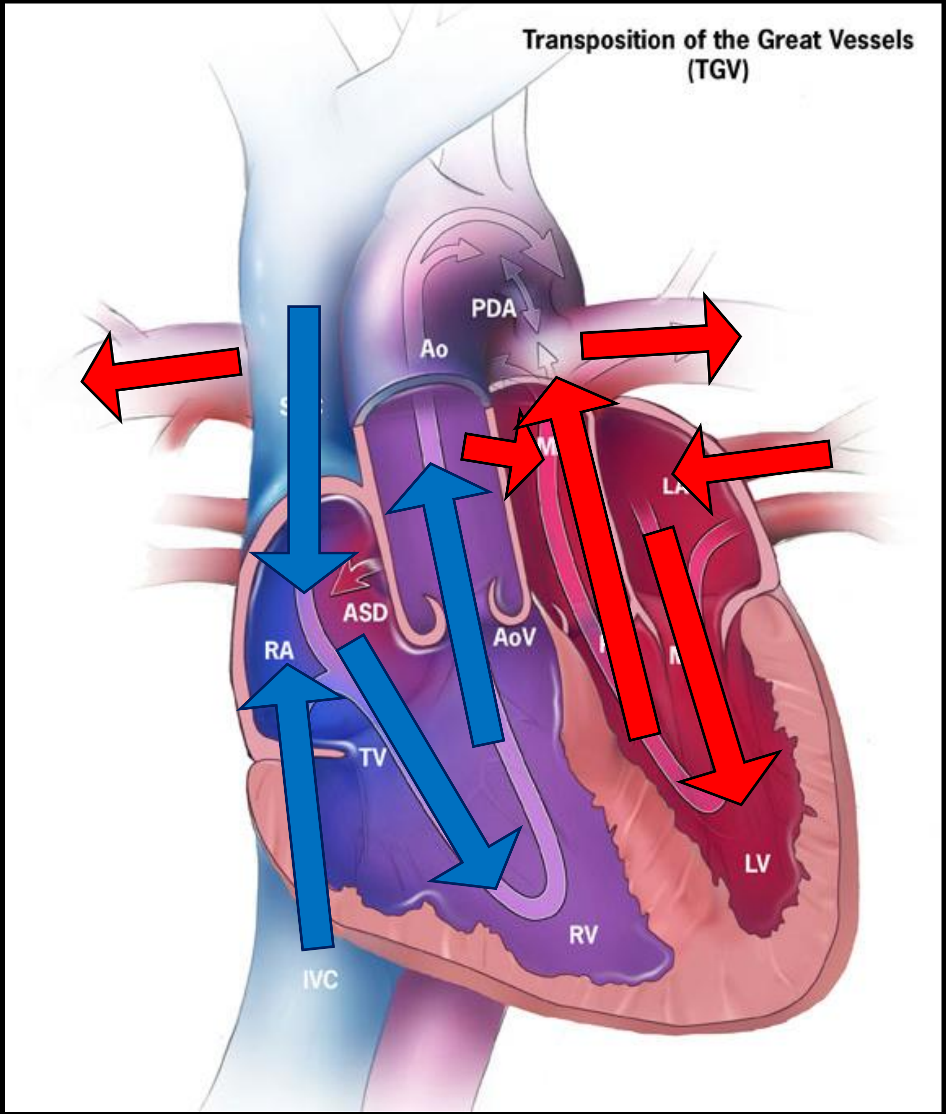


Image source: Neonatology Today 2010;5(8):1–7.



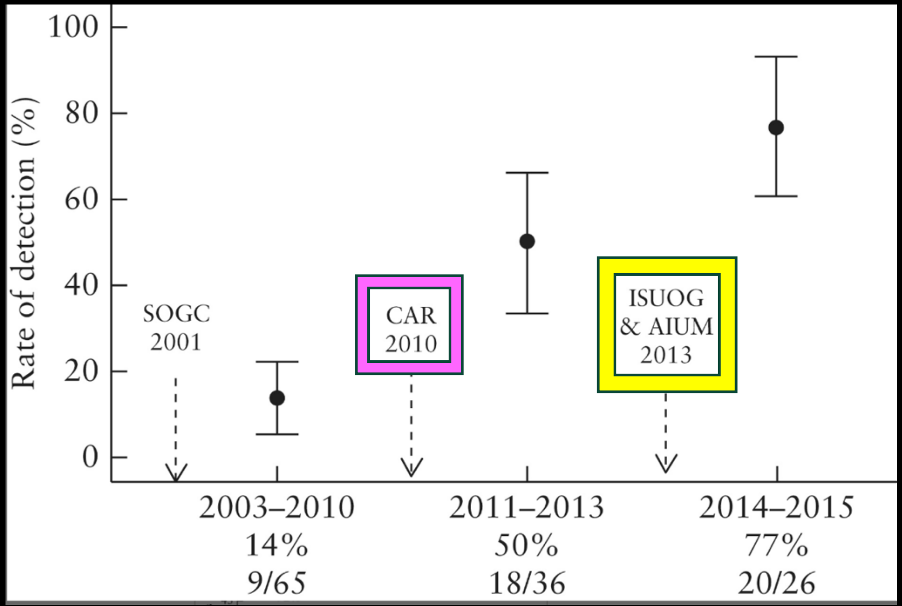
- Without communication between two parallel circuits, babies born with this physiology become **progressively cyanotic** which can lead to cardiac arrest
- Atrial shunting (Foramen ovale or ASD) is crucial for oxygenated blood to mix with deoxygenated blood to avoid profound cyanosis after birth
- Fetal physiology in DTGA is very stable due to placental circulation and foramen ovale

Prenatal Detection Rate

Van Velzen et al. Ultrasound Obstet Gynecol 2015; **45**: 320–325.
Everwijn et al. Prenatal Diagnosis 2018;38:951-957.

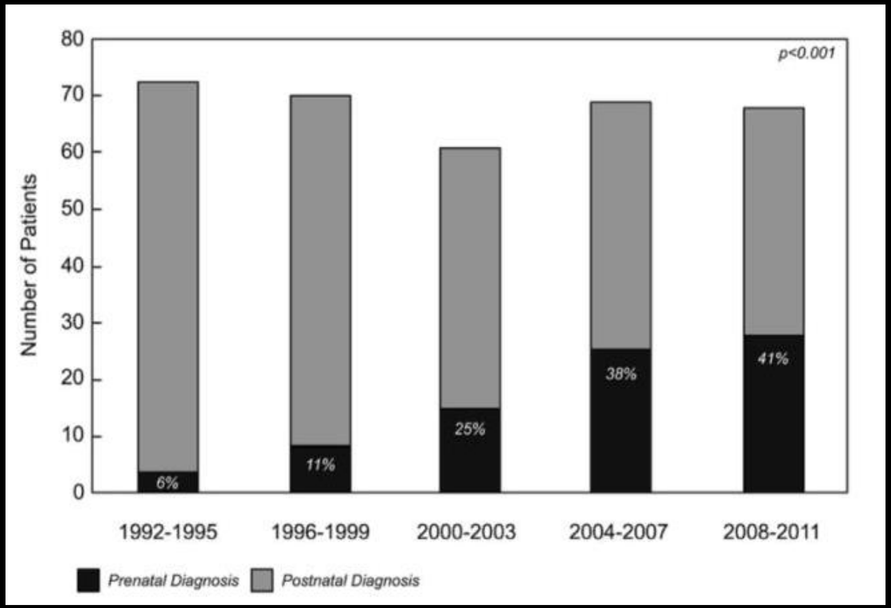
Center	Early Period					Overall Rate
Alberta	14% (2003-2010)	50% (2010-2013)	77% (2013-2015)			**37%**
Boston	6% (1992-1995)	11% (1996-1999)	25% (2000-2003)	38% (2004-2007)	41% (2008-2011)	**24%**
Netherlands	16% (2002-2006)	*cardiac screening program started 2007		41% (2007-2011)	82% (2012-2016)	****26%**

DTGA prenatal detection rate – Alberta,CA



Ravi et al. Ultrasound Obstet Gynecol 2018; **51**: 659–664

DTGA prenatal detection rate – Boston



Escobar Diaz et al. Ultrasound Obstet Gynecol. 2015 June ; 45(6): 678–682.

*2010 Canadian Association of Radiologists **add outflow tract view** to OB exam

*2013 AIUM requires outflow tract and suggests **3VV/3VTV** assessment to OB exam

*2012 Netherland national mandatory **3VV assessment** for standard anatomy scan

**2015 study data only

Prenatal D-TGA rates are still only 50-65% in the US currently

Impact of Prenatal Detection of D-TGA

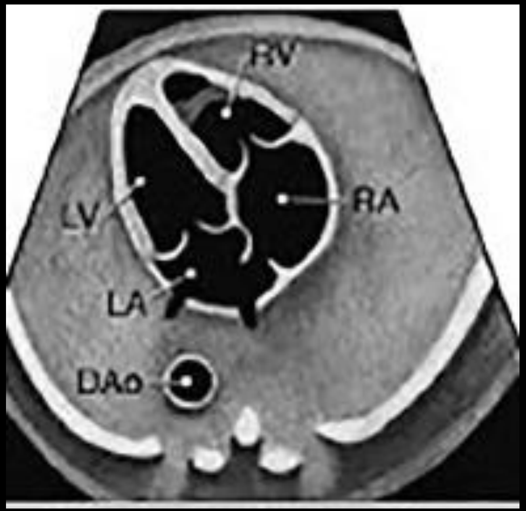
- Planned delivery at a tertiary care center for earlier access to balloon atrial septostmy (BAS) (Boston)
- Fewer needed mechanical ventilation (Boston)
- Higher O2 saturation at presentation (Netherlands)
- Normal renal function at presentation (Netherlands)
- Lower overall 1st-year and pre-op mortality (Netherlands)

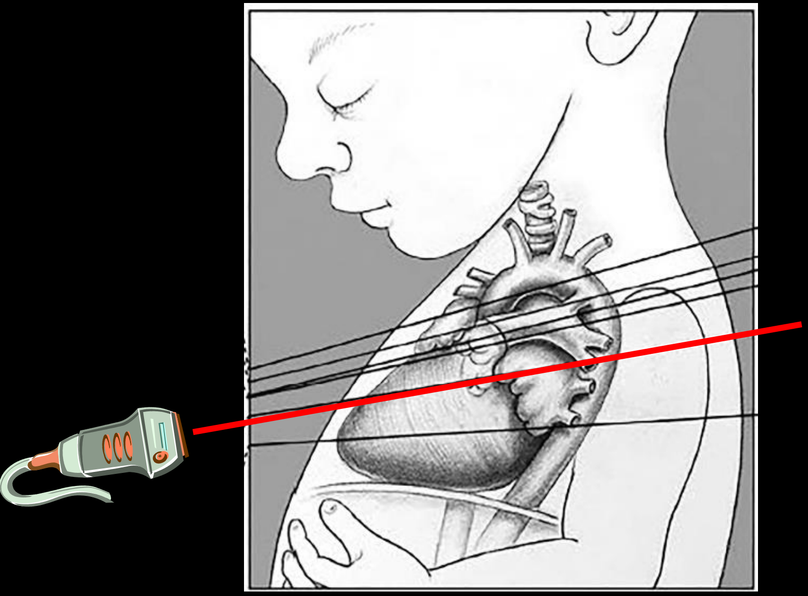
Escobar Diaz et al. Ultrasound Obstet Gynecol. 2015 June ; 45(6): 678–682.
Van Velzen et al. Ultrasound Obstet Gynecol 2015; **45**: 320–325.

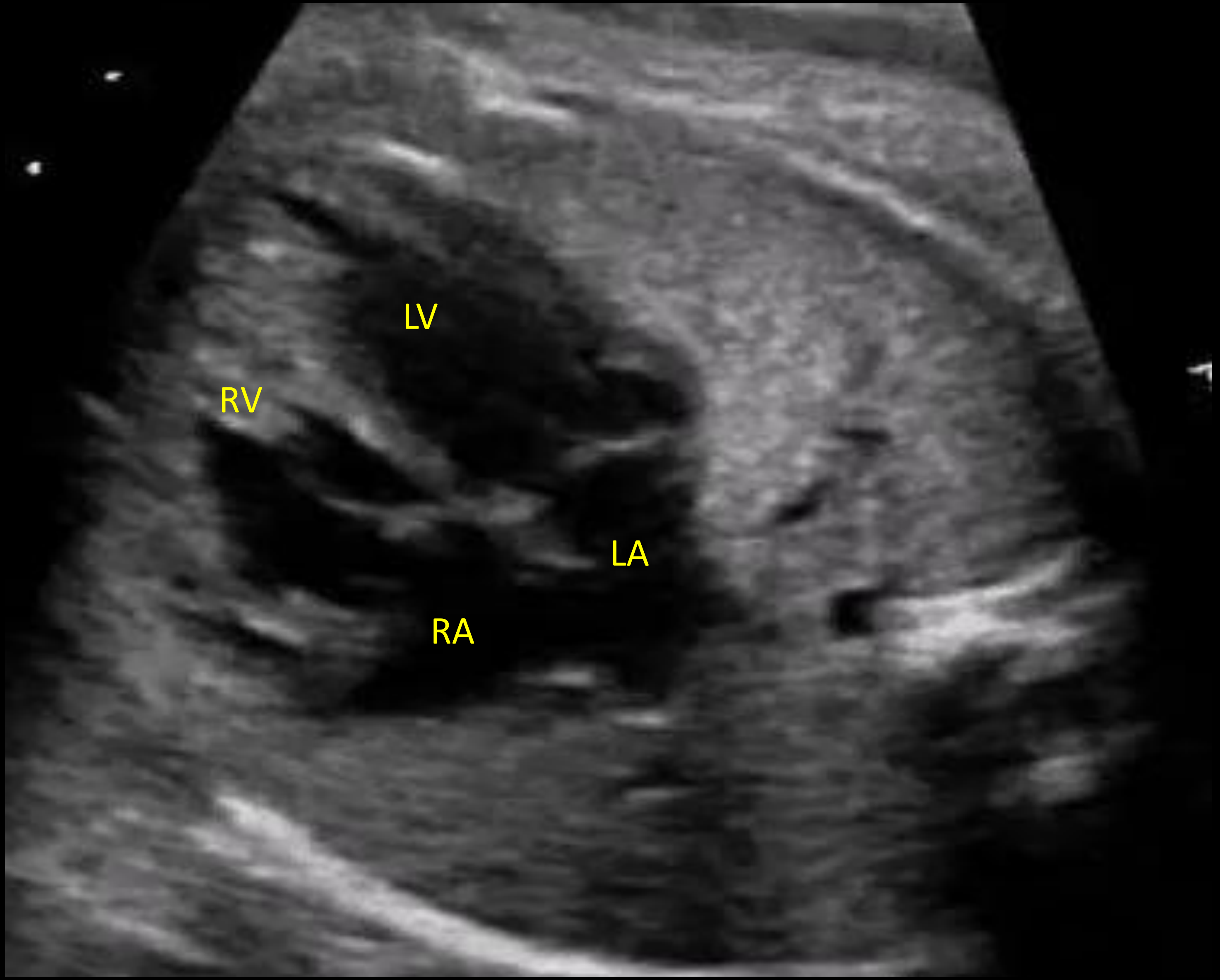
Fetal Echo Detection and Evaluation and Treatment Considerations for D-TGA

Typically, normal 4 chamber view

Apical 4 chamber (A4C)



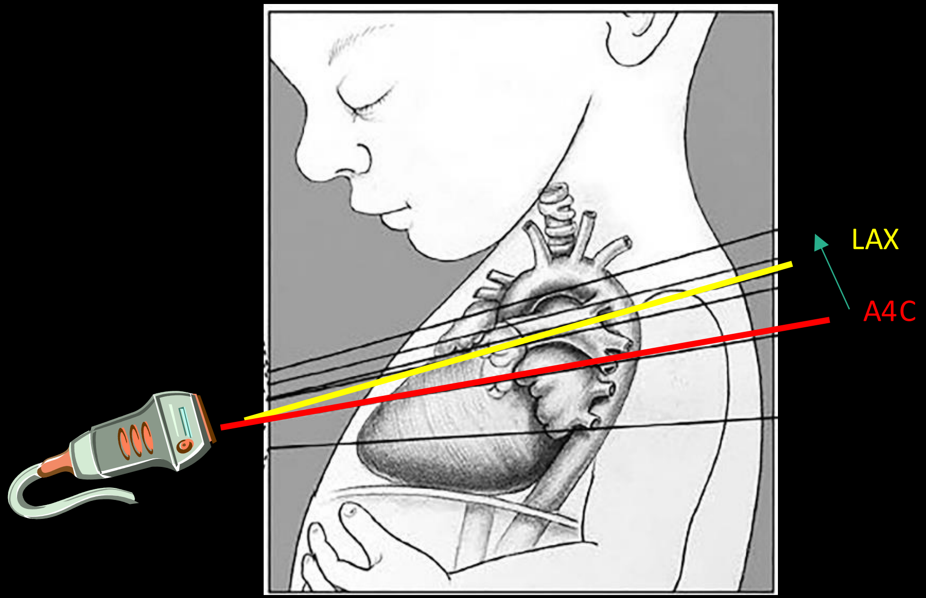


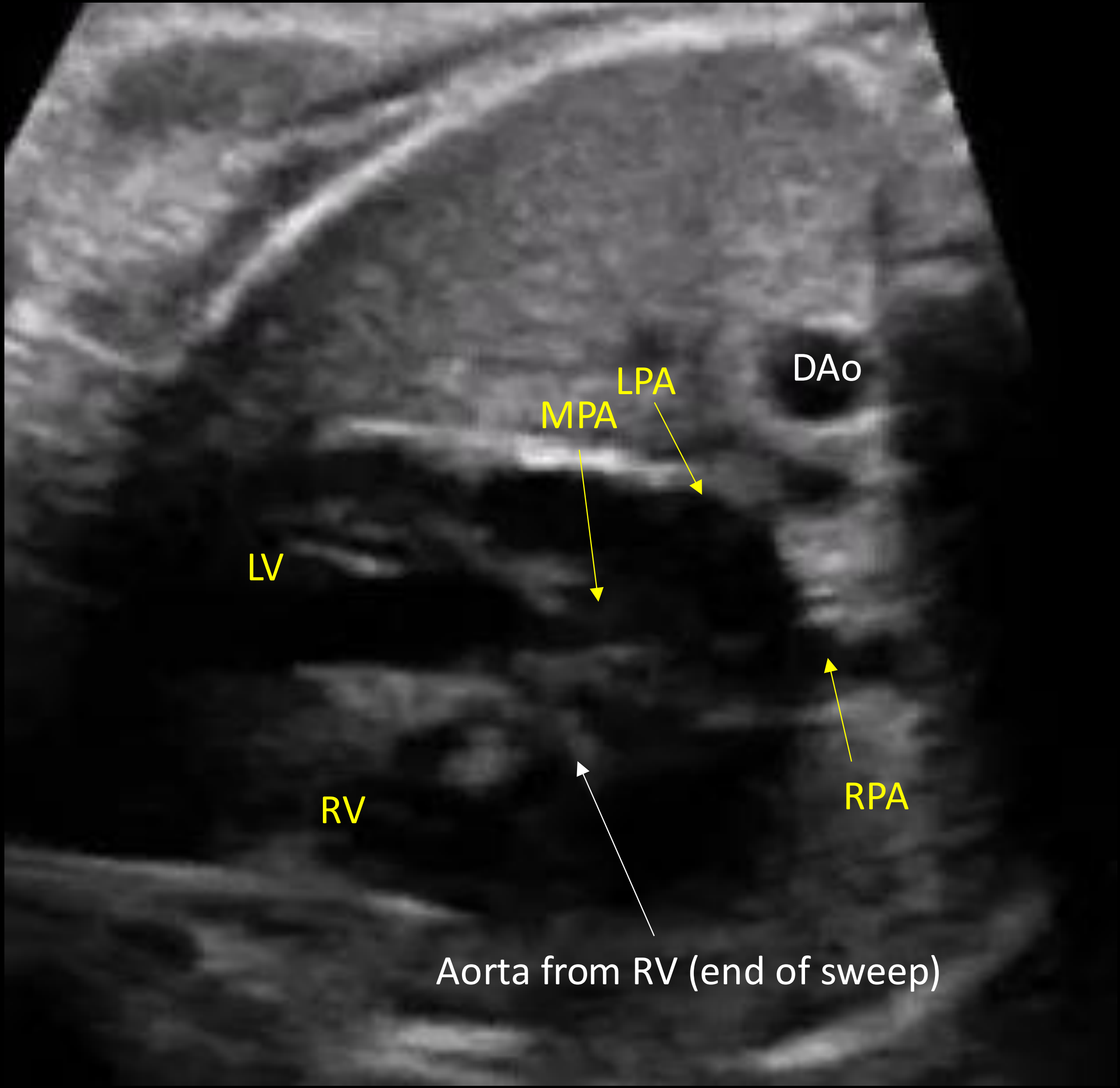


- Sweep up to from A4C to long axis view (LAX) to show LV to PA
- Anterior Ao arises from RV

Apical Long Axis View – LVOT

normal



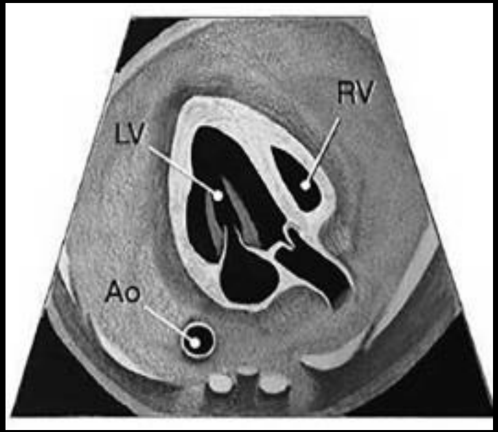


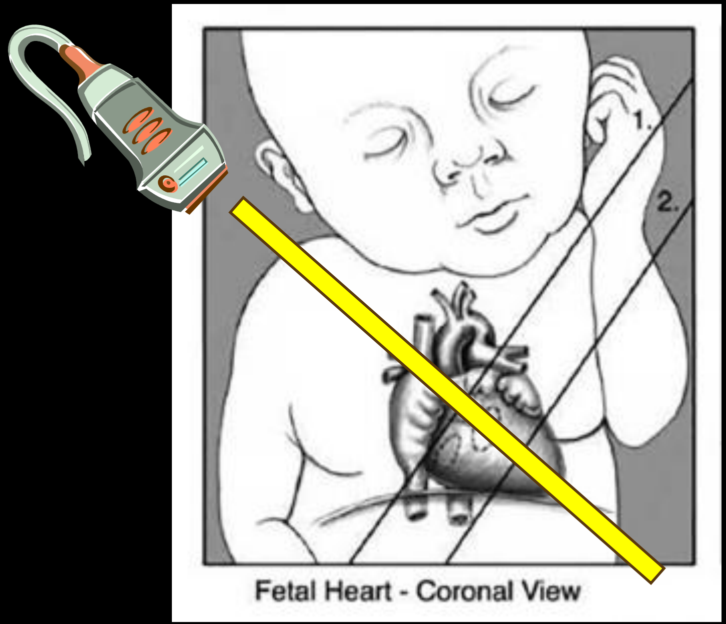
Parallel great vessels course

- Usually do not see both vessel simultaneously in this view
- Great vessels would normally cross, but do not in DTGA

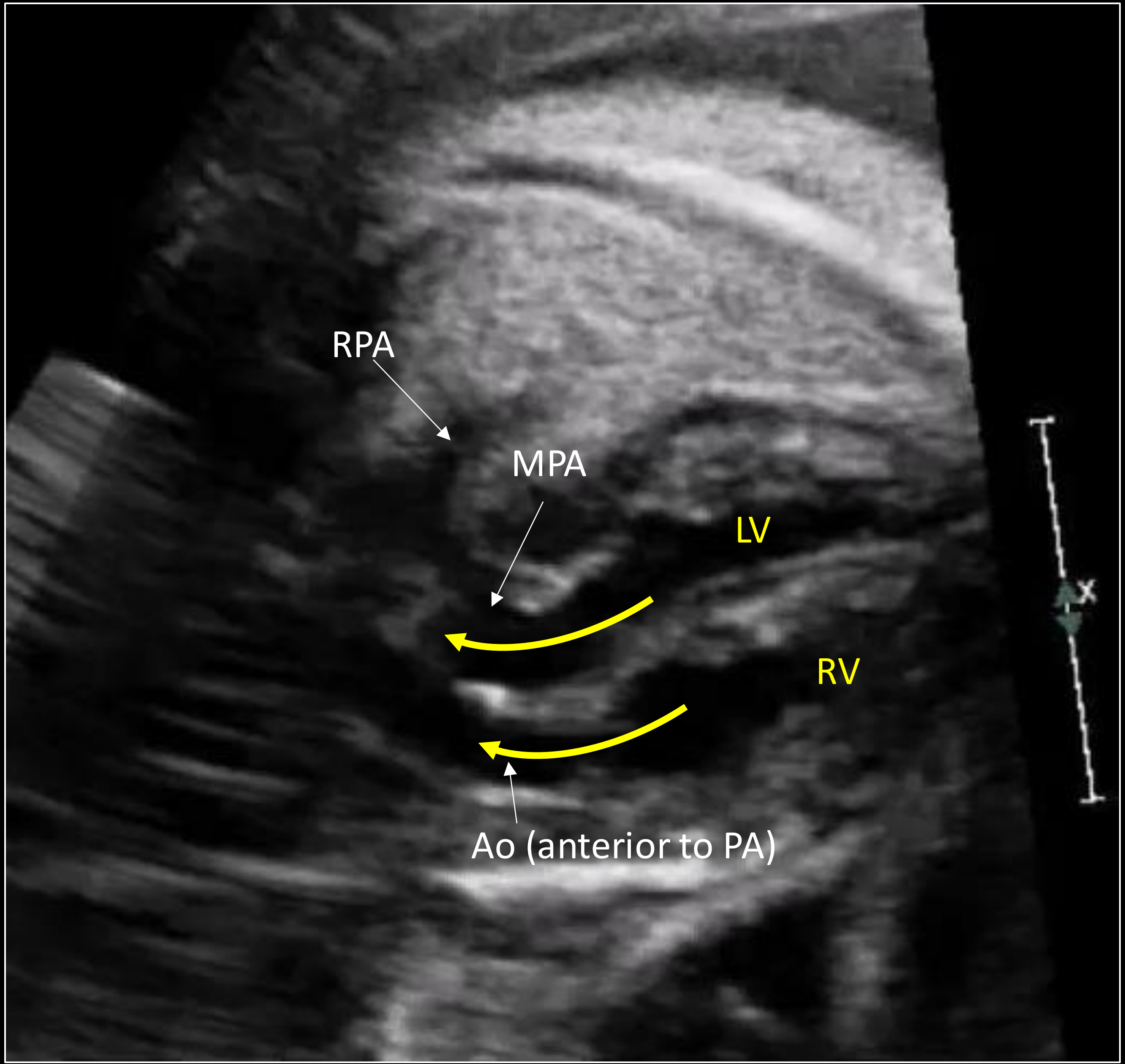
Parasternal LAX

normal



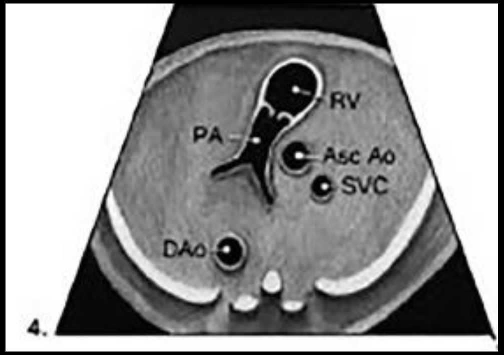


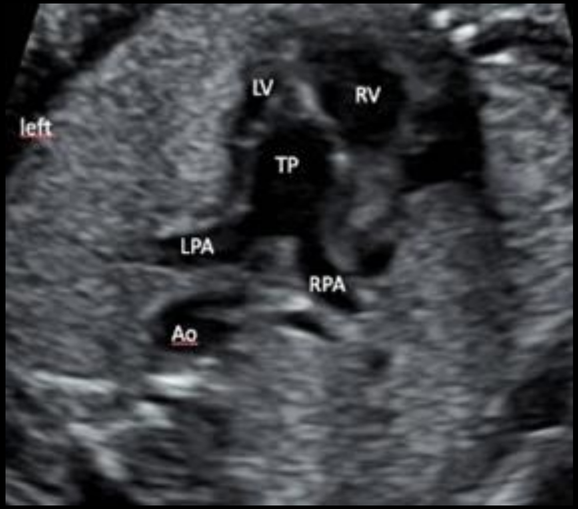
Oblique view LAX

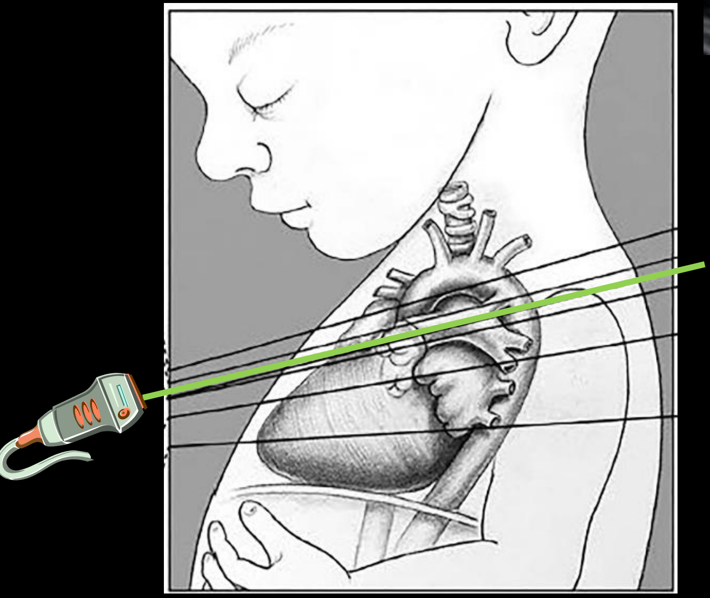


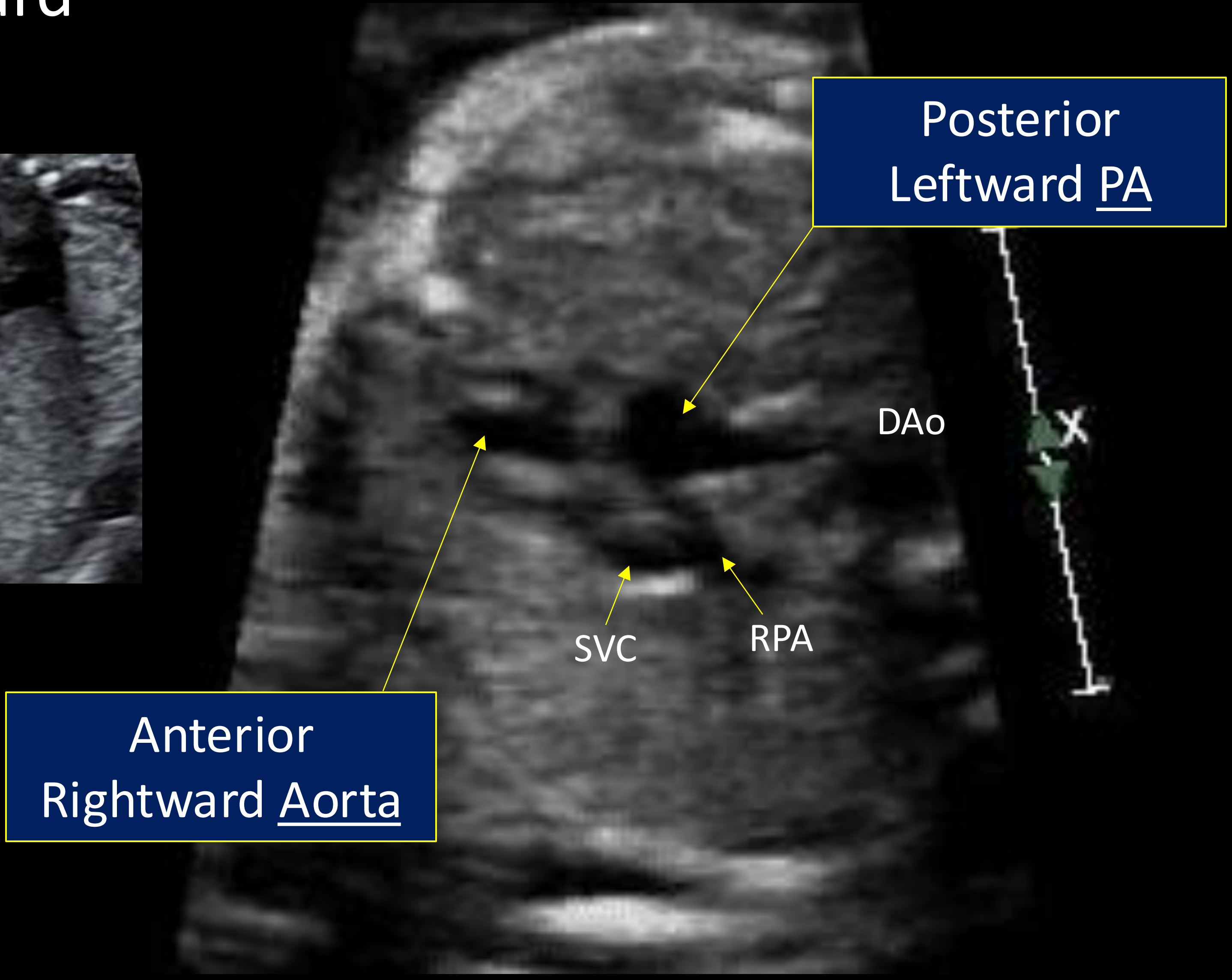
Aorta is anterior and rightward of the pulmonary artery

normal





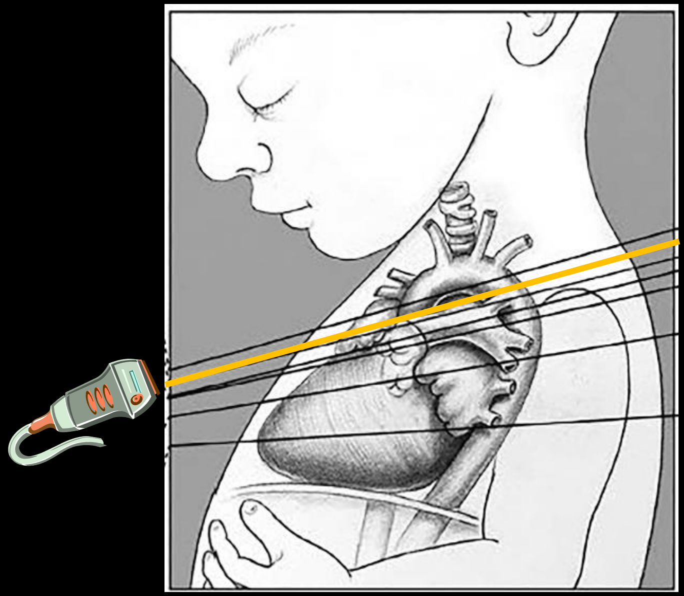


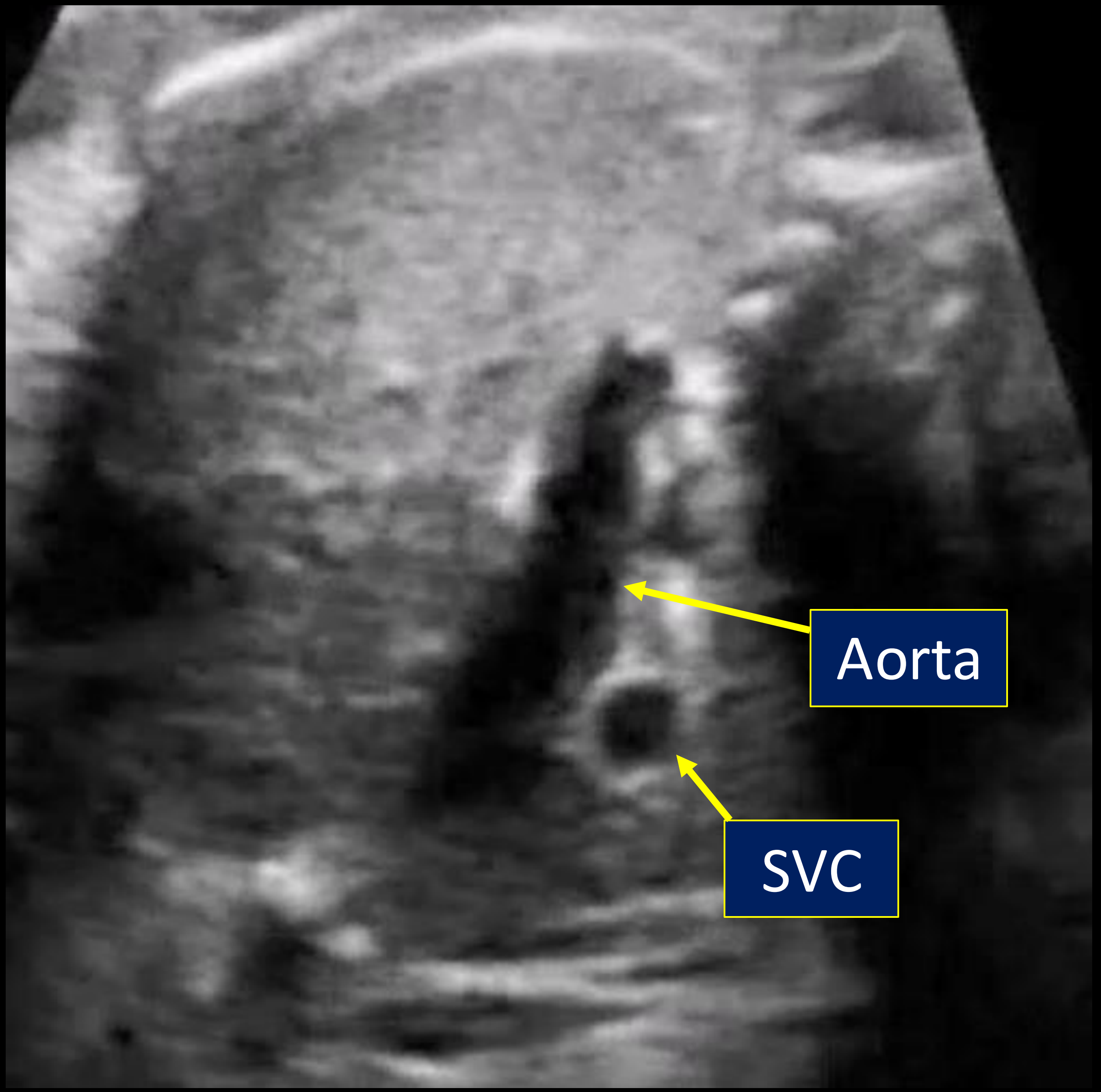


- 3VVT shows a single artery (transverse aorta)
- Ductal artery lies under the transverse aorta
- SVC lies to the right of the aorta

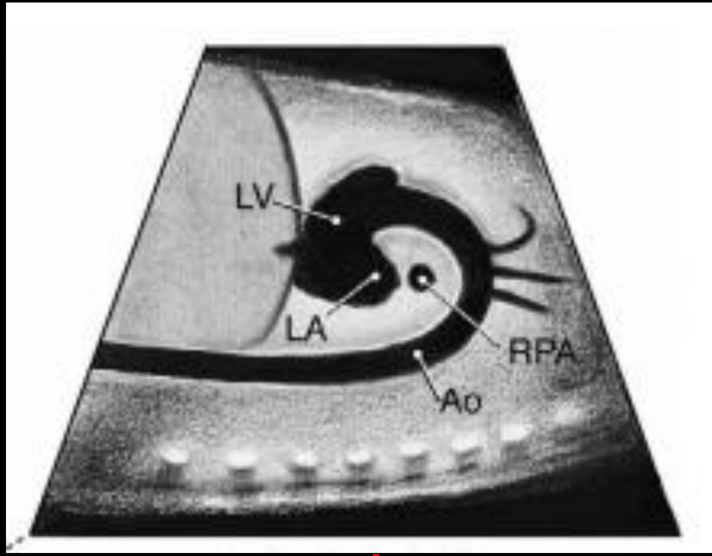
3VT View

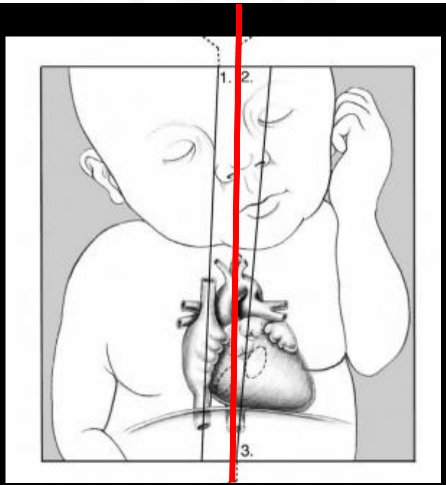
normal

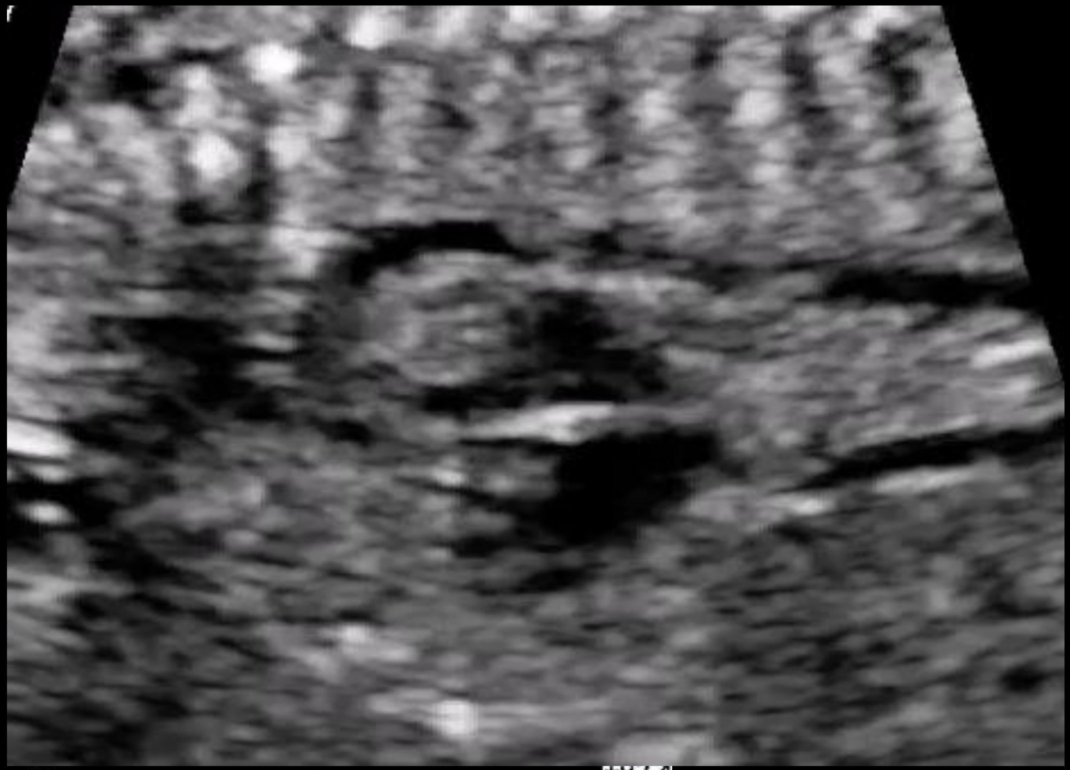


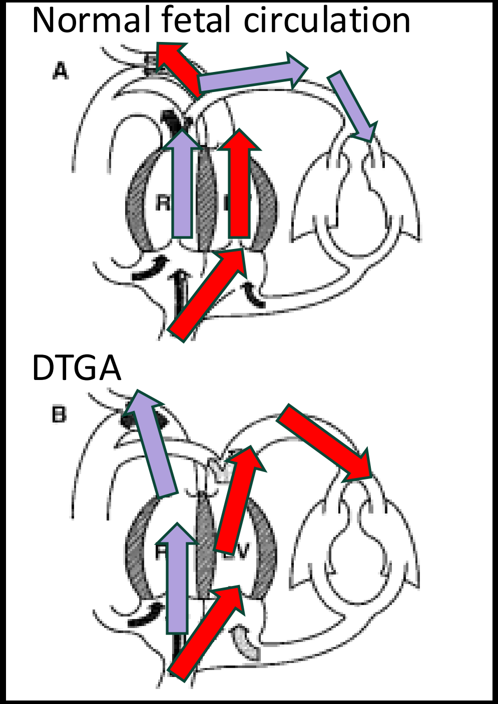


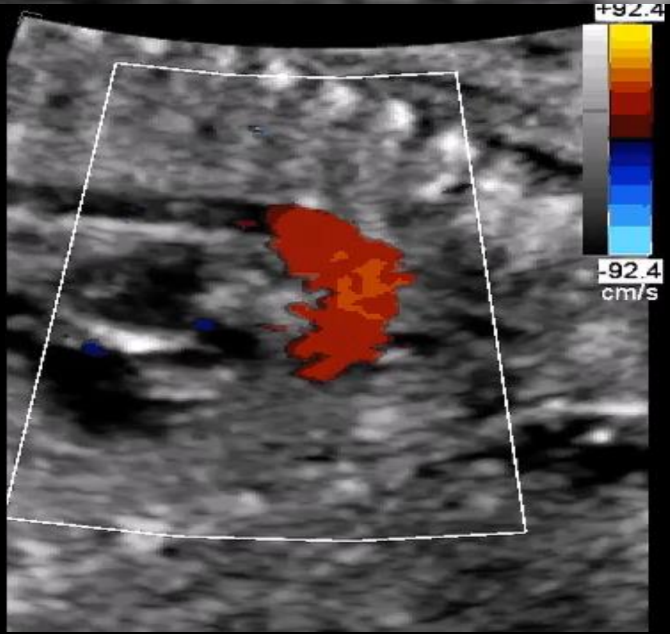
Elongated aortic arch arising from RV in sagittal view











- Bidirectional shunting due to lower pulmonary artery resistance due to greater oxygen delivery to the pulmonary arteries
  - Higher 02, decreased PVR from vasodilation
- Can also be associated with restrictive atrial septum

Postnatal physiology and intervention

It is critical for atrial shunting to occur to mix oxygenated and deoxygenated blood in two parallel circuits ensuring systemic oxygenation

Prevalence for balloon atrial septostomy (BAS) is 27.5%-65% before surgical repair (arterial switch)

Hamzah et al Pediatr Crit Care Med. 2020 Apr;21(4):324-331.

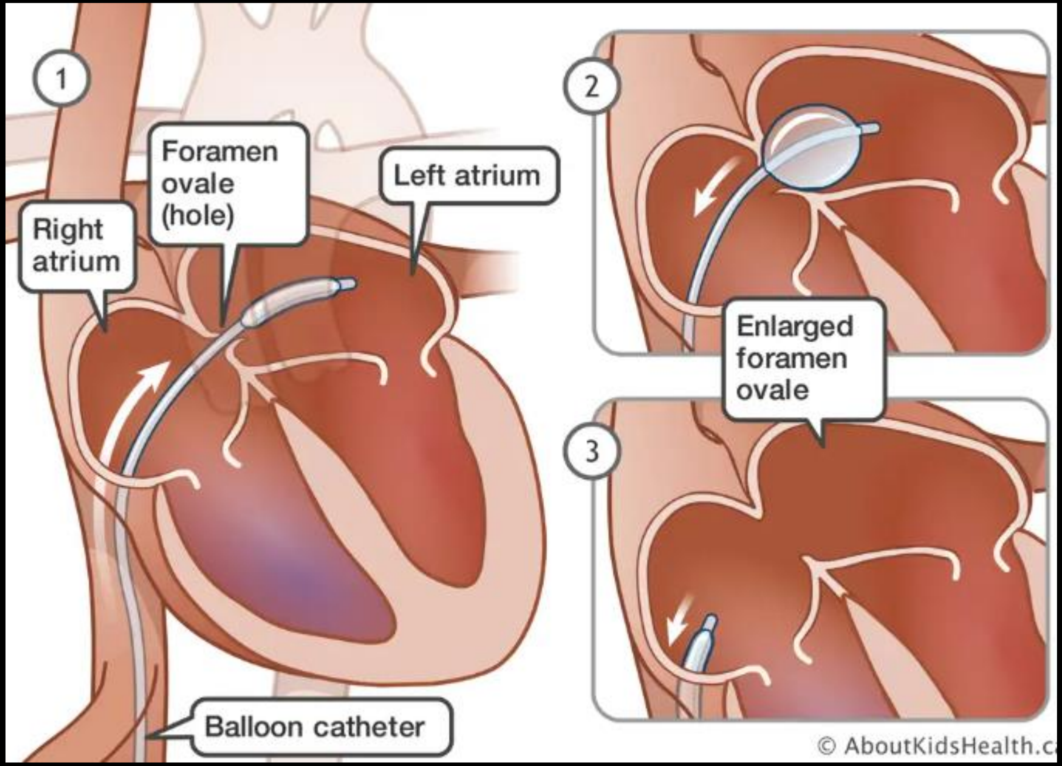


Image source: aboutkids.com

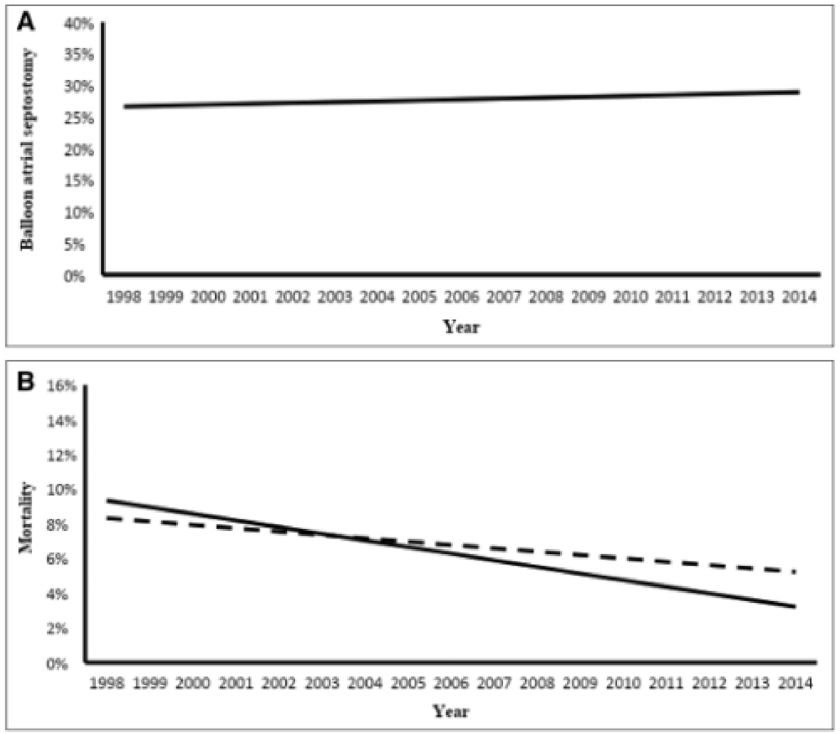


Figure 1. Balloon atrial septostomy (BAS) trend over the years (**A**) and mortality trends in BAS and no BAS groups (**B**). *Straight line* represents mortality trend in transposition of the great arteries (TGA) with BAS, whereas *dashed line* represents mortality trend in TGA without BAS.

Zaleski et al. Pediatr Cardiol. 2021 Mar;42(3):597-605

BAS have increased throughout the years due to increased prenatal detection

Mortality has also improved throughout the years with BAS

This is likely due to more prenatal dx and planning

The diversity of practices for managing d-TGA with BAS due differences in hospital systems. Some centers have built-in capabilities with CICU and cath lab access, others perform bedside procedures, and some require transport to another facility for intervention.

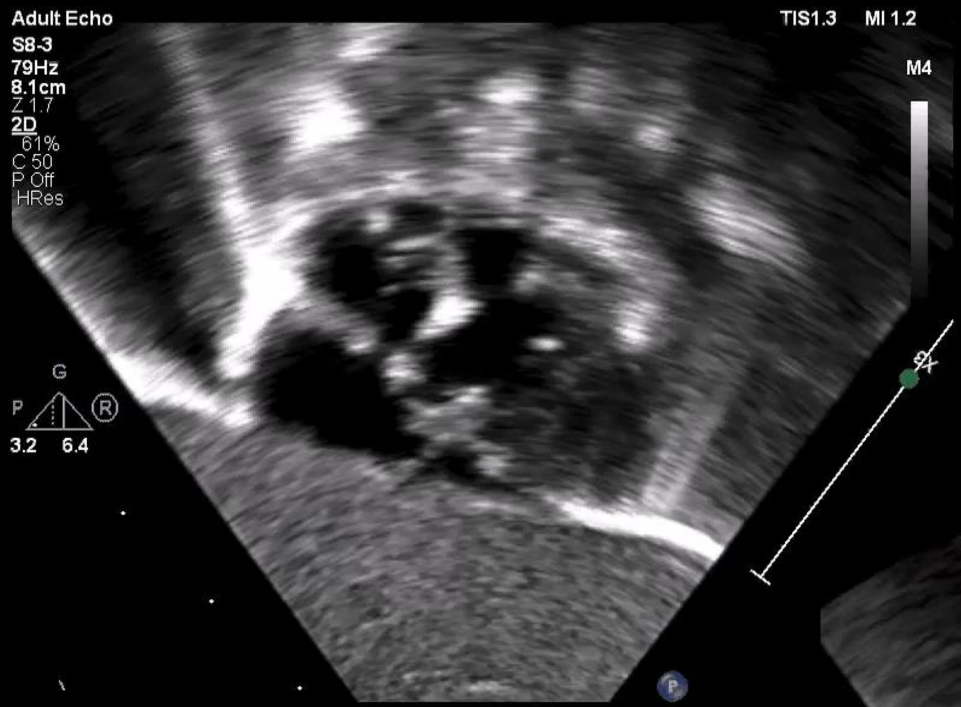
Can We Predict a Restrictive Atrial Septum in D-TGA?

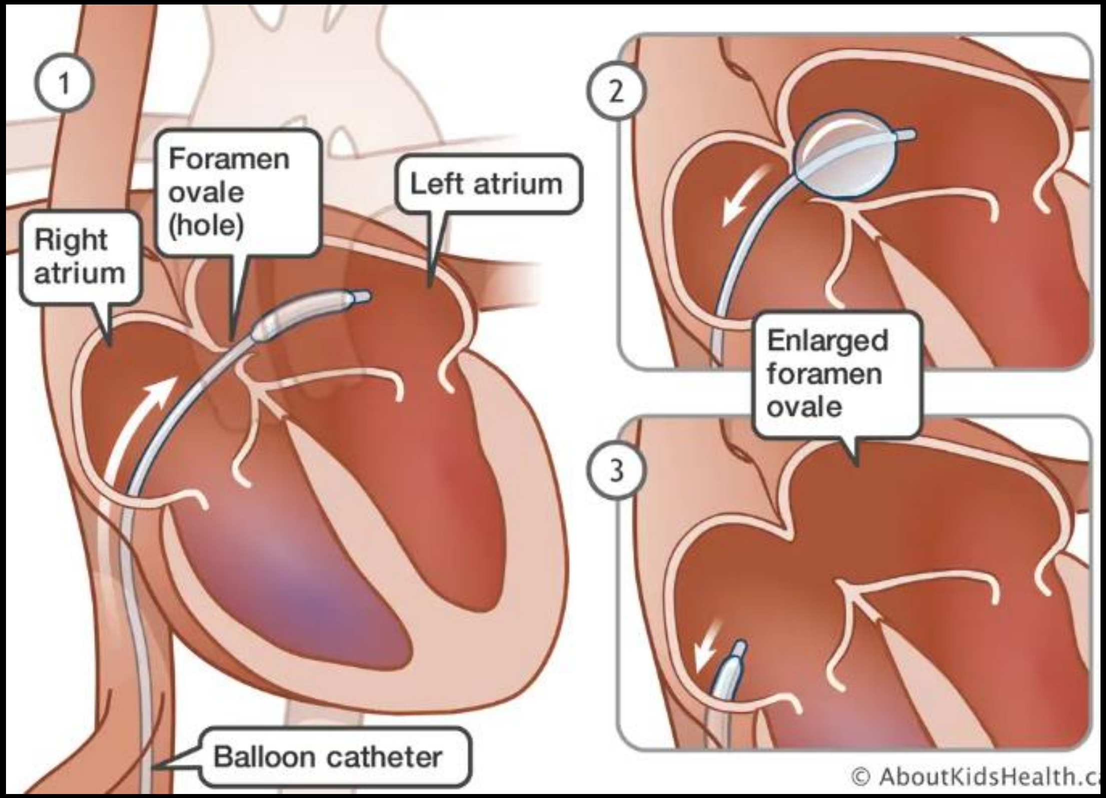
Author	Hypermobile Septum	Fixed Septum	Aneurysmal Septum	Small FO Indexed to TSL	Small DA	Abnormal DA Flow
Maeno et al	Yes	Yes	Yes	NA	Yes	Yes
Tuo et al	Yes	Yes	Yes	NA	NA	No
Jouannic et al	Yes	Yes	Yes	NA	NA	Yes
Punn and Silverman	Yes	No	NA	NA	NA	Yes
Ayzen and Rychik	NA	NA	No	NA	NA	NA
Vigneswaran et al	No	Yes	No	Yes	NA	NA

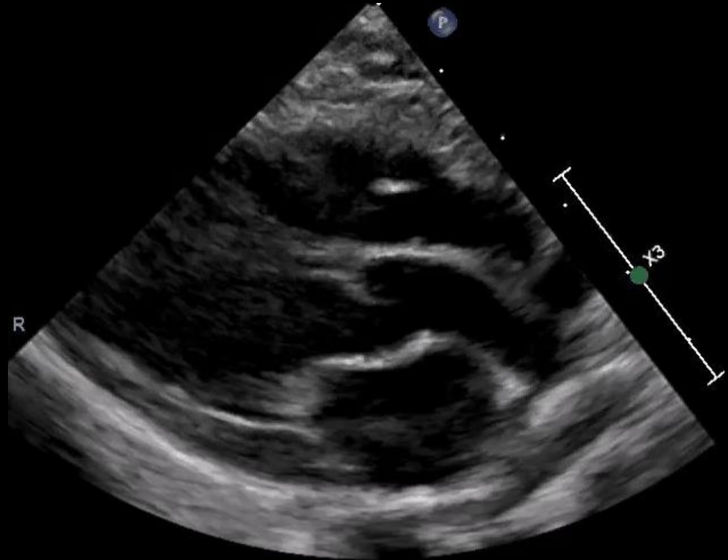
- *Take home point – NEVER TRUST A TRANSPOSITION!*

Fetal Heart Society

Fetal anatomy and physiology and associations with fetal and perinatal outcomes in D-transposition of the great arteries with intact ventricular septum, a multicenter, multi-arm, prospective study







Allows oxygenated LA blood to flow into deoxygenated RA blood -> delivered to systemic circulation through the RV

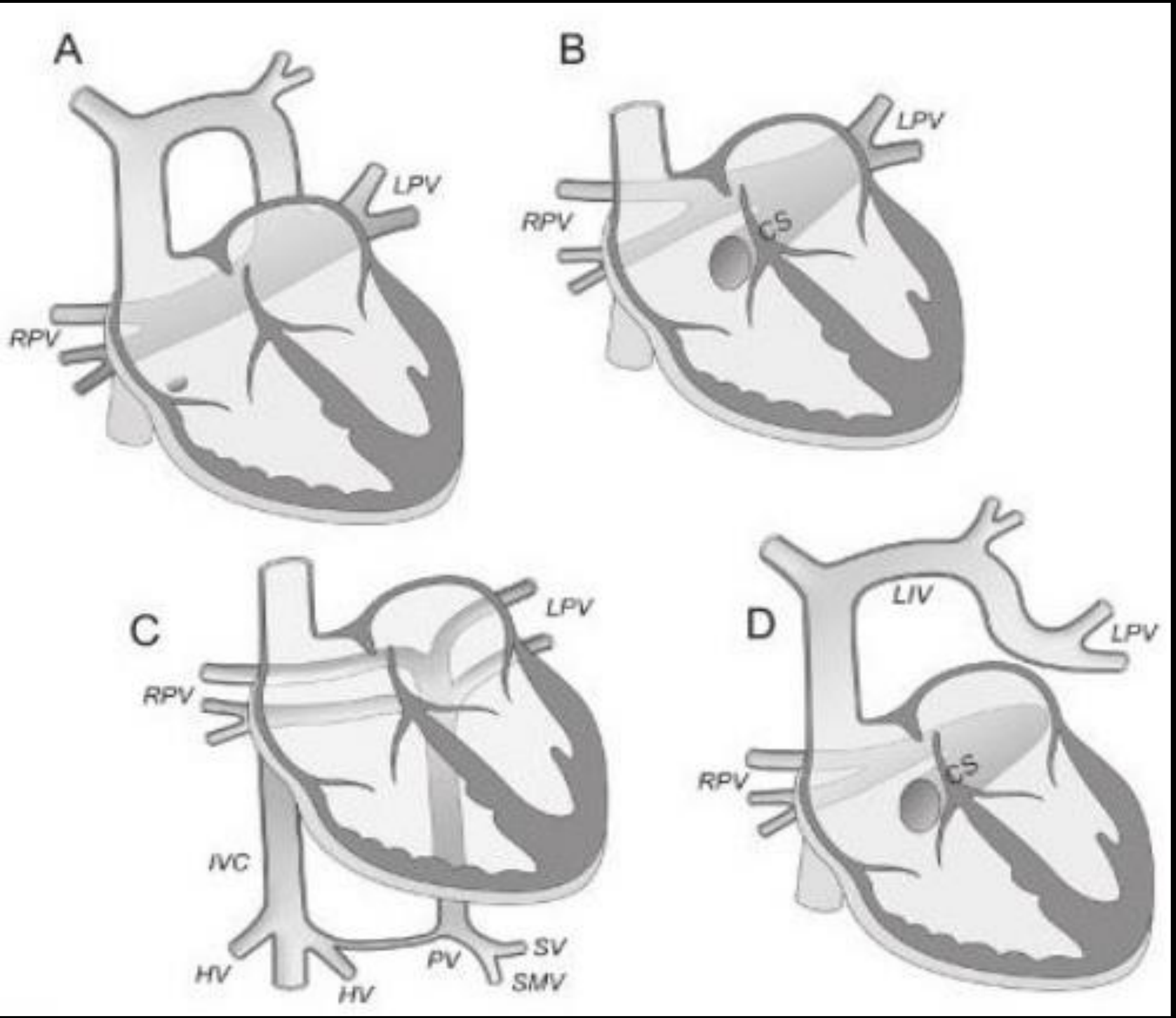
Summary for Tips for Imaging D-TGA

- Typically a normal 4 chamber view with exception of associated VSD
- Sweep up to Apical long axis view shows PA arising from LV
- Further superior sweep shows AO arising from RV
- Parallel vessels best obtained from oblique view of the heart from fetal right shoulder to left hip
- 3VT shows single large vessel (transverse AO) and SVC to the right
- Sagittal/longitudinal view shows AO arising anteriorly with elongated arch

Total Anomalous Pulmonary Venous Return (TAPVR)

Total Anomalous Pulmonary Venous Return (TAPVR)

- Accounts for 0.5-1.5% of CHD
- 5th most common cyanotic CHD
- 2/3 of cases are isolated
- 1/3 with additional CHD
  - Heterotaxy, HLHS, CAVC, Tri Atresia, Pulm Atresia, TGA, TOF, DORV, Vascular rings, CAT, and COA



Prenatal Detection of TAPVR

- CHD with **lowest prenatal detection** rate in United States demonstrated in several studies

Quartermain et al. Pediatrics 2015

Prenatal Detection Rates by Fundamental Diagnosis

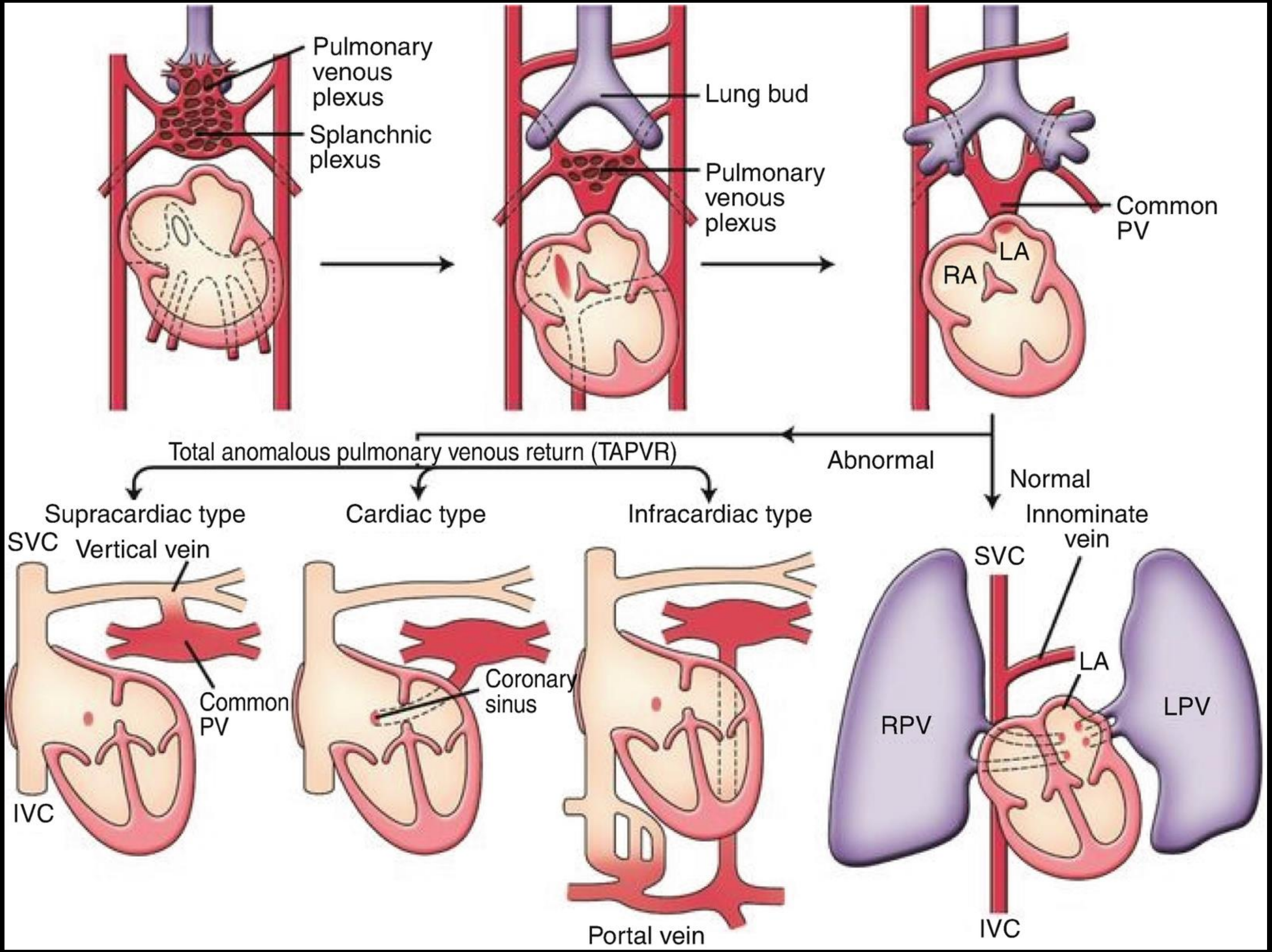
Fundamental Diagnosis	Total N	N with Prenatal Detection (%)
TAPVC	1359	123 (9.1%)
Ventricular septal defect	4706	577 (12.3%)
Isolated Arch Obstruction	3901	841 (21.6%)
Aortic Stenosis	318	81 (25.5%)
Arch Obstruction with VSD	1174	311 (26.5%)
Tetralogy of Fallot	3359	895 (26.6%)
TGA-IVS	1898	530 (27.9%)
TGA-VSD	1257	463 (36.8%)
Atrioventricular septal defect	3172	1295 (40.8%)
Pulmonary Stenosis and Pulmonary atresia-IVS	1056	433 (41.1%)
Truncus Arteriosus	761	314 (41.2%)
TOF with APV Syndrome	200	82 (41.3%)
Double-Outlet Right Ventricle	1227	539 (43.9%)
Congenitally Corrected TGA	127	57 (44.9%)
Pulmonary Atresia-VSD	1082	500 (46.2%)
Tricuspid Valve Disease *	262	147 (56.1%)
Single Ventricle, other	2362	1482 (62.7%)
Hypoplastic Left Heart Syndrome	3153	2125 (67.4%)

Development of TAPVR

Normal pulmonary vein development occurs when pulmonary venous complex grows from the primitive lungs and forms common pulmonary vein (CPV).

Common pulmonary vein then joins the back of the left atrium and ultimately is incorporated into back wall forming 4 individual pulmonary veins. In addition, connection to the systemic veins is also lost.

Failure for the common pulmonary vein to connect to the LA results in varying forms of anomalous venous return



TAPVR Types

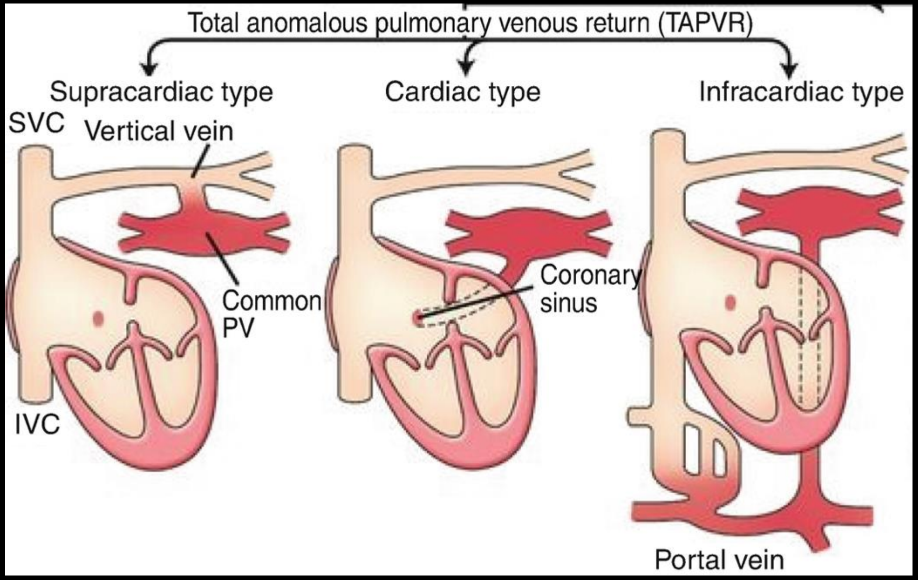
Darling Classification

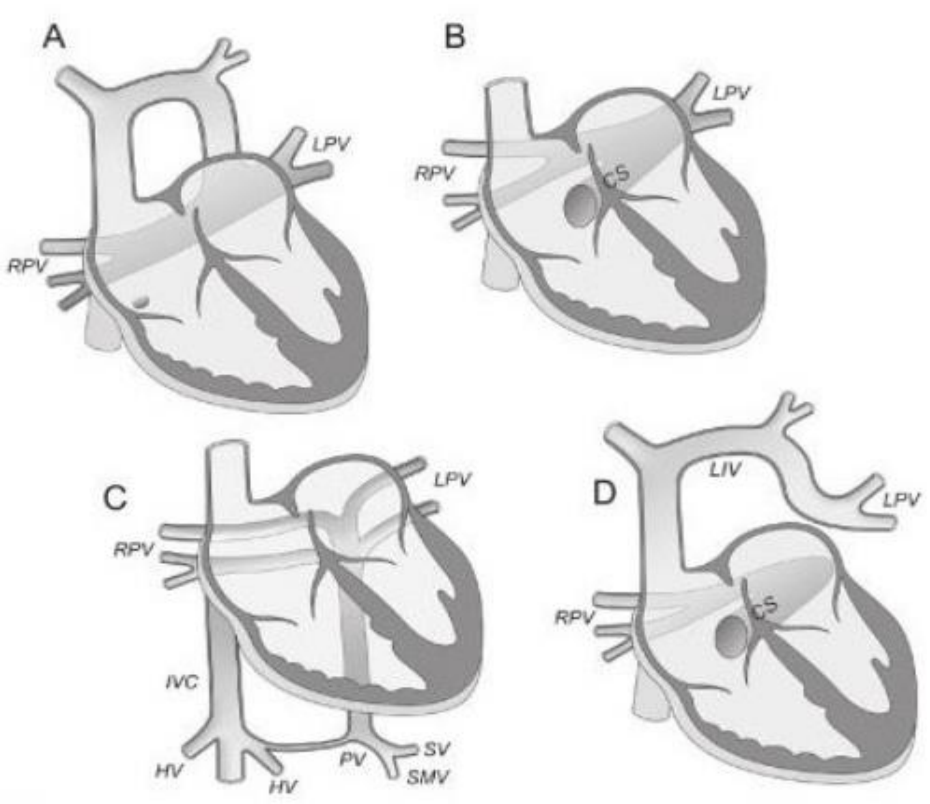
A. Supracardiac type I: PVC drains via VV to innominate vein

B. Intracardiac type II: PVC drains to CS or RA directly

C. Infracardiac type III: PVC drains via VV to IVC, hepatics, DV

D. Mixed type IV: drainage to 2 or more sites but none to LA

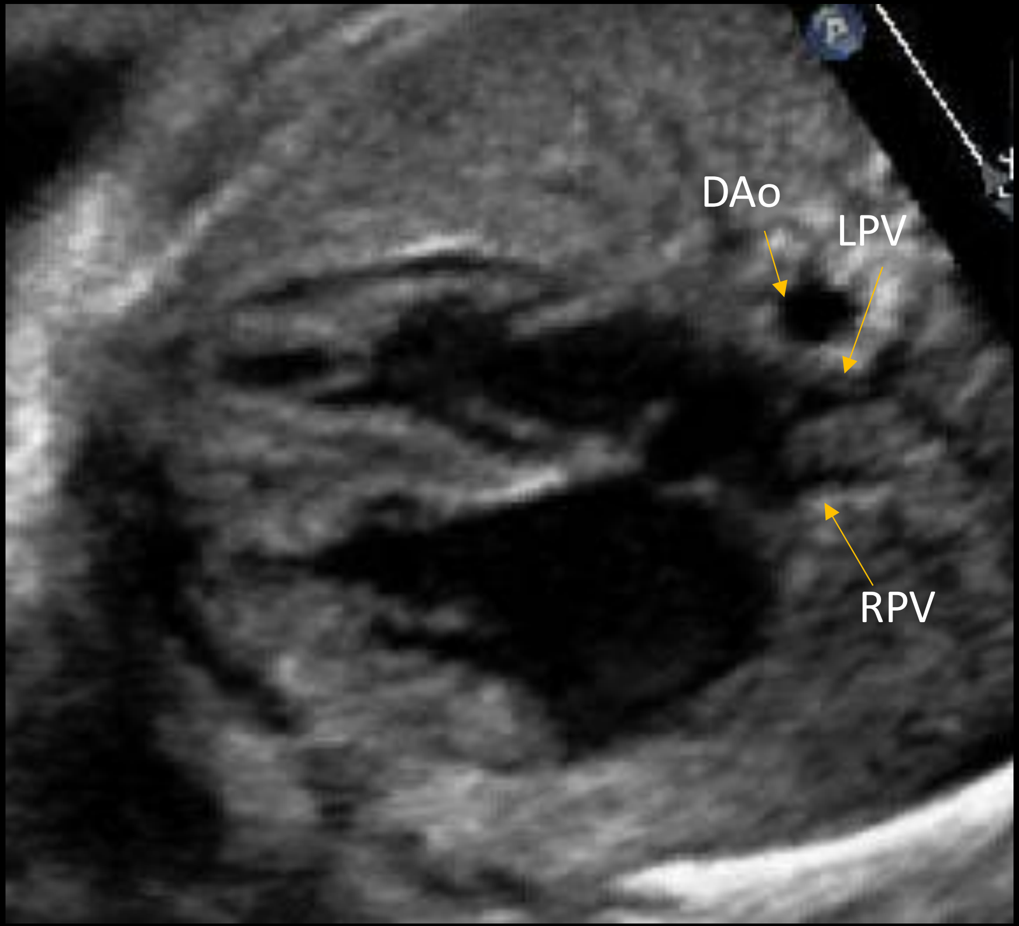




Fetal Echo Detection and Evaluation

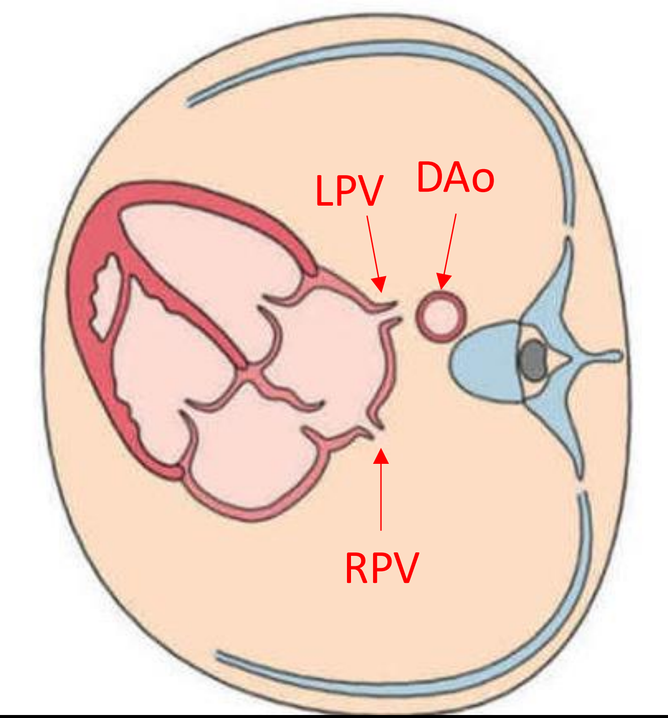
Treatment Considerations for Total Anomalous Pulmonary Venous Return (TAPVR)

Normal Left Atrium Landmarks

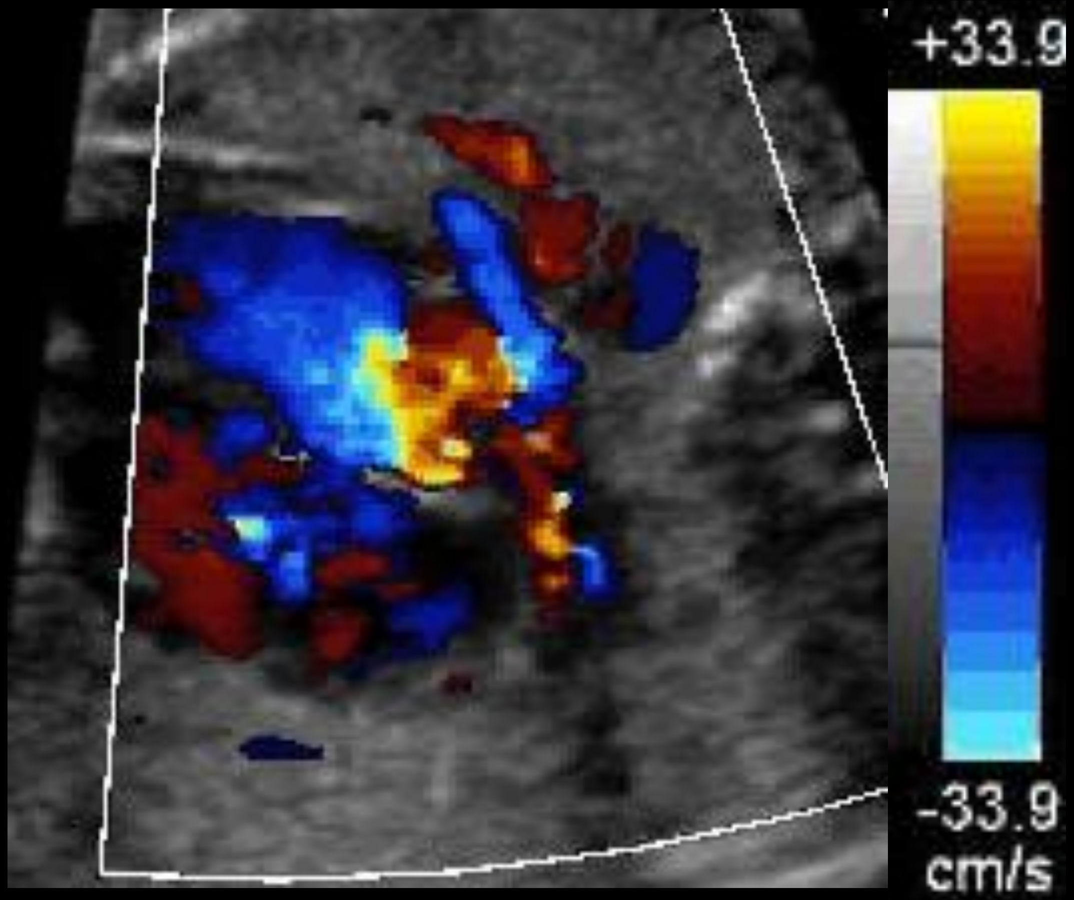


Irregular posterior left atrial wall

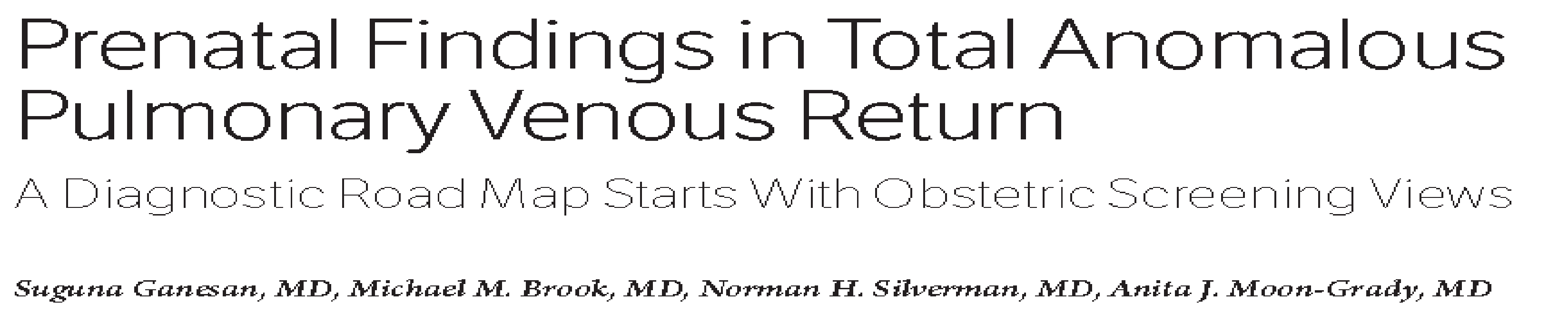
Left atrial wall closely approximated to the descending aorta



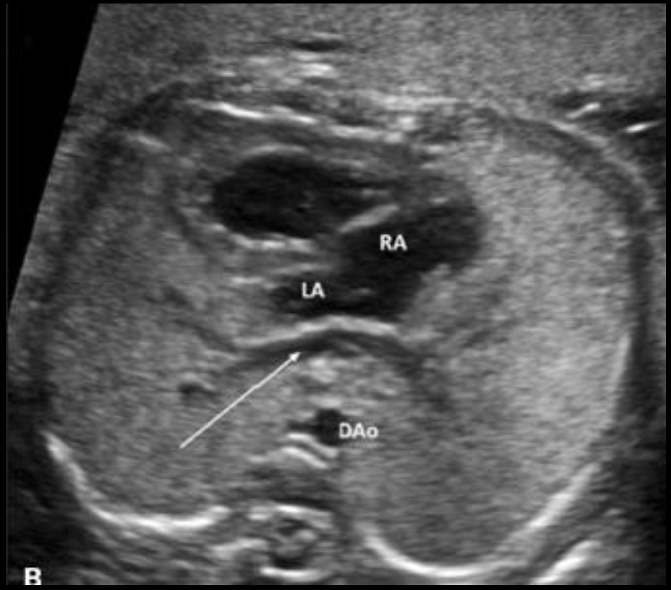
Normal Pulmonary Venous Color Flow



- Drop Nyquist limit <35 cm/sec
- Demonstrate at least one vein draining from each lung
- Finding and differentiating upper and lower veins can be challenging due to fetal resolution



Clues to the Diagnosis



- Smooth walled LA
- PV confluence behind LA – “twig sign”
- Abnormal PV Doppler waveform (if obstructed)

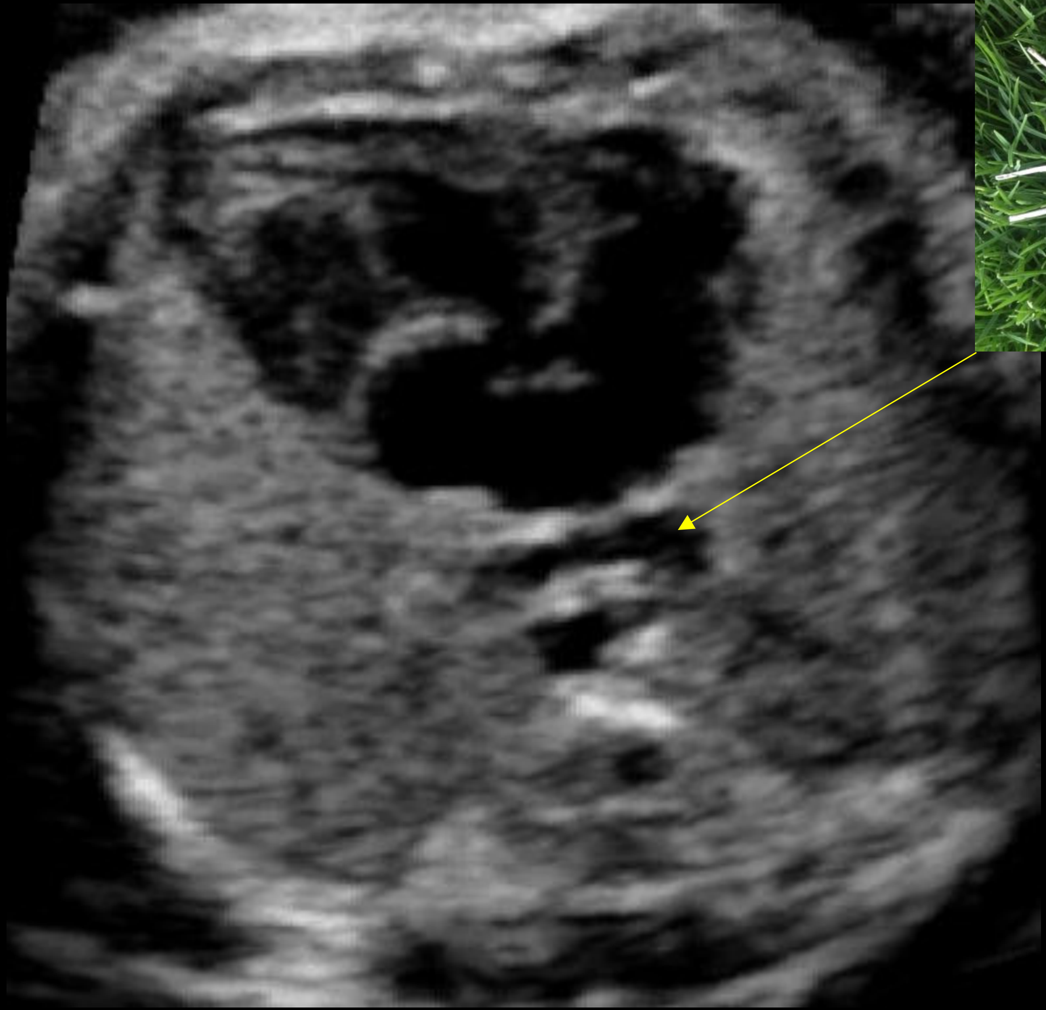
J Ultrasound Med 2014; 33:1193–1207

Prenatal Findings in Total Anomalous Pulmonary Venous Return

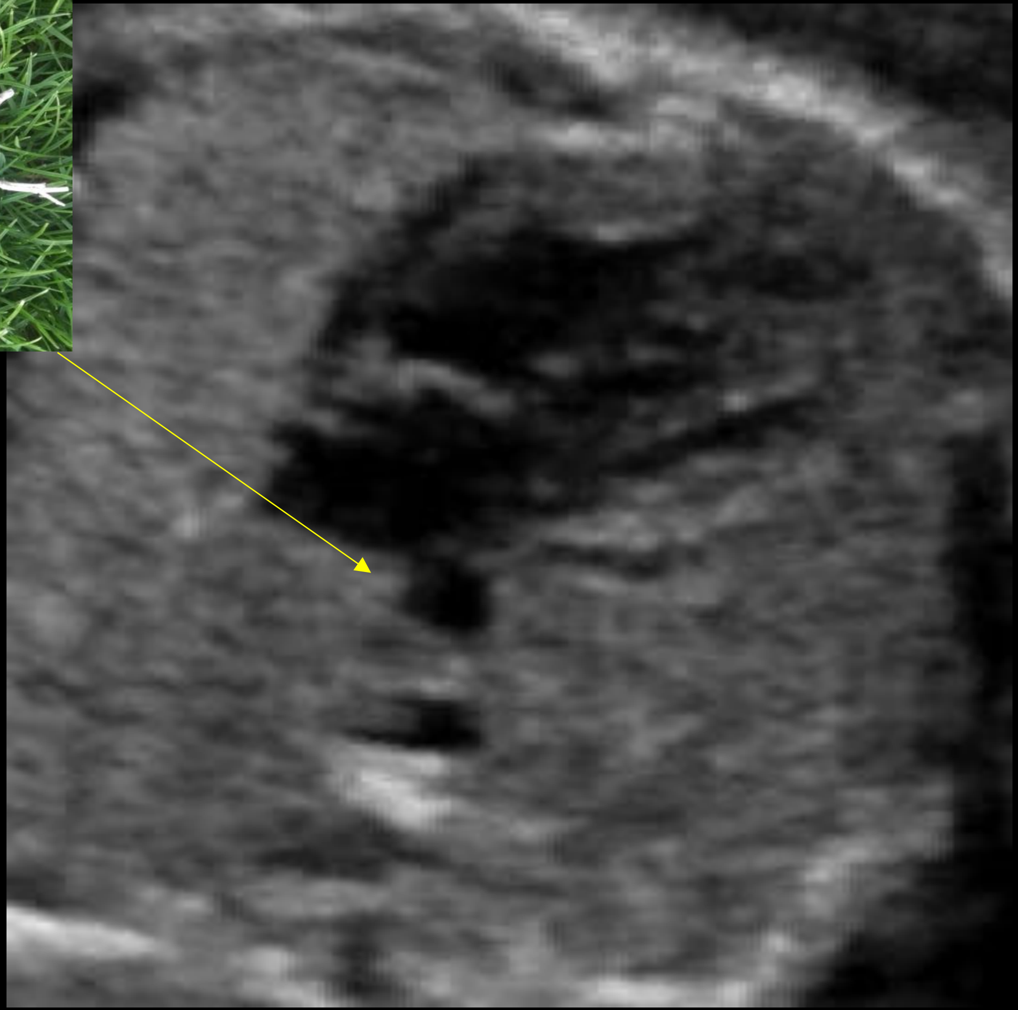
A Diagnostic Road Map Starts With Obstetric Screening Views

Suguna Ganesan, MD, Michael M. Brook, MD, Norman H. Silverman, MD, Anita J. Moon-Grady, MD

Clues to the Diagnosis



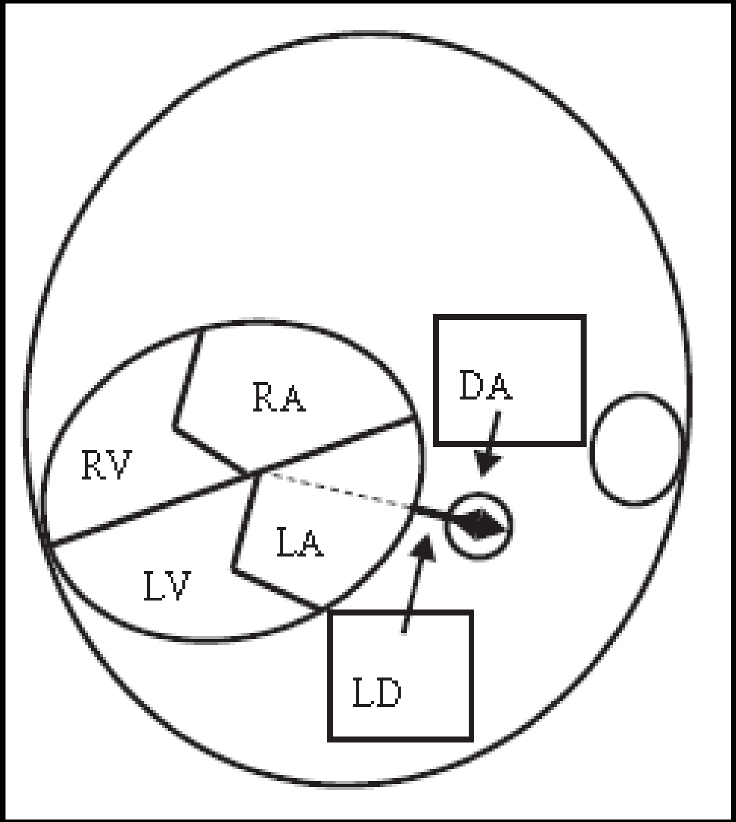
“twig” sign

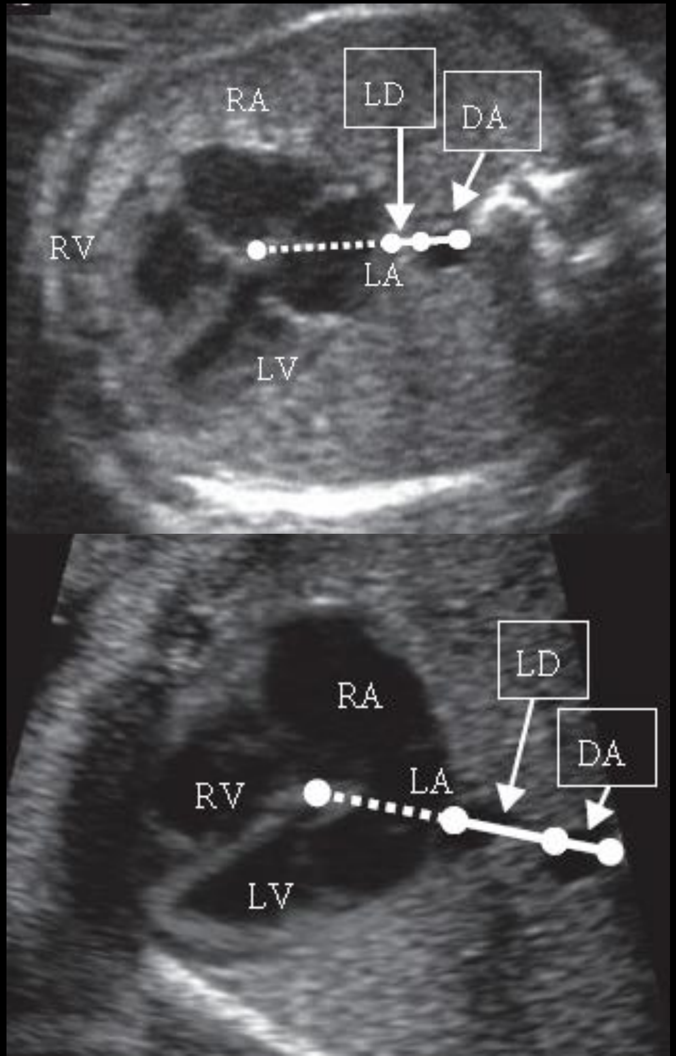


‘Post-LA space index’ as a potential novel marker for the prenatal diagnosis of isolated total anomalous pulmonary venous connection

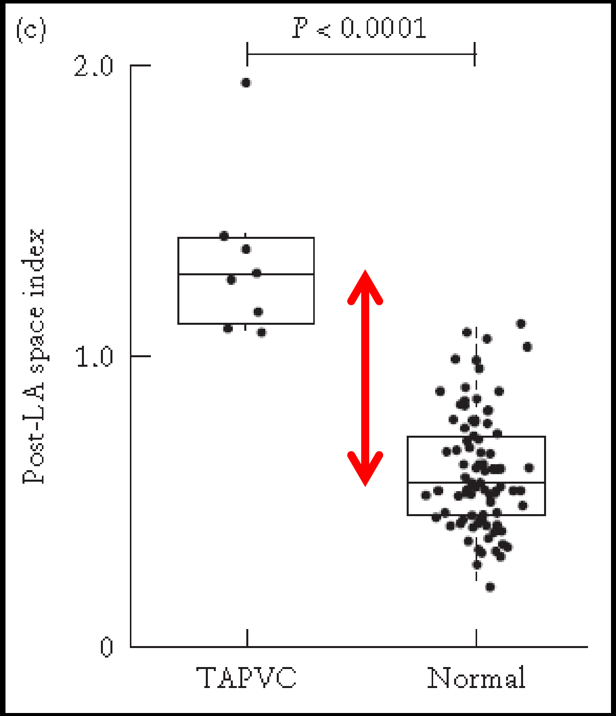
Clues to the Diagnosis

“Post-LA space index” = LD/DA





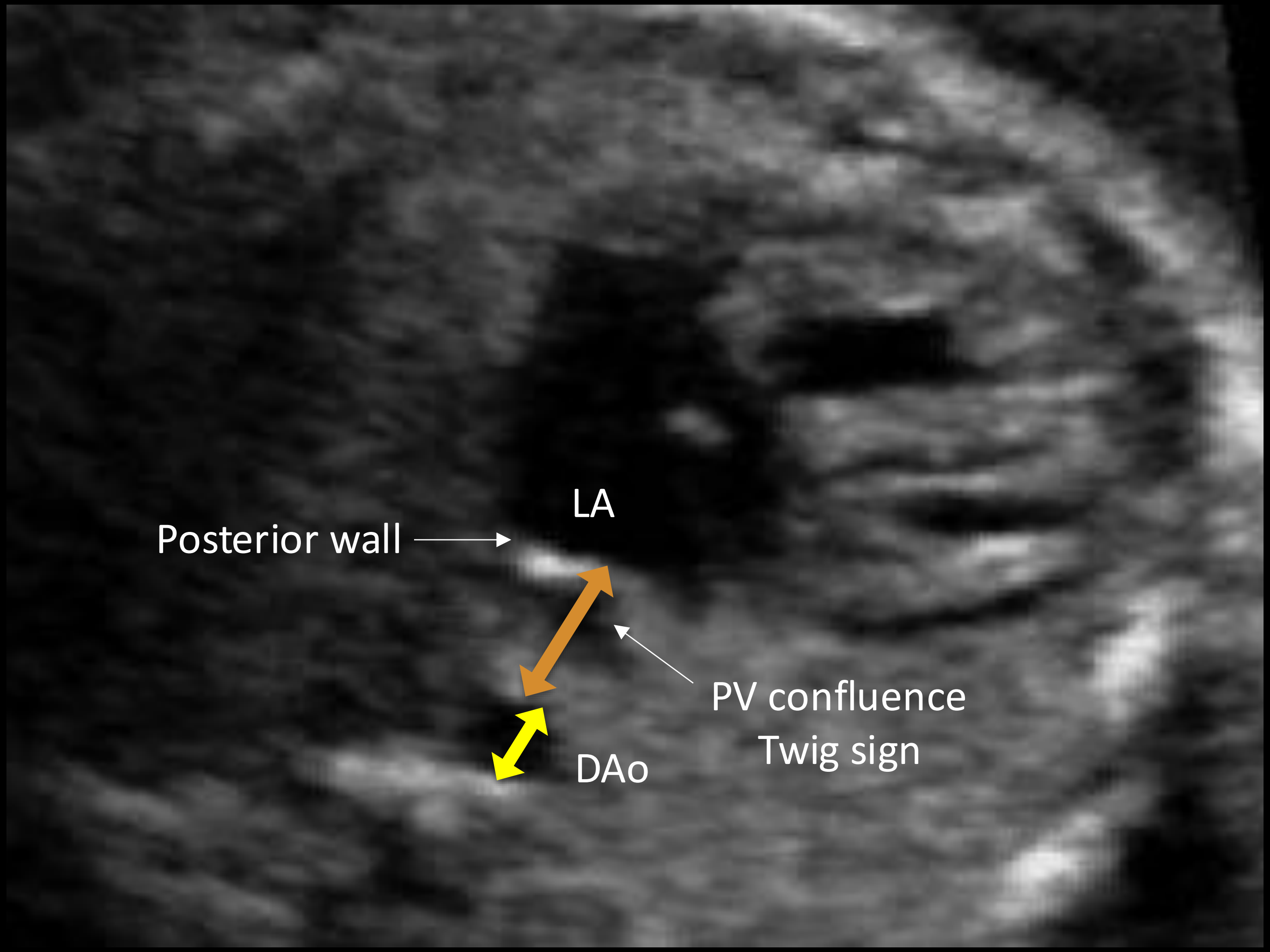
Index of >1.27 100% sensitive and 99% specific



TAPVR=TAPVC

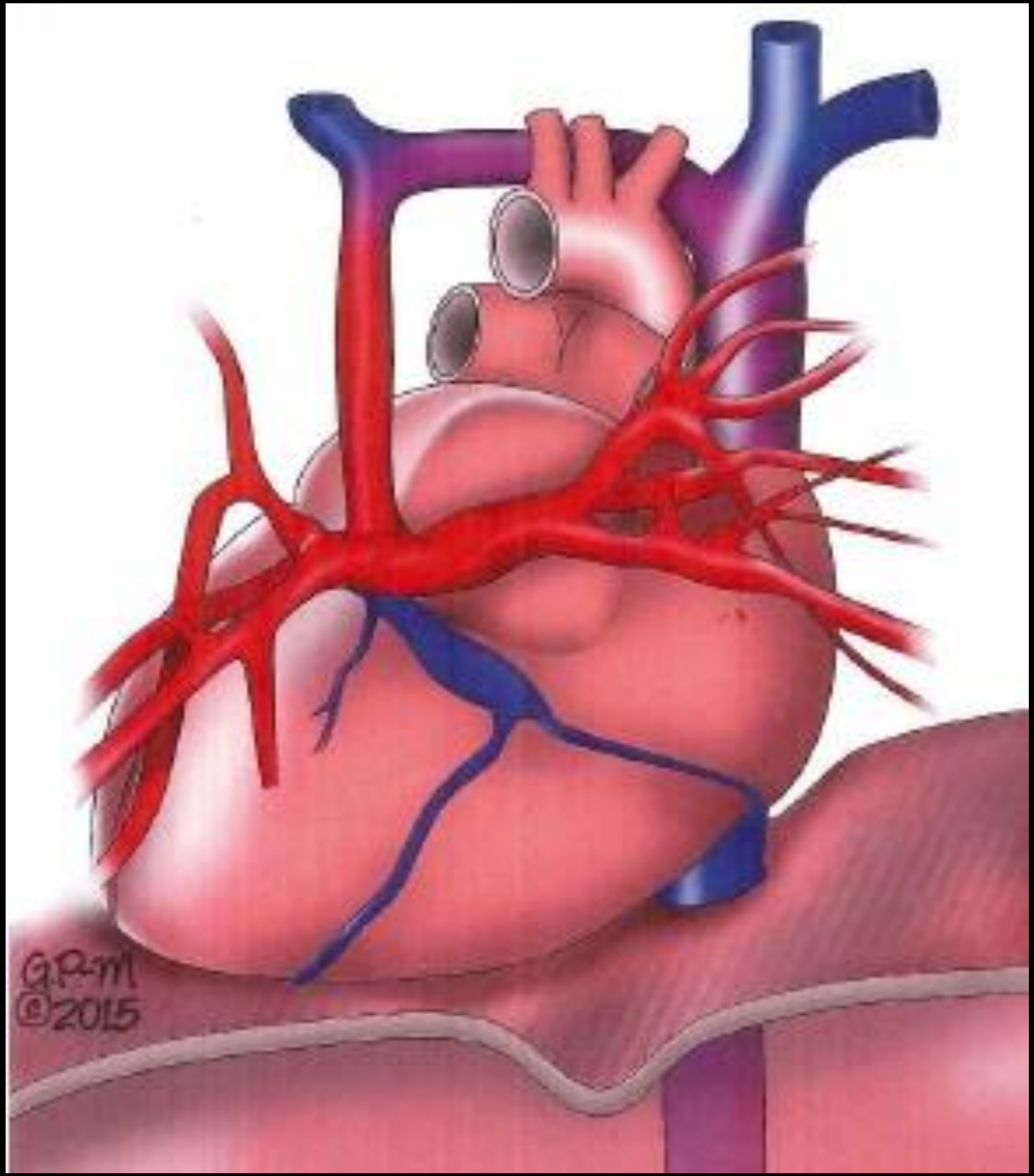
‘Post-LA space index’ as a potential novel marker for the prenatal diagnosis of isolated total anomalous pulmonary venous connection

Clues to the Diagnosis



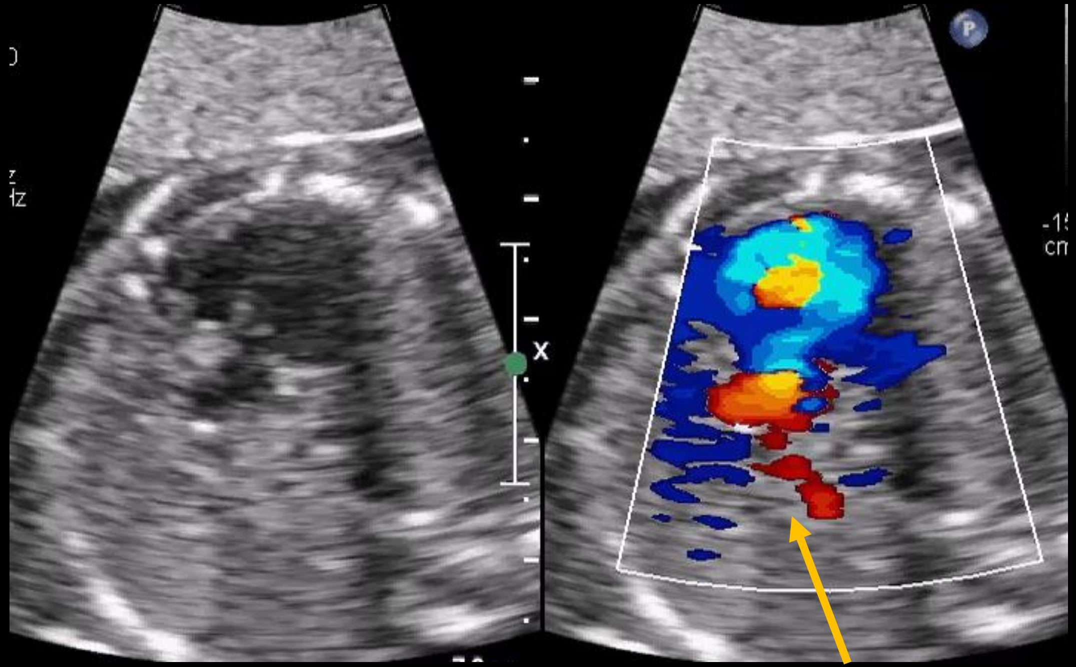
Supracardiac TAPVR: Type I

Most common type of TAPVR



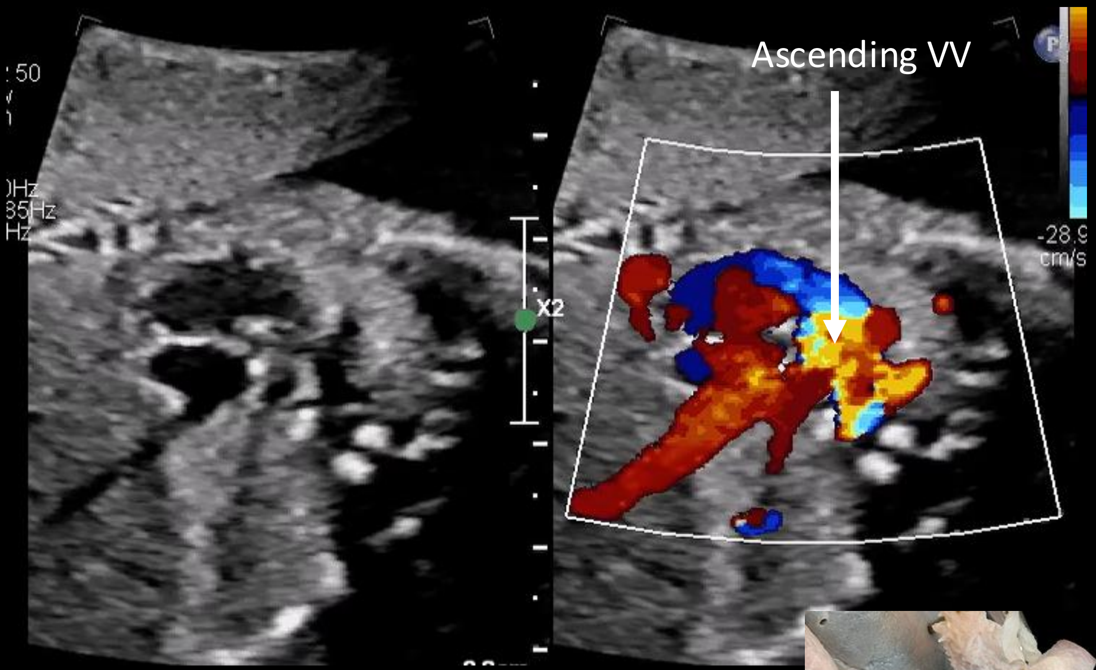
Supracardiac TAPVR

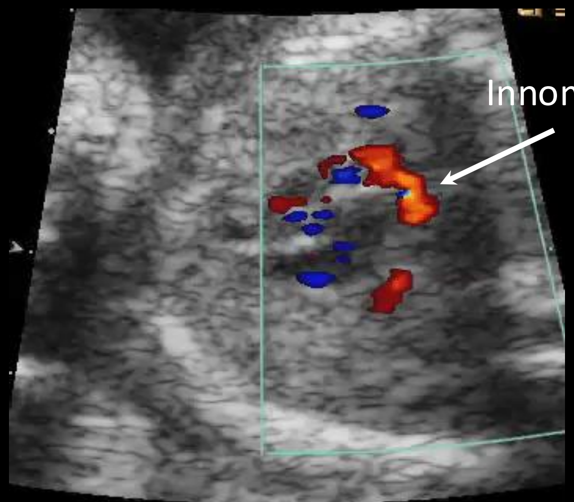
Pulmonary veins drain into confluence near posterior LA

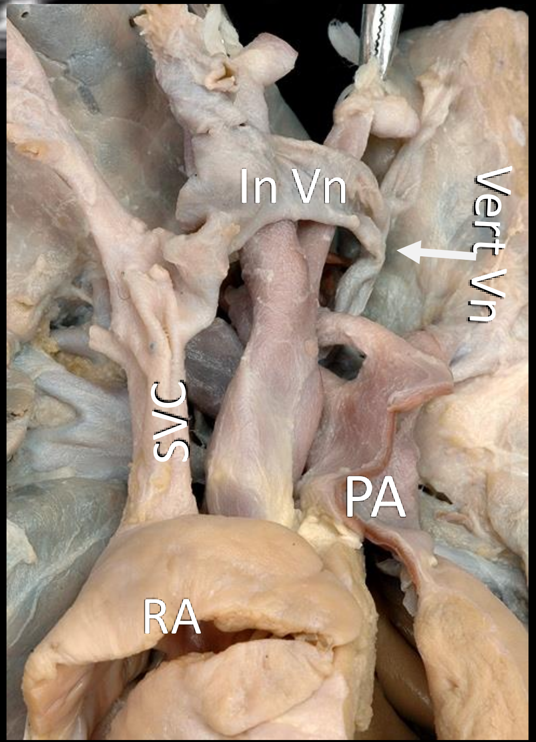


PVs to Confluence

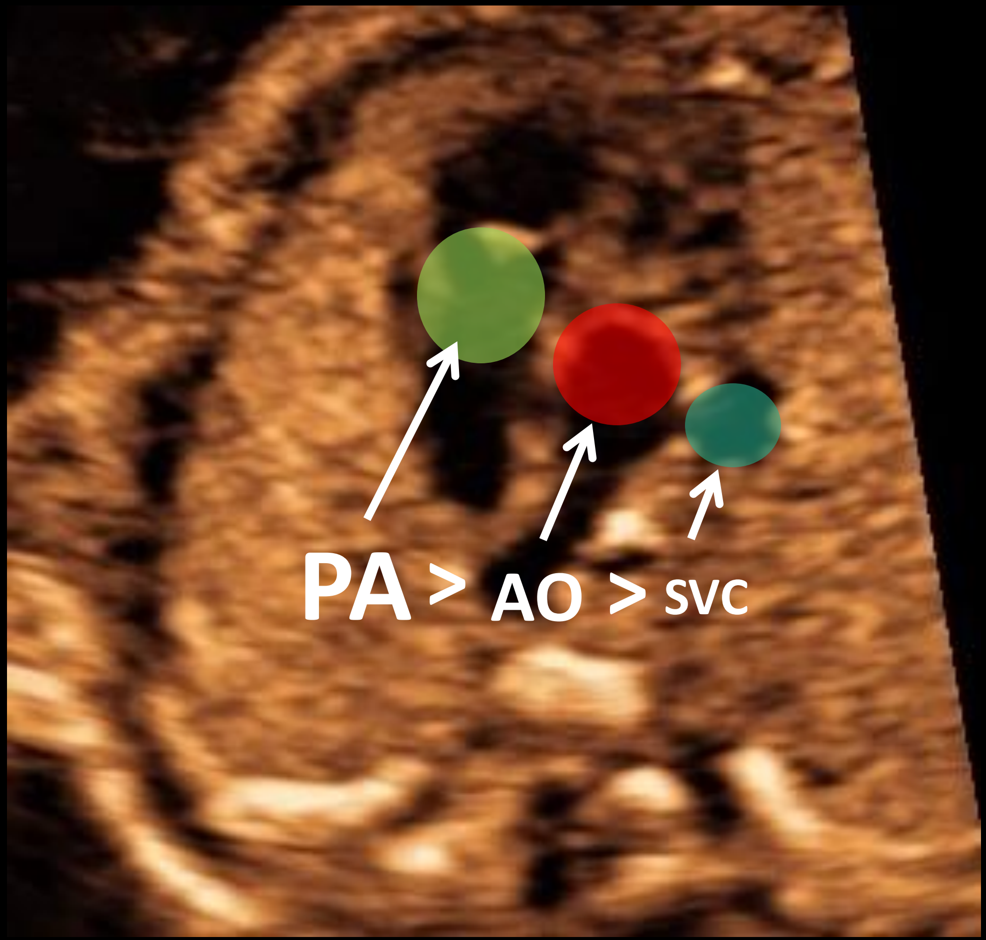
Vertical vein draining superiorly in Innominate vein



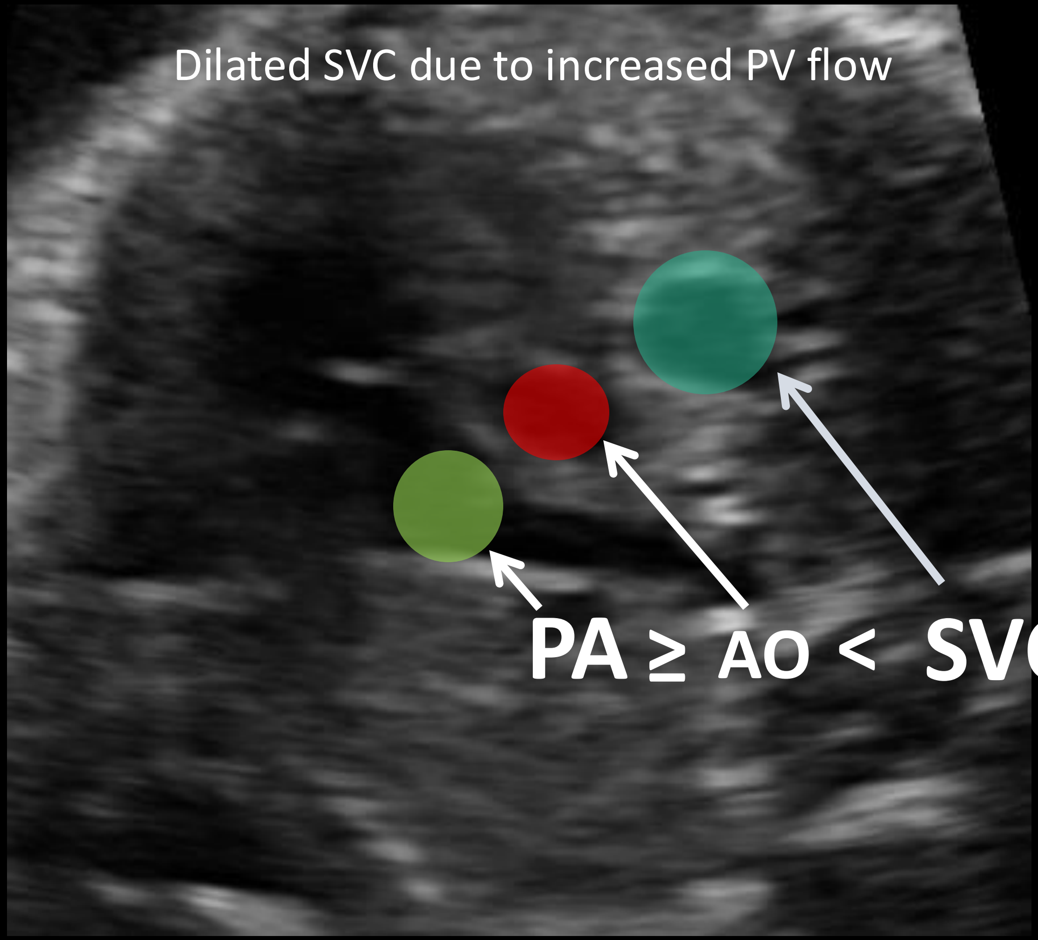




Supracardiac TAPVR – 3VV



Normal

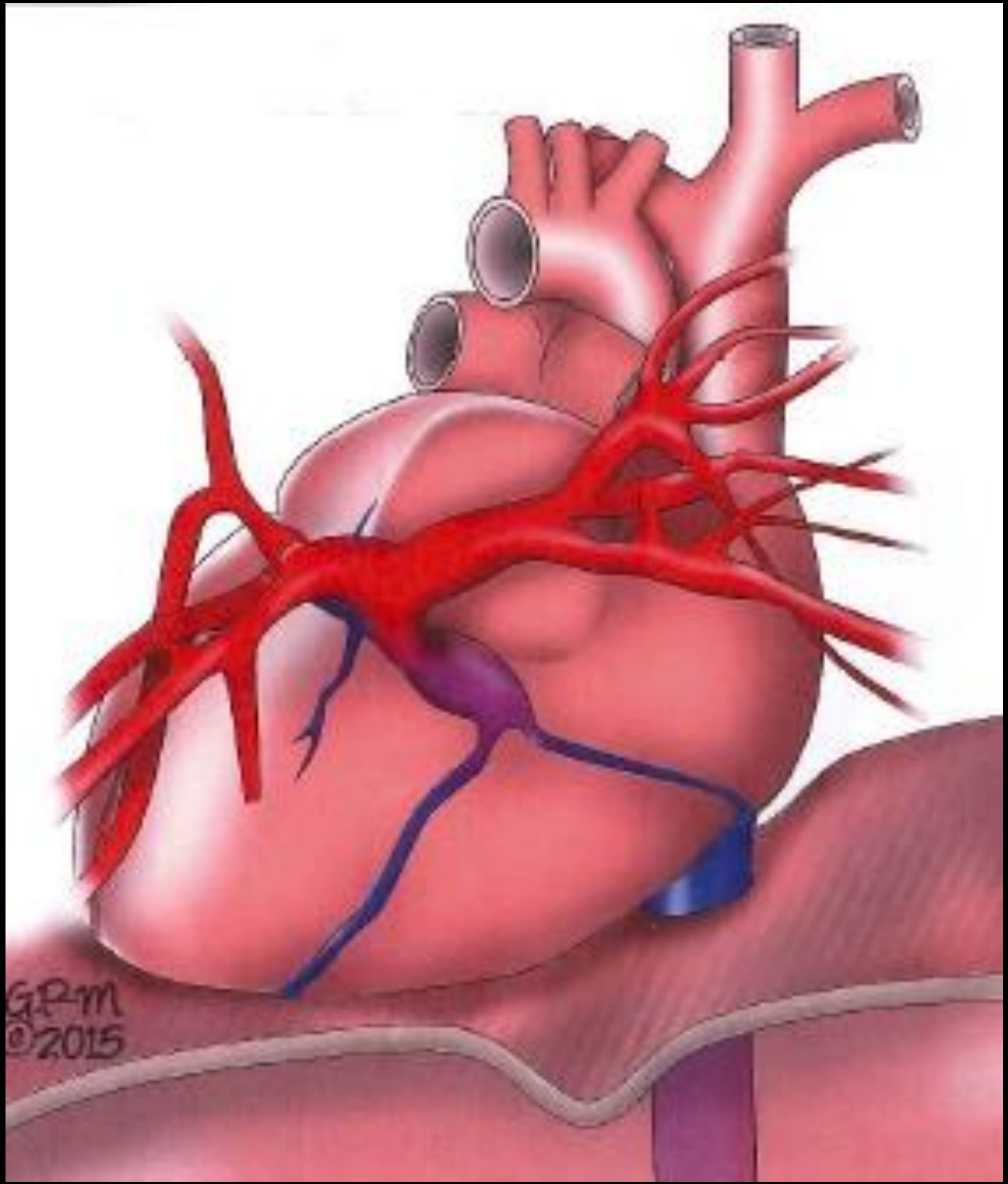


Supracardiac TAPVR

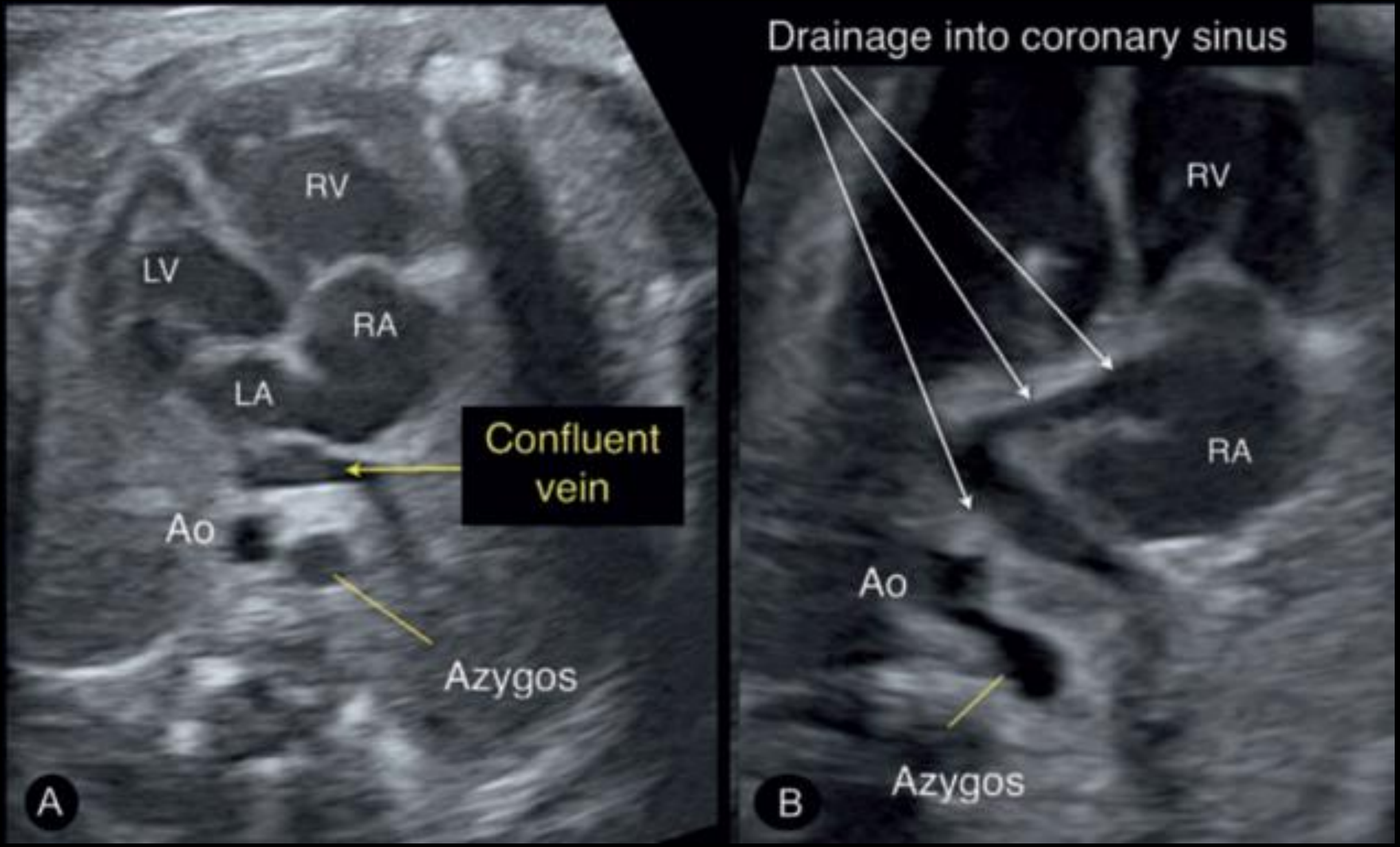
Cardiac TAPVR: Type II

Direct RA drainage more common in right isomerism

Also common to have pulmonary confluence drain to coronary sinus



Cardiac TAPVR to Coronary Sinus: Type II



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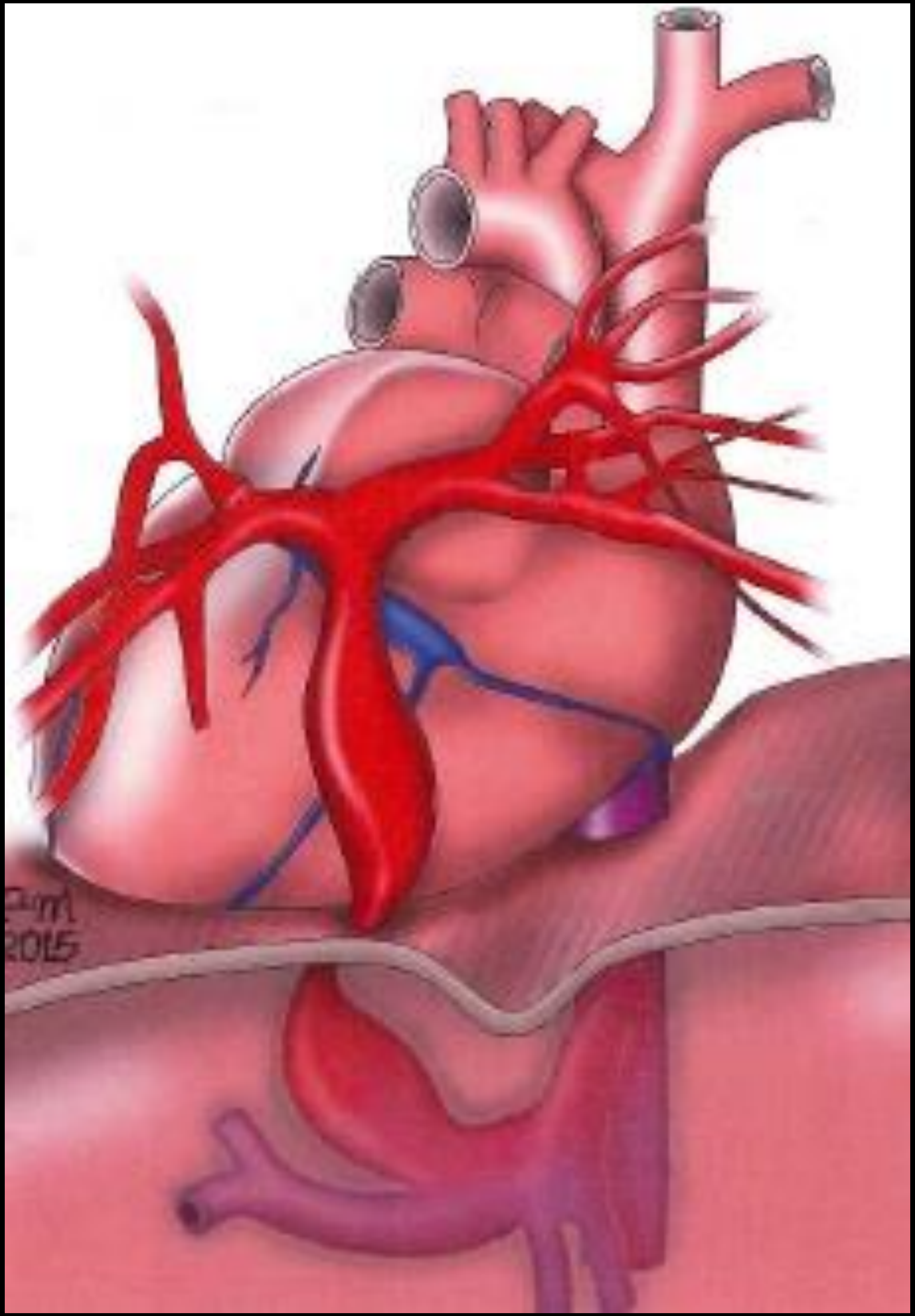
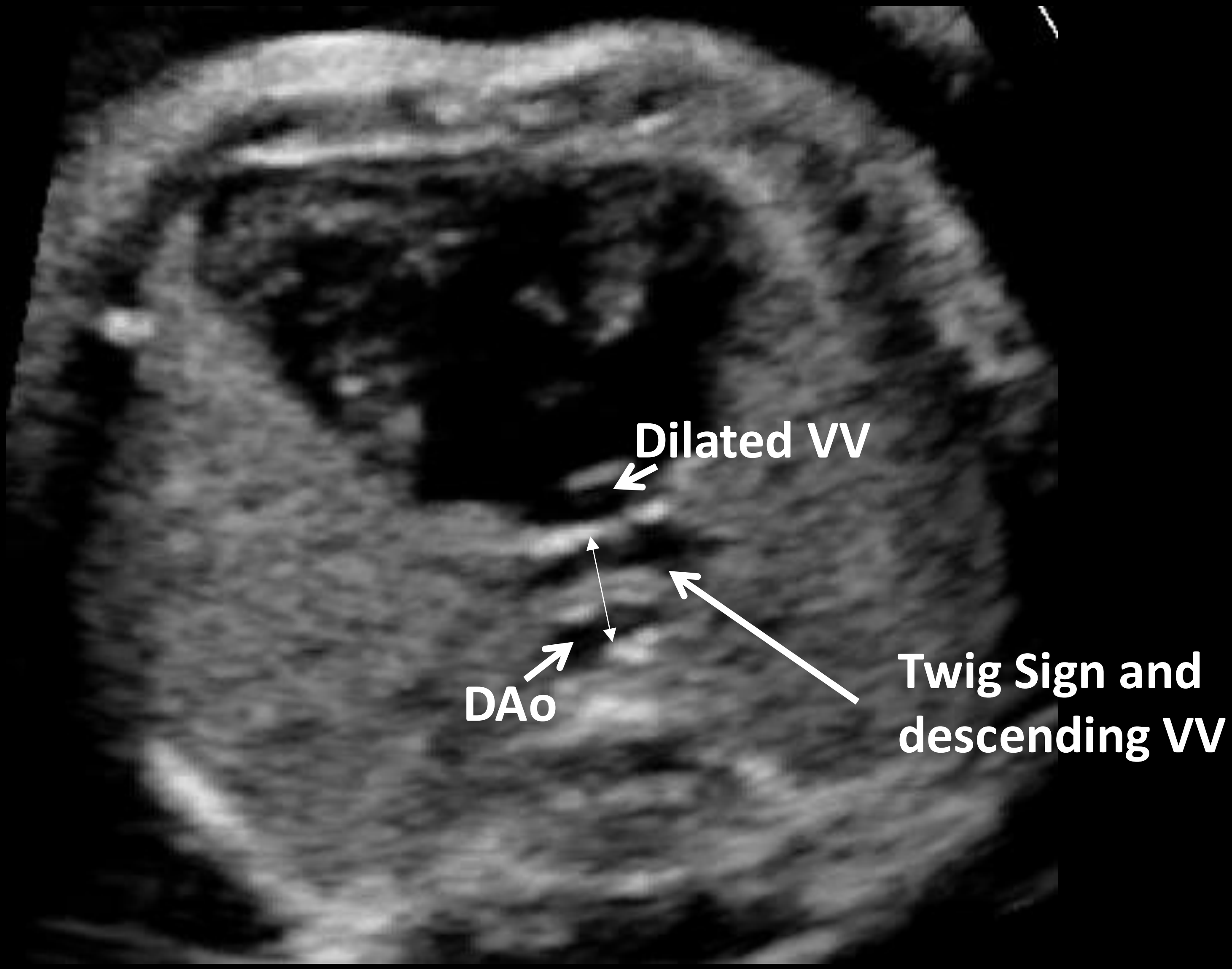
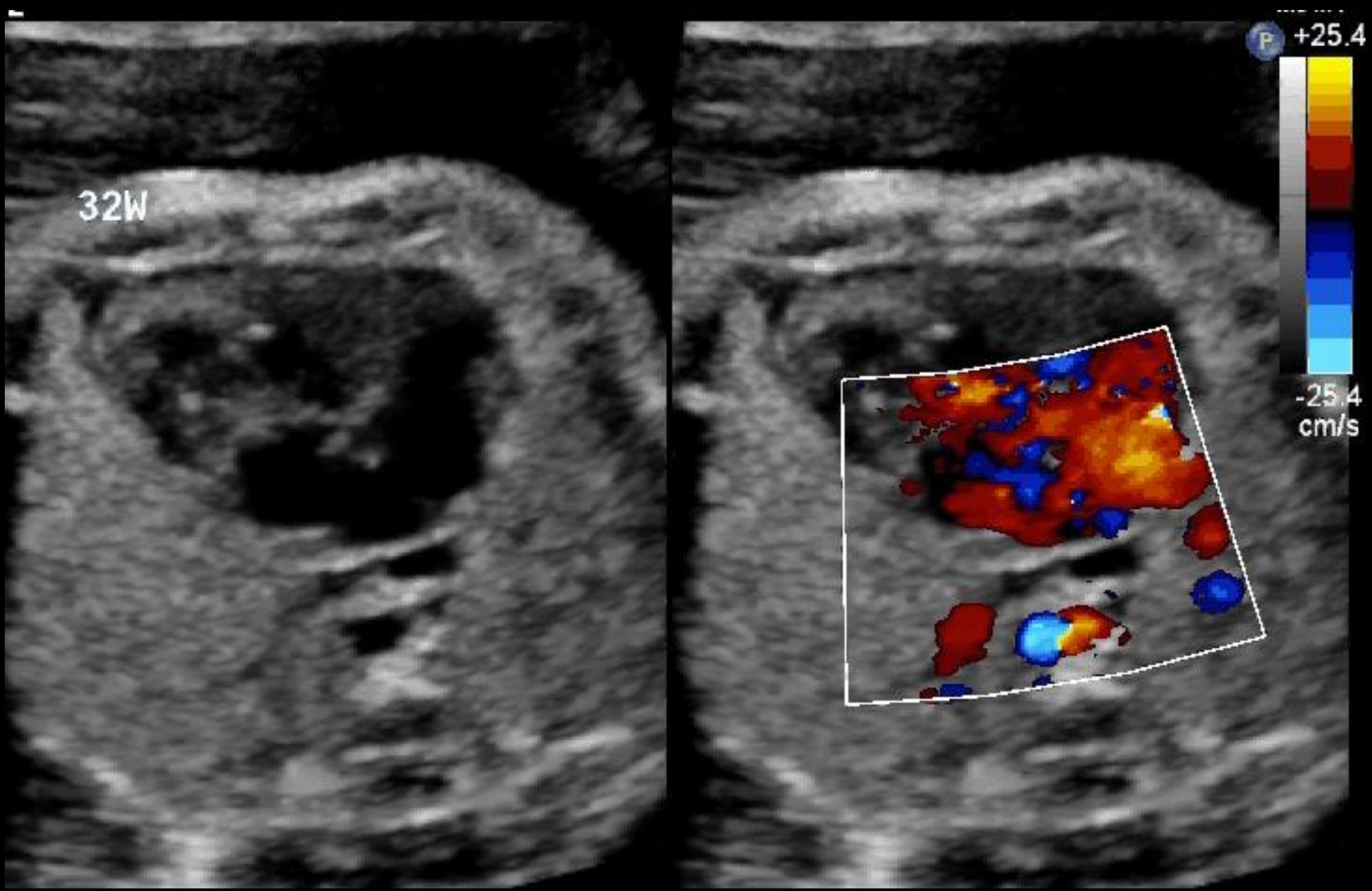
Infracardiac TAPVR: Type III

PV obstruction is the rule

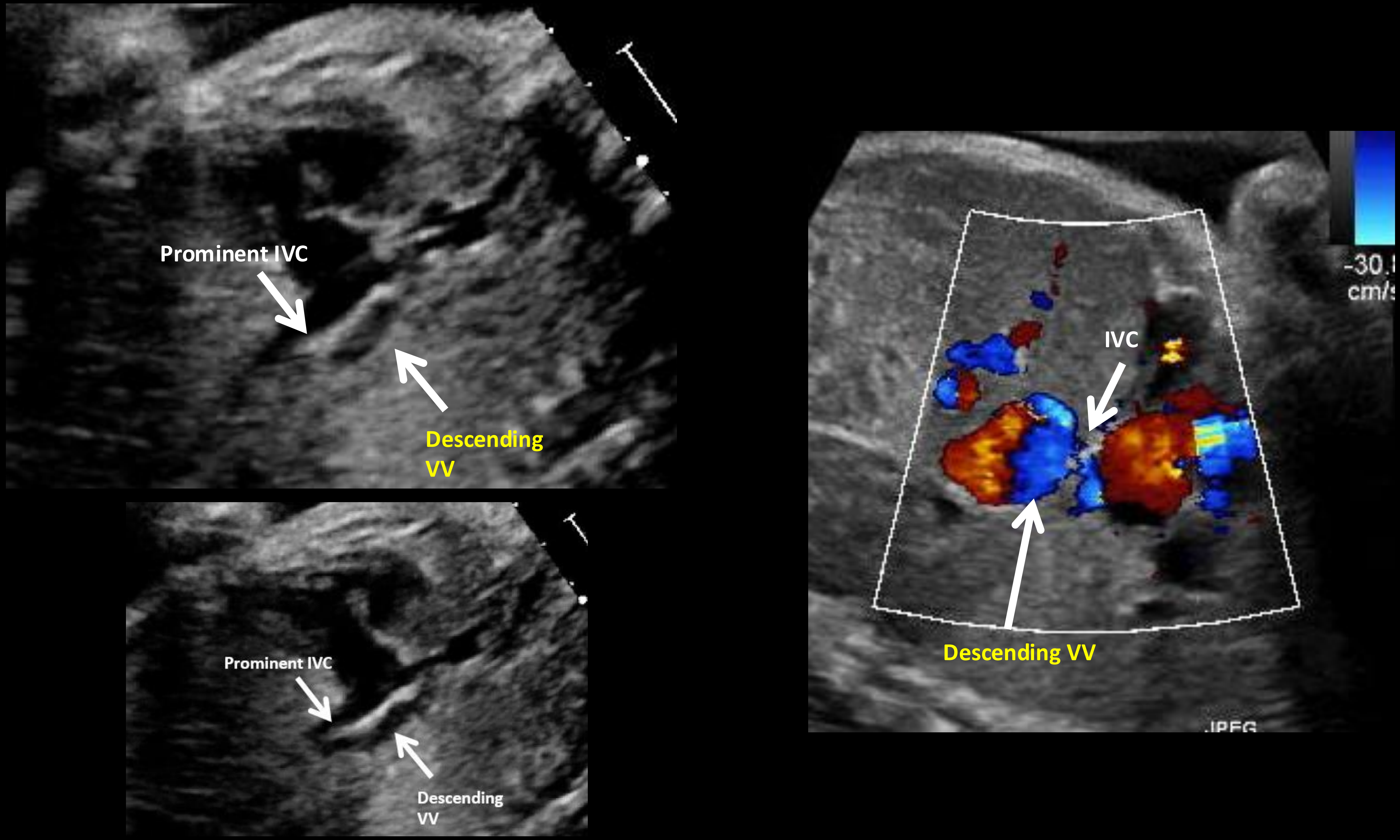


Infracardiac TAPVR: Type III

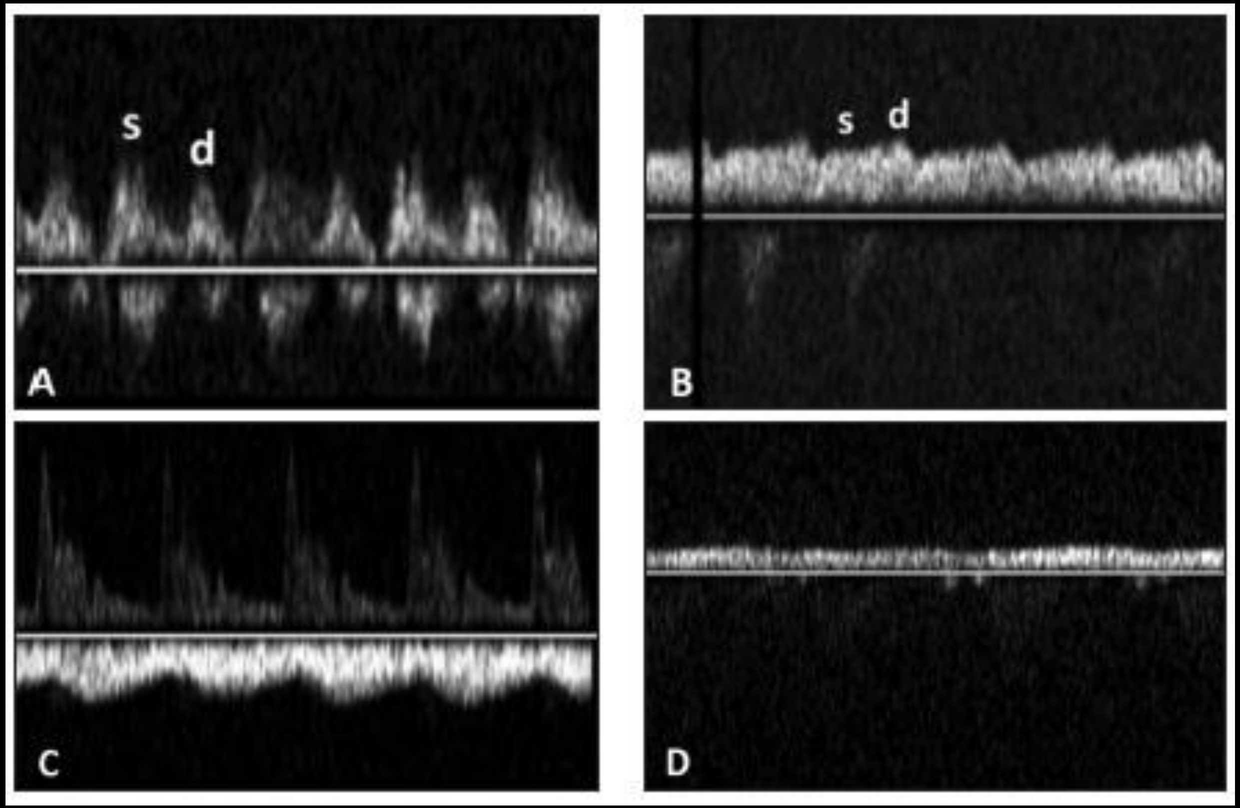




Vertical veins descends inferiorly and drains into hepatics and IVC.
Very dilated vein at obstruction site with swirling flow


Prominent IVC
Descending VV
Prominent IVC
Descending VV
IVC
Descending VV

Predicting Postnatal Outcome



A. Pseudonormal with abnormal s & d but normal pulsatility
B. Abnormal biphasic with decreased pulsatility
C. Monophasic but pulsatile
D. Low velocity monophasic, continuous

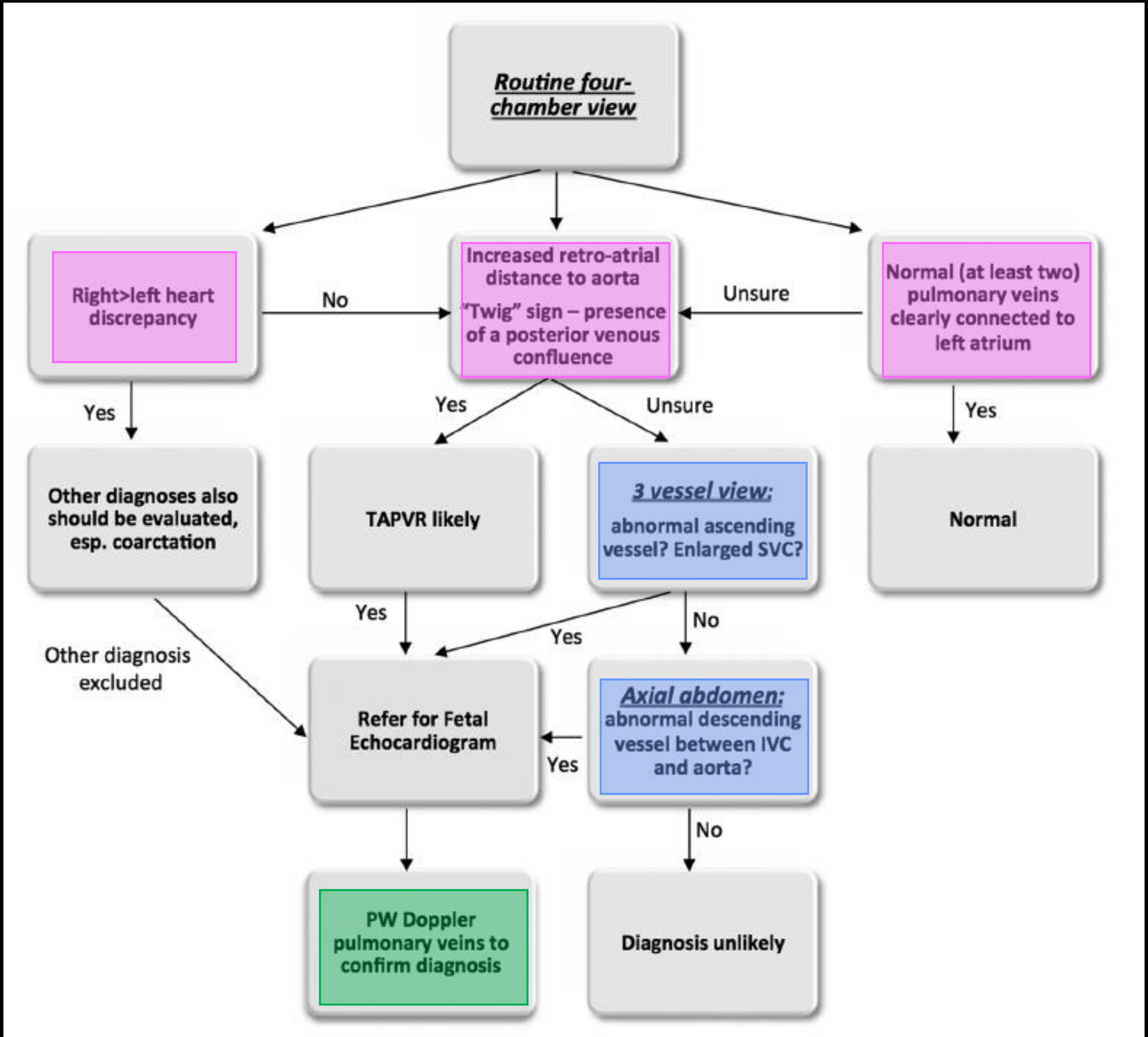
Sonographic Clues to Diagnosis

- <u>RV > LV</u> and <u>RA > LA</u> disproportion
- Presence of <u>pulmonary venous confluence</u>-Twig sign
  - <u>Separation of posterior LA wall</u> and the <u>descending aorta</u>
- Lack of normal pulmonary venous drainage to the LA
- Presence of <u>VV draining</u> cephalad or caudad
  - Three Vessel/Sagittal View-> supracardiac
  - Abdominal Axial/Sagittal View -> infracardiac
- <u>Asymmetric</u> SVC and IVC size
- <u>Dilated coronary sinus</u> (cardiac type)
- Use low Nyquist (scale) to pick up the low flow state
- Abnormal <u>pulmonary venous Doppler</u> pattern

TAPVR: Perinatal Management

- Prognosis dependent on severity of pulmonary venous obstruction and associated cardiac disease
- TAPVR with obstruction
  - Specialized high risk delivery planning
  - Prenatal consultations
  - Neonatology at delivery
  - Rapid access surgical team, consider presence in the DR
  - Emergent surgery after birth – surgery is the only treatment

Approach for screening OB ultrasound and confirming TAPVR diagnosis



Ganesan et al, JUM 2014

Thank you

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