

# ***SITUS AMBIGUOUS***

**SITUS**  
**(thoracic-abdominal)**

# What is it?

- also known as heterotaxy or heterotaxia
- is a rare congenital defect in which the major VISCERAL organs (heart, lungs, liver, spleen and bowels) are distributed abnormally within the CHEST and ABDOMEN.
- there's a disruption of the left and right axis differentiation (in early development) resulting in a variety of cardiac and noncardiac anomalies

# *What are the effects?*

The early disruption of cardiac development typically causes right or left atrial isomerism, which often results in discordance among cardiac segments

Right and left isomerism have different patterns of cardiac defects, which can be observed through combinations of anatomic lesion of various cardiac segments

Atrial isomerism is rare, occurs in approximately 1 per 10,000- 40,000 live births, about 3-6% of all CHD.



# Heterotaxy

derived from Greek (*hetero*, meaning “different”, and *taxy*, meaning “arrangement”).

the normal asymmetry of the thoracic and abdominal organs is lost

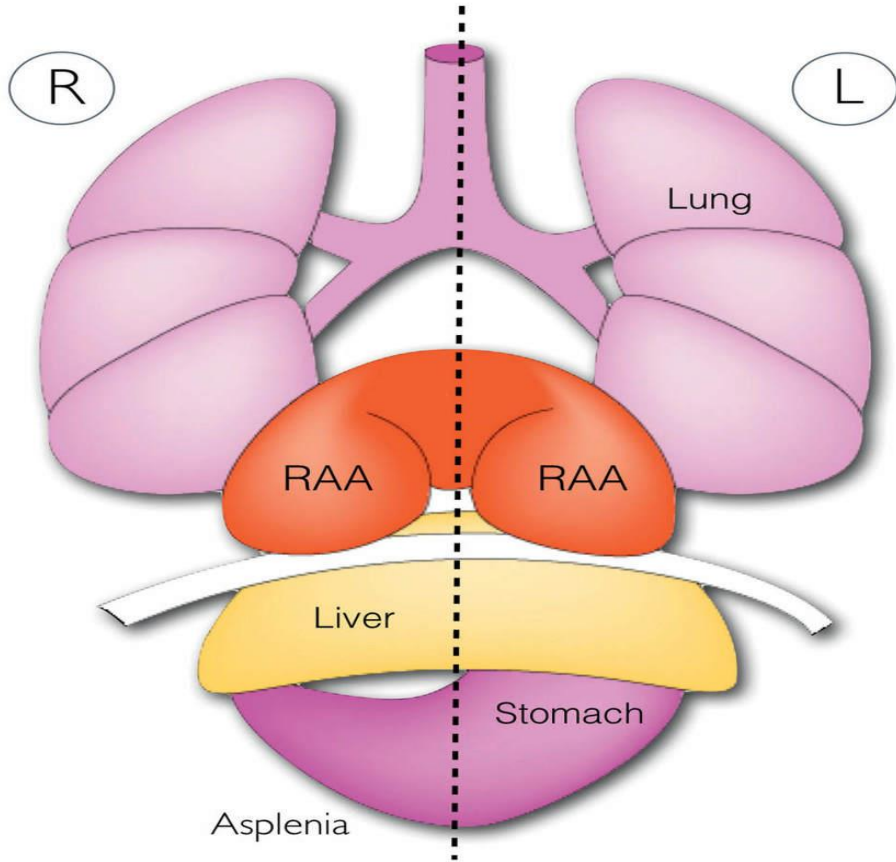
50-70%---levocardia

25-50%---dextrocardia

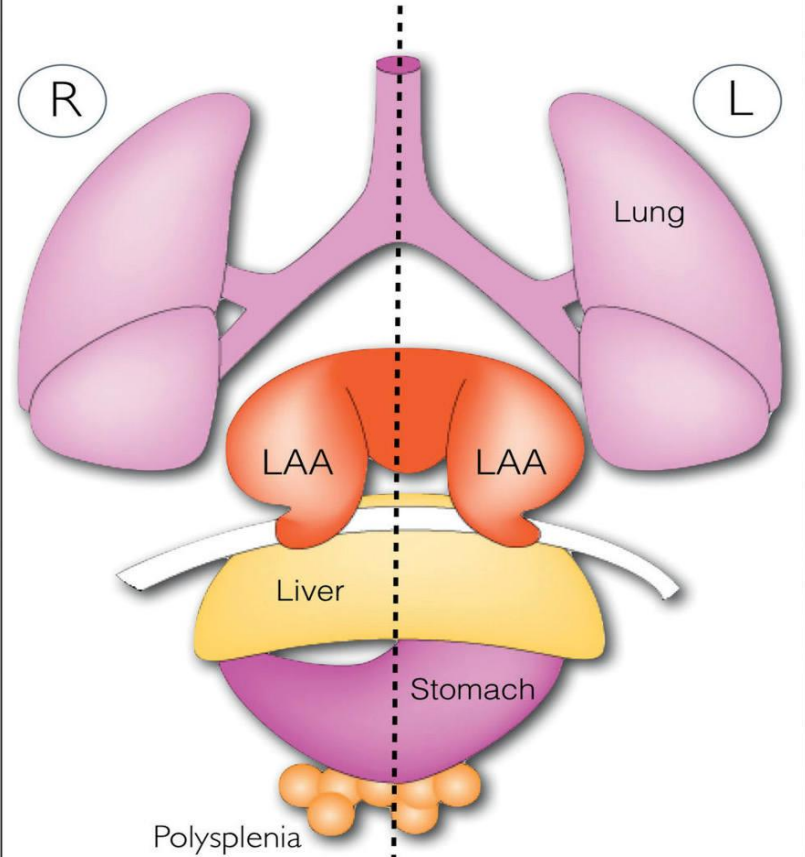
5-10% ---mesocardia



# Right atrial isomerism



# Left atrial isomerism



# Heterotaxy

Patients with heterotaxy have organs with 2-right-sides or 2-left-sides, most commonly observed in relation to the atria of the heart.

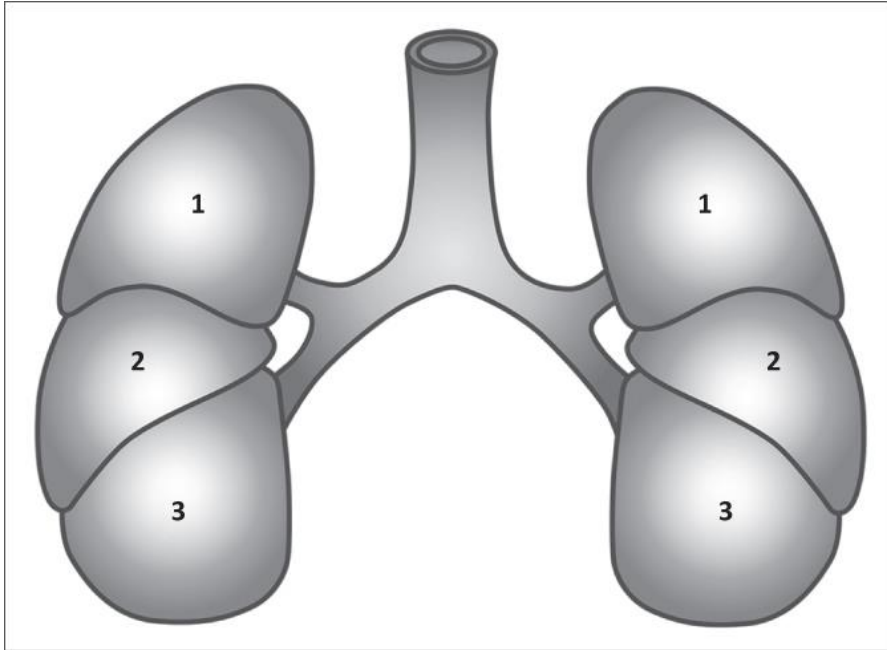
Heterotaxy syndrome

1. alters the structure of the heart

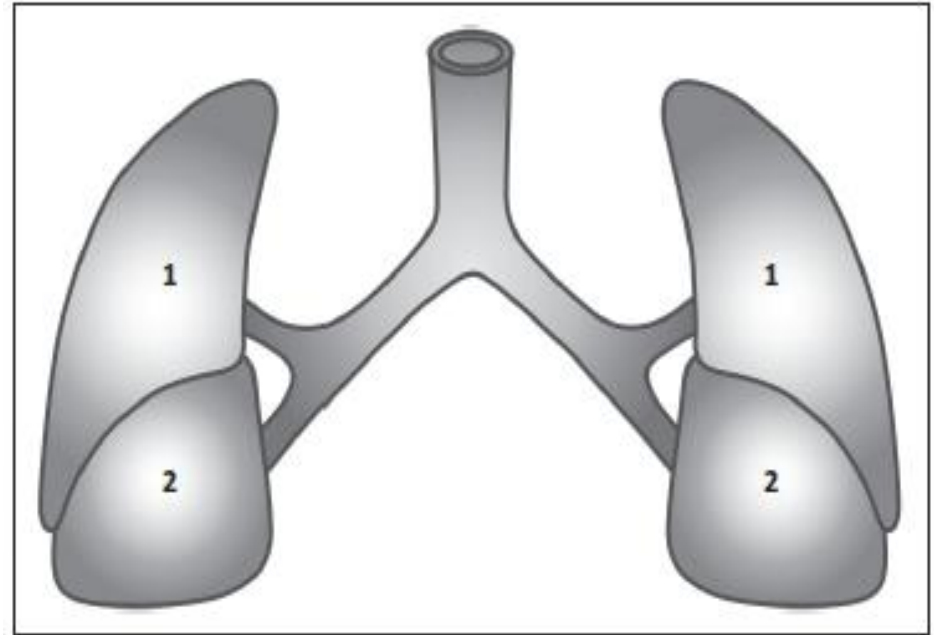
2. affects the structure of the lungs (such as the number of lobes in each lung and the length of the bronchi)

3. In the abdomen, the condition can cause a person to have no spleen (asplenia) or multiple small, poorly functioning spleens (polysplenia).

# *Bronchial anatomy in Heterotaxy*



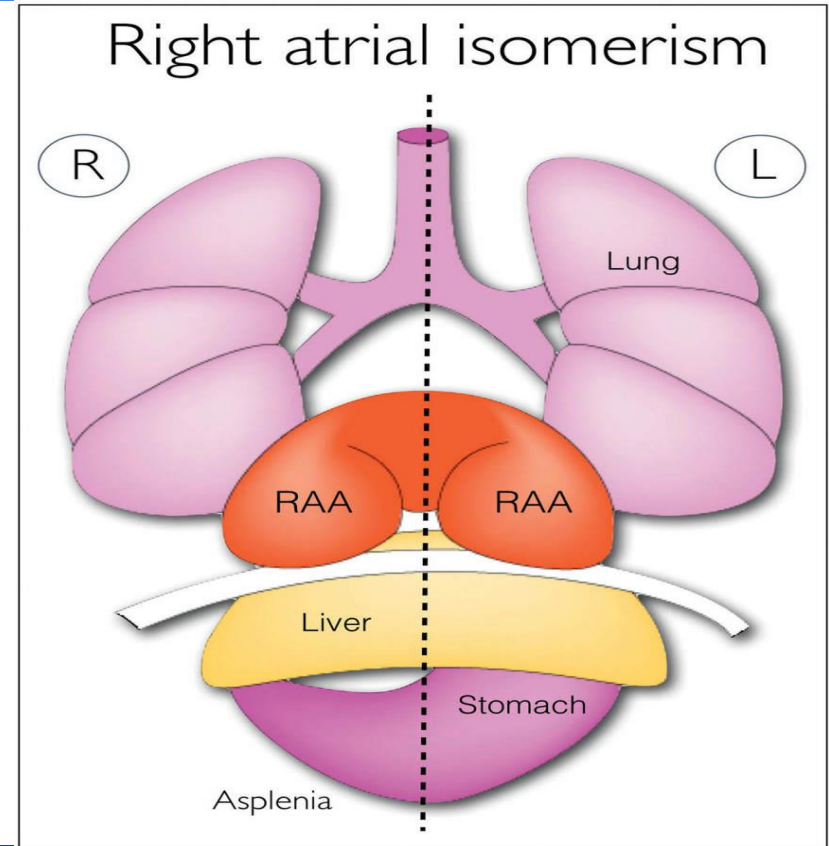
**FIGURE 12:** Illustration of trilobed lungs with eparterial bronchus in right isomerism.



**FIGURE 13:** Illustration of bilobed lungs with hyperterhal bronchus in left isomerism.

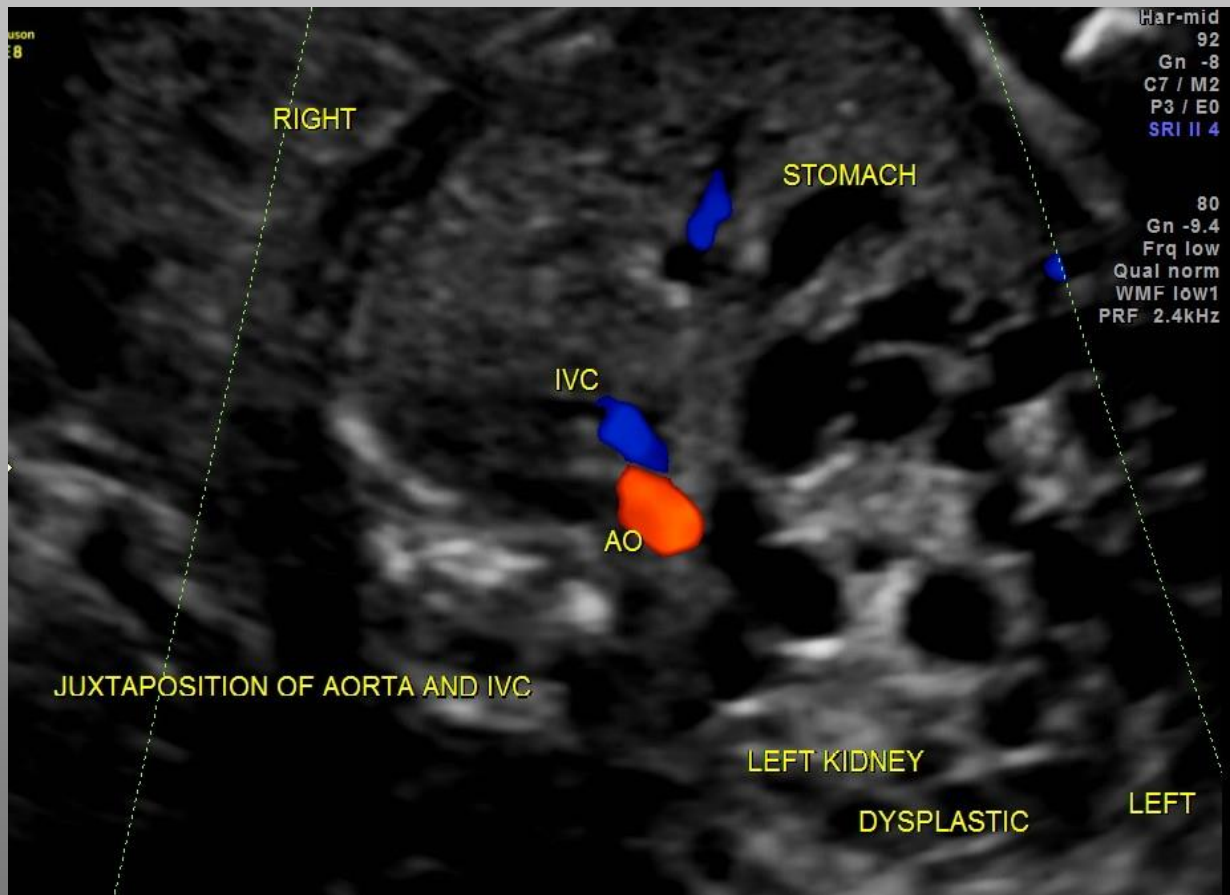
# Right Atrial Isomerism

Liver	Symmetrical and horizontal
Stomach	The middle, right, or left-sided
Spleen	No spleen- asplenia

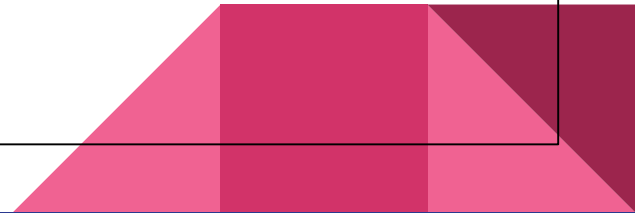


# Right atrial isomerism

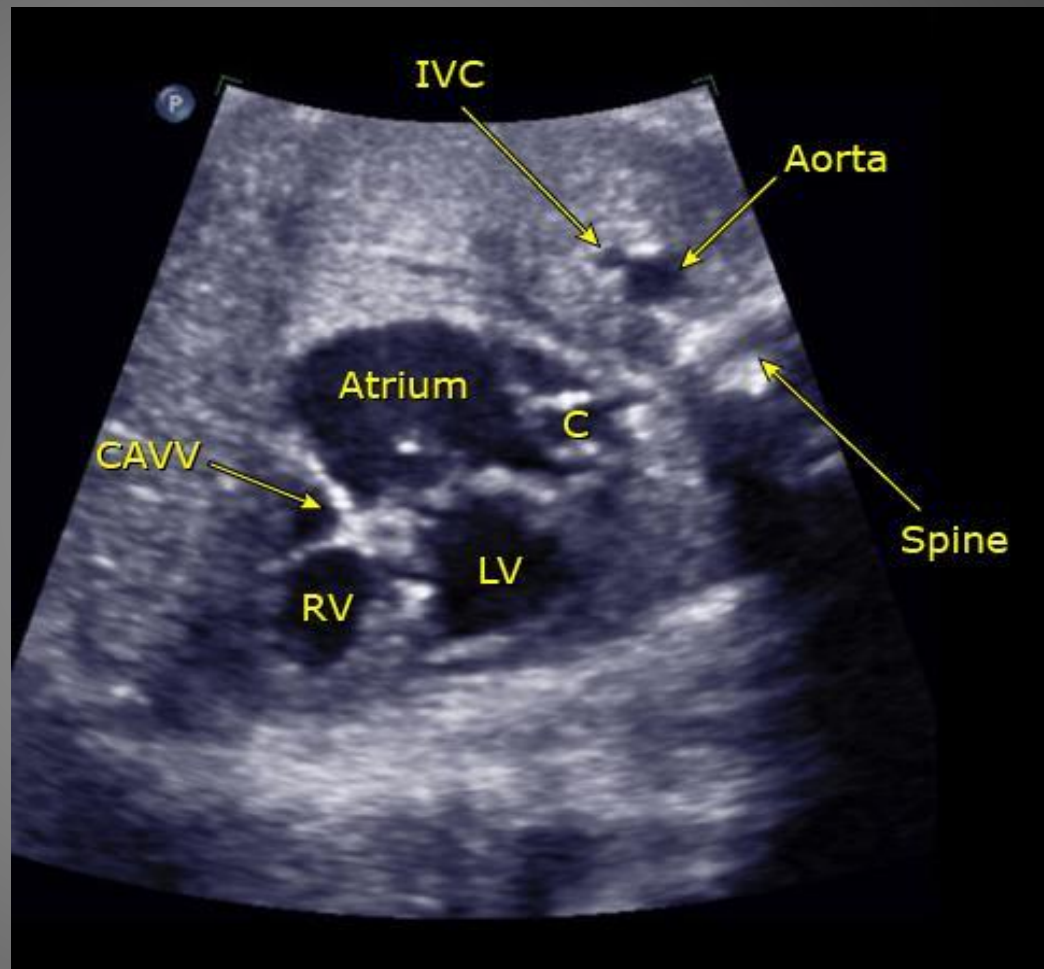
Bronchi	Both short/ eparterial
Lungs	Both trilobed
SVC	Bilateral: one to each atrium
Coronary Sinus	Absent
IVC	Present; can connect to either atria, but is always on same side as DAo
SA node	Bilateral with one dominant- SVT
Atria	Bilateral right atria with 2 right atrial appendages
AV valves	common
Ventriculoarterial connection	Malposed (~95%); commonly discordant (~5% DORV)
Pulmonary outflow obstruction	Severe pulmonary stenosis or atresia in >90%
Pulmonary veins	Extracardiac TAPVD



[https://www.uptodate.com/contents/images/PEDS/91439/Fetal\\_echo\\_R\\_atri\\_iso\\_video.mp4](https://www.uptodate.com/contents/images/PEDS/91439/Fetal_echo_R_atri_iso_video.mp4).

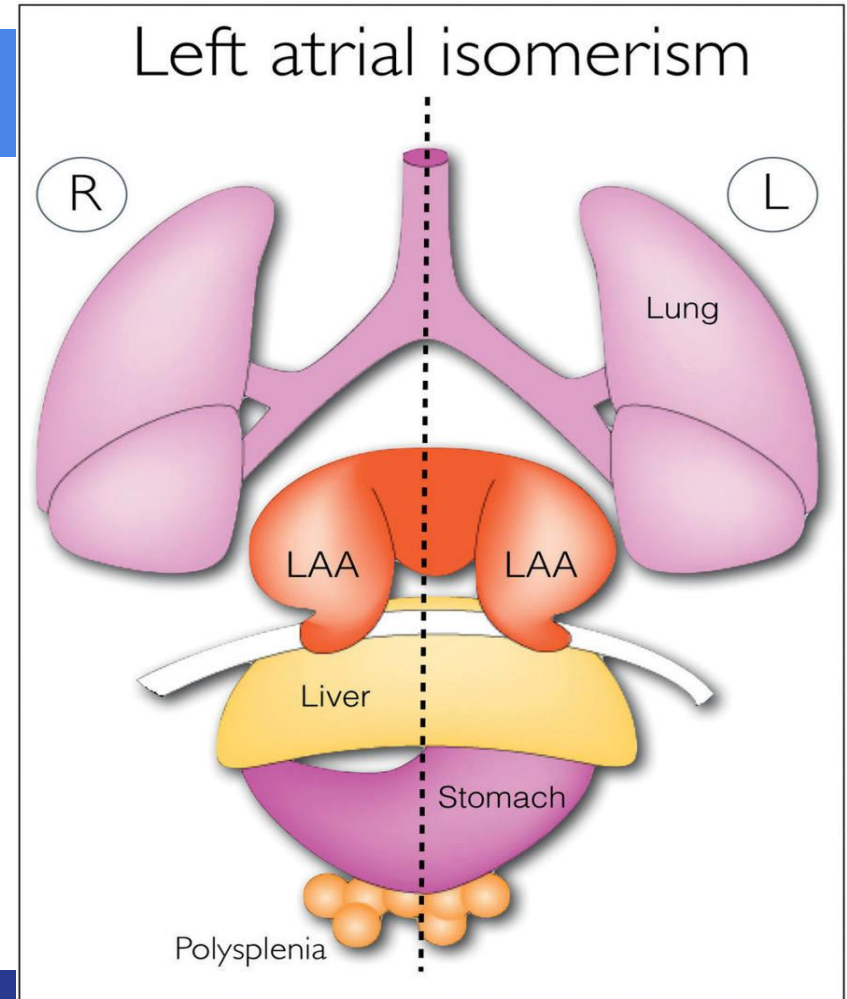


# Right Isomerism



# Left Atrial Isomerism

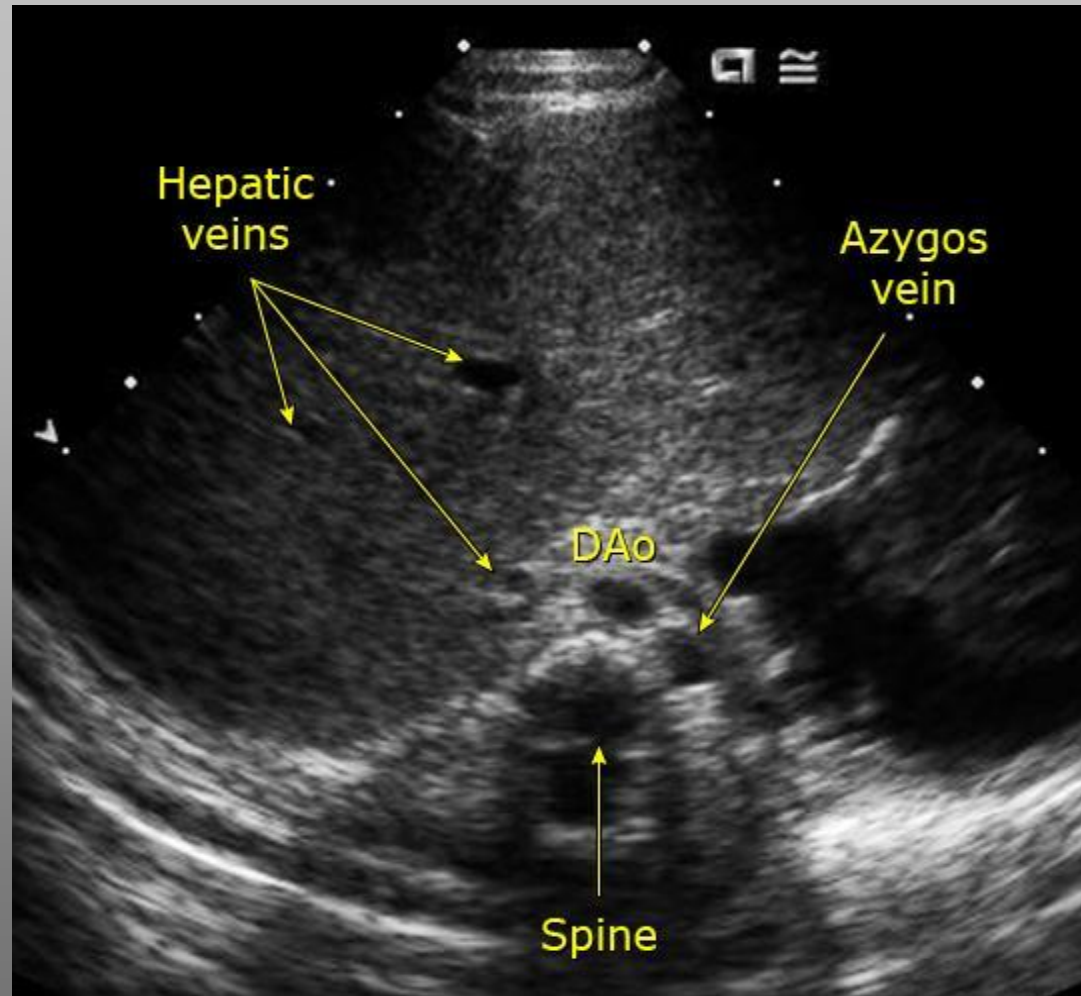
Liver	Bilobed, often to the left side
Stomach	Usually right-sided
Spleen	Polysplenia

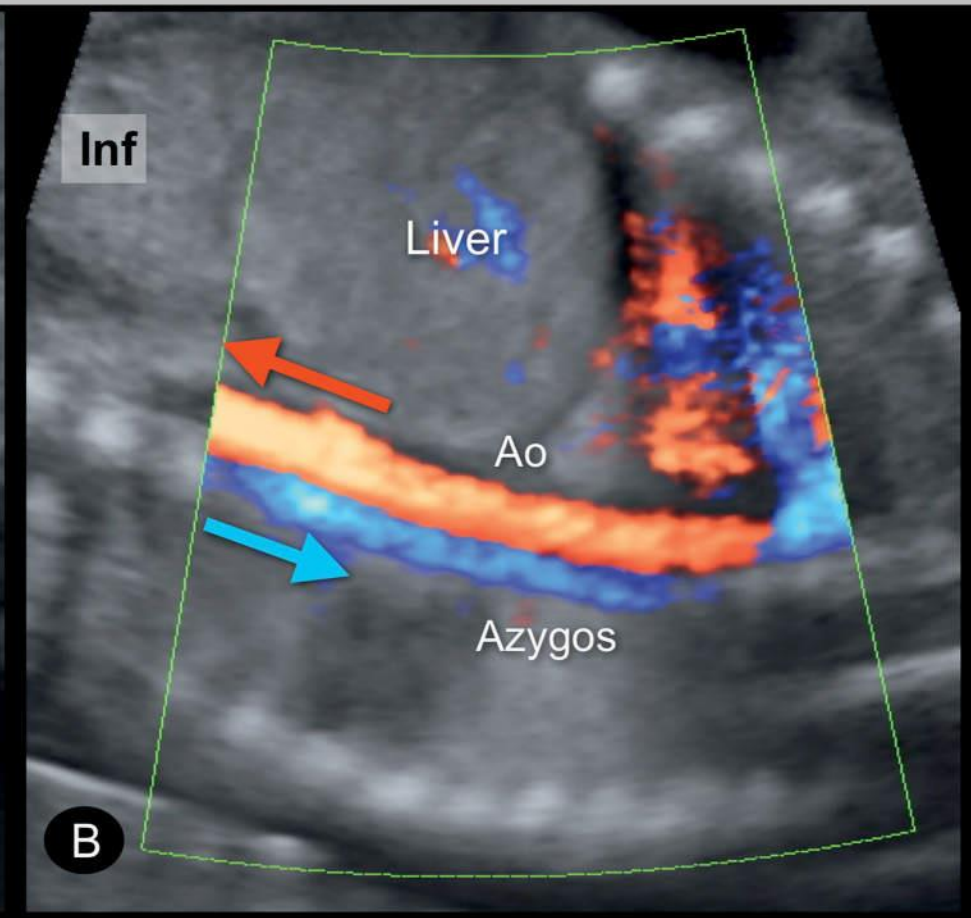
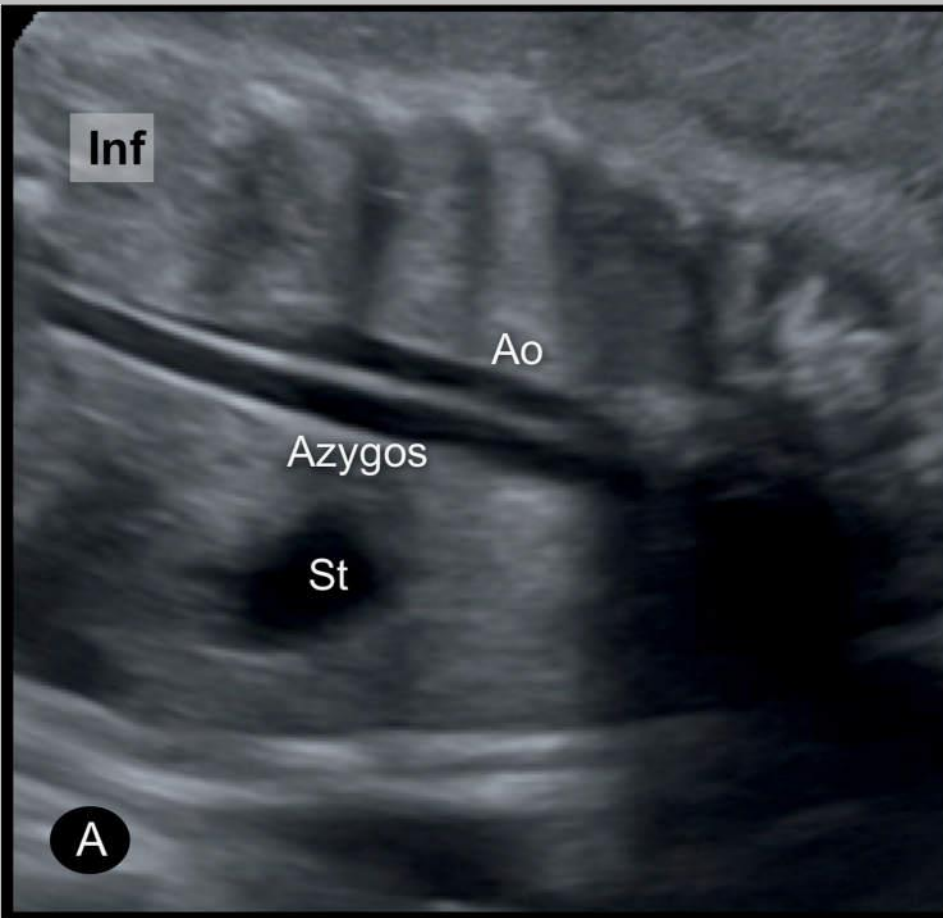


# Left atrial isomerism--(LAI)

Bronchi	Both long and hyparterial
Lungs	Bilateral bilobed
SVC	May be bilateral: one to each atrium (can be via coronary sinus)
Coronary Sinus	Generally present
IVC*	Usually interrupted with azygos continuation
Atria*	Bilateral left atria
SA node*	Absent; complete ht block/bradycardia
AV valve	2 or AVSD type
Ventricles	D-looping
Great arteries	Generally normal, occasional DORV
Left-sided obstruction	Common (MS, AS, CoA)

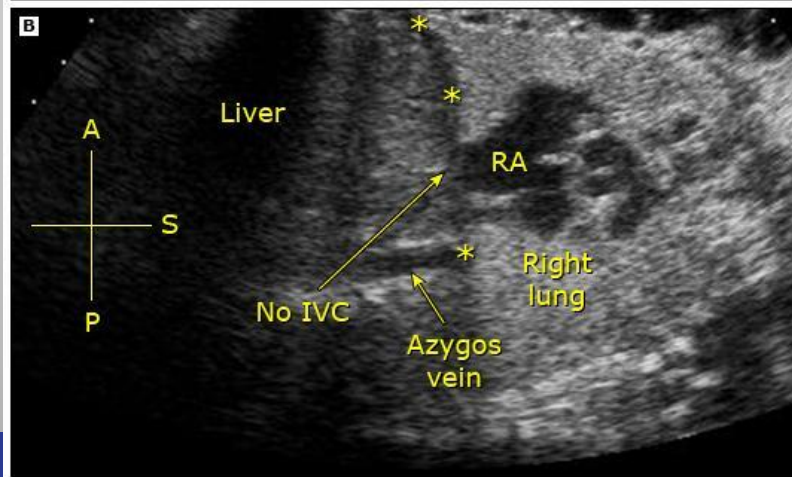
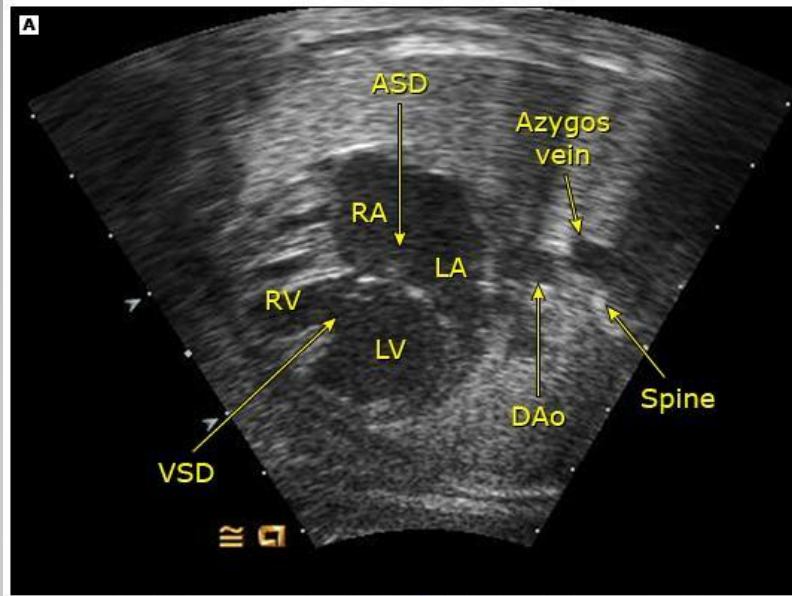
LAI





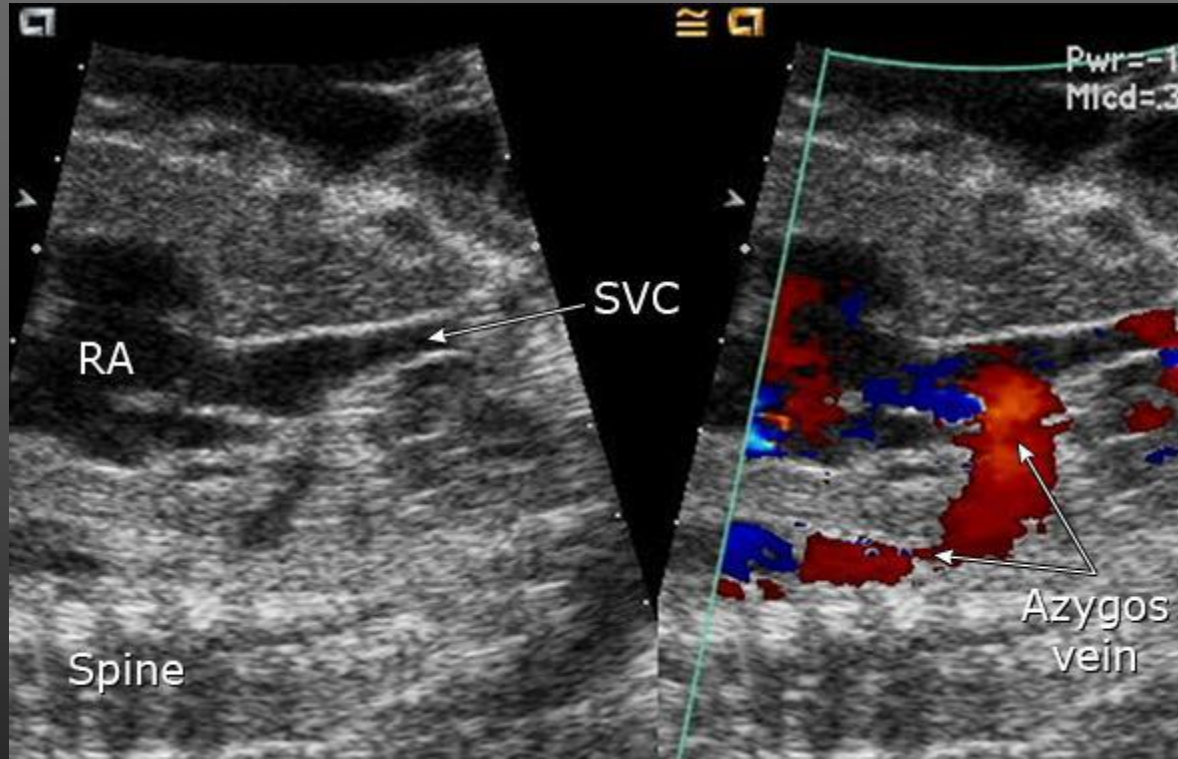
# LAI

(Fetal)



# LAI

(Fetal)





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thanks for listening!