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BMUS Obstetric Study Day 2023 - Cardiac Recap and Interesting Cardiac Cases

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Yorkshire & Humber Congenital Heart Disease Network

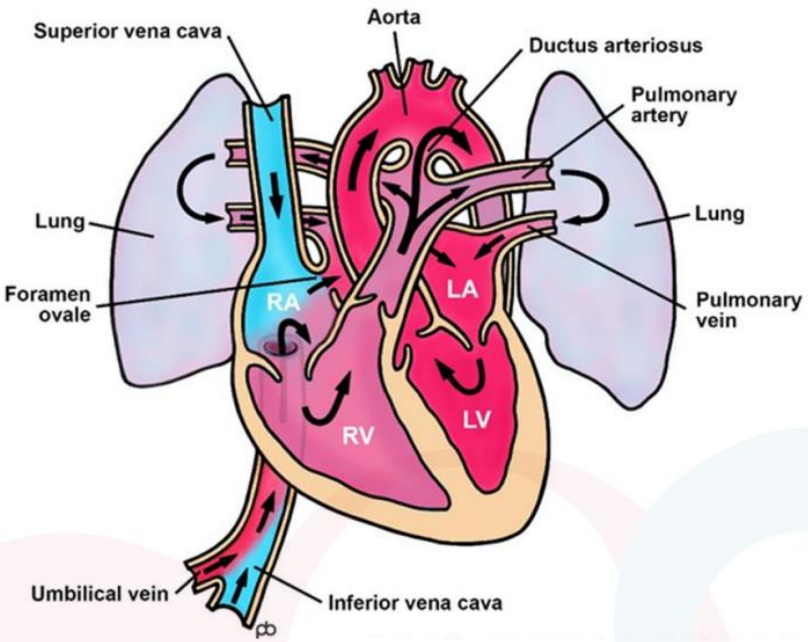
Objectives

- Indications for Fetal Echocardiography
- Normal Heart and Circulation
- Standard screening views
- Structural abnormalities for each screening view
- Rhythm Abnormalities

Indications for Fetal Echocardiogram

Suspected Abnormalities	Maternal Indications	Family History
CHD suspected at anomaly scan/unable to obtain normal views	Maternal CHD/heart block	1st degree relative with CHD/heart block
Fetal Arrhythmia (sustained HR <120 or >200 bpm)	Metabolic Disorders (poor control in early pregnancy)	History of cardiomyopathy
Increased NT - ≥3.5mm in first trimester or ≥6mm at mid trimester	Exposure to known teratogens	
Extracardiac anomalies associated with CHD	Confirmed TORCH infections	
Chromosomal anomalies associated with CHD	Autoimmune antibodies – Anti Ro/La	
Hydrops/pericardial or pleural effusion	NSAID medication in later pregnancy	
Risk of heart failure – TTTS, fetal anaemia, tumours etc		

- Fetal circulation



- Postnatal circulation

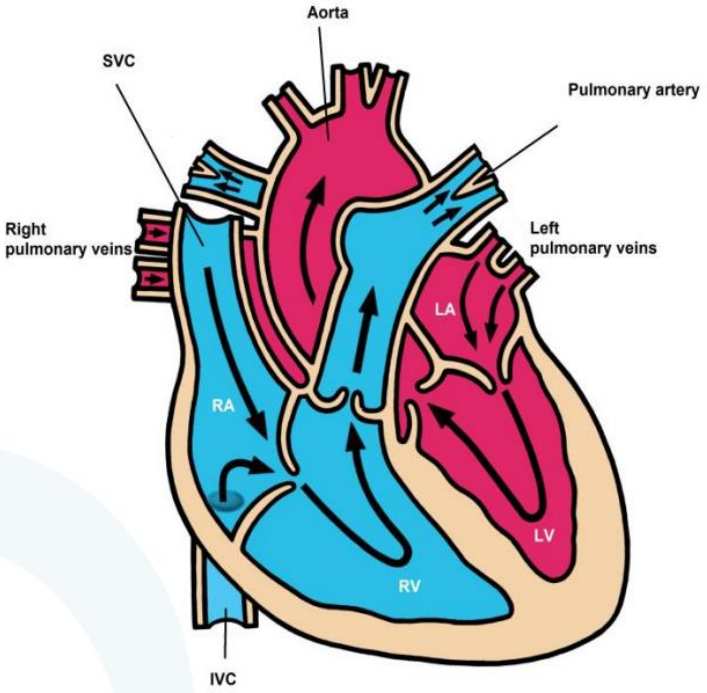
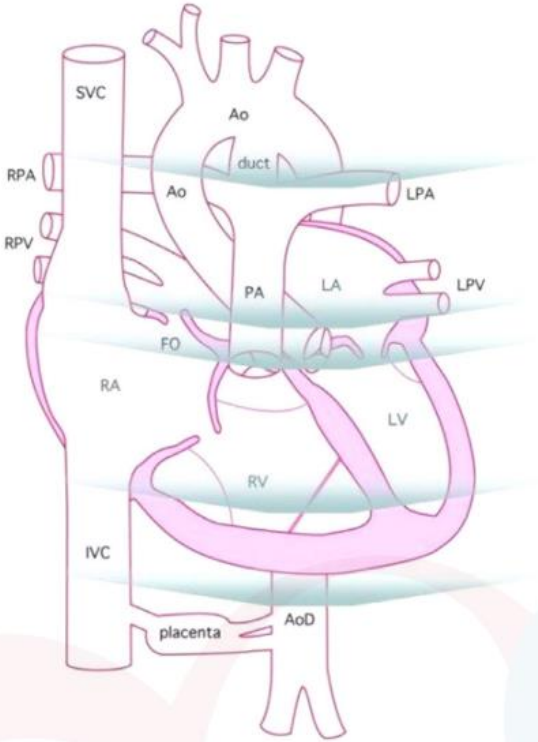
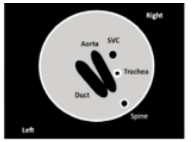


Illustration by P. Brown, Medical Illustration Department, Leeds Teaching Hospitals

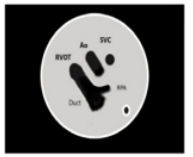
Screening Planes of the Fetal Heart



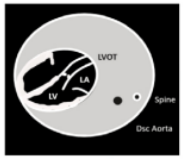
View 5: **3VT**



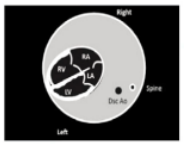
View 4: **RVOT**



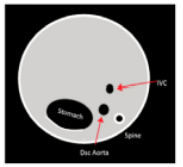
View 3: **LVOT**



View 2: **4 Chamber**



View 1: **Situs**



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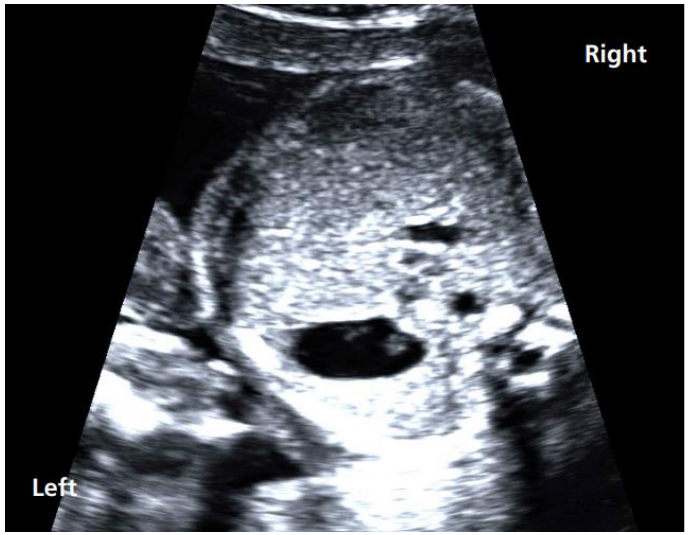
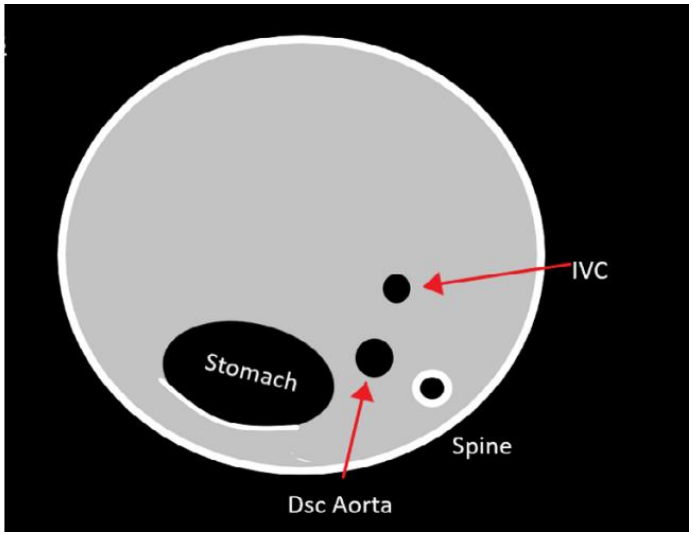
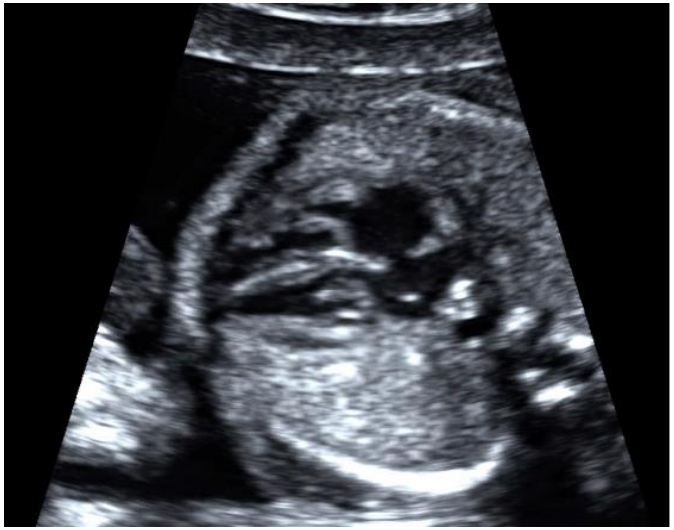
Situs View

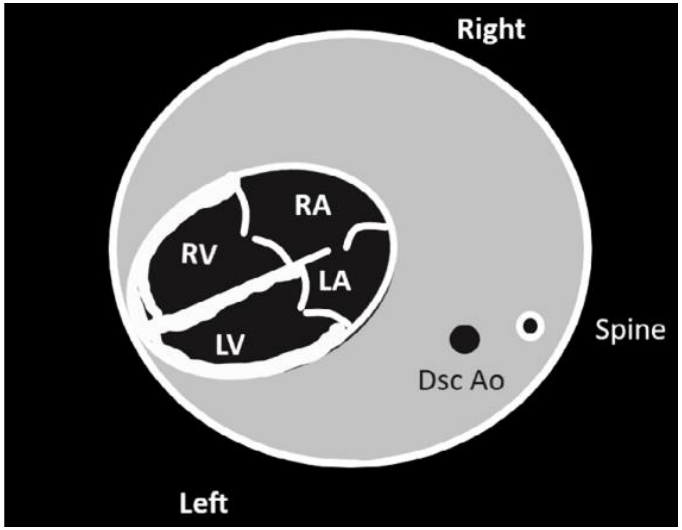




Key Points
Determine left from right from fetal position
Ensure stomach left sided
Ensure abdominal aorta to left of spine
Ensure IVC anterior and to right of spine

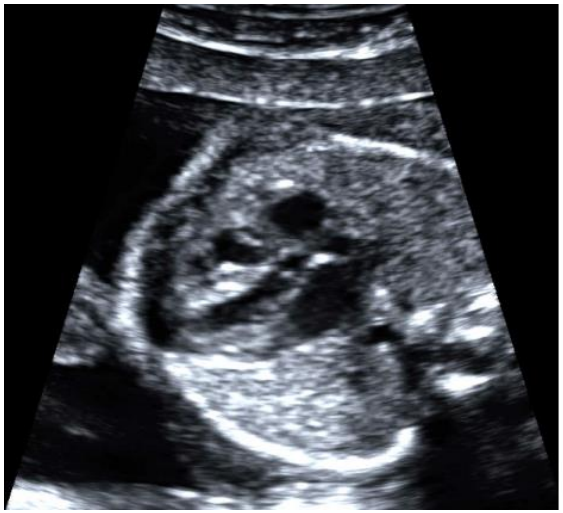
4 Chamber View

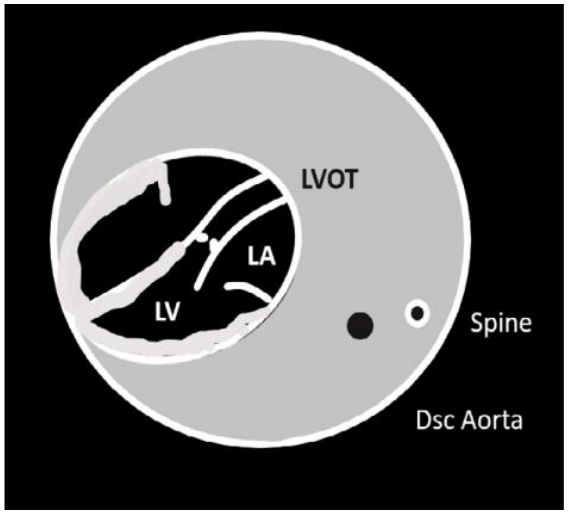


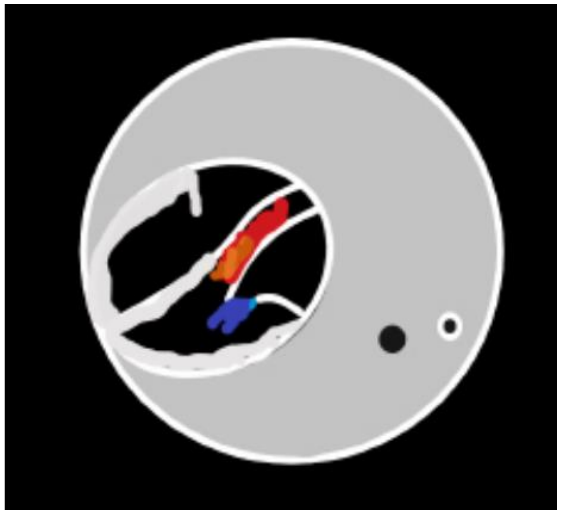


Key Points
Apex leftwards at 45^0 angle and occupies < ½ thorax
Balanced atria and ventricles – moderator band in RV
Primum Septum present with TV offset to apex
Ventricular septum intact
Normal heart rate, regular rhythm, good contractility
<2mm pericardial effusion

LVOT View

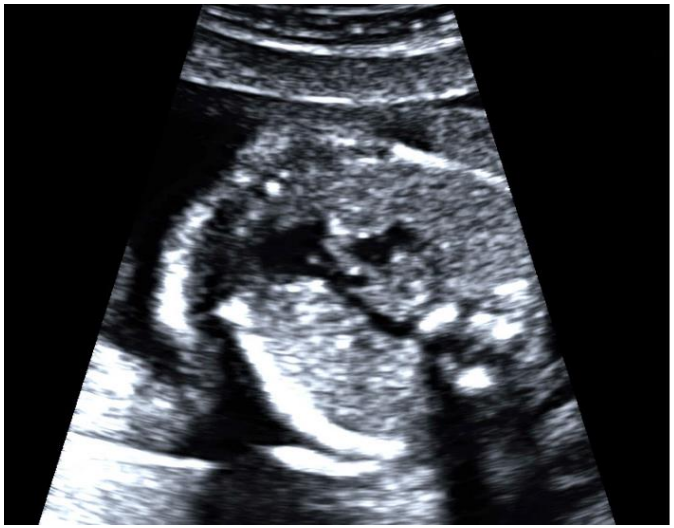


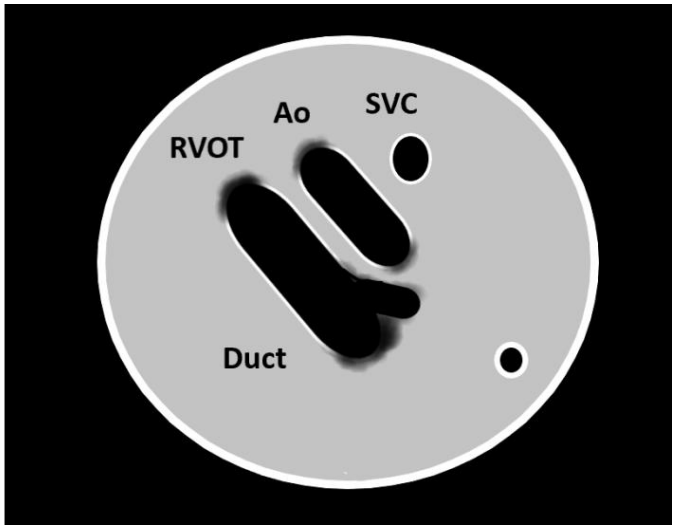




Key Points
Aorta committed to the posterior LV
Continuity of ventricular septum to aortic valve
LV outflow directed towards right shoulder

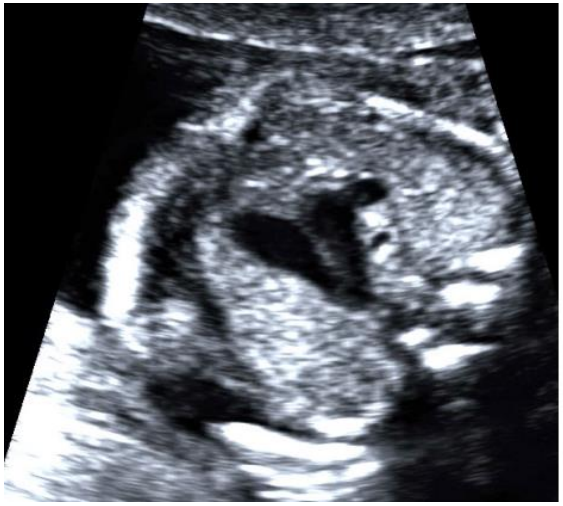
RVOT View

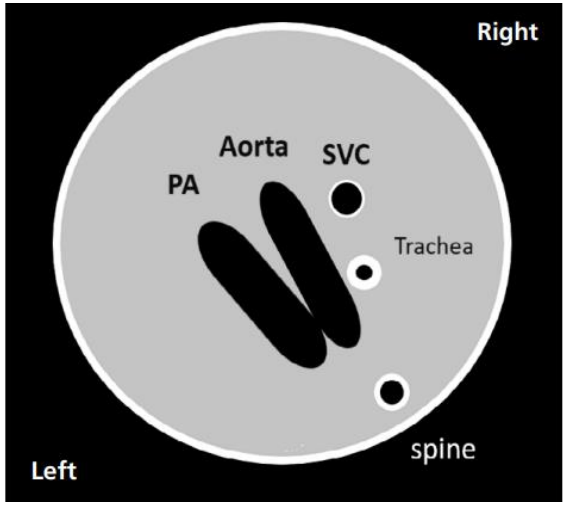


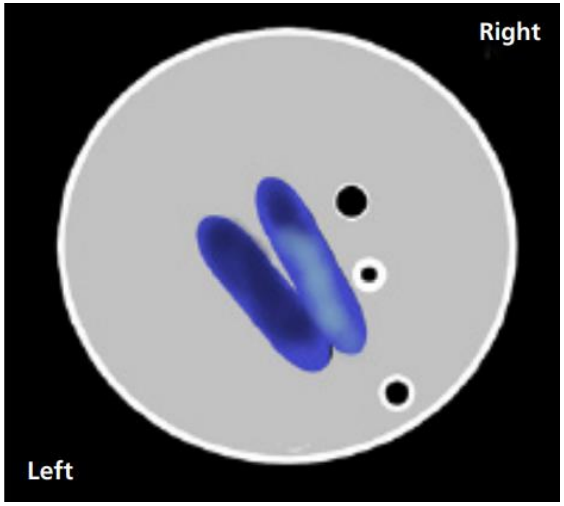


Key Points
RVOT arises anterior to LVOT
Outflow tract originating from the anterior RV
RVOT directed directly back towards spine
Normal crossover at 90^0 to the LVOT

3VT View







Key Points
PA/Duct > Aorta > SVC
Duct and transverse arch come together to form a V
Aorta to left of trachea
Flow in both duct and aorta towards spine

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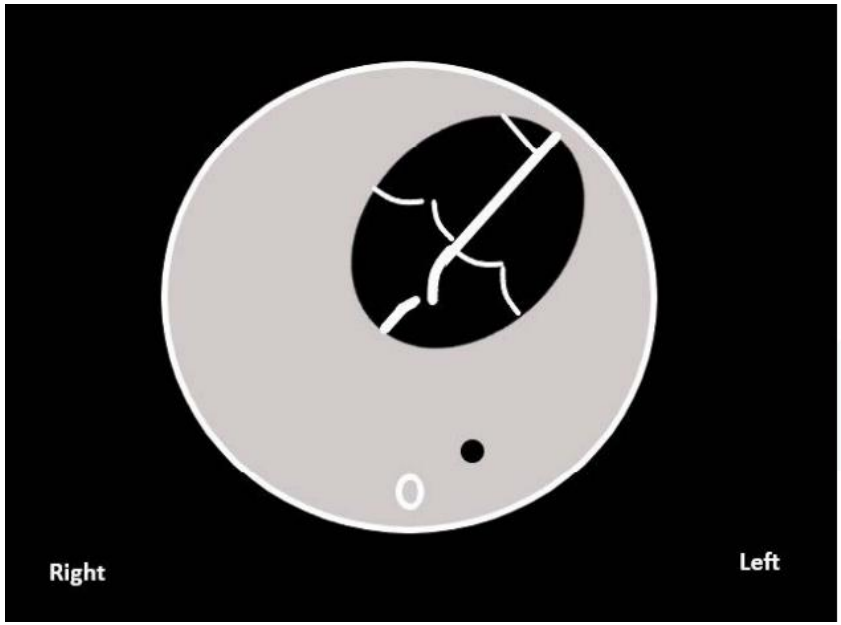
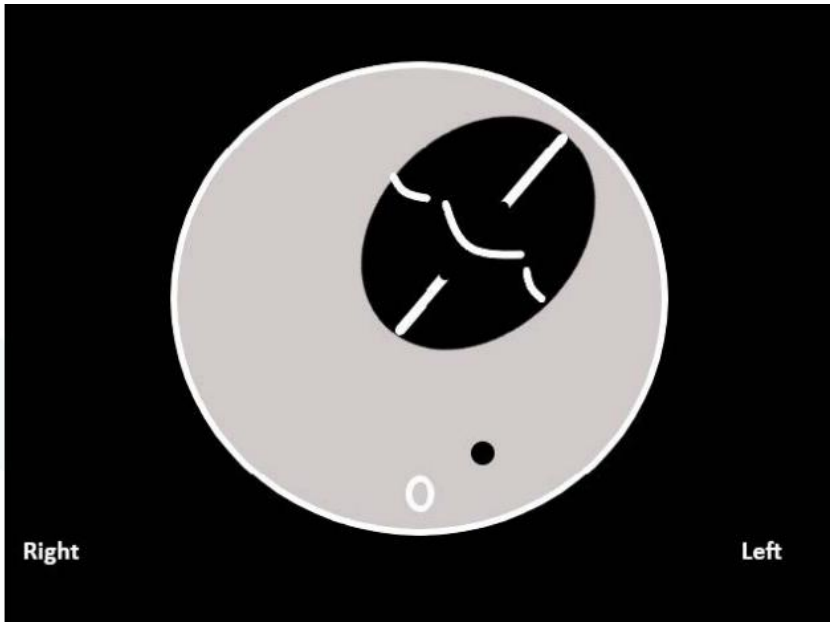
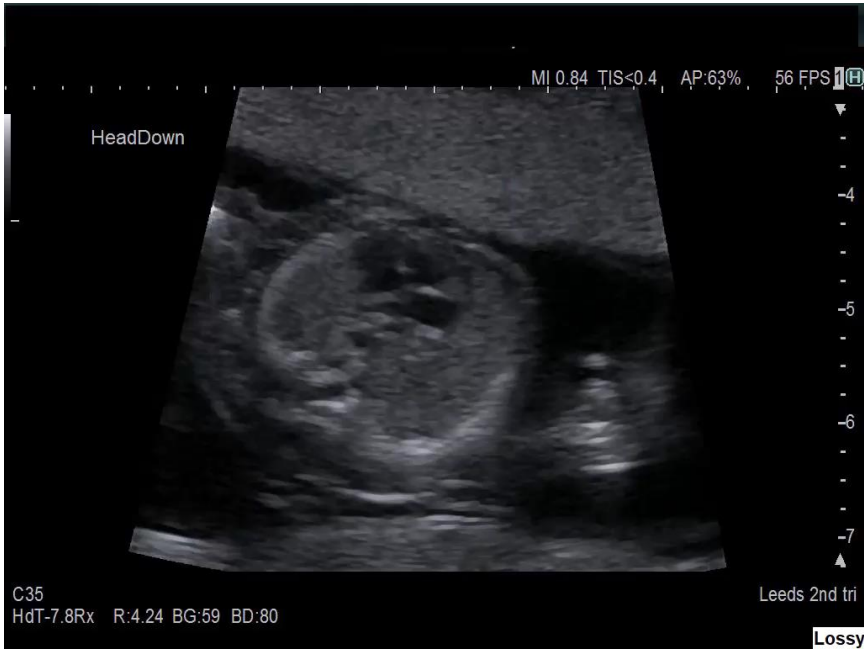
Abnormalities of 4 Chamber View – Atrioventricular Septal Defects





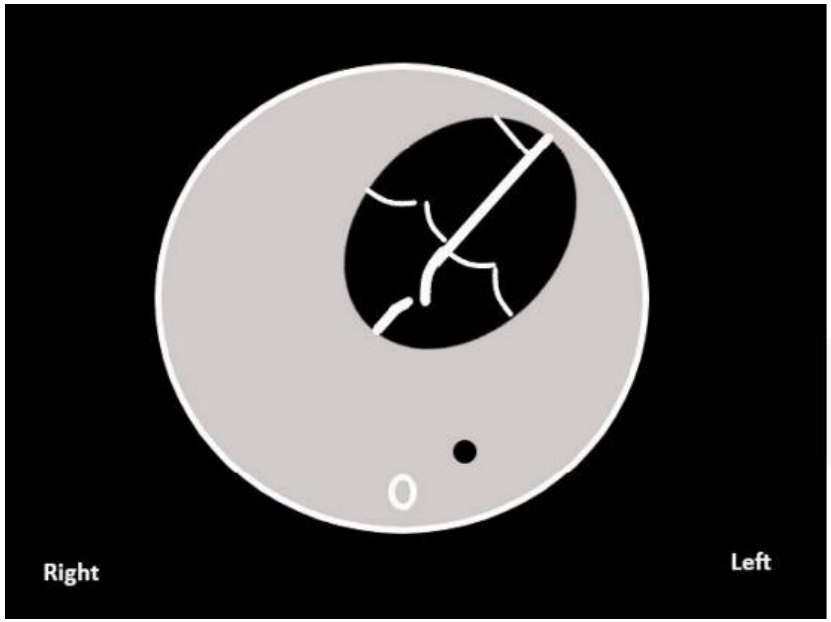
Atrioventricular Septal Defects - AVSDs

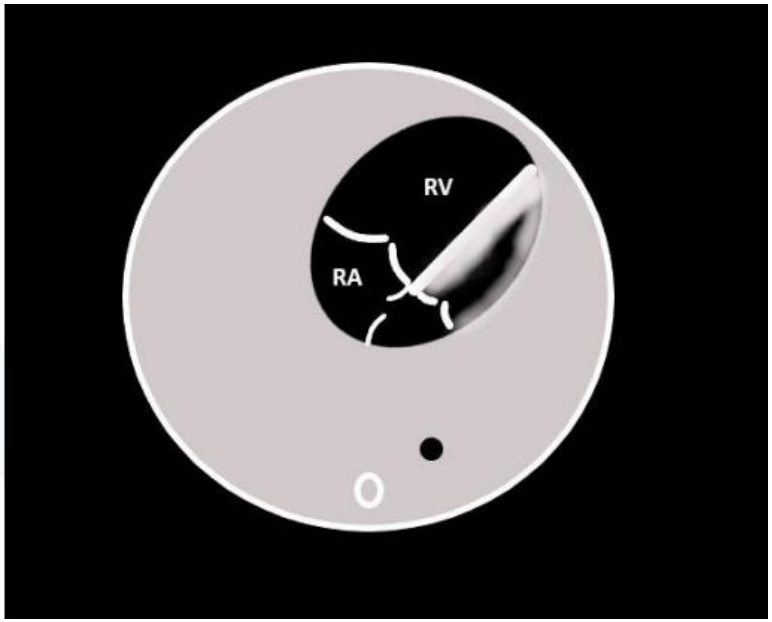
- Common abnormality with 4 chamber view
  - 3 in 10,000 live births [1]
  - Strongly associated with trisomy 21
    - In fetal series, incidence reported 50-90% [1]

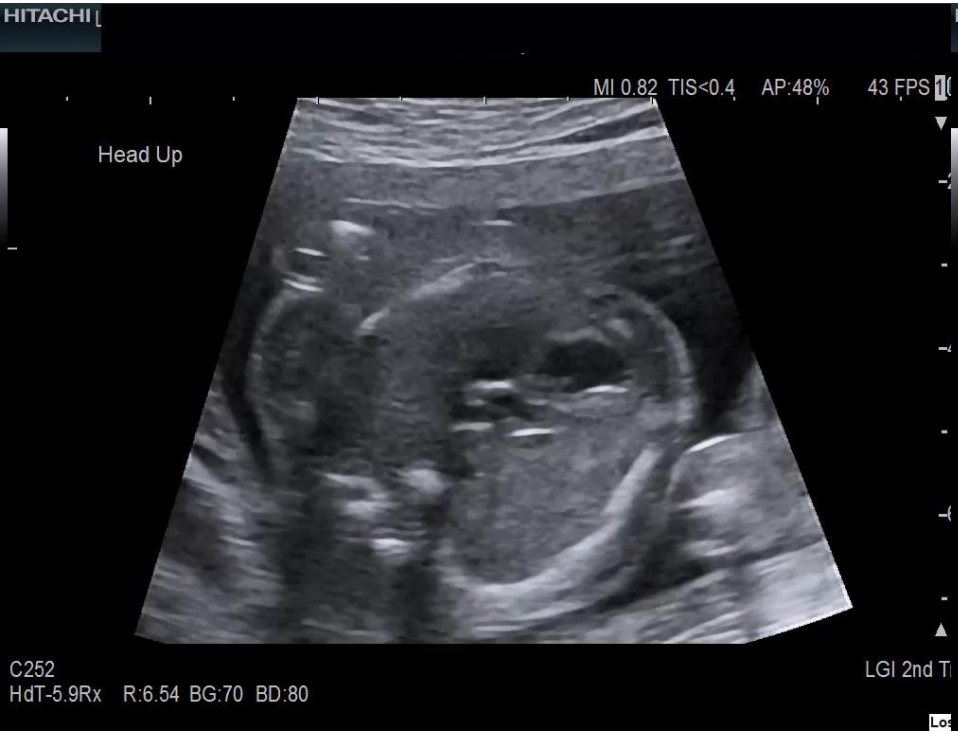


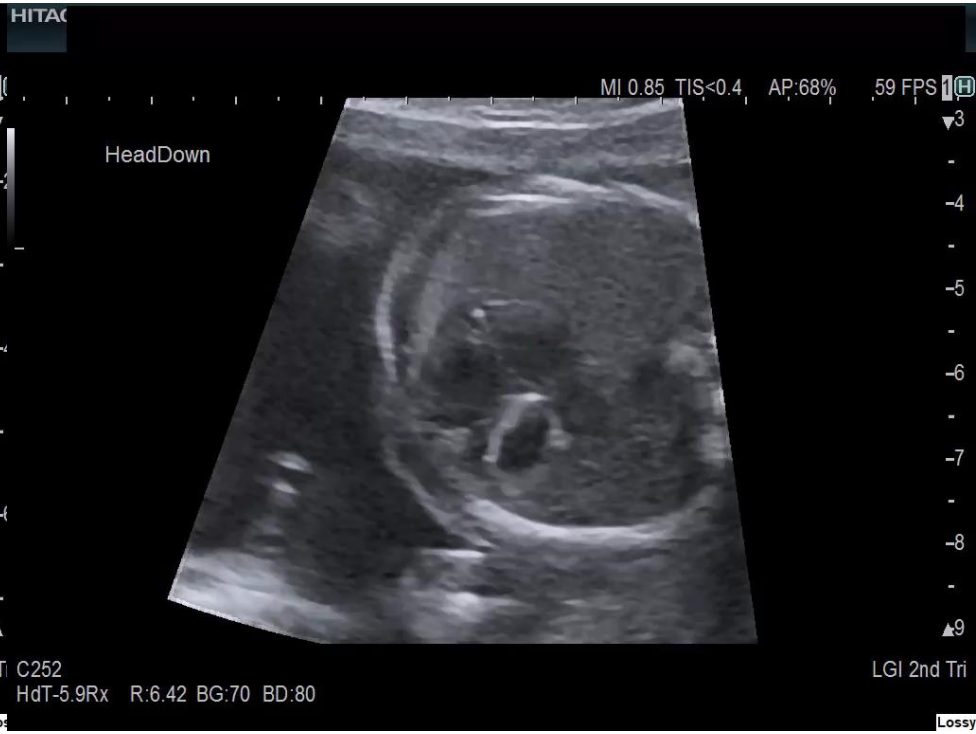
- Can occur in isolation or associated with:
  - Heterotaxy syndromes
  - Tetralogy of Fallot – 5%
  - DORV
  - Common arterial trunk
  - Left heart obstruction/LV hypoplasia (unbalanced AVSD)

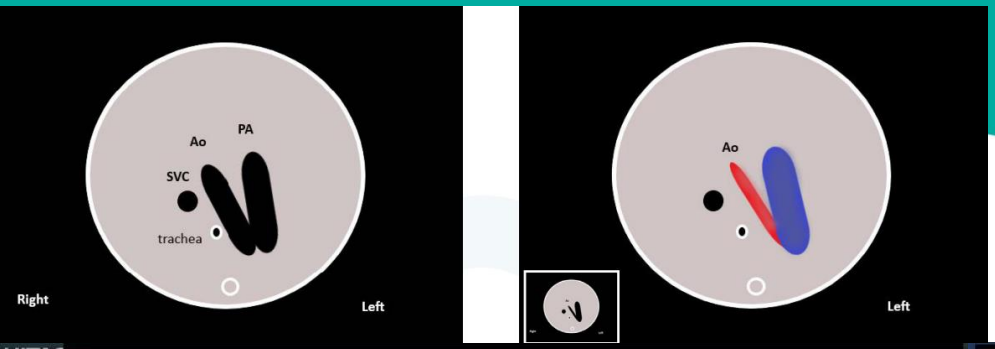
Abnormalities of 4 Chamber View – Hypoplastic Left Heart Syndrome

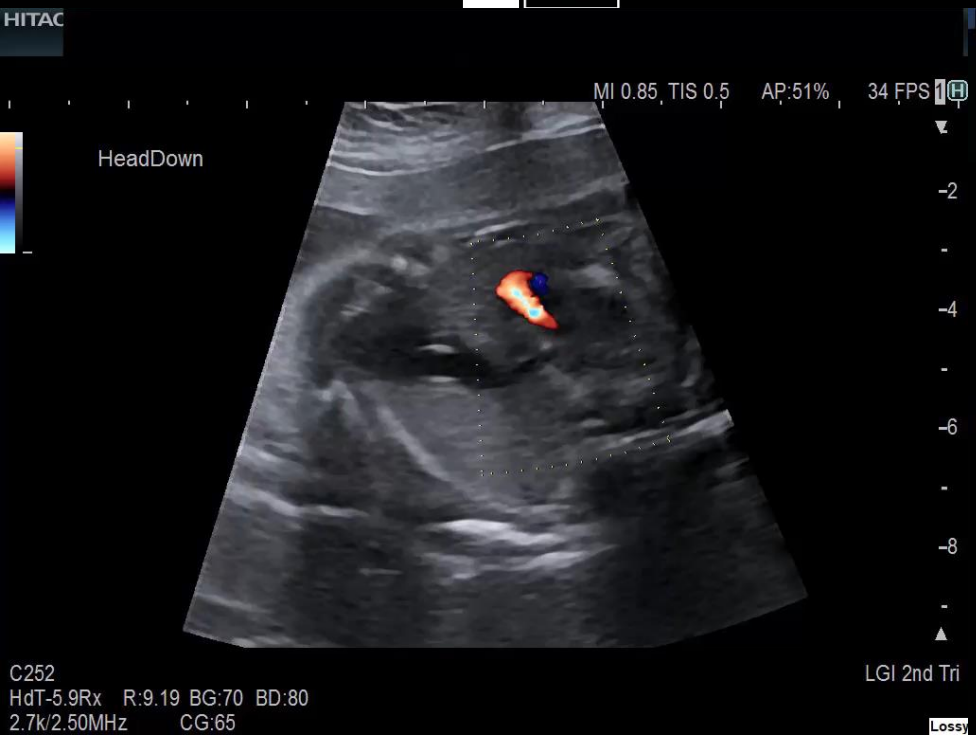


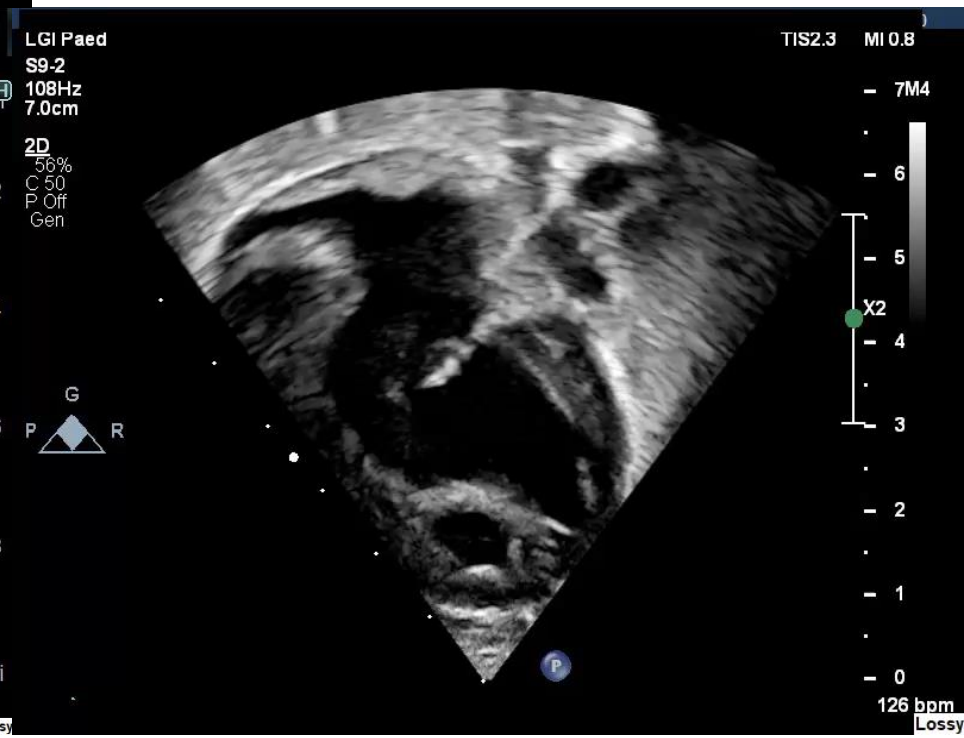



HITACHI
MI 0.82 TIS<0.4 AP:48% 43 FPS
Head Up
C252
HdT-5.9Rx R:6.54 BG:70 BD:80
LGI 2nd T


HITAC
MI 0.85 TIS<0.4 AP:68% 59 FPS
HeadDown
C252
HdT-5.9Rx R:6.42 BG:70 BD:80
LGI 2nd Tri
Lossy







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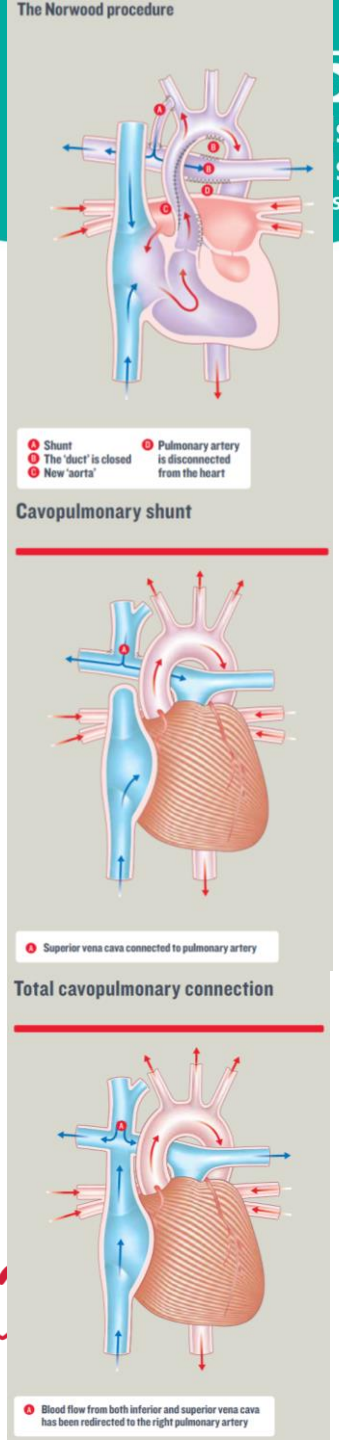
Hypoplastic Left Heart Syndrome

- Unable to achieve biventricular circulation
- Requires multiple stages palliative surgeries
  - Norwood Procedure in infancy
  - Cavopulmonary shunt 4-6 months of age
  - Fontan completion 3-5years of age
    - Multiple cardiac catheterisations and possible surgeries in the interim
- UK National Data
  - 55-60% chance of survival to Fontan completion/5yrs
- 15% risk of death prior to 1st stage surgery [2]



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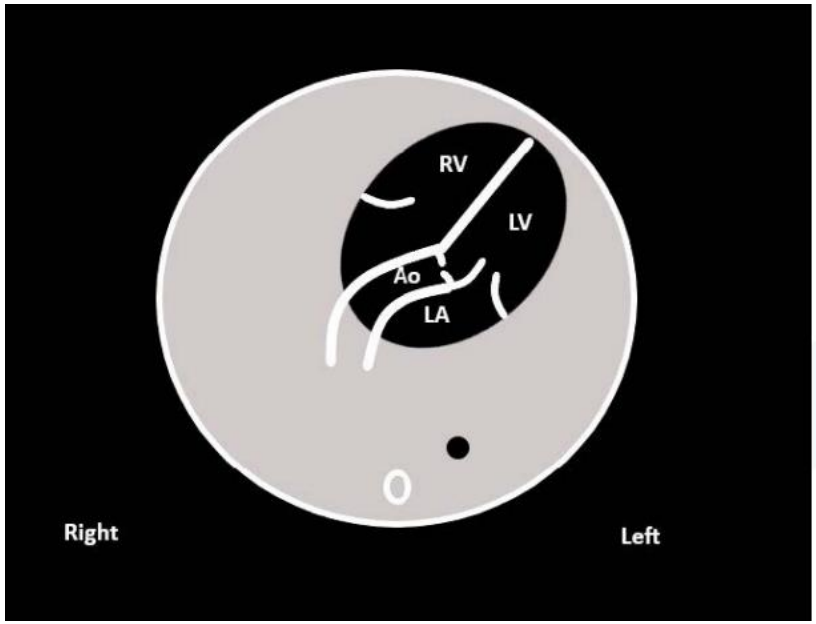
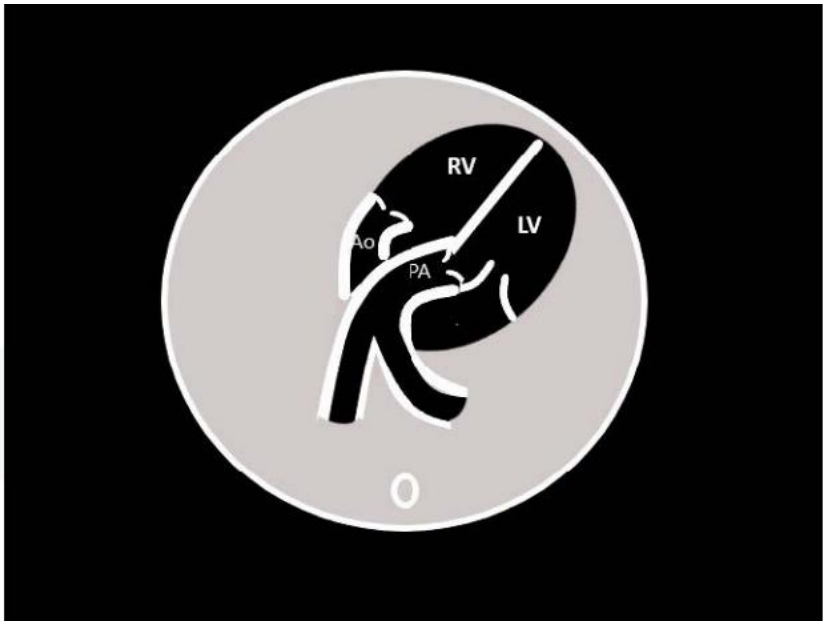
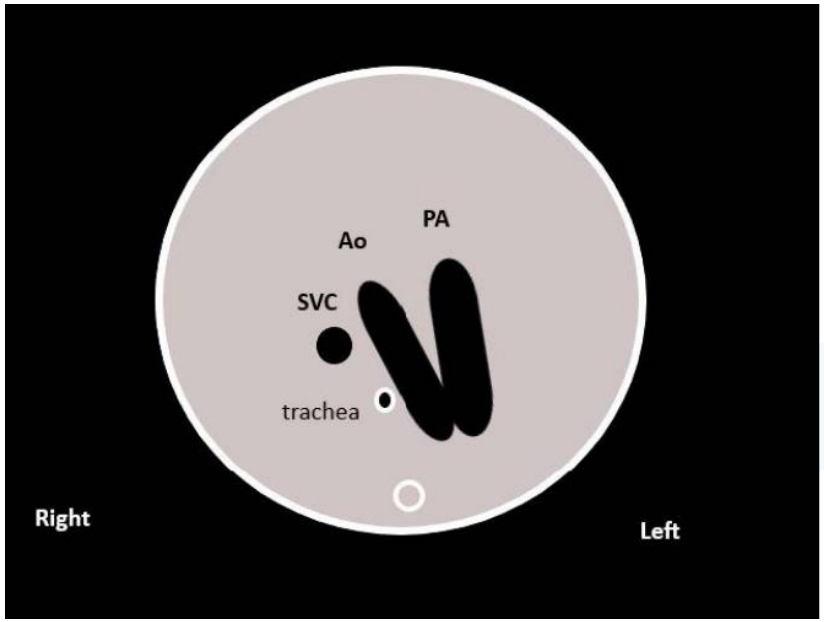
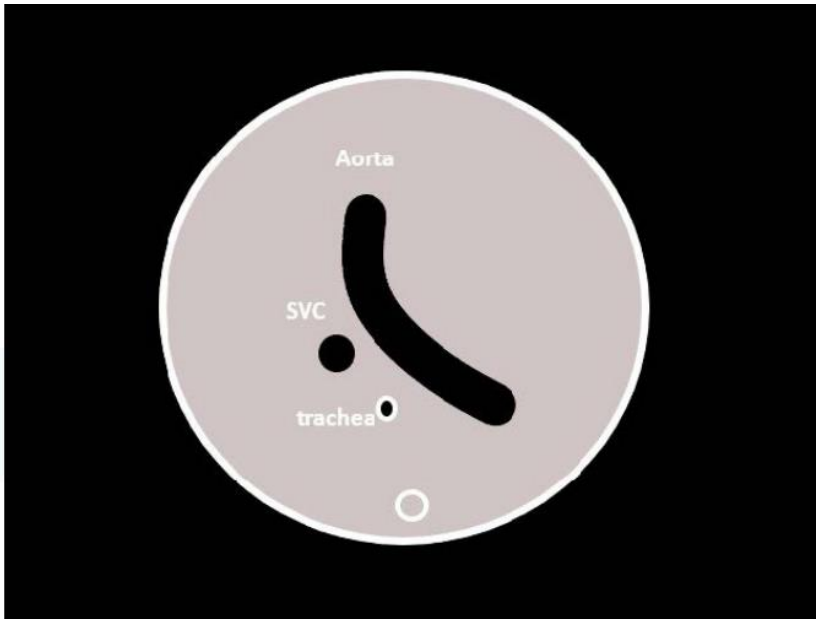
Abnormalities in LVOT/RVOT views – Transposition of the Great Arteries





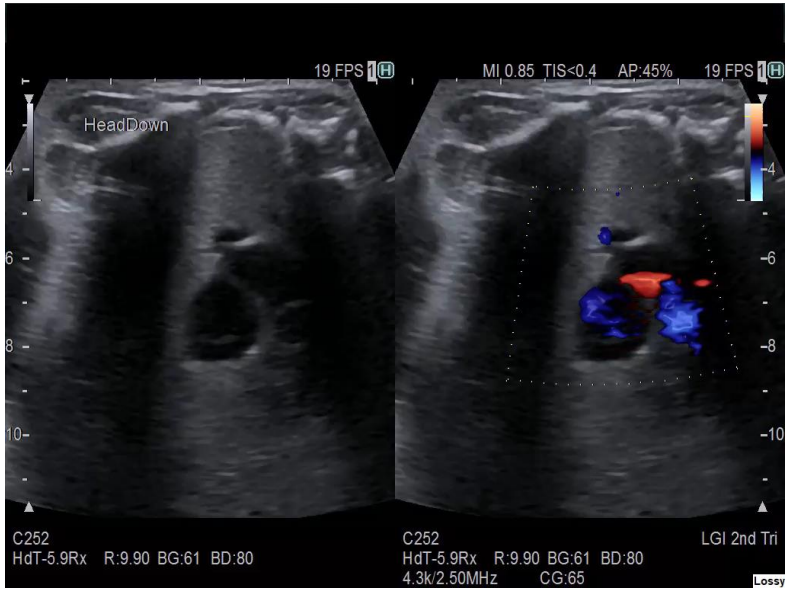
Transposition of the Great Arteries





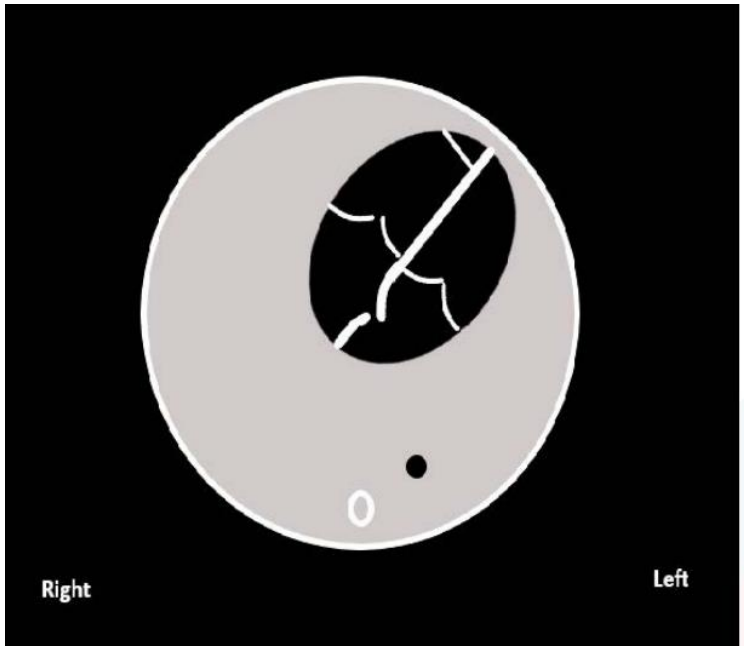
Transposition of the Great Arteries

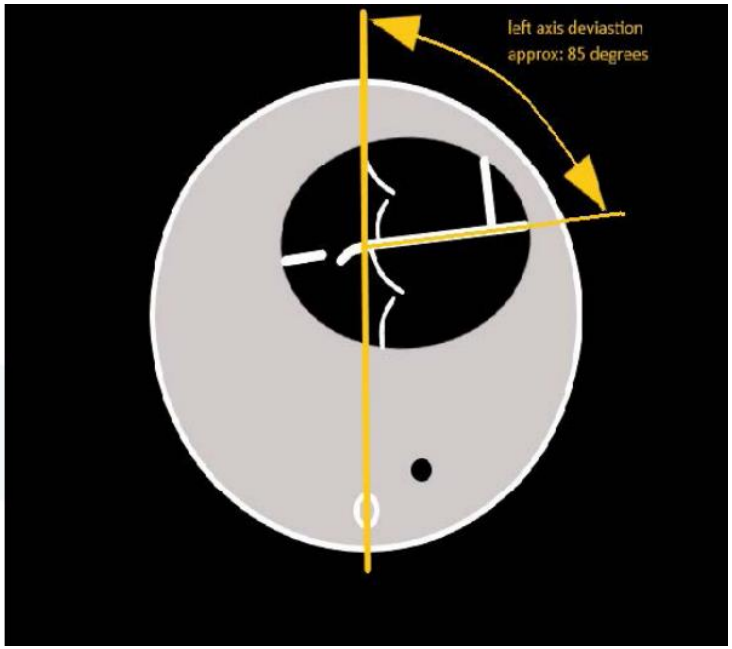
- 3.5 of 10,000 live births [1]
- 5-7% of all CHD
- 2nd most common cyanotic lesion
- 'Simple TGA' – no VSD
  - Very rare to have chromosomal abnormalities



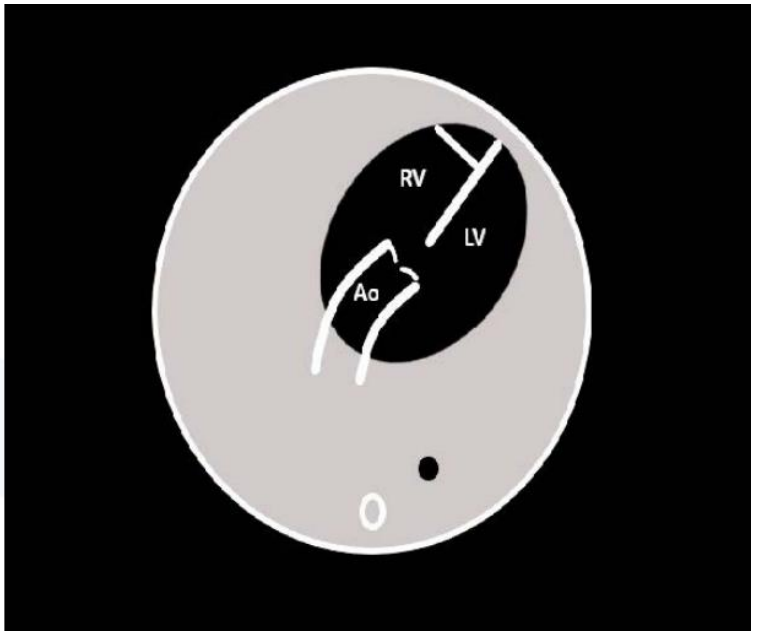
- In absence of other major structural abnormalities will require arterial switch operation in first few weeks of life
  - Up to 50% require a balloon atrial septostomy prior to surgery due to inadequate mixing

Abnormalities in LVOT/RVOT views – Tetralogy of Fallot





Tetralogy of Fallot



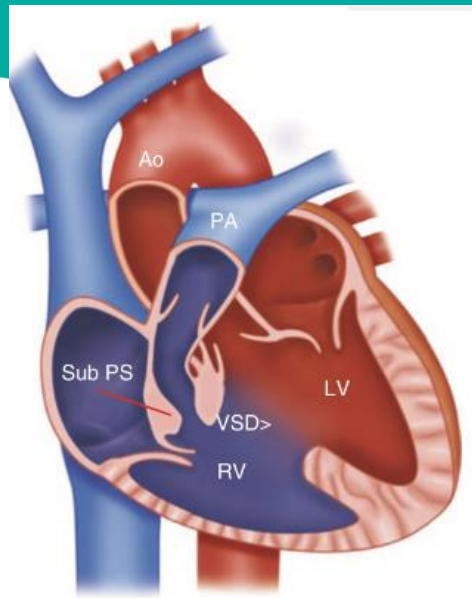
Tetralogy of Fallot

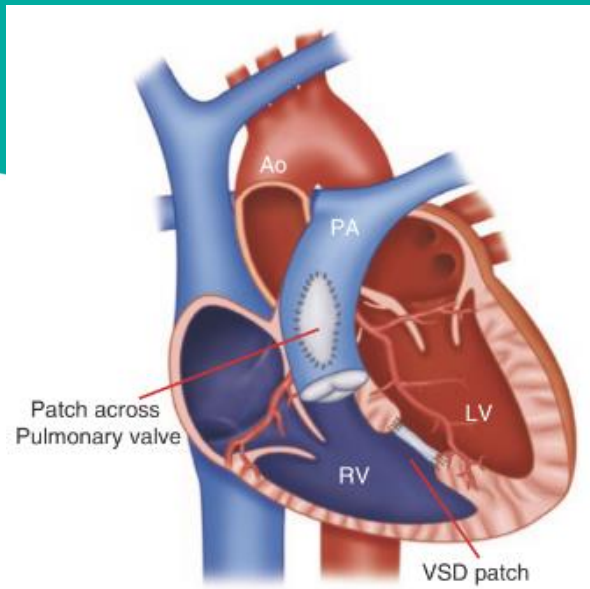
- 4 Chamber view – Left axis deviation
- LVOT view – Sub aortic VSD with aortic override
- RVOT view – Pulmonary stenosis (degree varies)
- 3VT view – PA smaller than arch and can be associated with right aortic arch

Tetralogy of Fallot

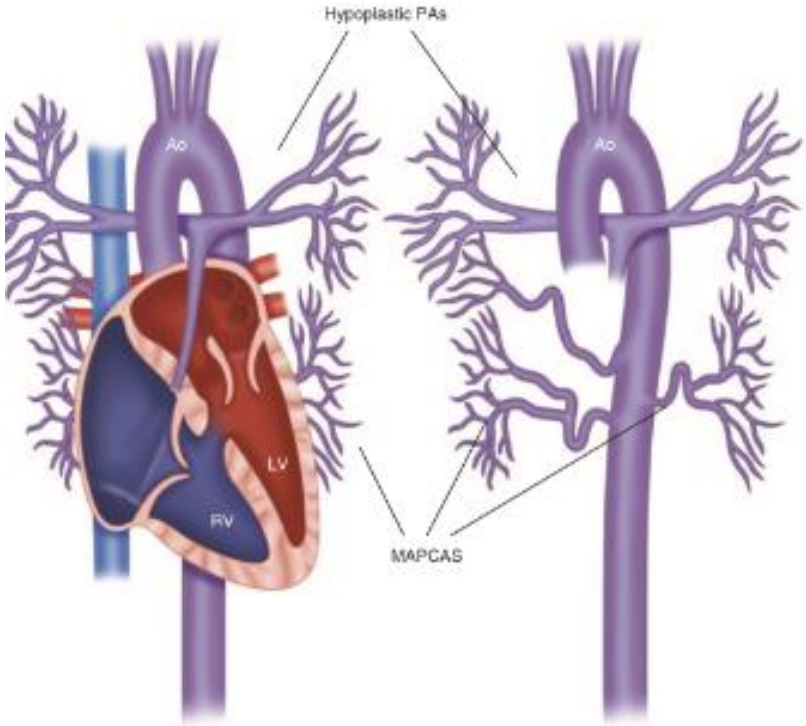
- Most common cause of cyanotic congenital heart disease
  - Prevalence 3-6 per 10,000
- Genetic/chromosomal associations in 30%
  - Most common Trisomy 21 and 22q11 deletion
- Varying degree of pulmonary obstruction
  - Minimal stenosis – act like a VSD
  - Pulmonary atresia – duct dependent pulmonary circulation
  - Absent pulmonary valve

Tetralogy of Fallot with PS – Transannular patch surgical repair





Pulmonary Atresia, VSD, MAPCAs – would require unifocalisation surgery



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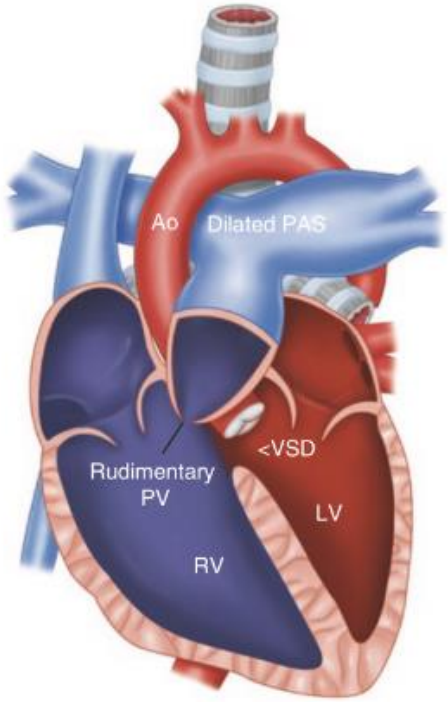
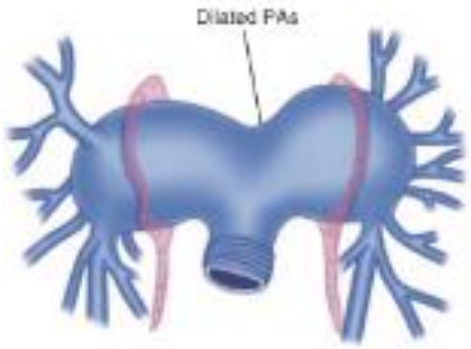
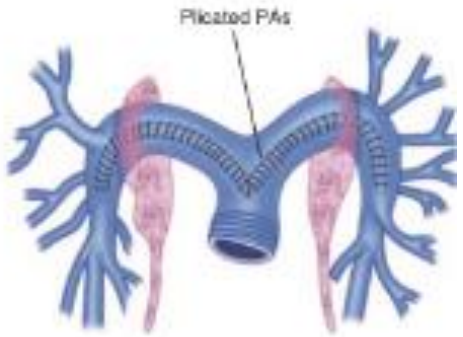
Tetralogy of Fallot –
Absent Pulmonary Valve







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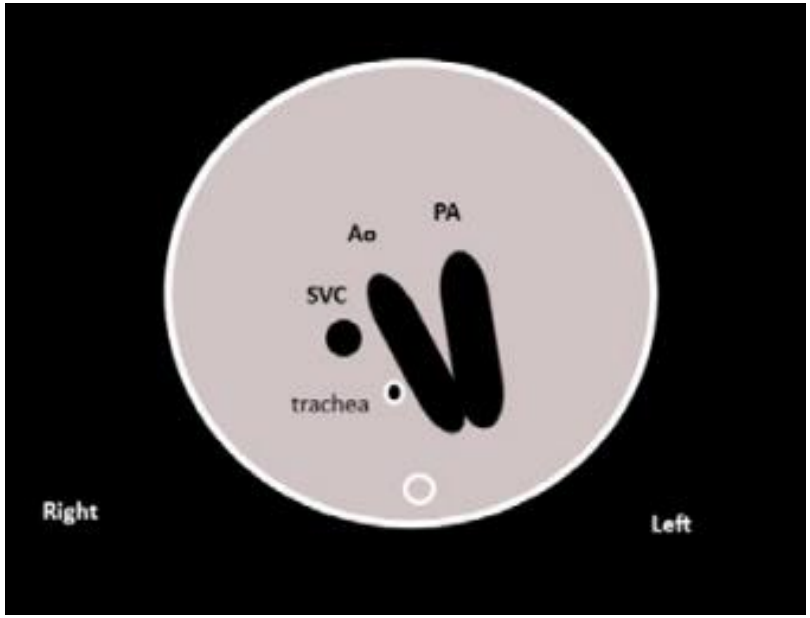
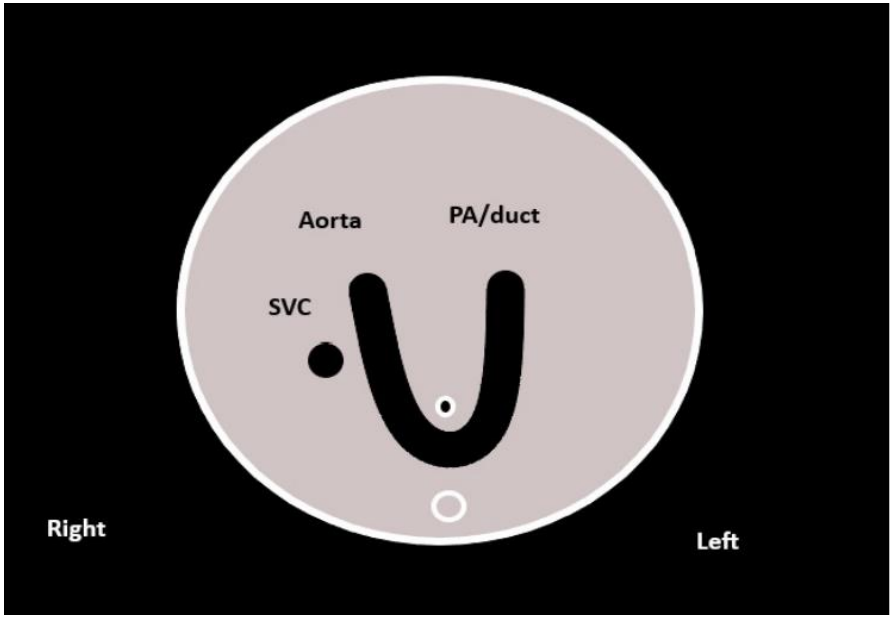
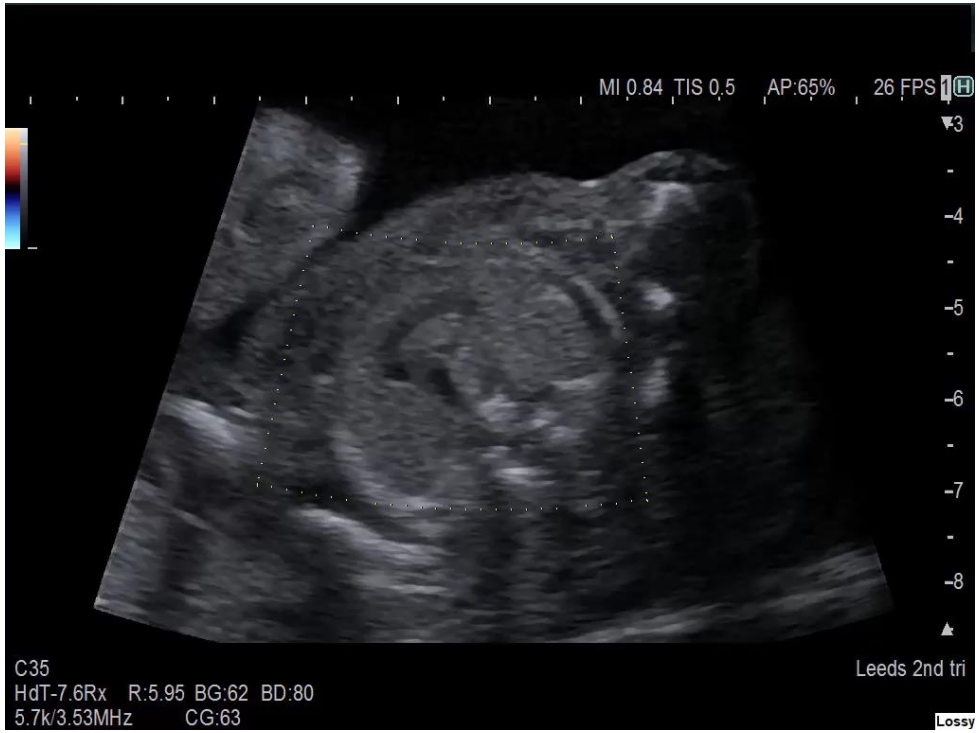
Abnormalities in 3VT view – Right Aortic Arch





Right Aortic Arch

- Aorta passes to right of trachea and oesophagus and ductus arteriosus to left
  - Encircle central structures
  - Risk of compression to these structures
- Most common branching pattern is with aberrant left subclavian artery – 86% in recent review [3]
- Symptoms occur in 25-30%
- Genetic associations in 8% [4]



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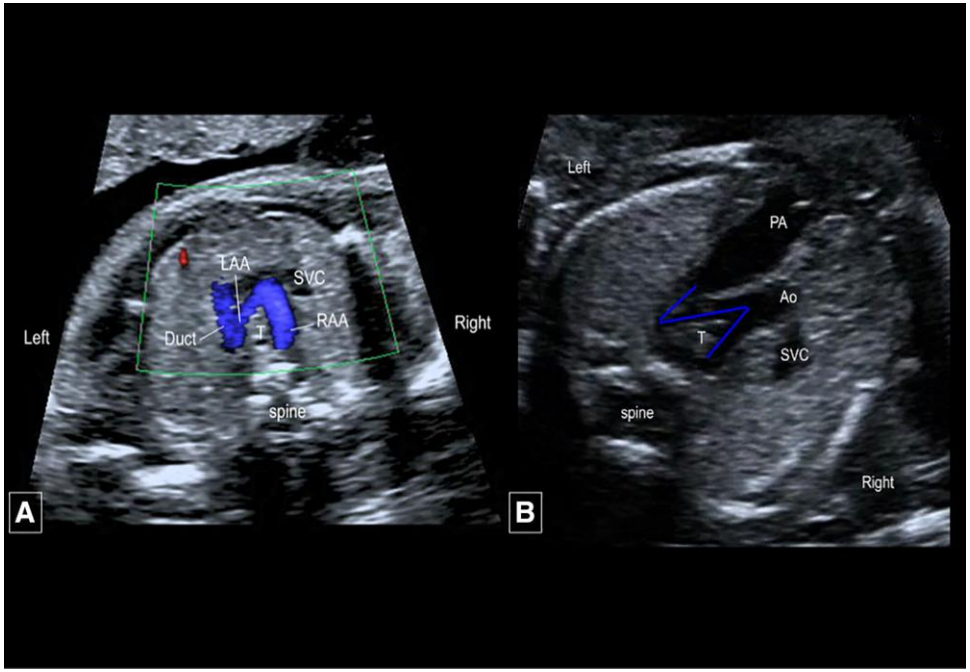
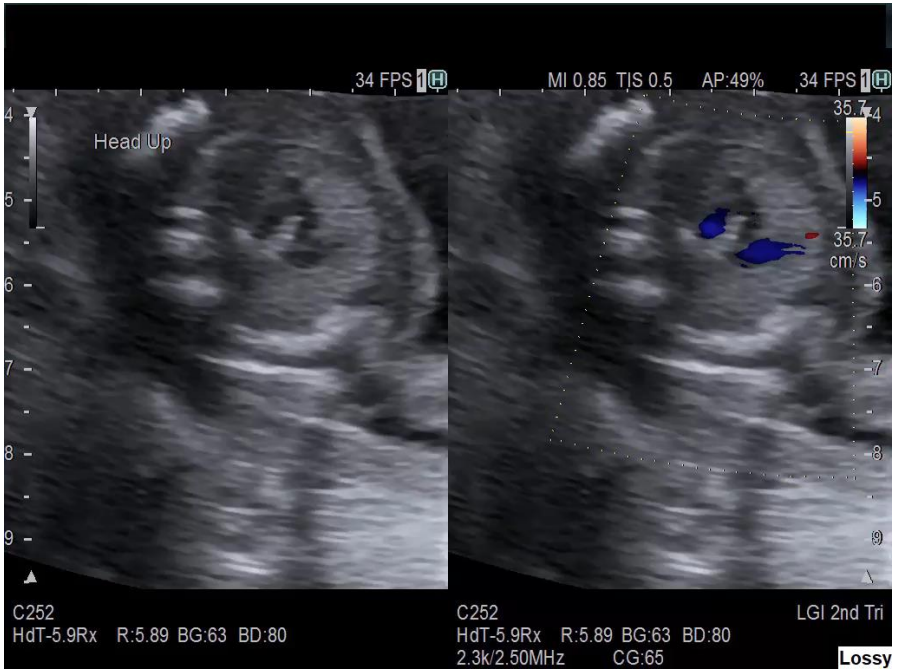
Double Aortic Arch





Van Poppel MPM, Zidere V, Simpson JM, Vigneswaran TV. Fetal echocardiographic markers to differentiate between a right and double aortic arch. Prenat Diagn. 2022 Apr;42(4):419-427. doi: 10.1002/pd.6104. Epub 2022 Jan 28. PMID: 35060138

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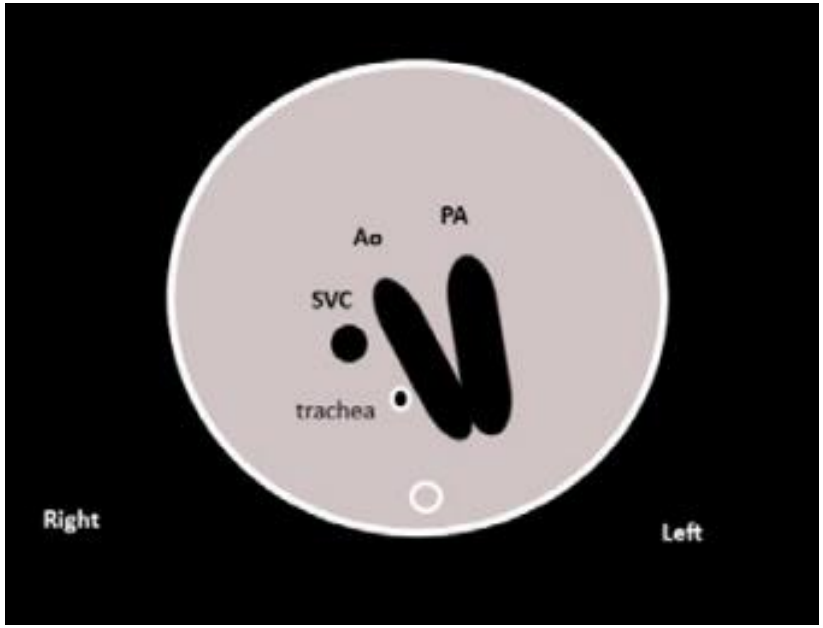
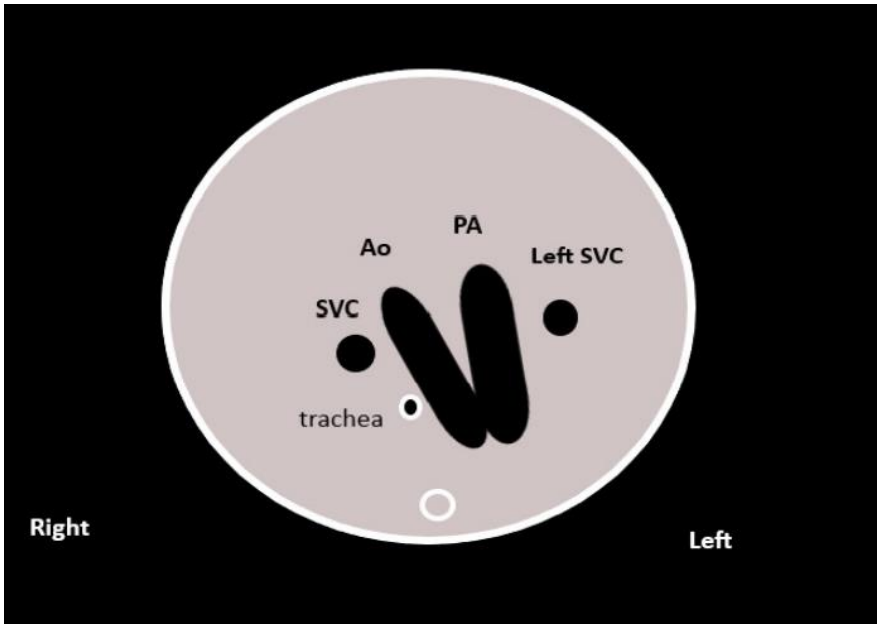
Abnormalities in 3VT view – Bilateral SVCs

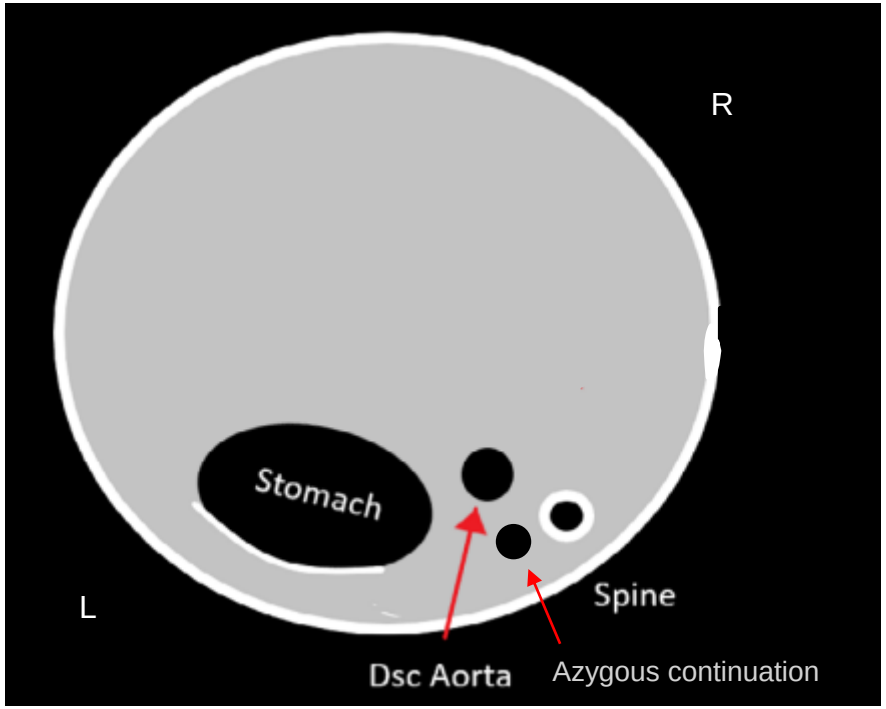


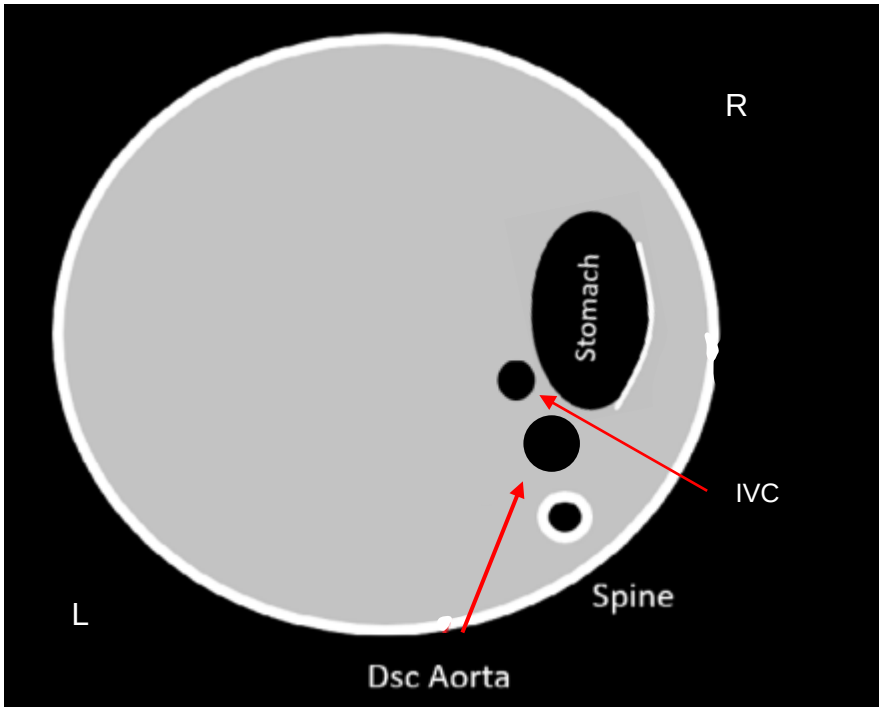


Bilateral SVCs

- Persistent left SVC
  - Left SVC drains into the coronary sinus which therefore appears dilated
  - Right SVC drains in normal position into the right atrium
- Possible association with coarctation of the aorta
- Otherwise usually of no cardiac concern if isolated

Abnormalities of Situs View





Laterality Disorders/Isomerism/Heterotaxy Syndromes

- During development complex series of signalling arrangement to delineate left from right
  - Poorly understood
- Abnormalities with this causes laterality disorders
  - Proliferation of left or right sided structures
  - Absence of the opposite sided structures

Laterality Disorders/Isomerism/Heterotaxy Syndromes

	Usual Atrial arrangement	Left Atrial Isomerism	Right Atrial Isomerism
Pulmonary Veins	To left atrium	Usually normal	Anomalous drainage
Systemic Veins	Single right SVC & IVC	Absent IVC with azygous continuation	Often bilateral SVCs and IVC same side as aorta
Intracardiac	Normal	Can be normal. Otherwise variable	Abnormal – common are AVSD, DORV, Pul stenosis/atresia, TGA, apex left or right
Liver	Right sided	Midline	Midline
Spleen	Left sided	Polysplenia	Asplenia
Bowel	Stomach left	Stomach right with possible malrotation	Stomach right with possible malrotation
Rhythm	Sinus	CHB/slow sinus	Normal

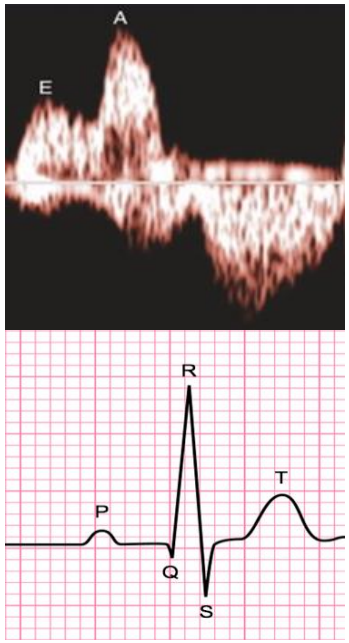
Laterality Disorders/Isomerism/Heterotaxy Syndromes

- All laterality disorders from 1980-2017 [5]
- 177 LAI, 100 RAI
- Major structural heart disease in all RAI and 91% of LAI. 40% LAI had CHB
- If CHB present, median postnatal survival was 0.2 months

5 year Survival	1980-1989	1990-1999	2000-2009	2010-2017
Left Isomerism	0%	54%	53%	75%
Right Isomerism	0%	62%	59%	67%

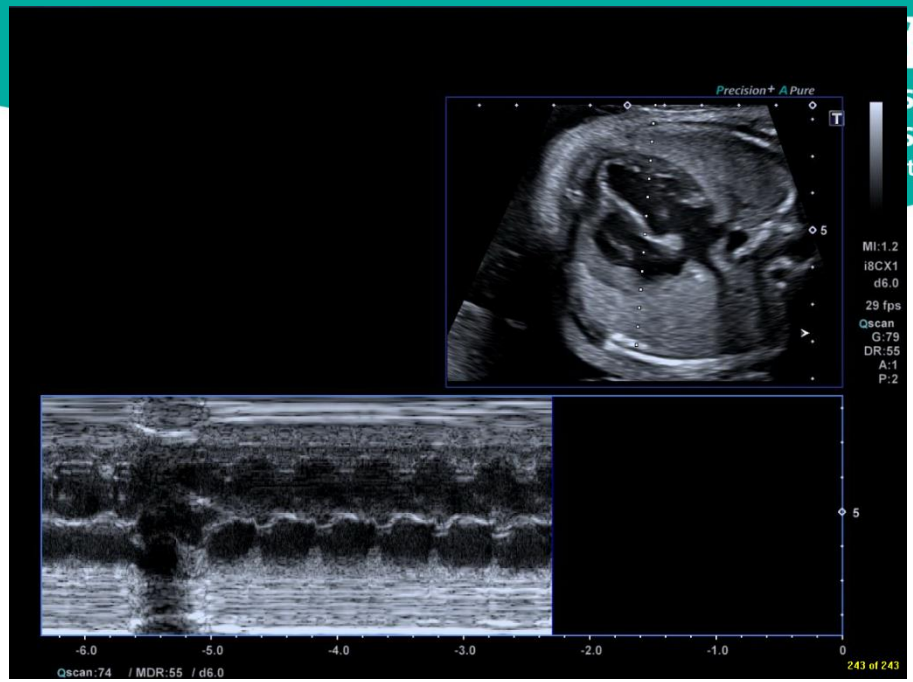
Assessing Fetal Rhythm

- Uses mechanical events to ascertain temporal relationship between electrical events
  - I.e Atrial and ventricular contractions

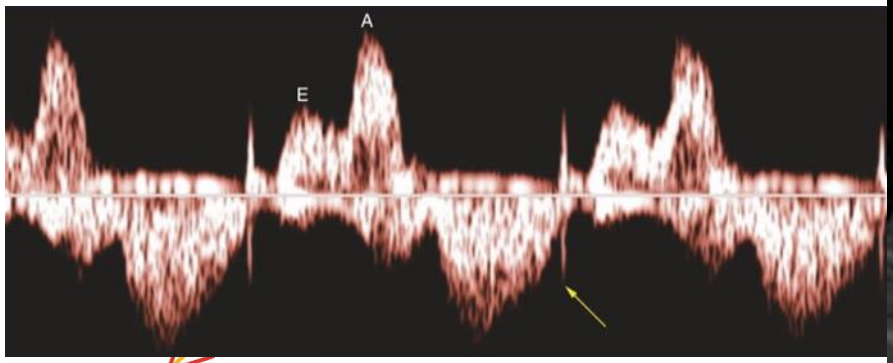


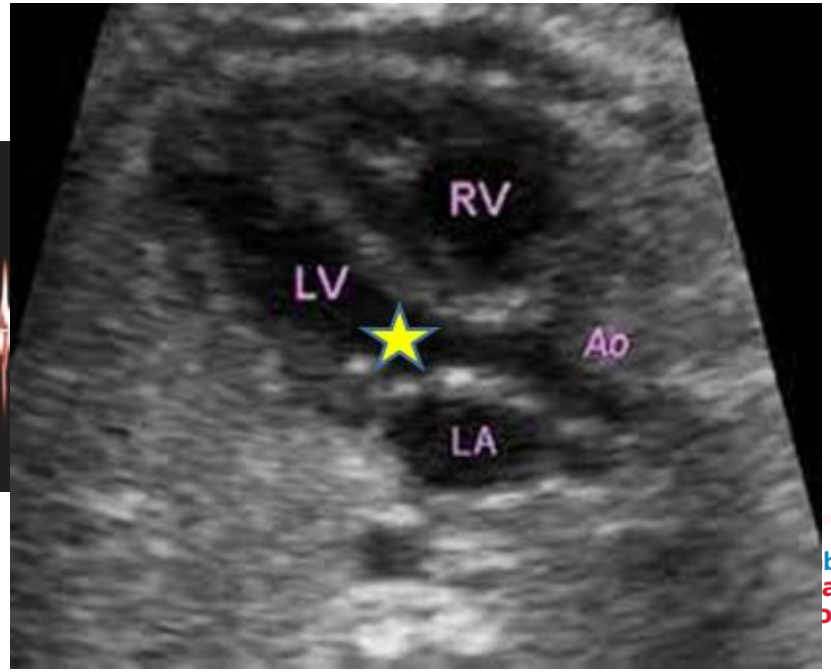
Techniques

- M Mode



- LV inflow/outflow





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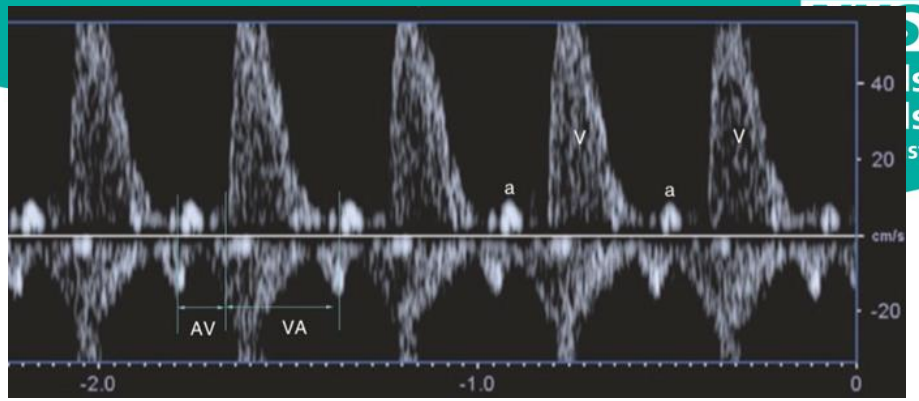
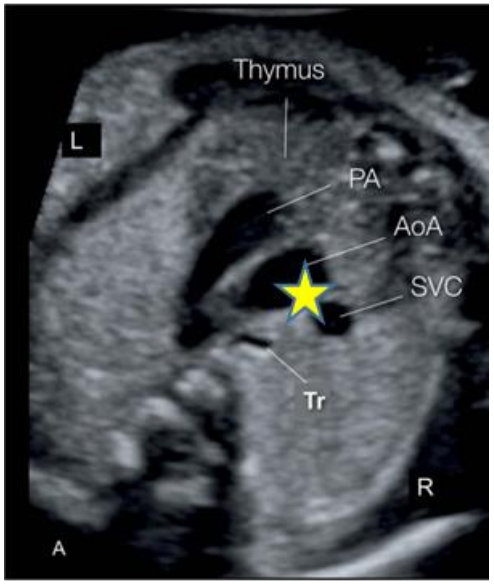
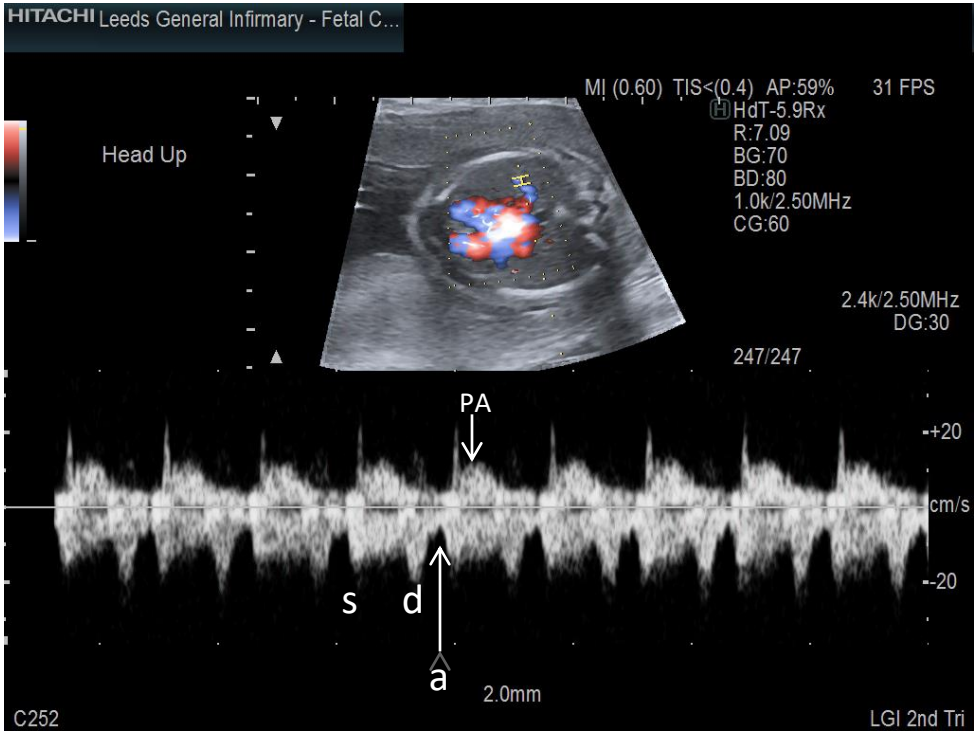
Techniques

- SVC/aortic flow
- Pulmonary vein/artery flow







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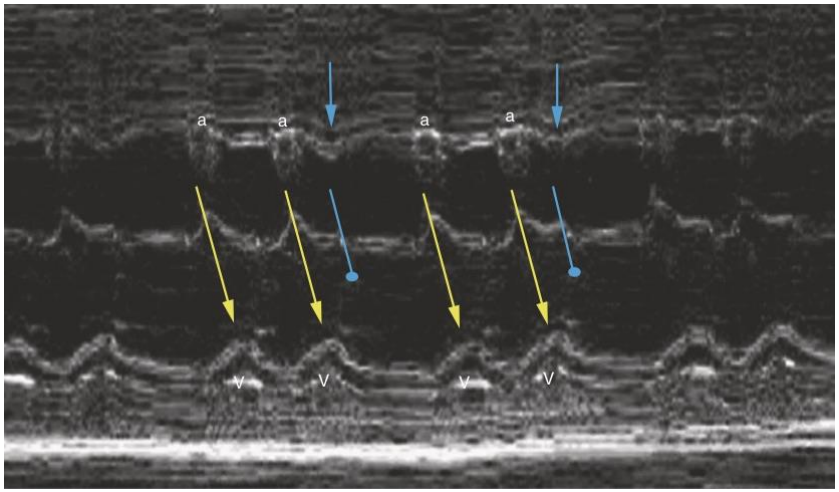
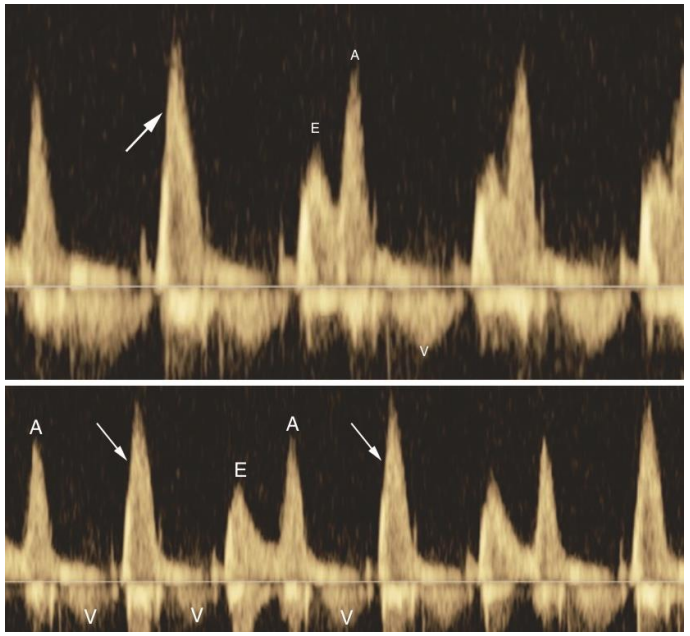
Atrial ectopics





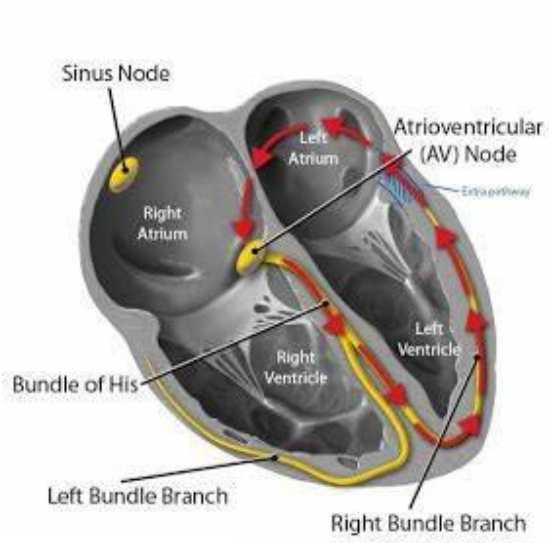
- No treatment required - most self resolve
- Small number (2-3%) go on to develop fetal tachycardia
- If >1 in 10 beats of ectopy, weekly/fortnightly local auscultation and referral if tachycardia present

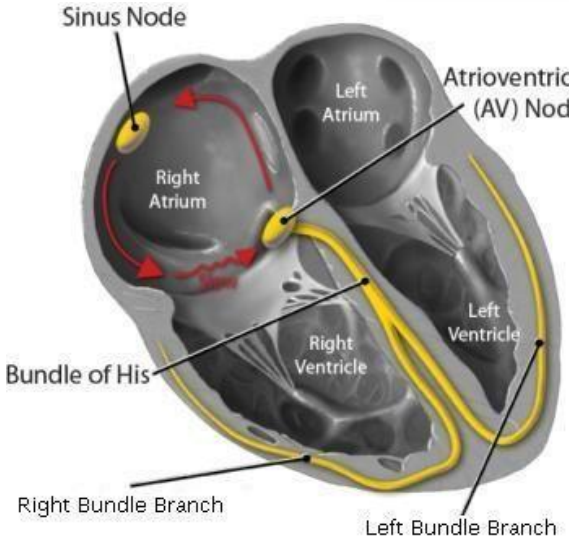
Fetal Tachycardias

- Sinus tachycardia
  - Consequence of another concern with fetus
    - Anaemia
    - Thyrotoxicosis
    - Fetal distress
    - Maternal fever
  - Rarely exceed 200bpm

Supraventricular Tachycardias

- Rate usually over 220bpm
- Most common re-entrant tachycardia
  - 1:1 relationship
- Atrial flutter
  - Loss of 1:1 relationship





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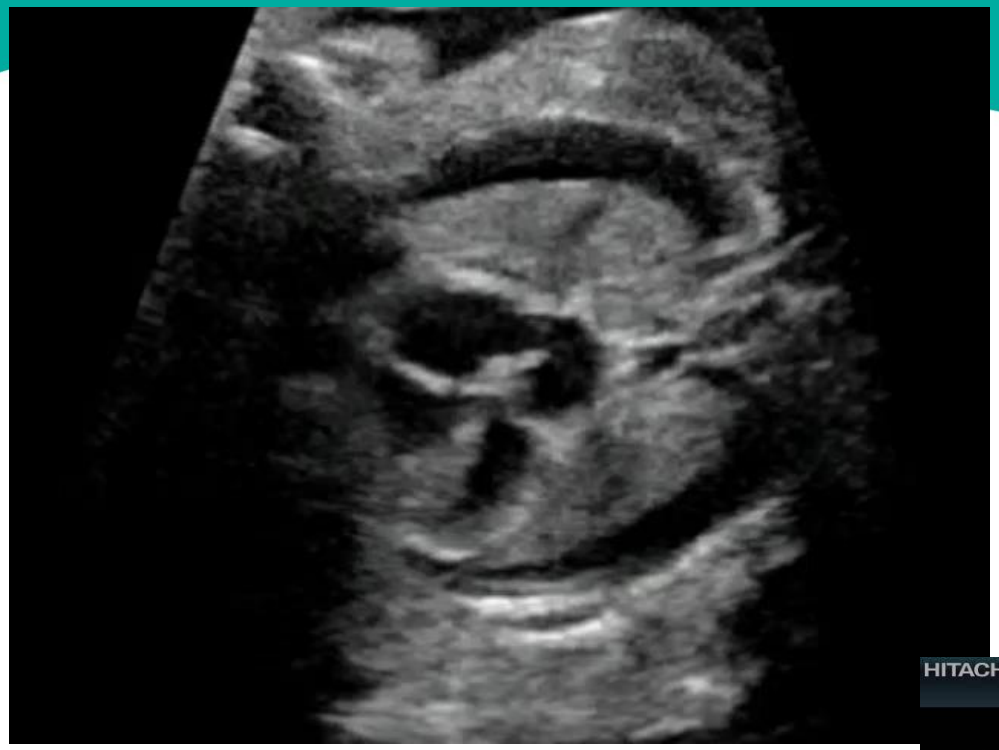

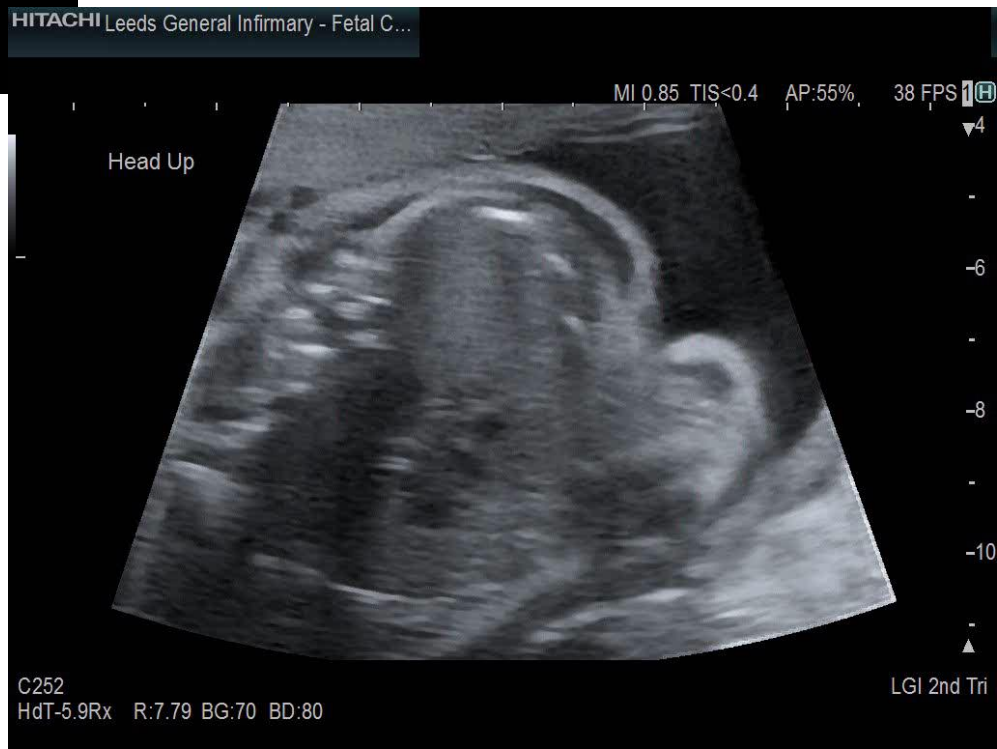

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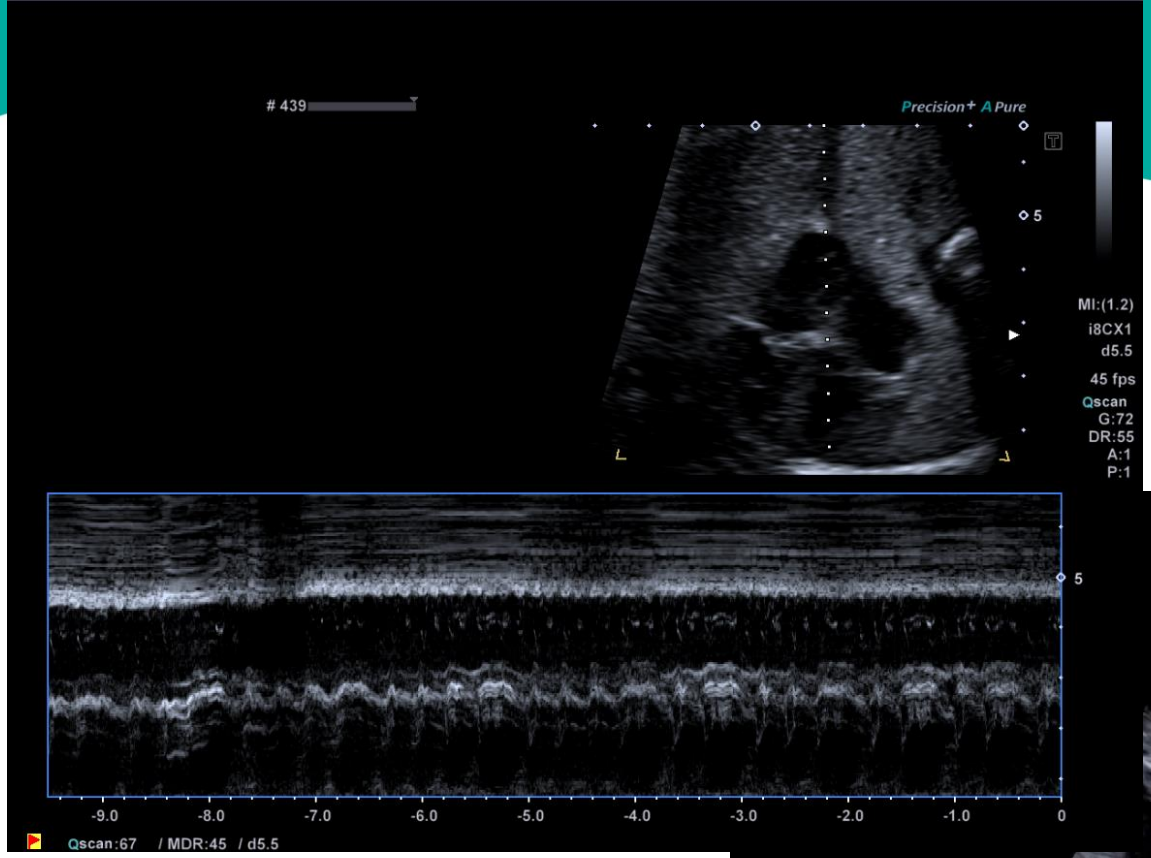

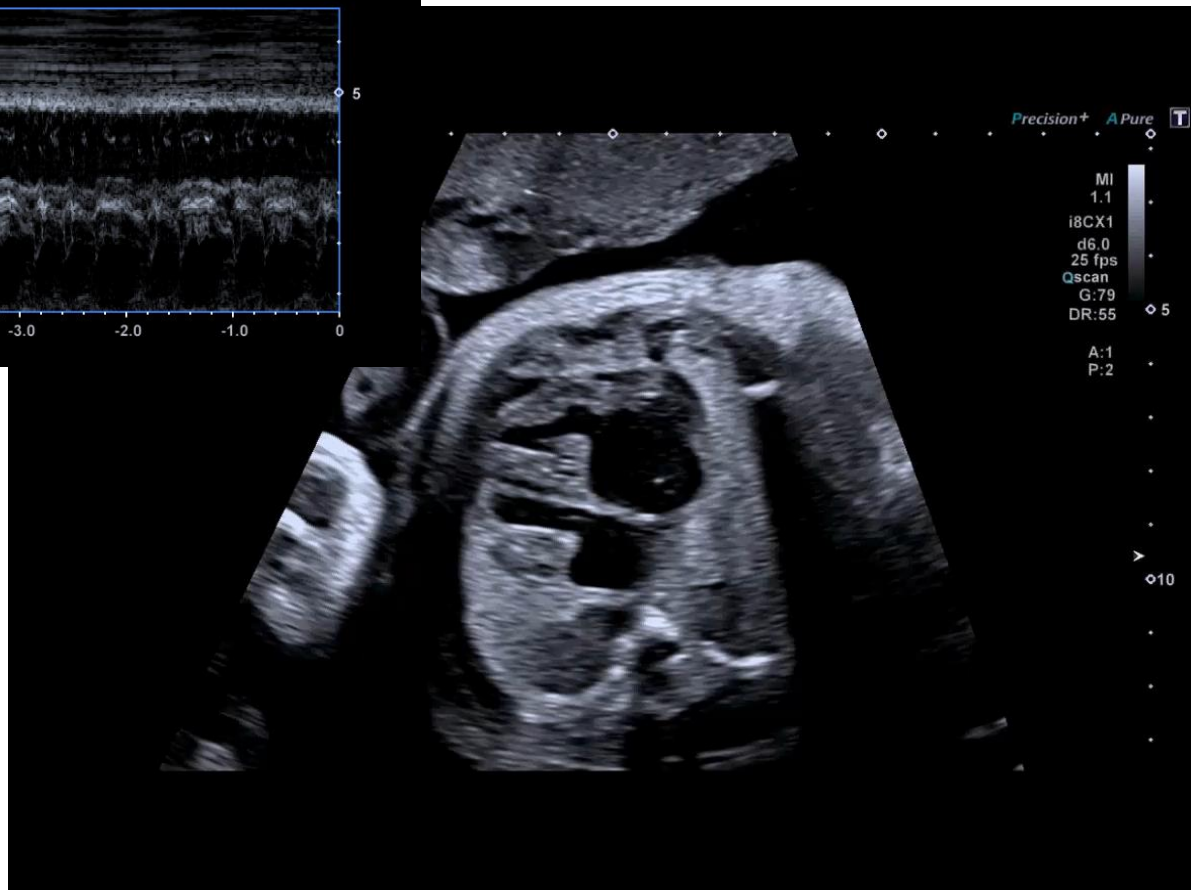

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Fetal Bradycardia – Complete Heart Block

- Can be associated with:
  - Maternal anti-Ro and anti-La antibodies
  - Left Atrial Isomerism
  - Congenitally Corrected Transposition of the Great Arteries (ccTGA)
- Maternal anti-Ro or anti-La present
  - 2% risk of congenital complete heart block
  - If previous pregnancy affected – risk increases to 20%

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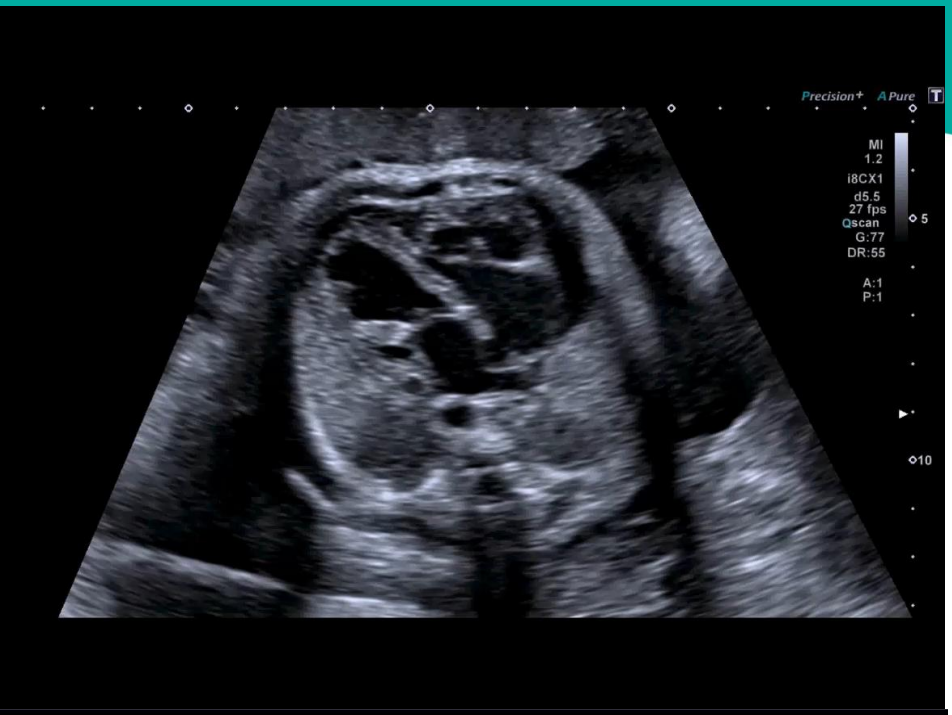

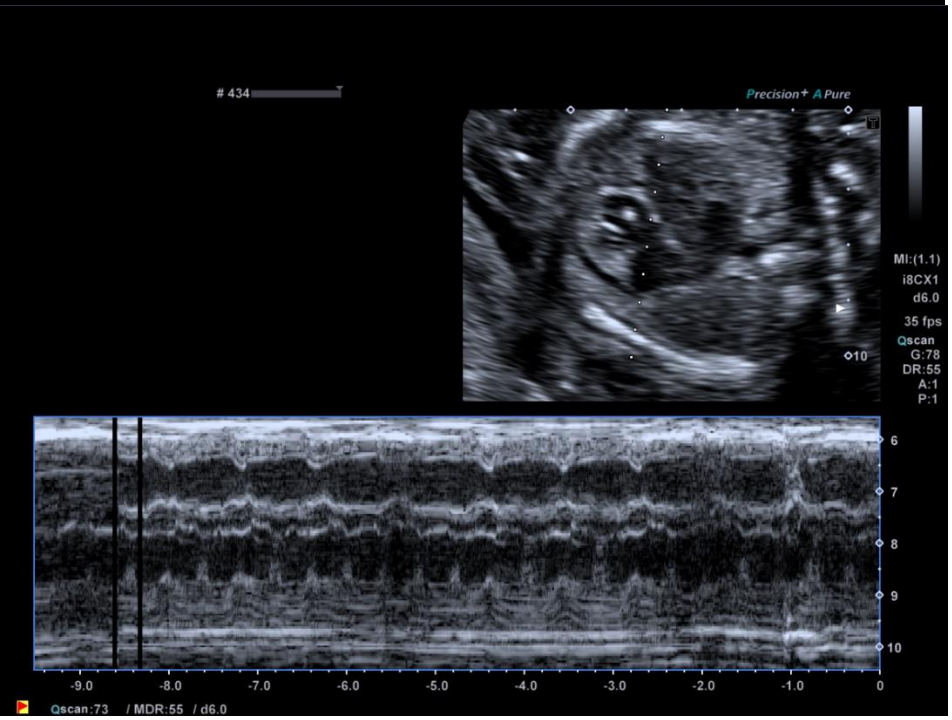

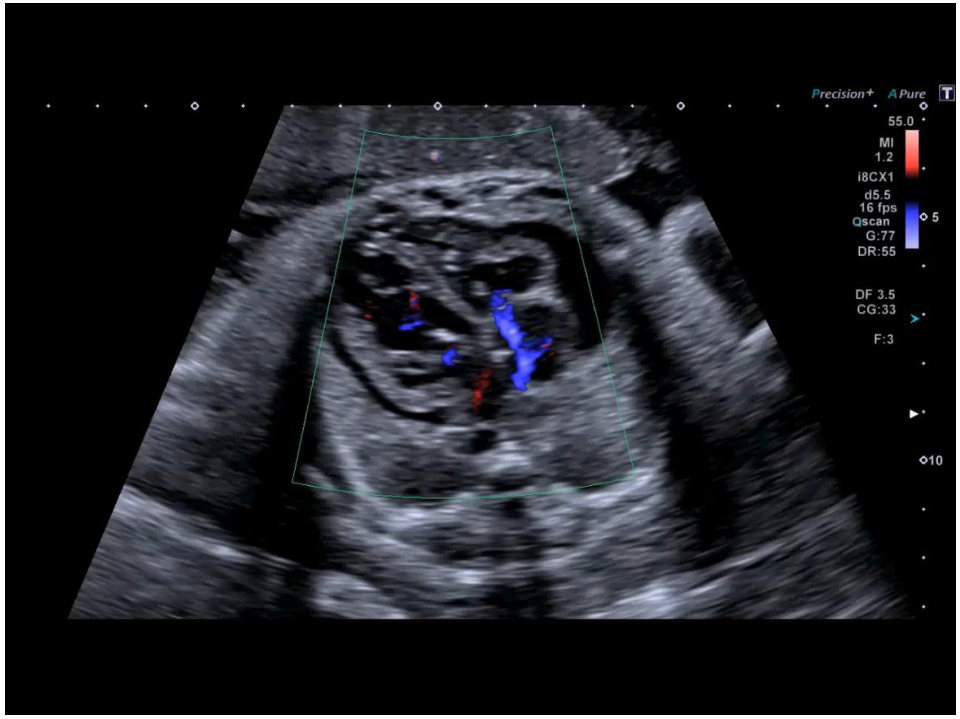

Acknowledgements and References

- Pictures from Leeds Teaching Hospitals and Yorkshire and Humber Congenital Heart Disease Network Fetal Heart Training Resource created by Sam Bainbridge and Jan Forster

- 1) Simpson J, Zidere V, Miller O et al. Fetal cardiology a practical approach to diagnosis and management. Springer
- 2)Lillitos P, Nolan O, Cave D et al. The fetal single ventricle journey to first postnatal procedure. A multicentre UK cohort study – to be published
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- 4) Seale A, Vigneswaran T, Hunter L et al. PRAADA: Prenatal Right Aortic Arch and Double Arch: A Population-based Study – to be published
- 5) Vigneswaran TV, Jones CB, Zidere V, Charakida M, Miller OI, Simpson JM, Sharland GK. Effect of Prenatal Laterality Disturbance and Its Accompanying Anomalies on Survival. Am J Cardiol. 2018 Aug 15;122(4):663-671. doi: 10.1016/j.amjcard.2018.04.040. Epub 2018 Jun 20. PMID: 29954599

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