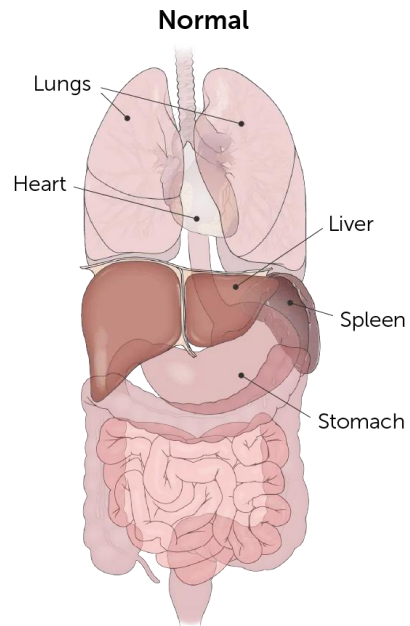
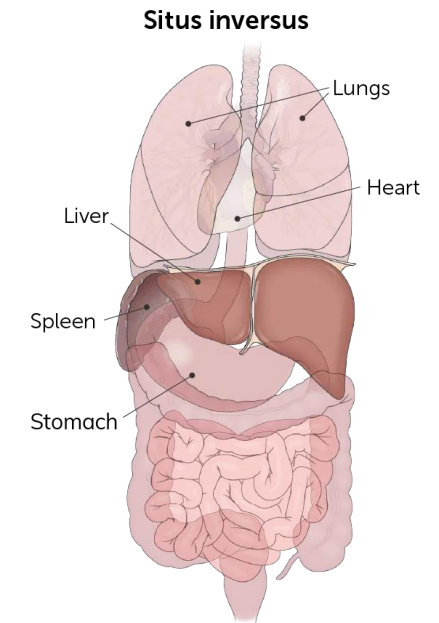


BASIC SITUS ASSESSMENT



DR MOHD AMIN ITAM
CONSULTANT PAEDIATRIC CARDIOLOGIST
HOSPITAL SULTAN IDRIS SHAH SERDANG



WHAT IS SITUS?

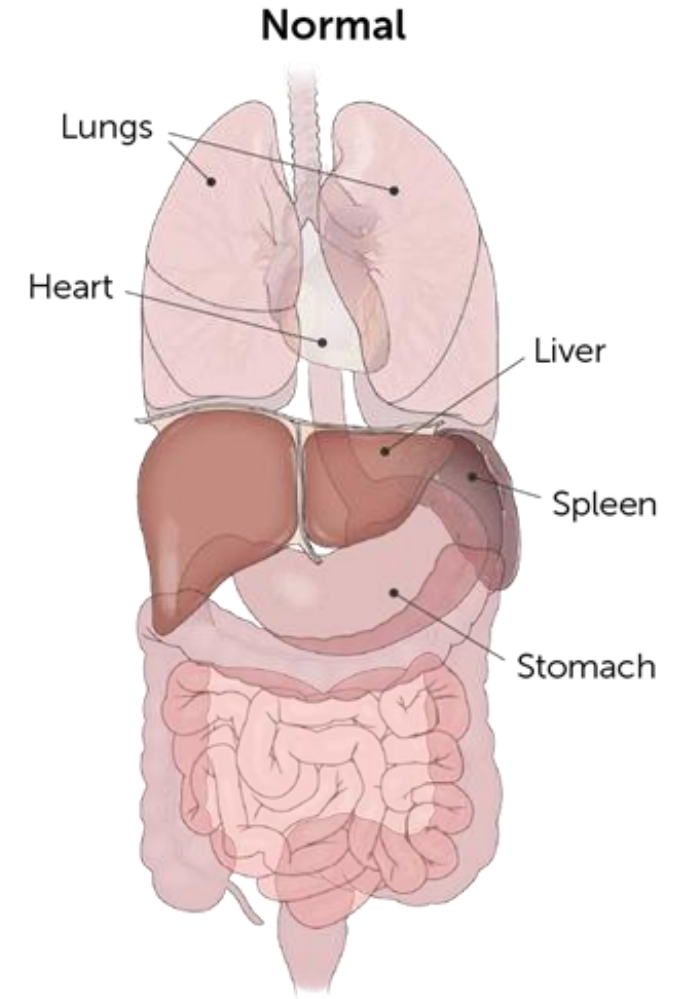
- **Visceroatrial situs** refers to the **position** and **configuration**:
 - *Cardiac atria*
 - *Tracheobronchial tree*
 - *Abdominal viscera.*
- **Situs solitus** (from Latin 'usual site') is referring to the ***normal position*** of thoracic and abdominal organs.

IS SITUS IMPORTANT ?

- Abnormalities of viscerocardiac situs are often associated with complex congenital heart disease.
- **Situs solitus** the risk of CHD is 0.8%.
- **Situs inversus totalis** the risk of CHD is 3–5%.
- **Situs ambiguous** has higher incidence of CHD
 - Right isomerism = CHD in 99–100% of cases.
 - Left isomerism = CHD in 75% of cases.

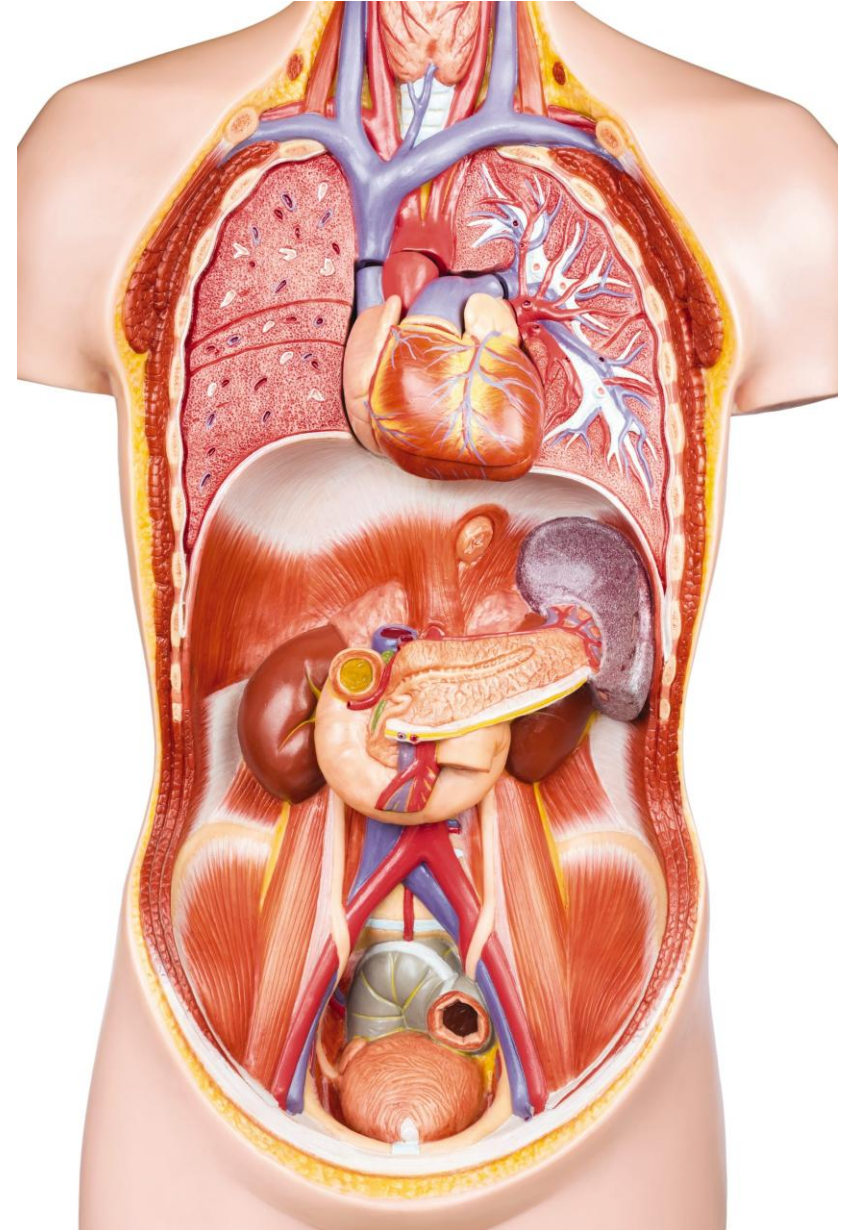
SITUS SOLITUS

- In humans, most of the thoracic and abdominal viscera are normally ***asymmetric and lateralized***.
- **Situs solitus** with levoverversion is the normal anatomic state, with
 - Trilobed right lung
 - Bilobed left lung
 - Morphologic right atrium is to the right
 - Morphologic left atrium is left-sided.
 - Normal intra abdominal organ arrangement

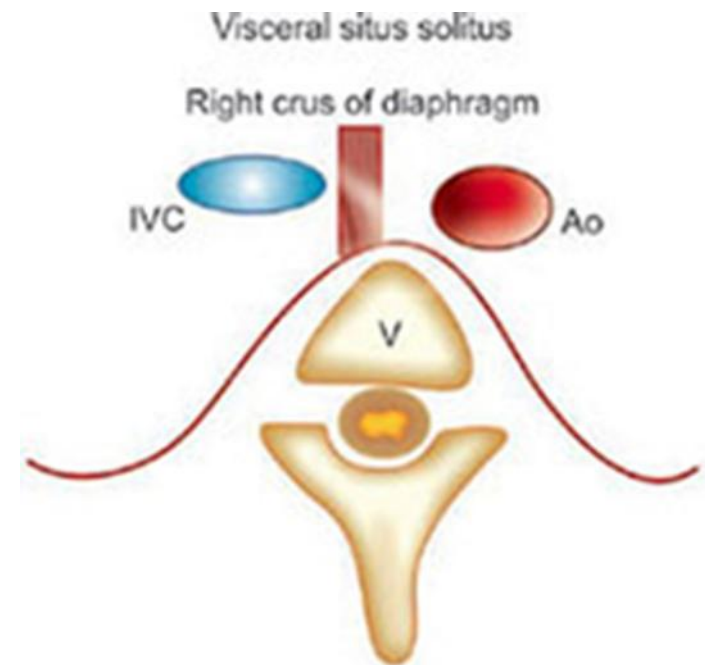


SITUS SOLITUS

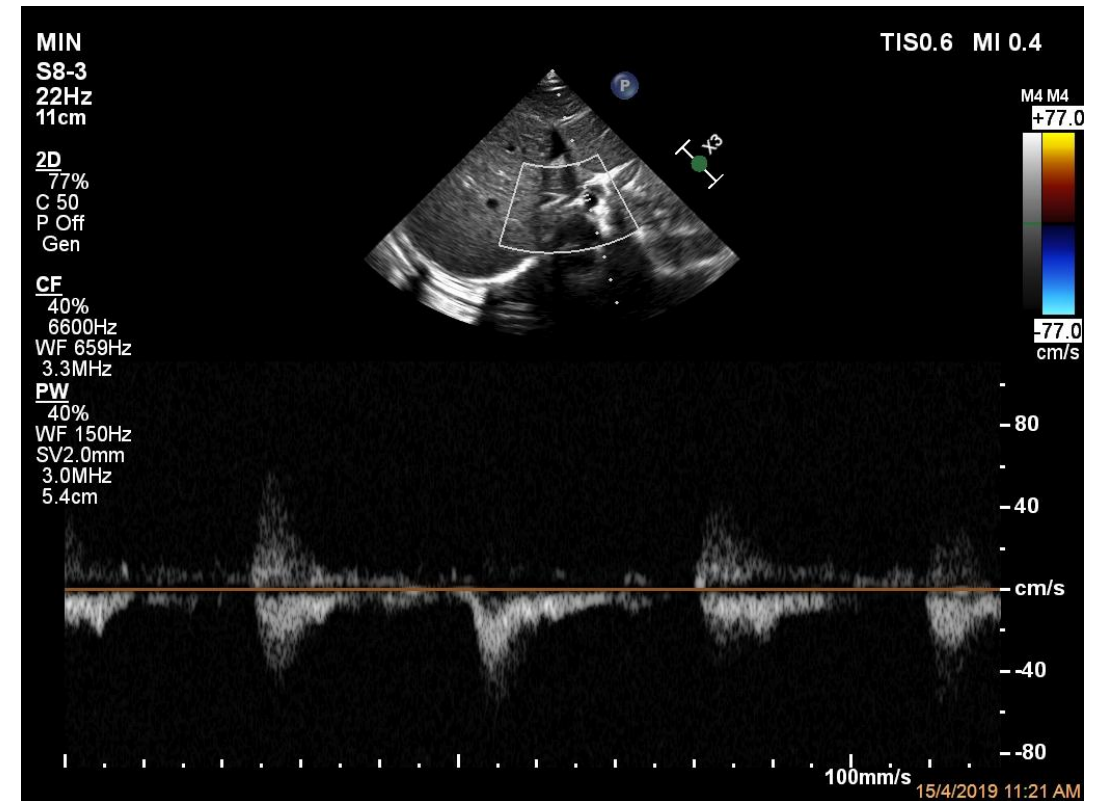
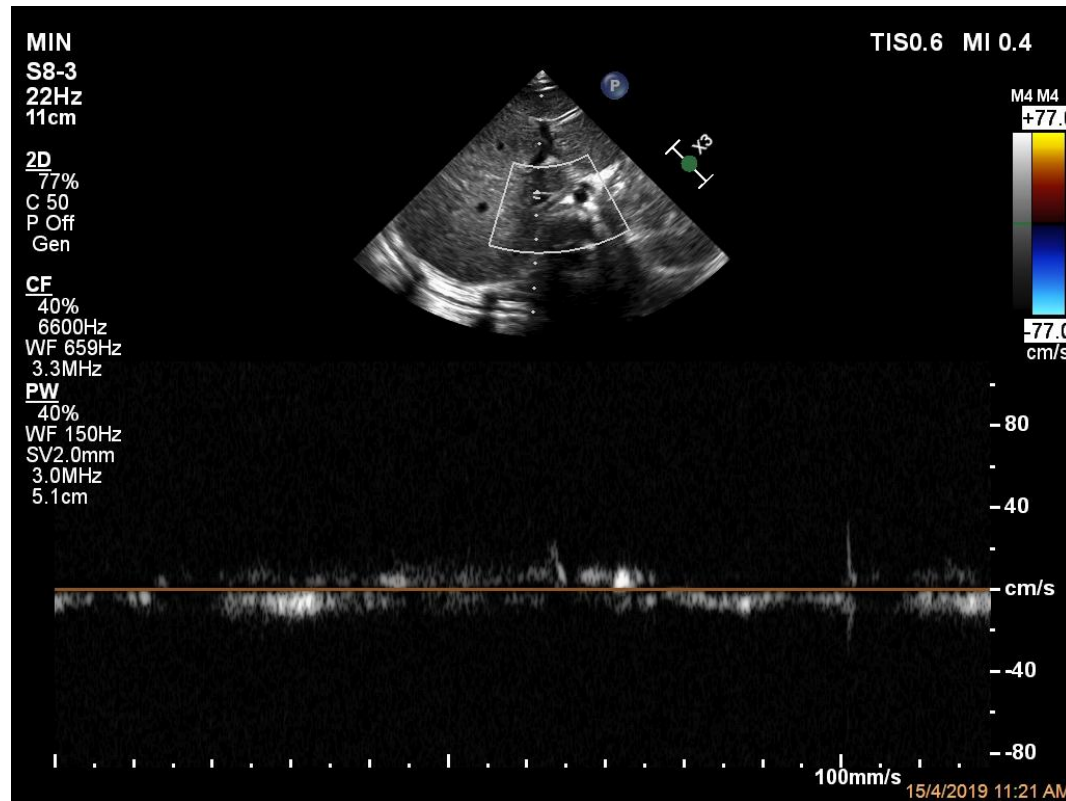
- Right-sided organs:
 - Trilobed lung
 - Liver
 - Gall bladder
 - Inferior vena cava
- Left-sided organs:
 - Bilobed lung
 - Stomach
 - Single spleen
 - Aorta



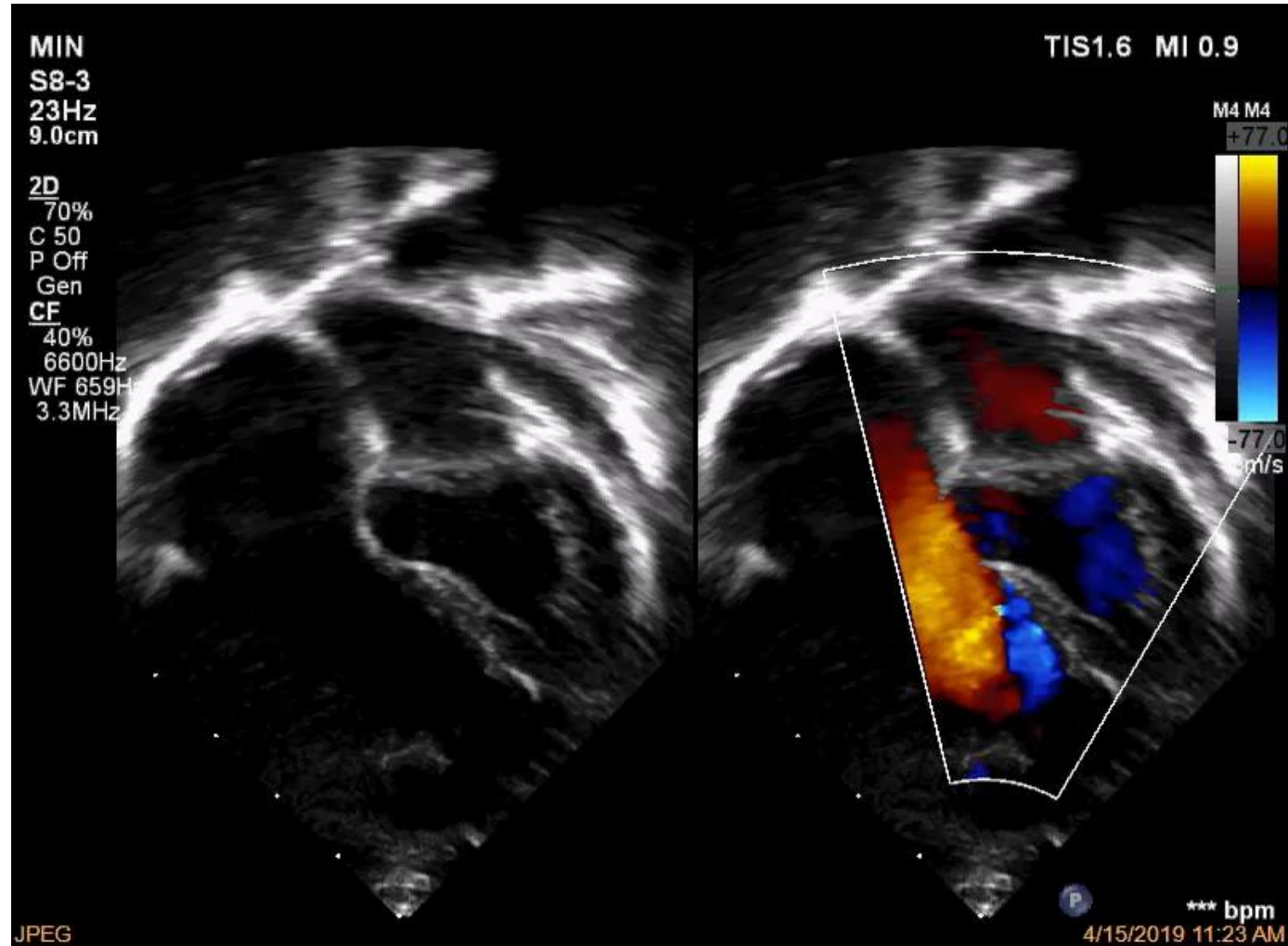
SITUS SOLITUS



SITUS SOLITUS



SITUS SOLITUS



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9.0cm

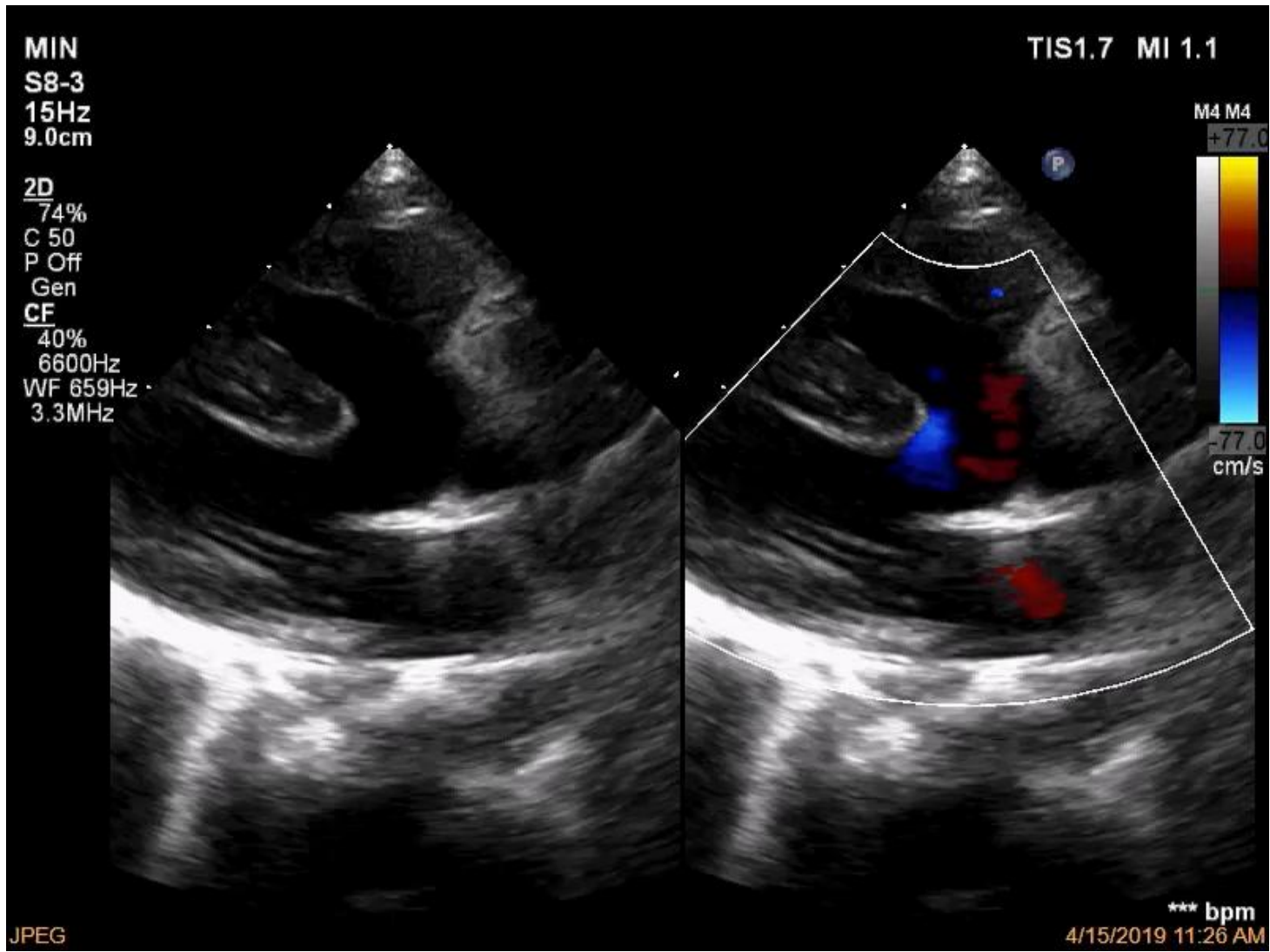
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70%
C 50
P Off
Gen
CF
40%
6600Hz
WF 659Hz
3.3MHz

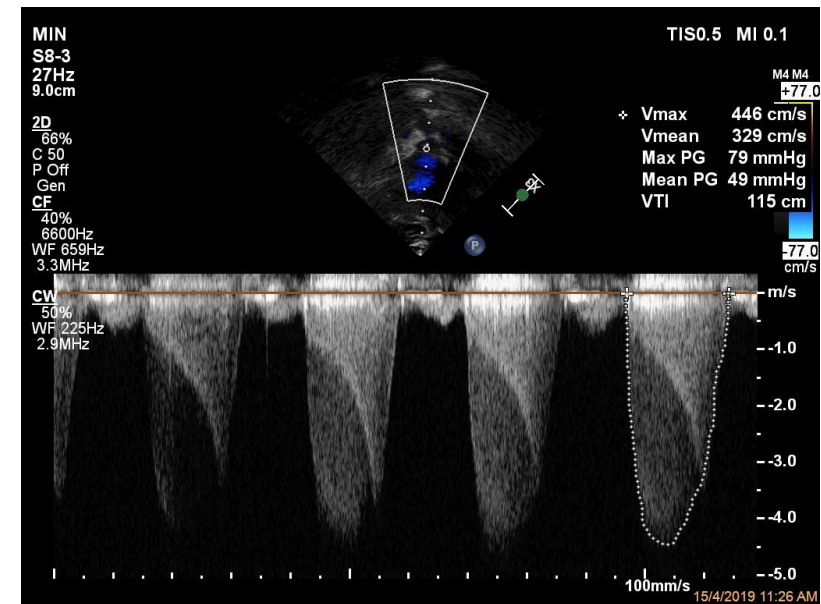
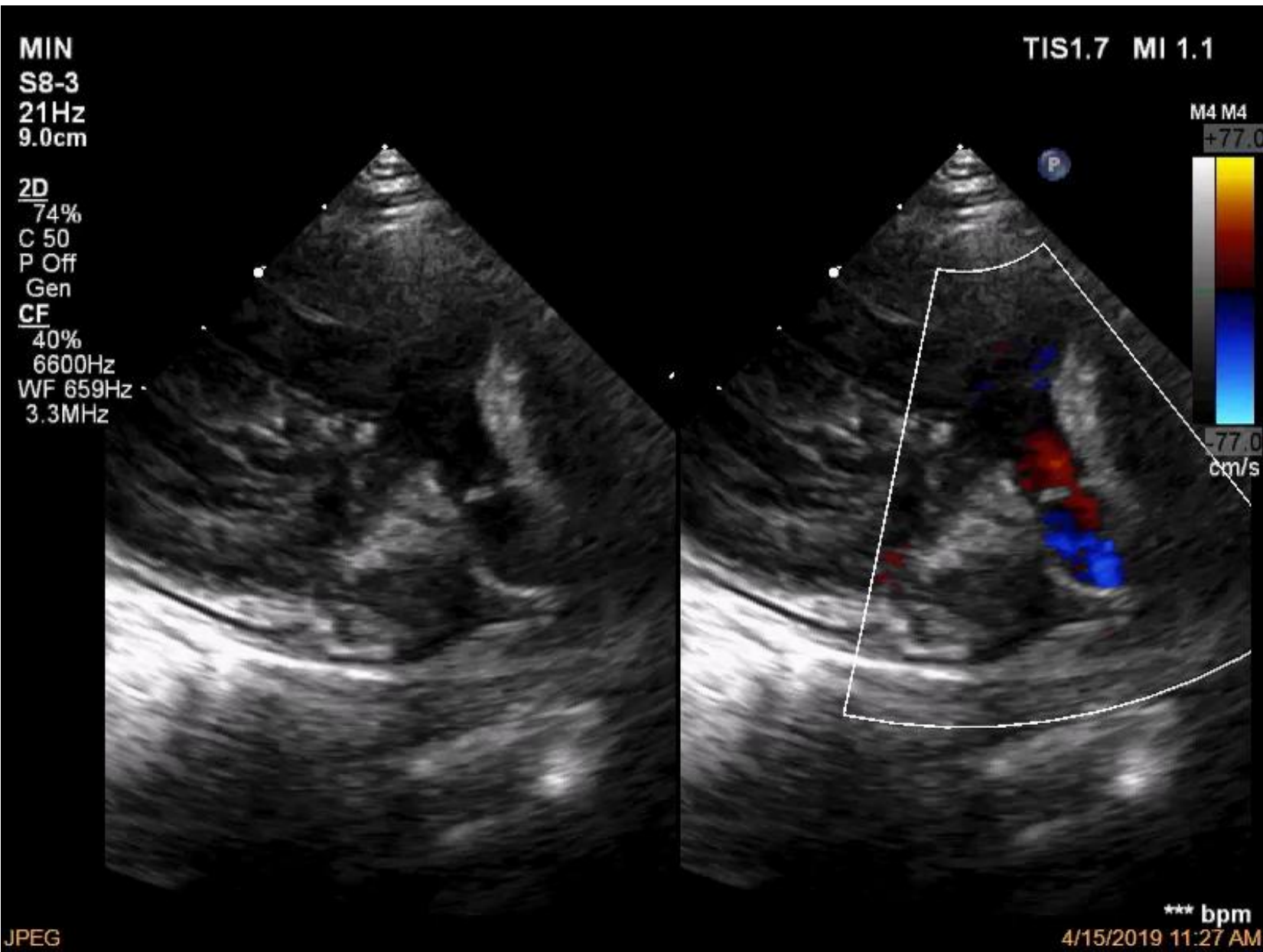
TIS1.7 MI 1.1

M4 M4
+77.0
-77.0
cm/s

JPEG

P *** bpm
4/15/2019 11:24 AM



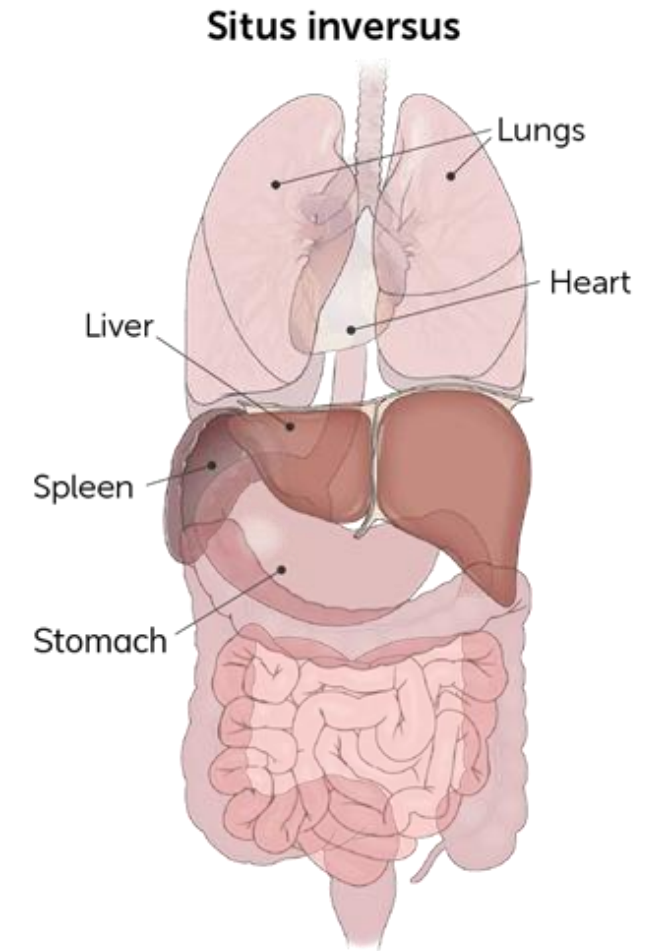


ECHOCARDIOGRAHY DIAGNOSIS

- Diagnosis:
- Situs solitus
- Tetralogy of Fallot
- Large PMVSD with outlet muscular extension
- Overriding of aorta
- Multilevel severe pulmonary stenosis
 - Infundibular, valvular and supra-ventricular stenosis
 - Hypoplastic pulmonary valve

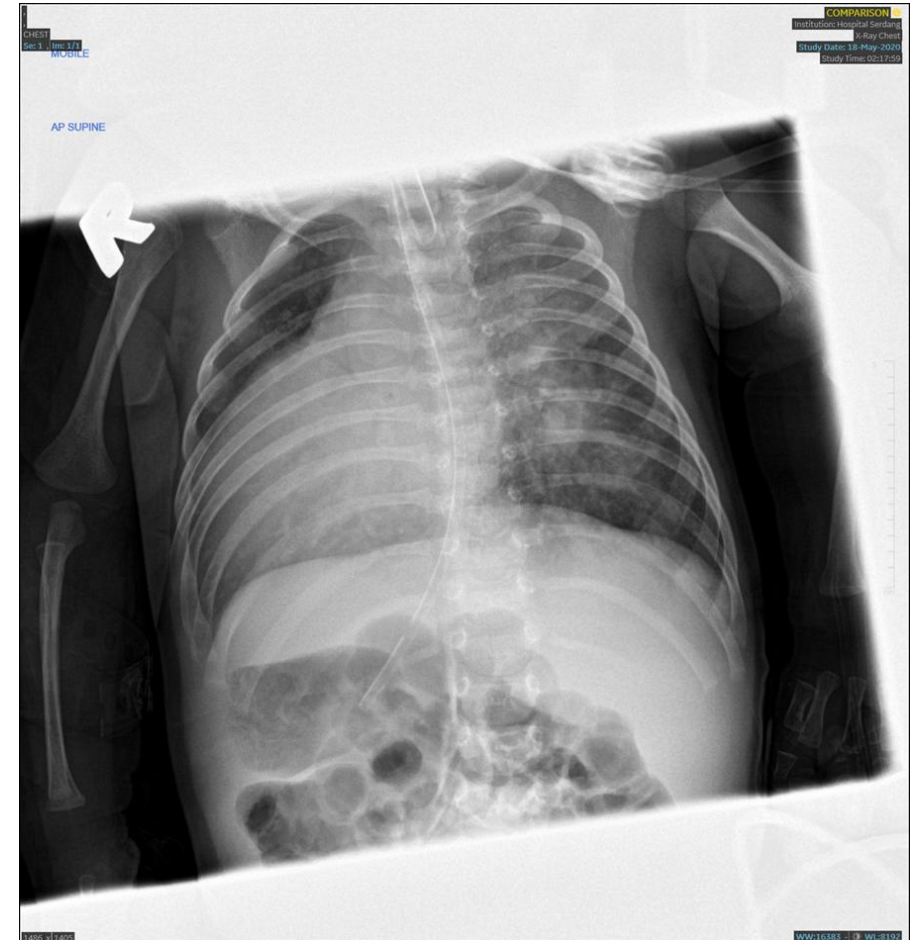
SITUS INVERSUS

- **Situs inversus** with dextroversion refers to a **mirror image of atrial situs solitus**.
 - Morphologic right atrium located to the left
 - Morphologic left atrium located on the right.
- **Mirror image** of **thoracic and abdominal organs**



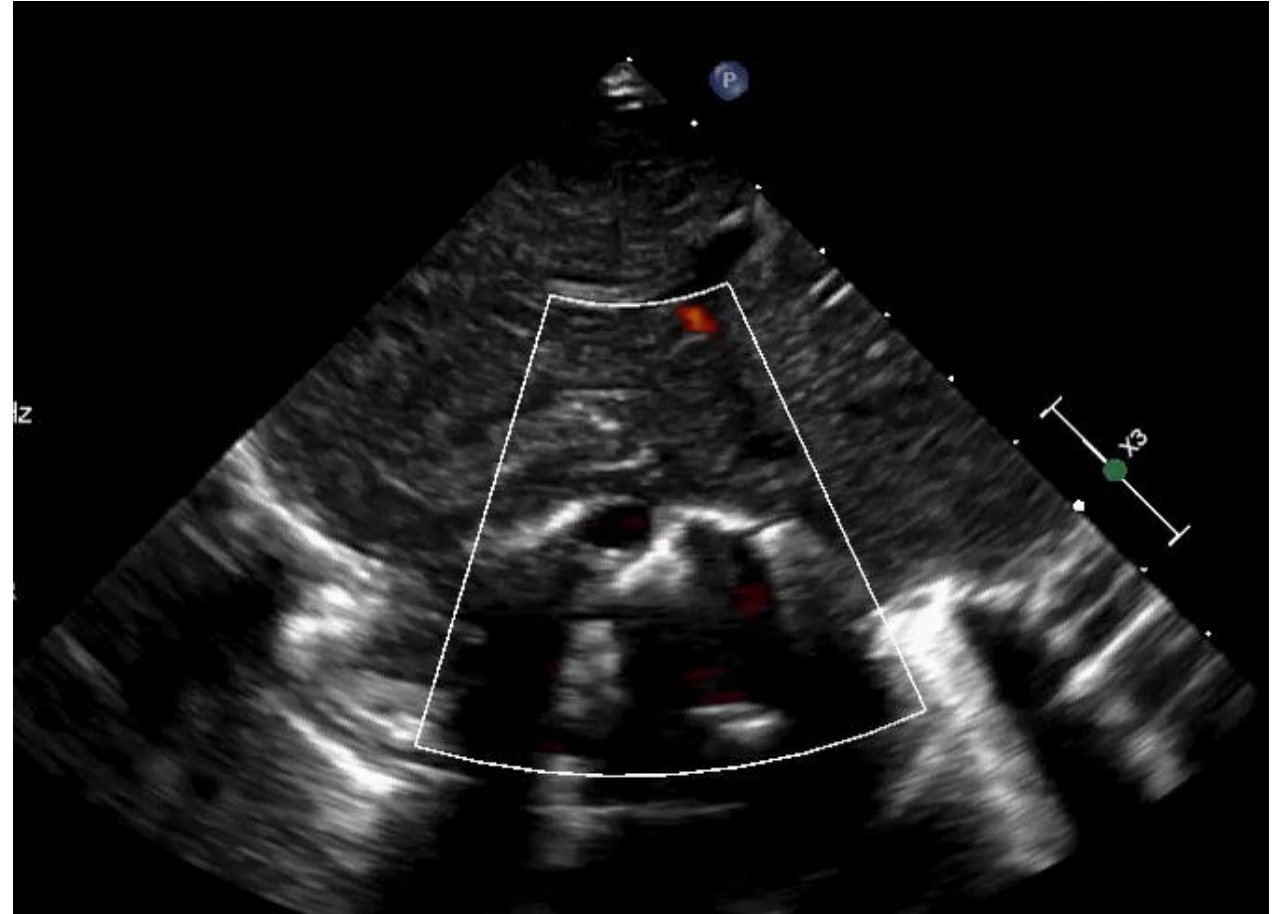
COMPLETE SITUS INVERSUS

- Dextrocardia
- Liver on the left
- Stomach on the right



COMPLETE SITUS INVERSUS

- Aorta on the right
- Stomach on the right
- IVC on the left
- Liver on the left



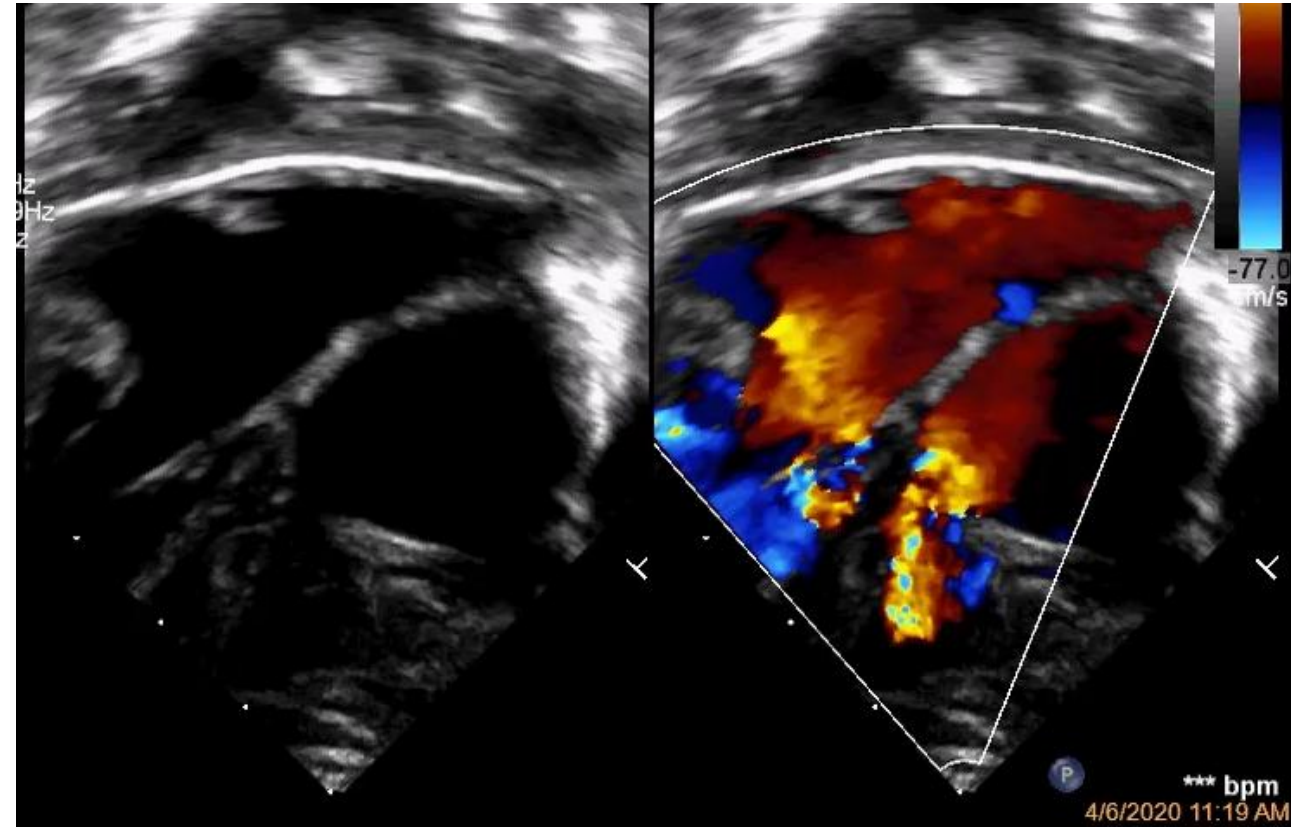
COMPLETE SITUS INVERSUS

- Dextrocardia
- Morphologic LA on the right
- Morphological LV on the right
- Morphology RA on the left
- Morphological RV on the left



COMPLETE SITUS INVERSUS

- Pulmonary veins draining to morphological LA on the right
- Small patent foramen ovale from LA to RA
- Mild mitral valve regurgitation

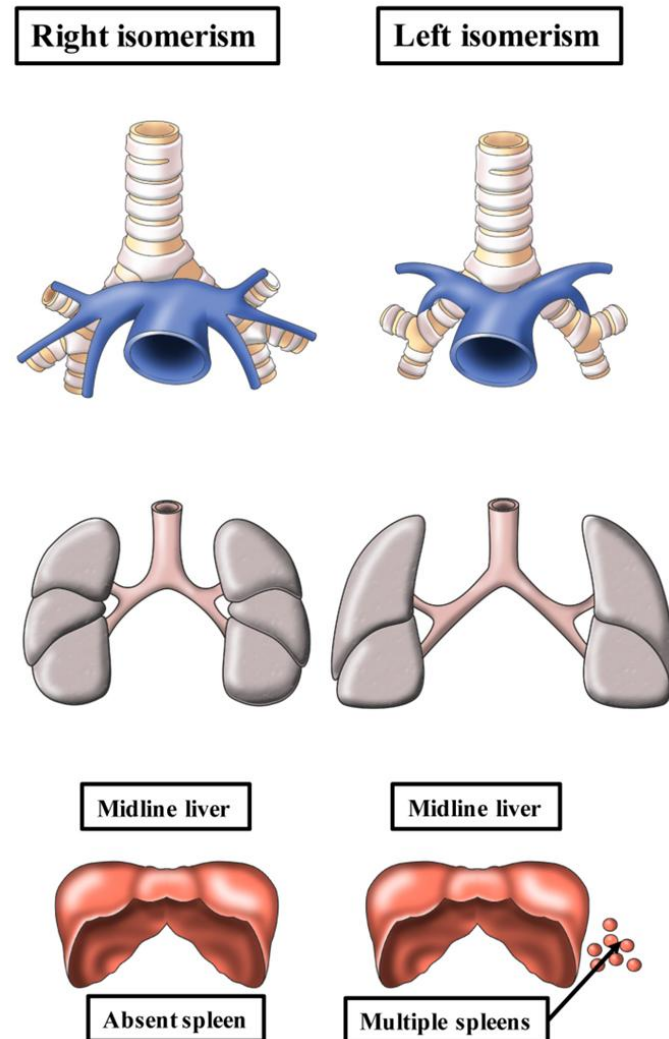


ECHOCARDIOGRAPHY DIAGNOSIS

- Complete situs inversus
- Dextrocardia
- Small patent foramen ovale
- Mild mitral valve regurgitation

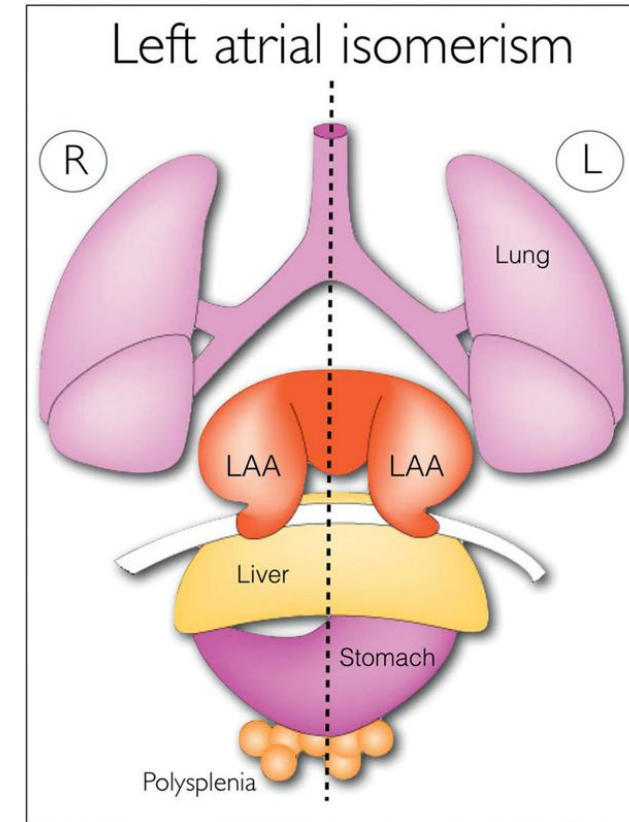
HETEROTAXY SYNDROME

- **Heterotaxy** derived from Greek (hetero: *different* and taxy: *arrangement*)
- Defined as and an *abnormal arrangement* of the internal thoracic and abdominal organs across the left and right axis of the body.
- Heterotaxy is caused by *disruption of left and right axis orientation* during early embryonic development



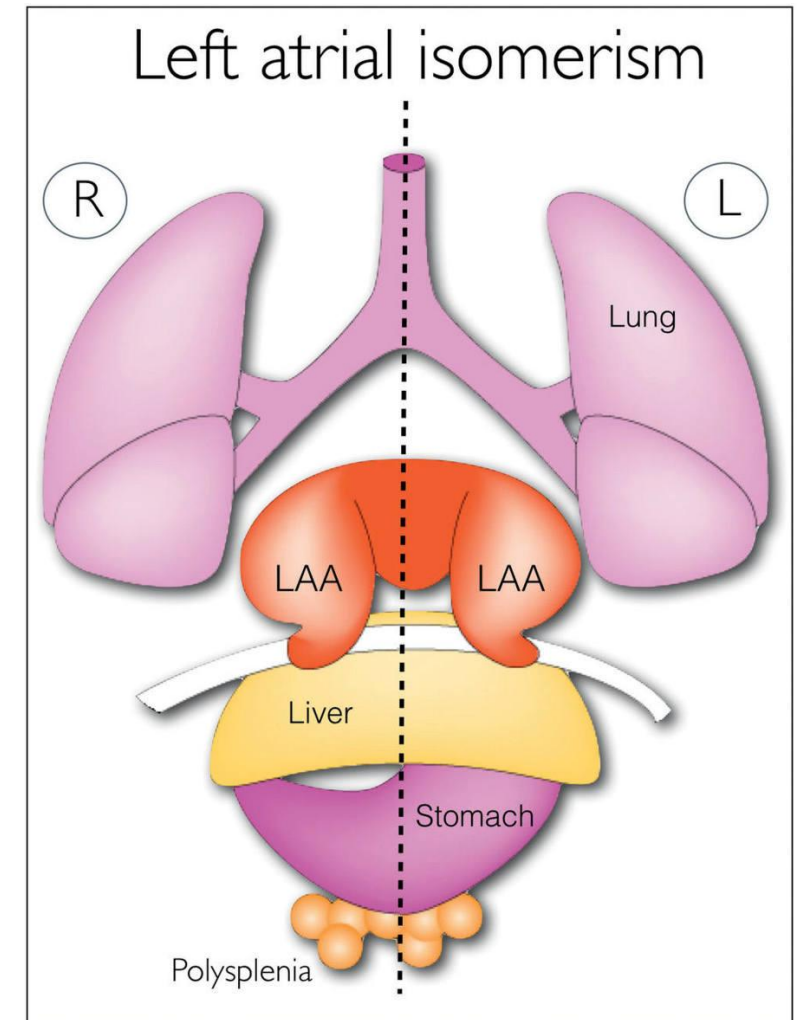
HETEROTAXY SYNDROME

- **Failure of normal lateralization** results in
 - **Abnormal bilateral symmetry** of normally asymmetric viscera and
 - **Duplication** of either right- or left-sided structures.
- Heterotaxy is also known as:
 - **Asplenia syndrome** (right isomerism) or
 - **Polysplenia syndrome** (left isomerism).
- Heterotaxy syndrome is caused by **genetic abnormalities**. These abnormalities may be inherited from one or both parents, or they randomly happen.
- Incidence 1 in 10,000 births
- A male to female ratio of 2:1.
- Heterotaxy accounts for approximately 3% of cases of CHD.



LEFT ISOMERISM

- Left Isomerism known as polysplenia syndrome.
- Multiple splenules without parent spleen.
- Azgous and hemiazygous continuation of the inferior vena cava.
- Bilateral hyparterial bronchus
- Bilateral bilobed lungs
- Bilateral left atria
- Midline/ transverse liver
- Intestinal malrotation



LEFT ISOMERISM ASSOCIATION

- **Congenital heart disease (>50%)**
 - *Especially non cyanotic* and less complex/severe than asplenia syndrome
 - *Abnormal pulmonary venous return* (70% PAPVR/TAPVR)
 - Dextrocardia (37%)
 - *Complete heart block*
- **Vascular**
 - *IVC interruption with azygos or hemiazygous continuation.*
 - Portal vein anomalies
 - Preduodenal portal vein

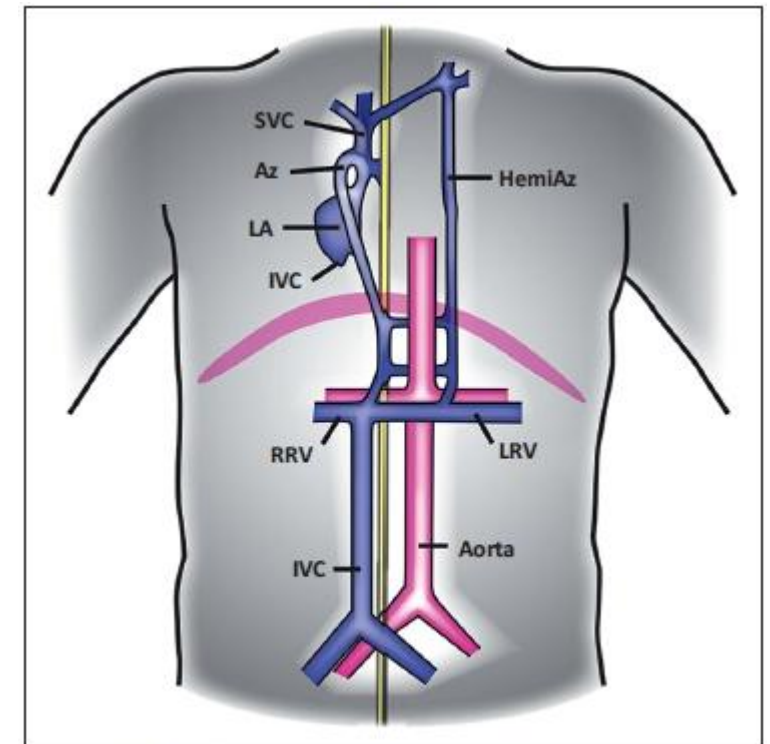
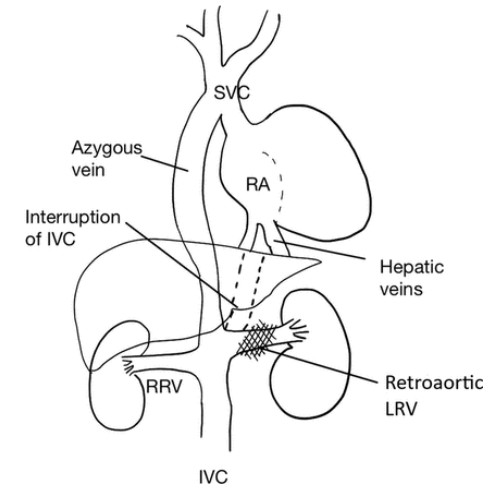
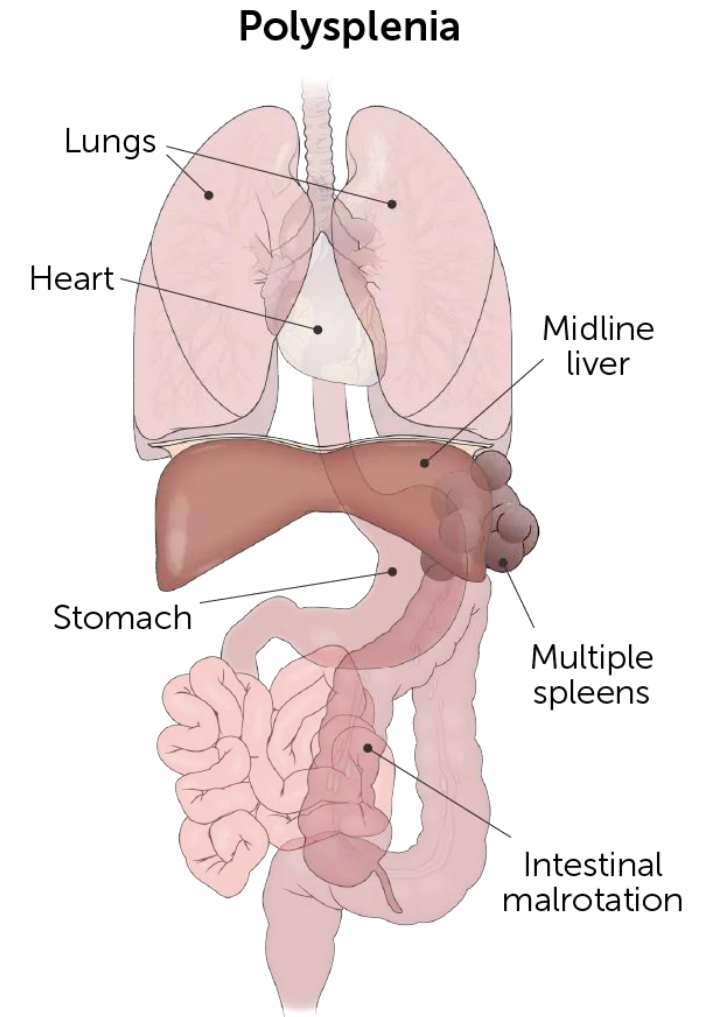


FIGURE 11: Illustration showing interrupted IVC continuing with azygos or hemiazygous vein. The aorta is located on the opposite side of the midline.

LEFT ISOMERISM ASSOCIATION

- Gastrointestinal
 - **Midgut rotation (80%)**
 - Semi annular pancreases/ dorsal pancreatic agenesis
 - Gall bladder agenesis (50%) and biliary atresia
 - Mobile cecum
 - Transesophageal fistula
- Genitourinary
 - Renal cyst
 - Renal agenesis
 - Ovarian cyst



LEFT ISOMERISM

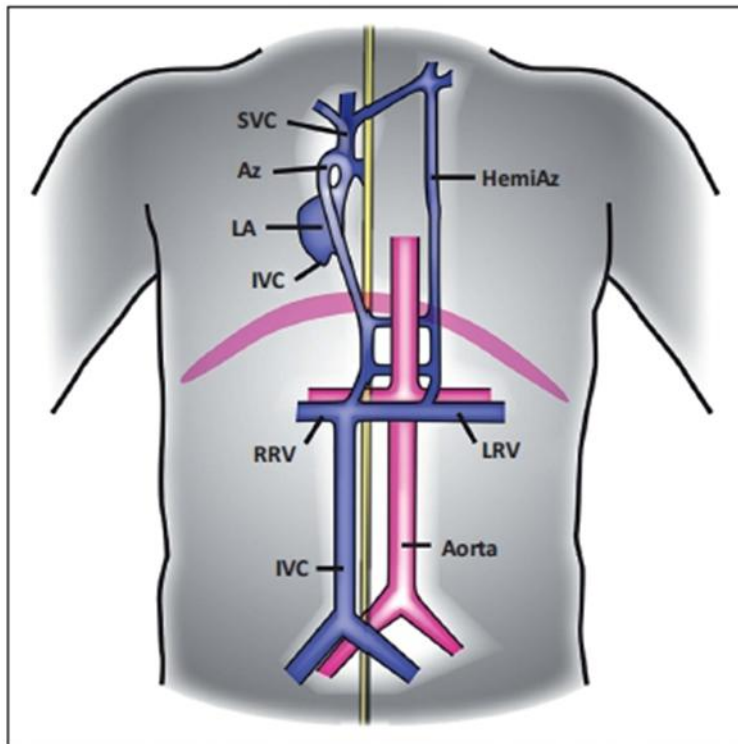
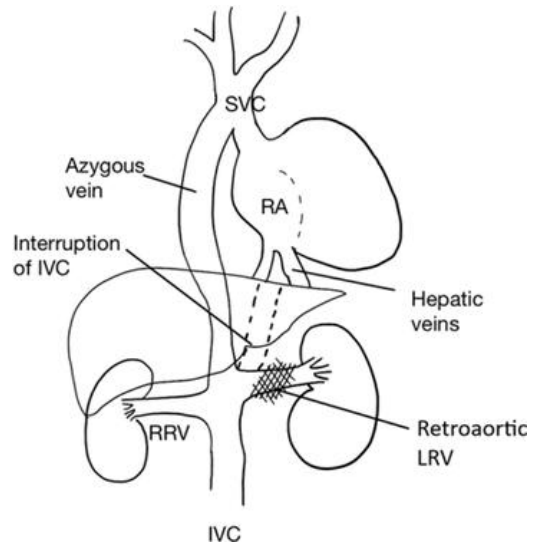


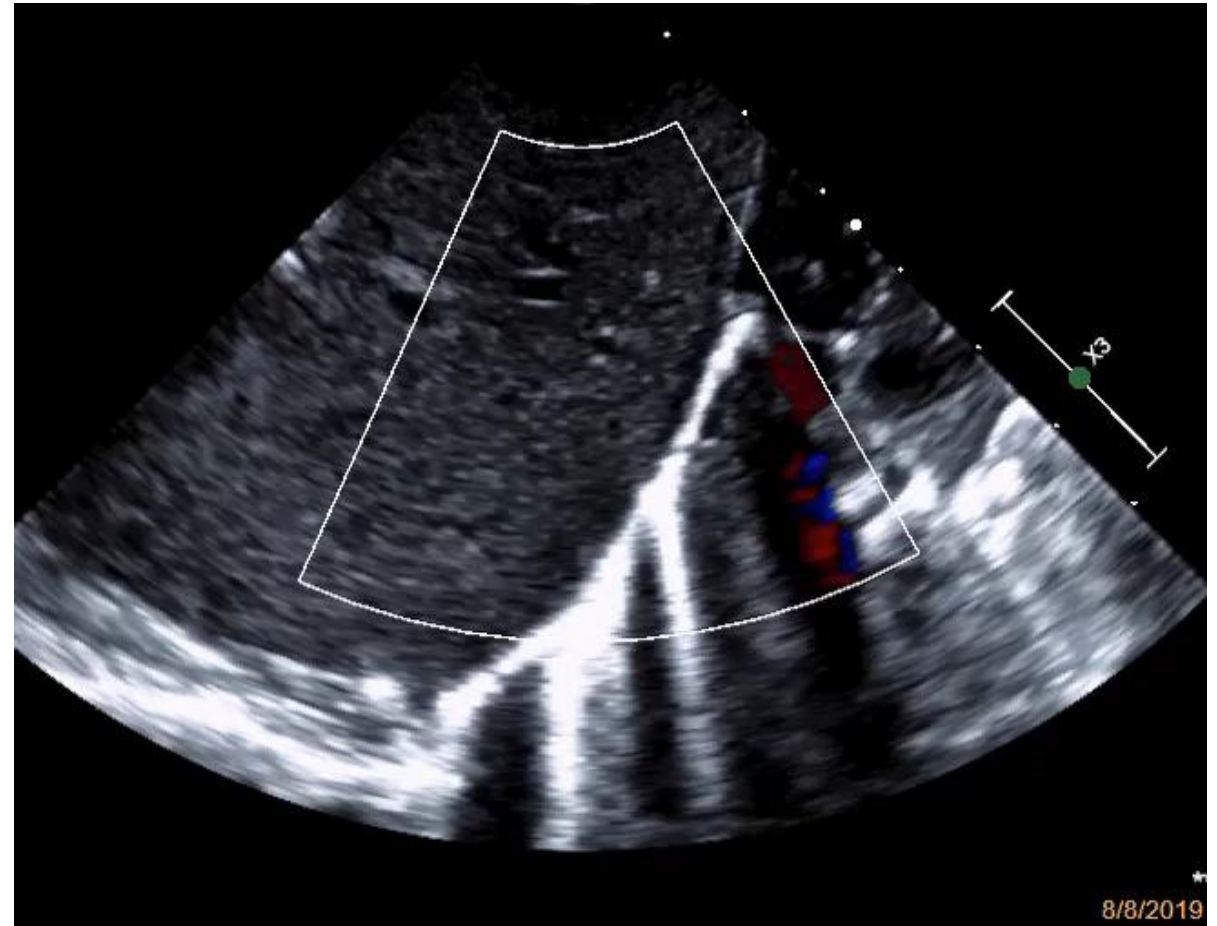
FIGURE 11: Illustration showing interrupted IVC continuing with azygous or hemizygous vein. The aorta is located on the opposite side of the midline.



LEFT ISOMERISM

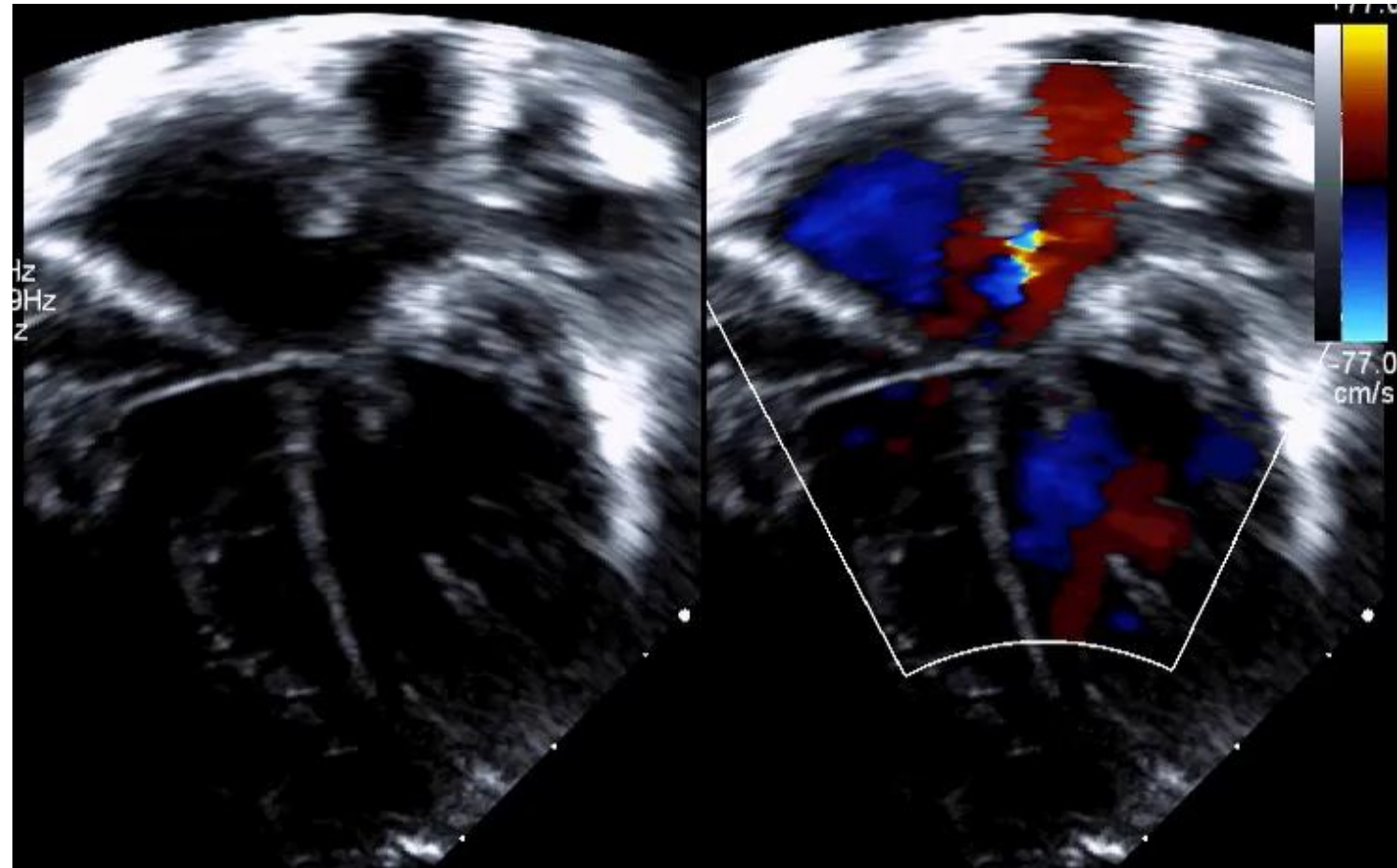


Absent of inferior vena cava/ interrupted IVC



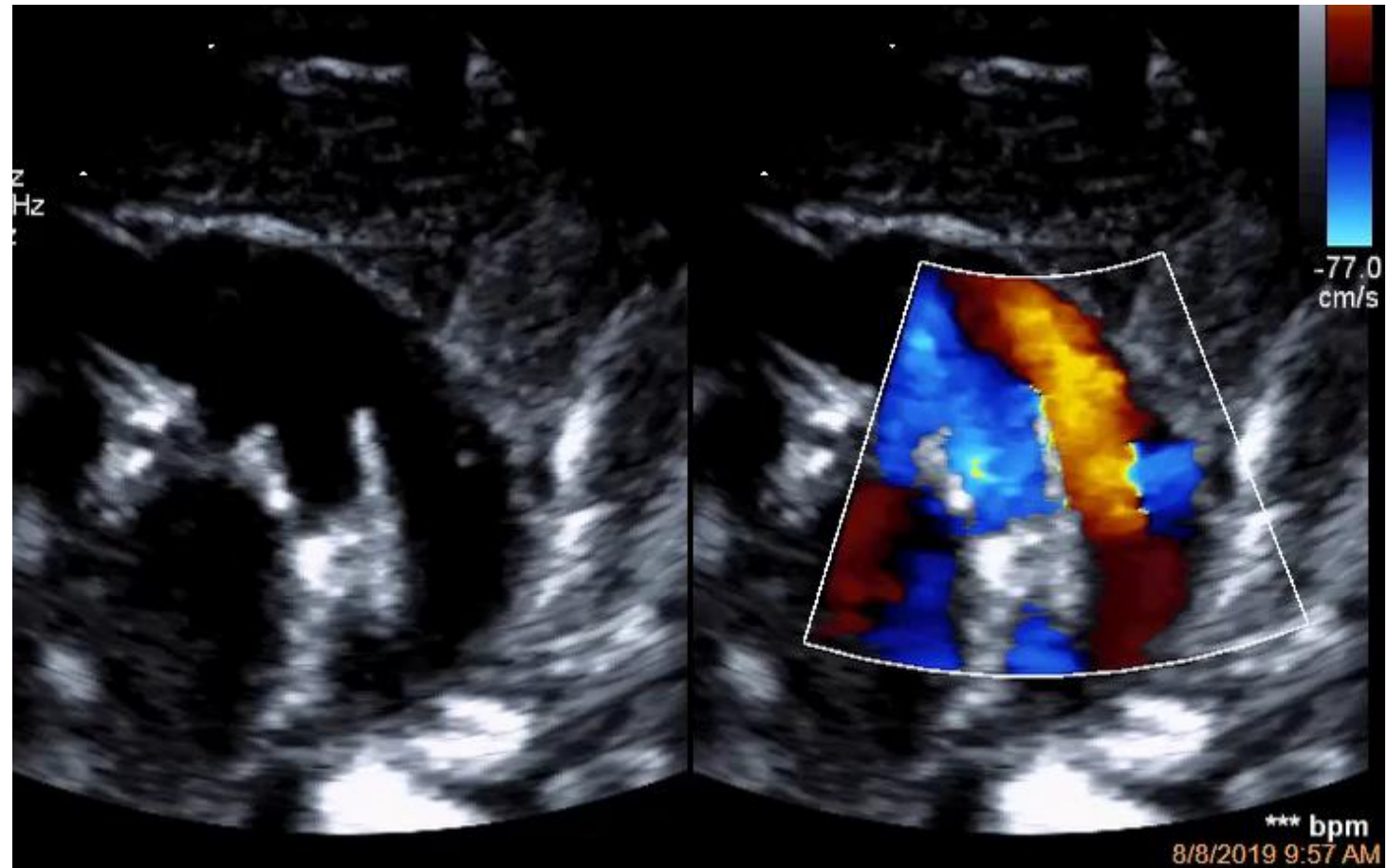
LEFT ISOMERISM

- Complete balanced atrioventricular septal defect
- Large primum atrial septal defect
- Restrictive inlet VSD
- Pulmonary veins draining to left sided atrium
- Mild left and right atrioventricular valve regurgitation



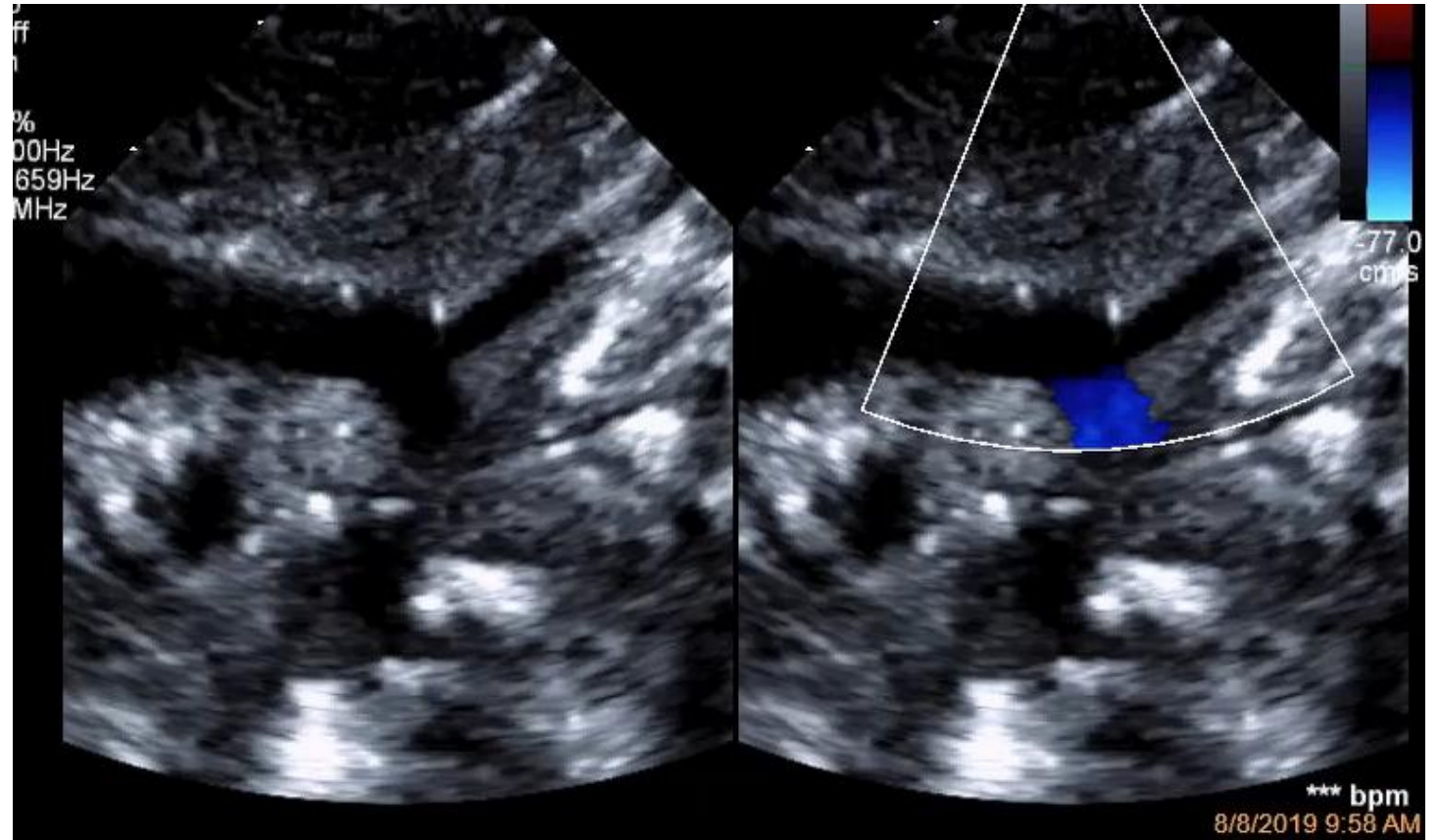
LEFT ISOMERISM

- Confluence good size branch pulmonary artery
- Large PDA with bidirectional shunt
- Turbulent flow at the proximal descending aorta



LEFT ISOMERISM

- Hypoplastic aortic arch
- Coarctation of aorta



ECHOCARDIOGRAPHY DIAGNOSIS

- Left atrial isomerism/ Left heterotaxy
- Complete balanced AVSD
- Large primum
- Restrictive/ small inlet VSD
- Mild hypoplastic aortic arch
- Coarctation of aorta
- Large PDA
- Pulmonary hypertension

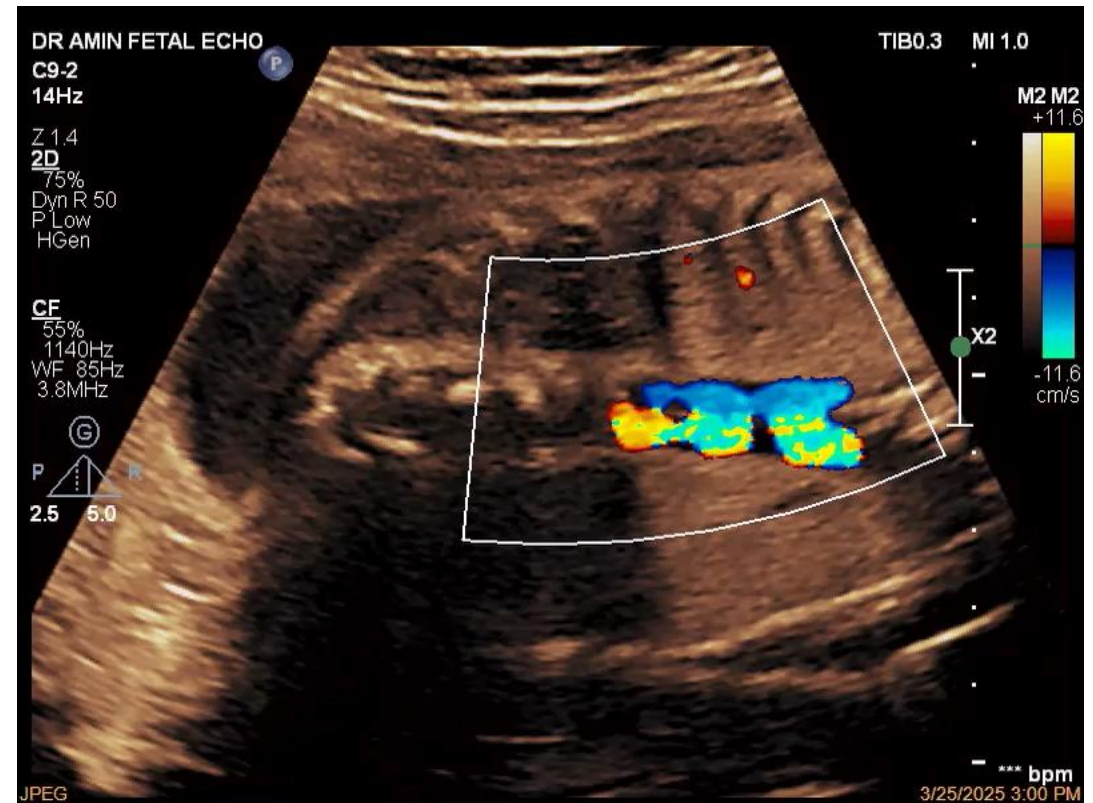
Fetal echocardiography

Left isomerism

Stomach on the right



Fetal echocardiography with left isomerism



DR AMIN FETAL ECHO

C9-2

87Hz

RS

2D

68%

Dyn R 47

P Low

HGen

TIB0.8

MI 1.1

M2



9.9cm

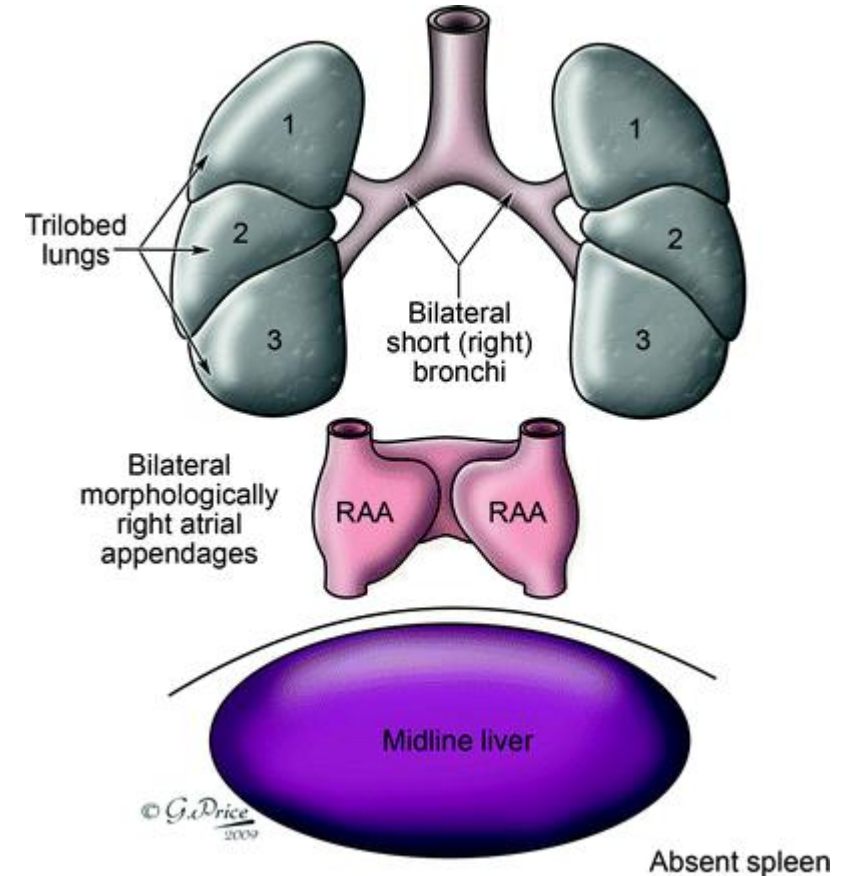
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3/25/2025 3:01 PM

JPEG

RIGHT ISOMERISM

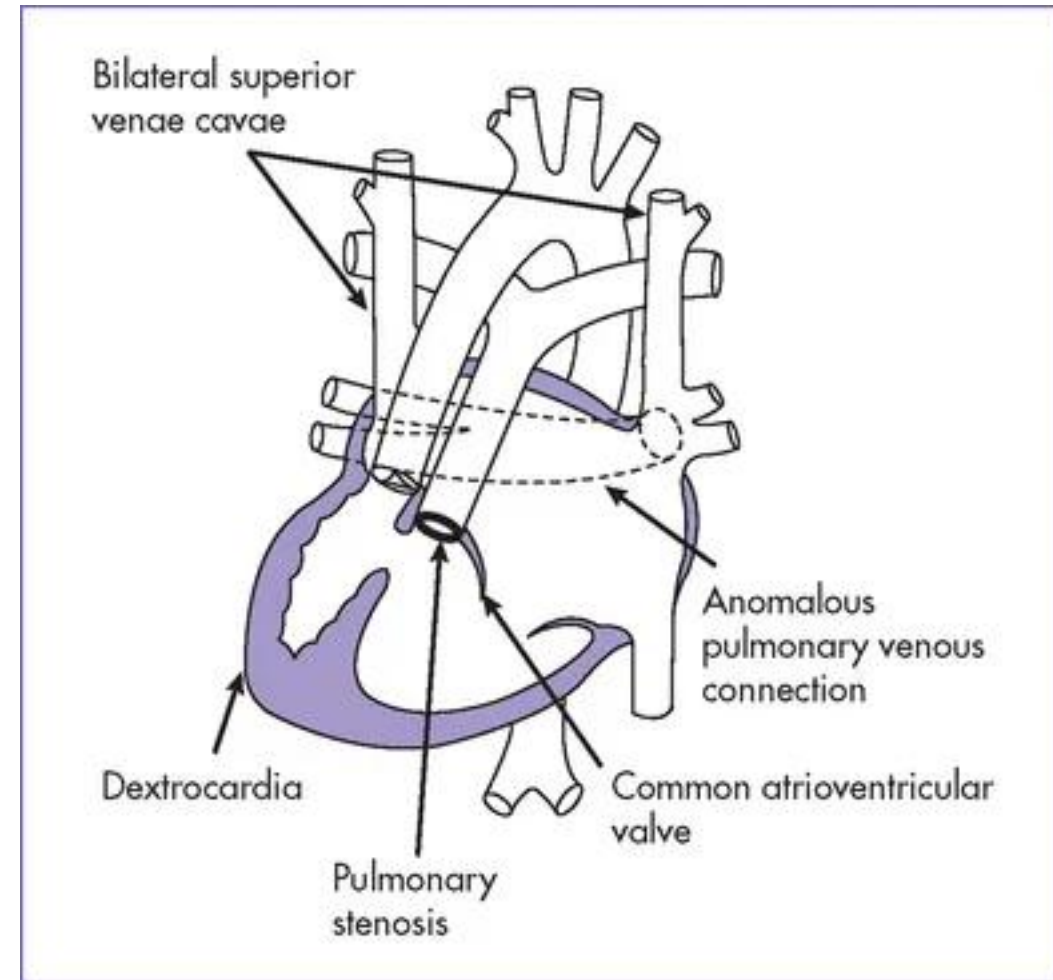
- Right isomerism known as asplenia syndrome
 - Severe cyanotic congenital heart disease
 - Absence spleen
 - Bilateral eparterial bronchi
 - Bilateral trilobed lungs
 - Bilateral right atrium
 - Midline /transverse liver
 - Intestinal malrotation



RIGHT ISOMERISM ASSOCIATION

- **Heart**

- Severe/complex congenital heart disease (50%) especially cyanotic congenital heart disease
- Total anomalous pulmonary venous
- Endocardial cushion defect (85%)
- Transposition of great artery (72%)
- Single ventricle (51%)
- Double outlet right ventricle



RIGHT ISOMERISM ASSOCIATION

- Vascular
 - Duplication of superior vena cava
 - Absent coronary sinus
 - Juxta position of the IVC in front (usually) of the abdominal aorta

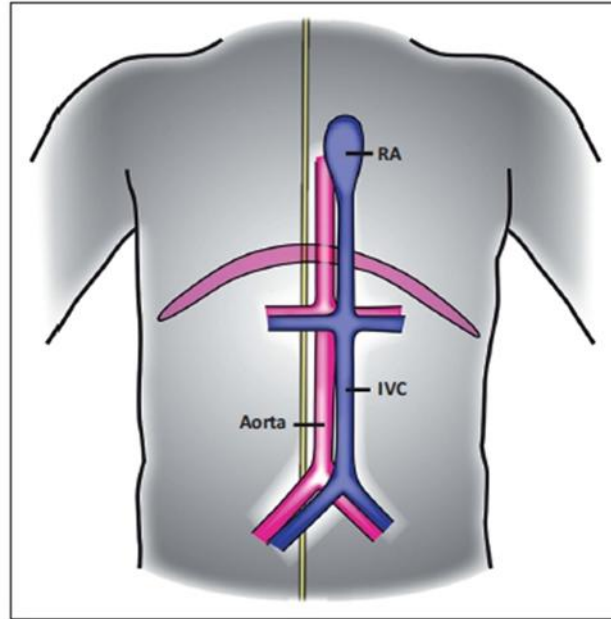
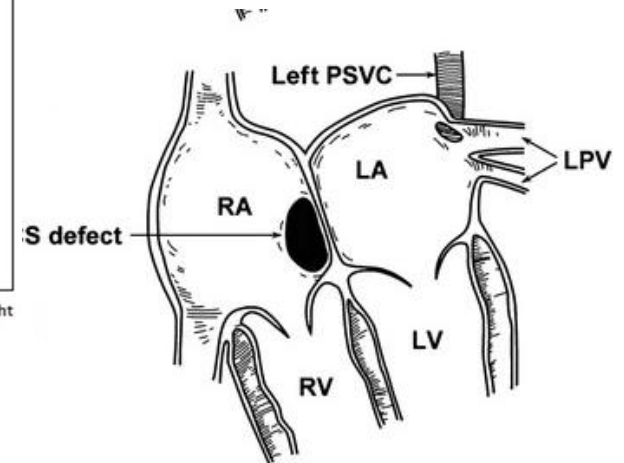
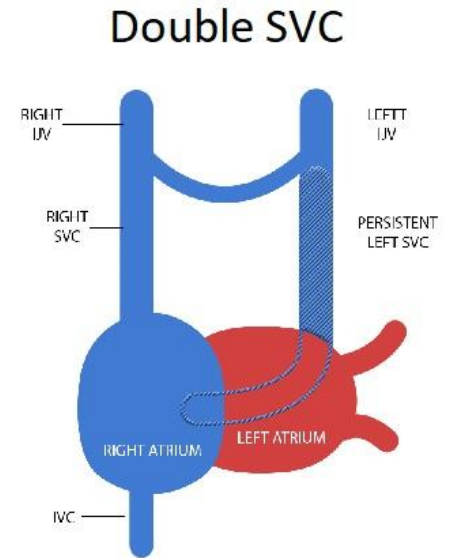


FIGURE 10: Illustration showing the position of the aorta and IVC in right isomerism.



RIGHT ISOMERISM ASSOCIATION

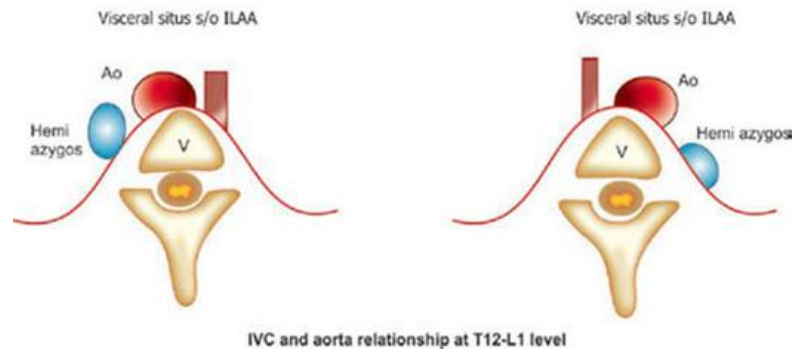
- **Gastrointestinal**

- Gall bladder agenesis
- Intestinal malrotation (up to 100%)
- Micro gastria
- Imperforated anus

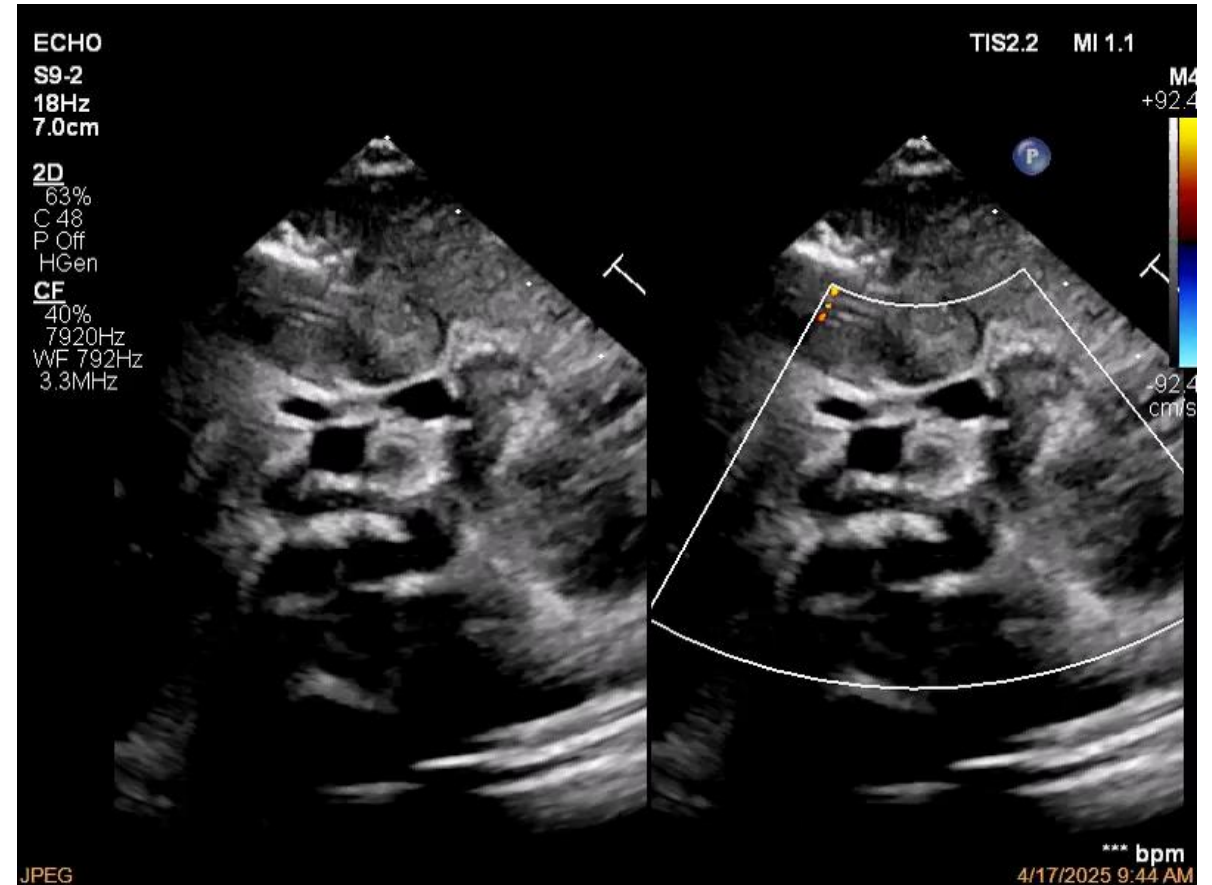
- **Genitourinary**

- Horseshoe kidney
- Fused/horseshoe adrenal gland or absent left adrenal gland
- Bicornuate uterus
- Bilobed urinary bladder.

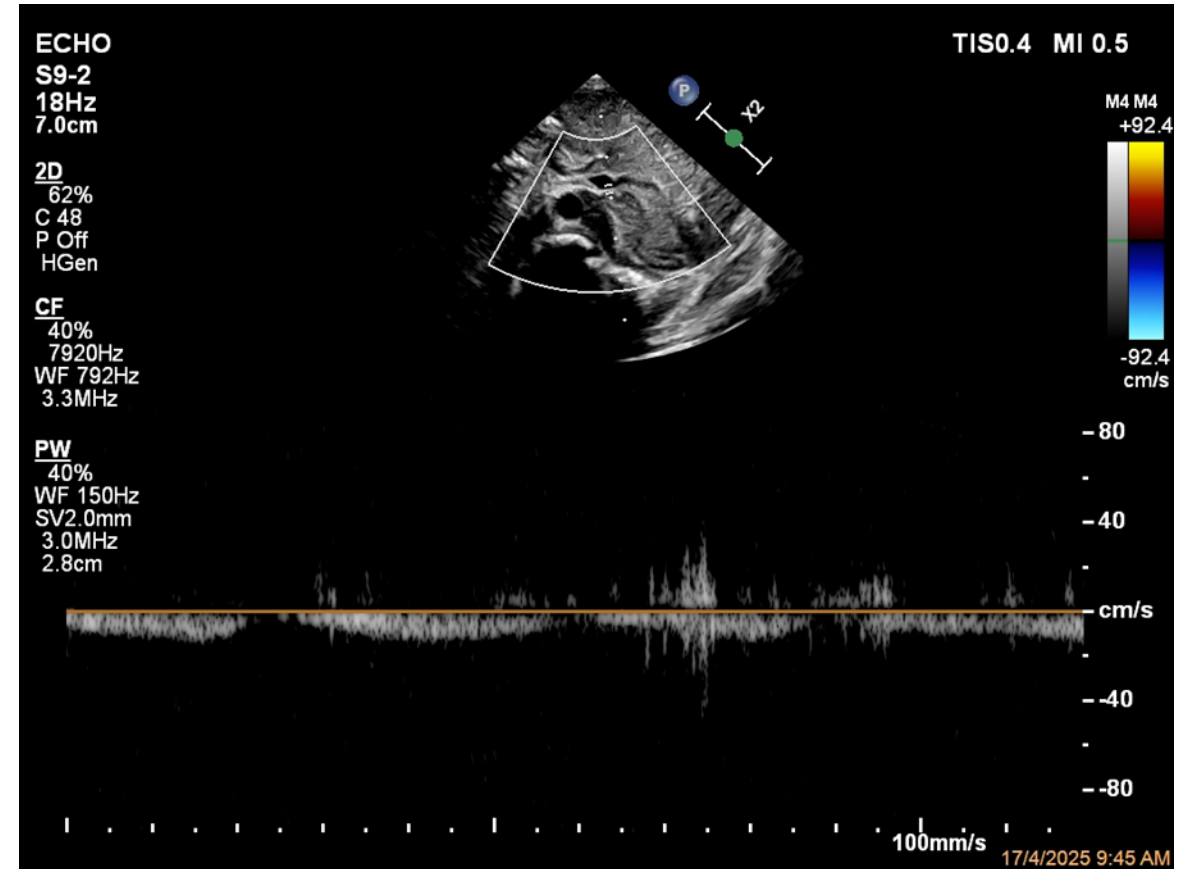
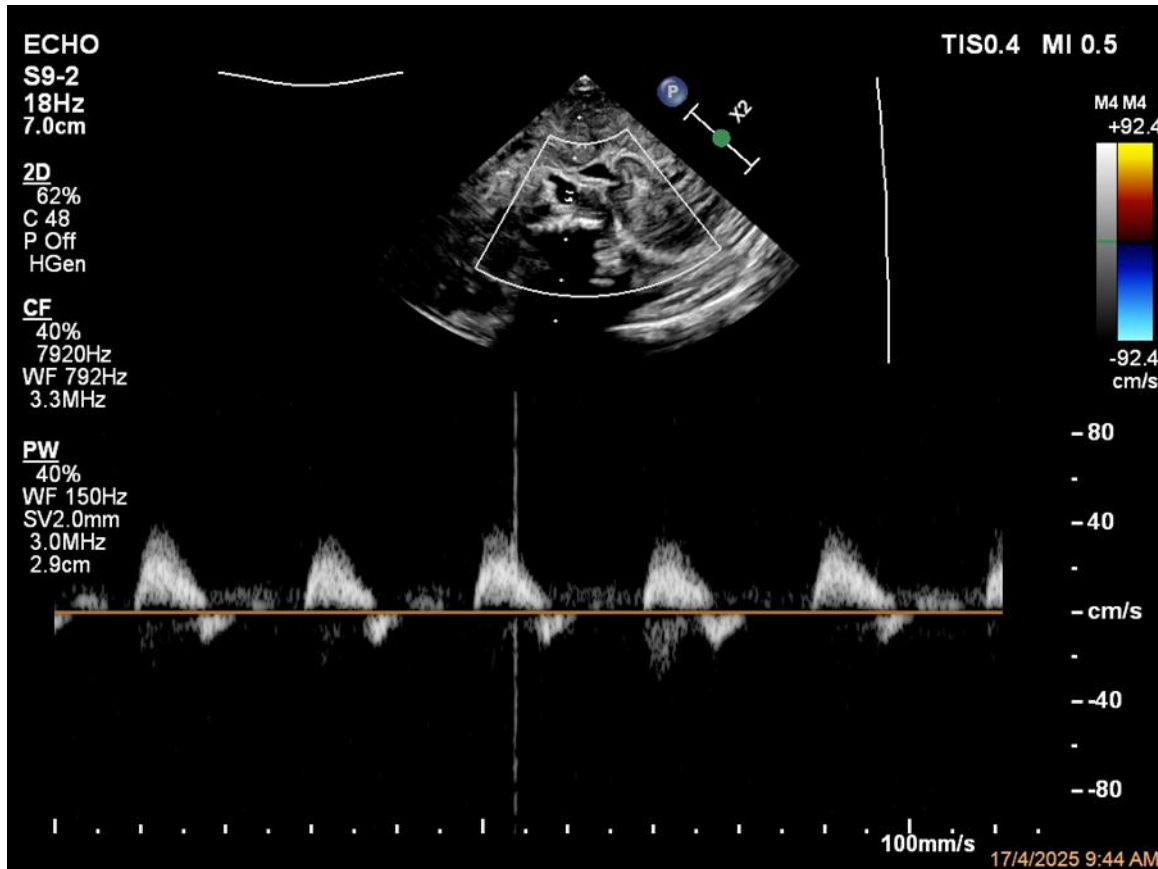
RIGHT ISOMERISM



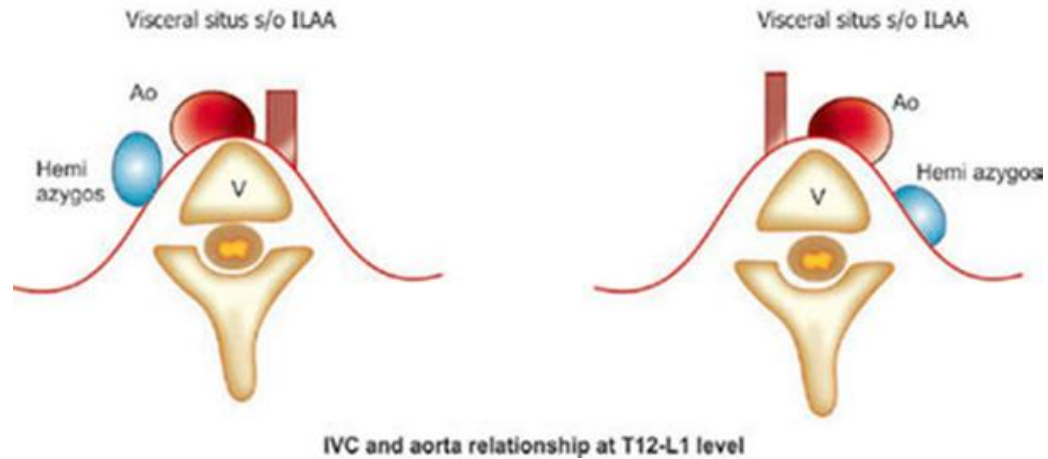
- Ipsilateral IVC and abdominal aorta



RIGHT ISOMERISM



RIGHT ISOMERISM

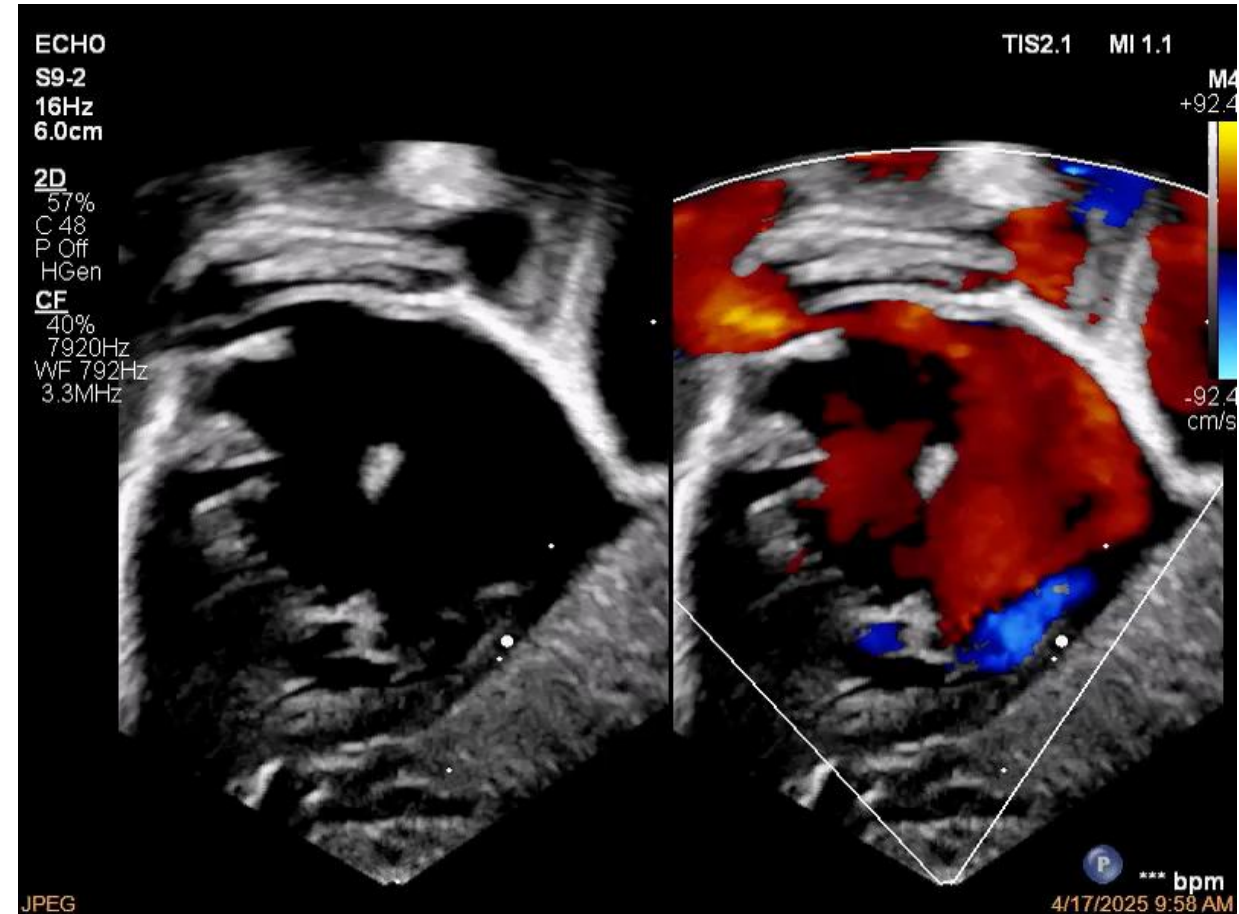


- Ipsilateral IVC and abdominal aorta



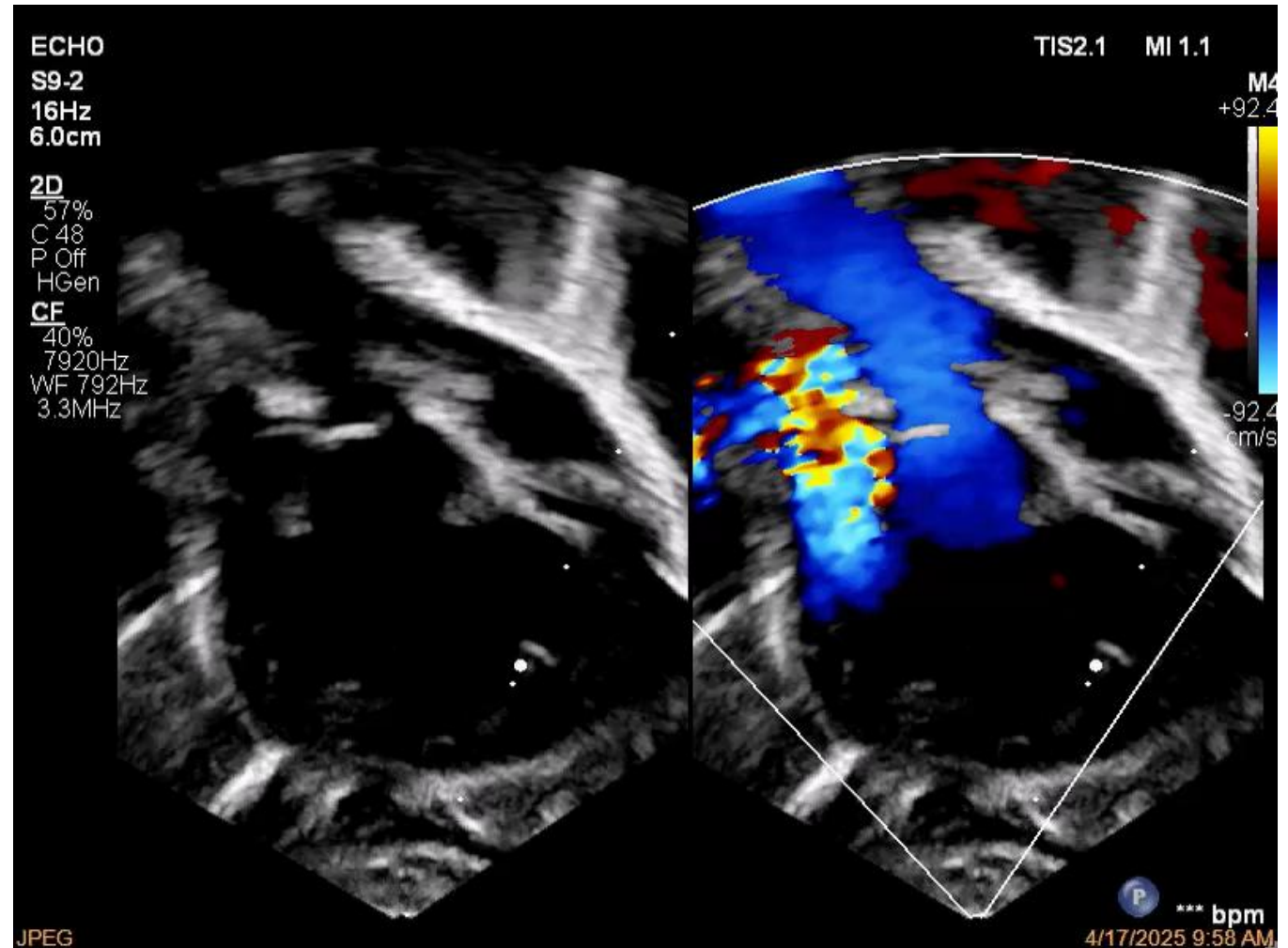
RIGHT ISOMERISM

- Dextrocardia
- Both atrium is morphologically right atrium
- Complete atrioventricular septal defect
- Abnormal pulmonary venous drainage



RIGHT ISOMERISM

- Double outlet right ventricle sub pulmonary ventricular septal defect
- Infundibular and valvular pulmonary stenosis



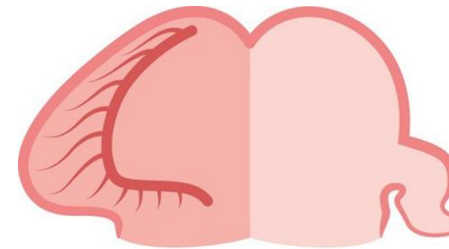
ECHOCARDIOGRAPHY DIAGNOSIS

- Right isomerism/ right heterotaxy
- Complete atrioventricular septal defect
- Double outlet right ventricle sub pulmonary ventricular septal defect
- Infundibular and valvular pulmonary stenosis
- Mixed anomalous pulmonary venous drainage

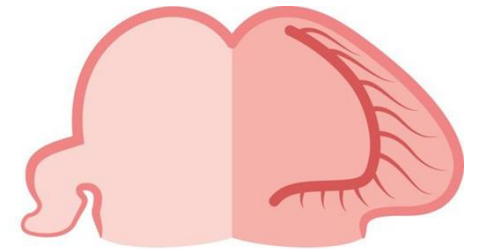
Determination of atrial solitus

Echocardiography

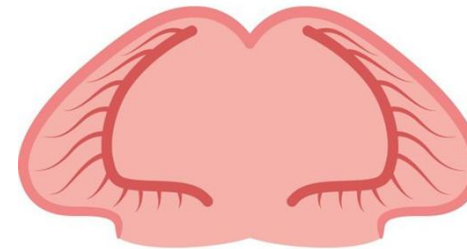
- Basic determination rely on the ability to discriminate **the right and left atrium** which depend on the morphologic characteristics of the atria and other features including:
 - **Internal and external anatomy of the atrial appendages**
 - **Connection of pulmonary and systemic venous drainage to the heart**
 - **Anatomy of the interatrial septum**



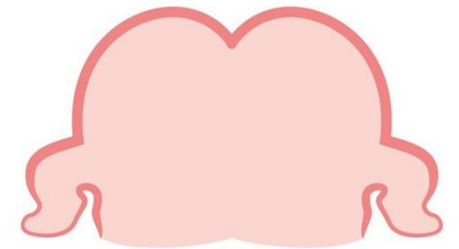
Usual



Mirror Imaged



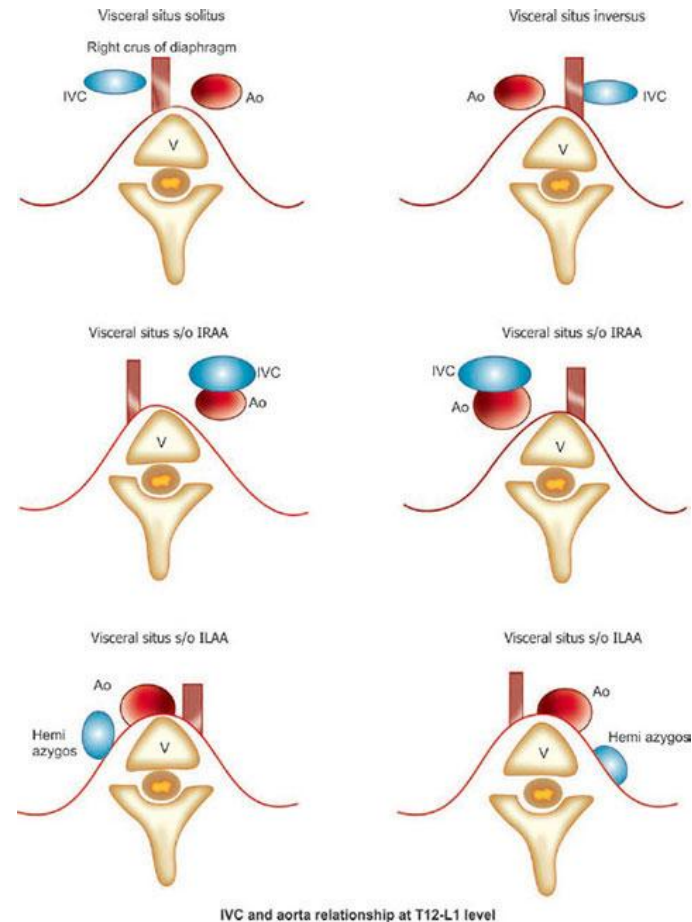
Right Isomerism



Left Isomerism

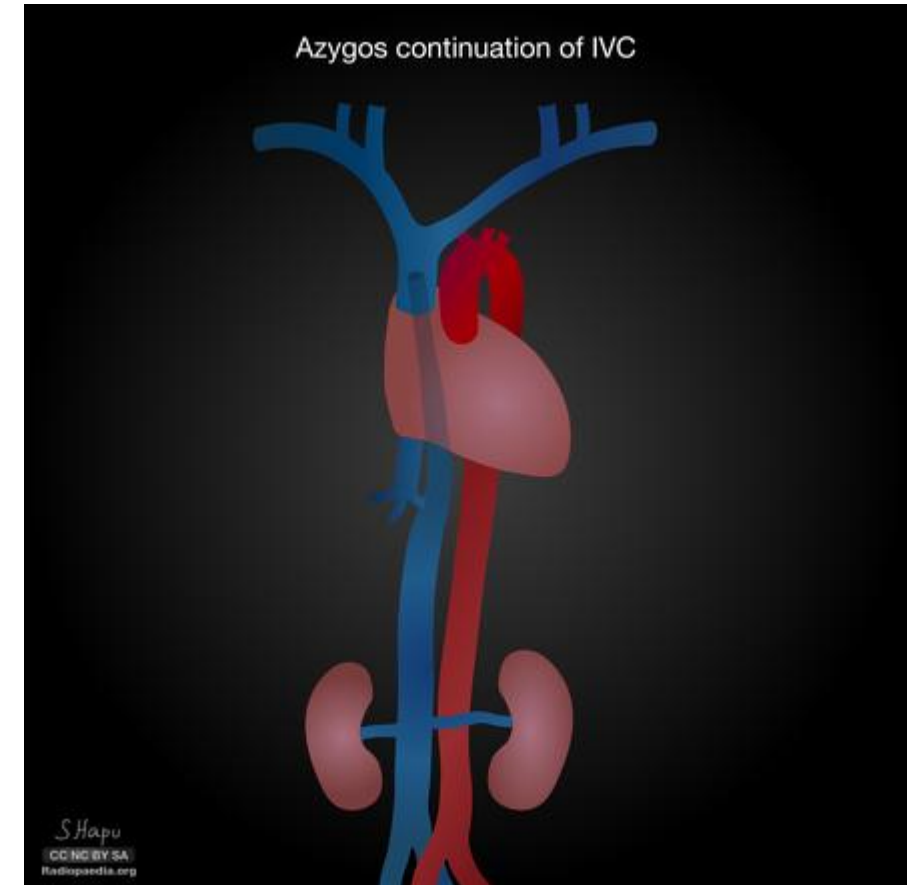
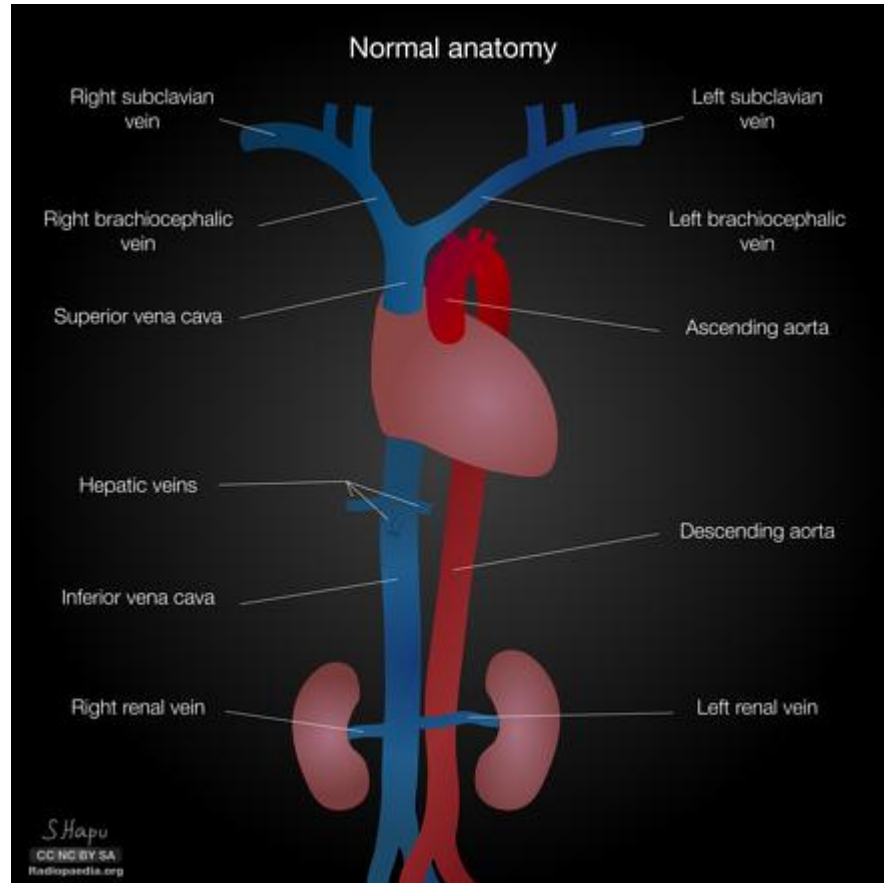
Determination of atrial solitus

Echocardiography: situs in subcostal view



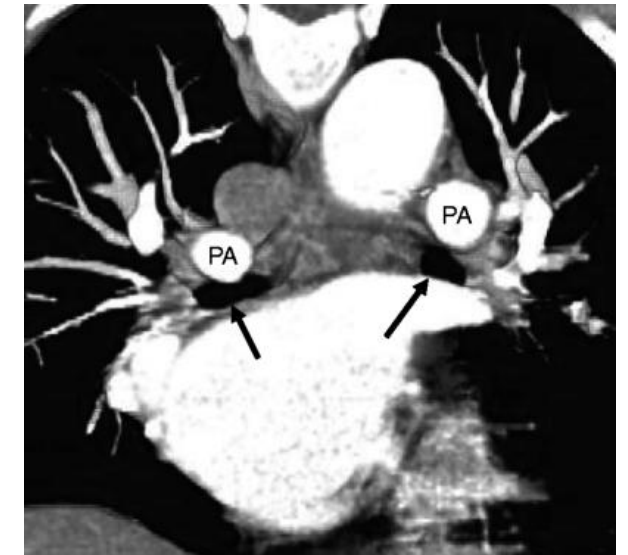
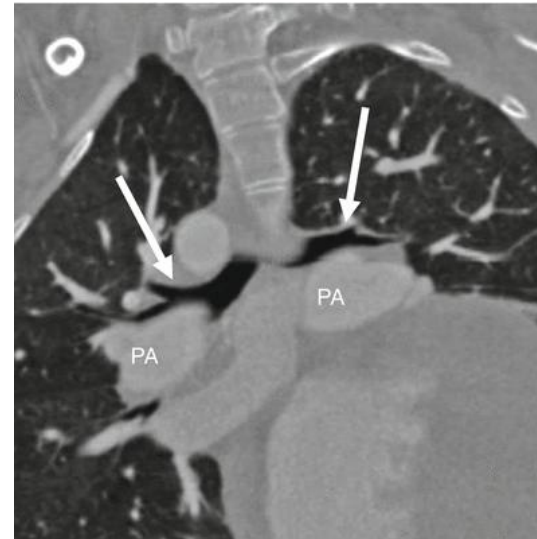
Determination of atrial solitus

Echocardiography



RECOMMENDED IMAGING

- Echocardiography
- Chest radiograph
- Abdominal ultrasound
- CT scan



CASE REPORT | Originally Published 26 August 2020 | [Check for updates](#)

Diagnosis of Heterotaxy Syndrome in a Patient With Multiple Congenital Cardiac Malformations Using Magnetic Resonance Imaging

Sani Namik Murat, MD, Damla Yalcinkaya, MD, Mikail Yarlioglu, MD, Hasan Yigit, MD, Ugur Bozkurt, MD, Ibrahim Etem Celik, MD, Fatih Oksuz, MD, Fatih Atik, MD, and Mustafa Duran, MD | [AUTHOR INFO & AFFILIATIONS](#)

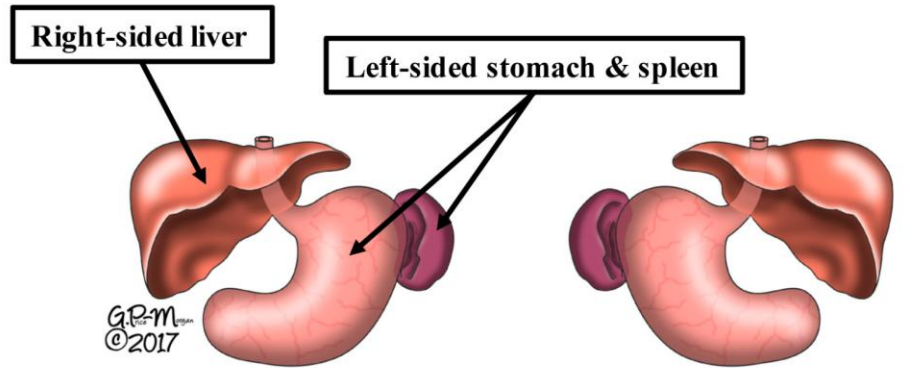
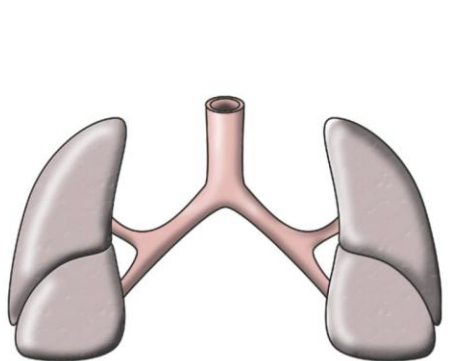
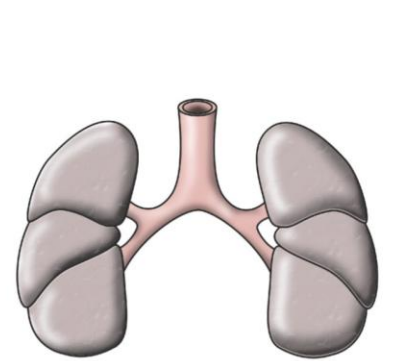
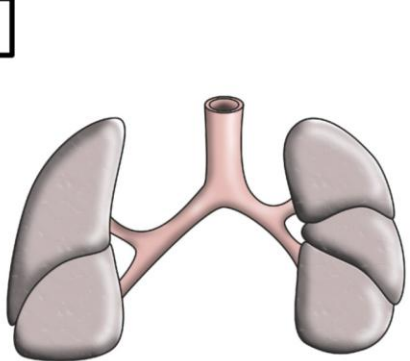
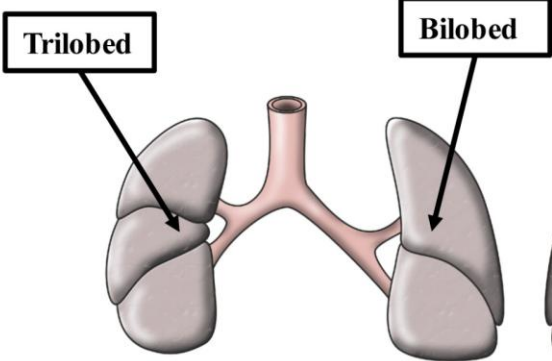
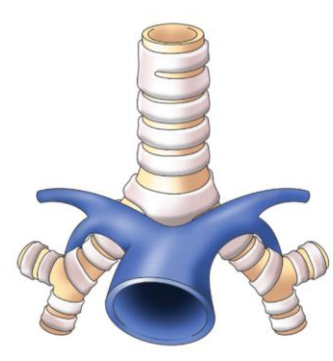
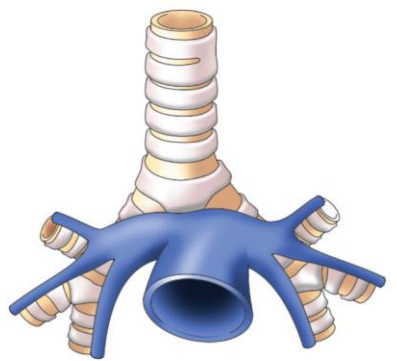
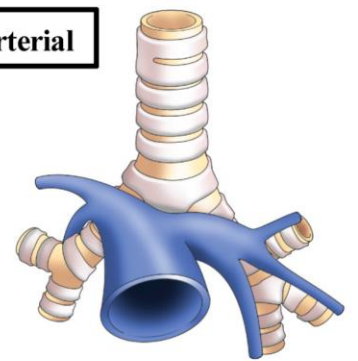
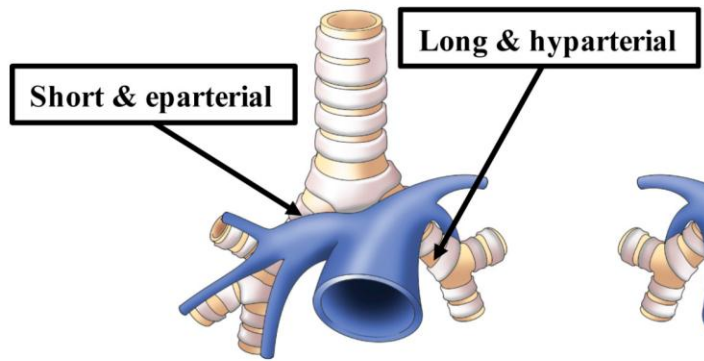
Circulation: Cardiovascular Imaging • Volume 13, Number 9 • <https://doi.org/10.1161/CIRCIMAGING.119.010307>

Usual arrangement

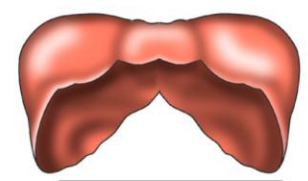
Mirror-imagery

Right isomerism

Left isomerism

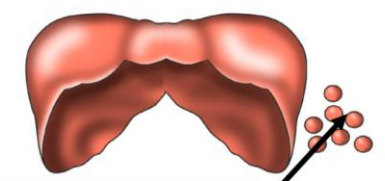


Midline liver



Absent spleen

Midline liver



Multiple spleens

CONCLUSION

- Determination of situs is crucial in echocardiography assessment.
- Situs solitus and situs inversus has lower association with congenital heart disease.
- Be prepared for difficult echocardiography assessment in heterotaxy heart as it is usually has more cardiac abnormalities lesions especially in right isomerism.



**THANK
YOU
FOR
YOUR
ATTENTION**

A stack of five light-colored wooden blocks with rough, hand-cut edges. The blocks are arranged in a slightly staggered, pyramid-like fashion. The top block is the widest, followed by a narrower one, then a wider one, then a narrower one, and the bottom block is the widest again. The text is printed in a bold, black, sans-serif font on each block. The stack sits on a dark wooden surface against a dark green background.