# Systemic Venous Anomalies

Jenna Schermerhorn, DO

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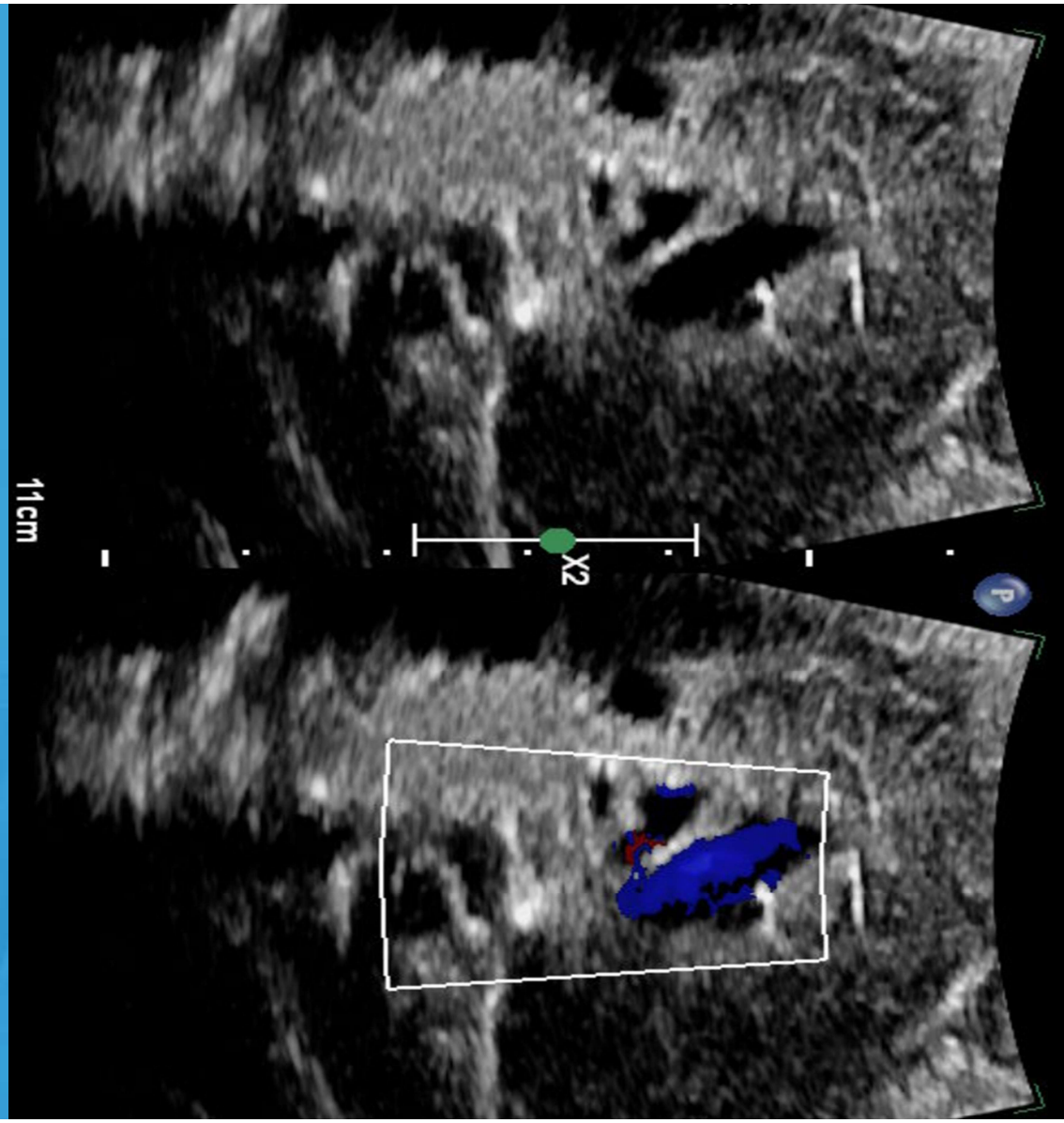


# Outline

Normal fetal venous anatomy and development
Persistent LSVC
Absent RSVC with persistent LSVC
Retroaortic innominate vein
Interrupted IVC
Agenesis of the ductus venosus
Persistent right umbilical vein
Umbilical Varix

# Disclosures

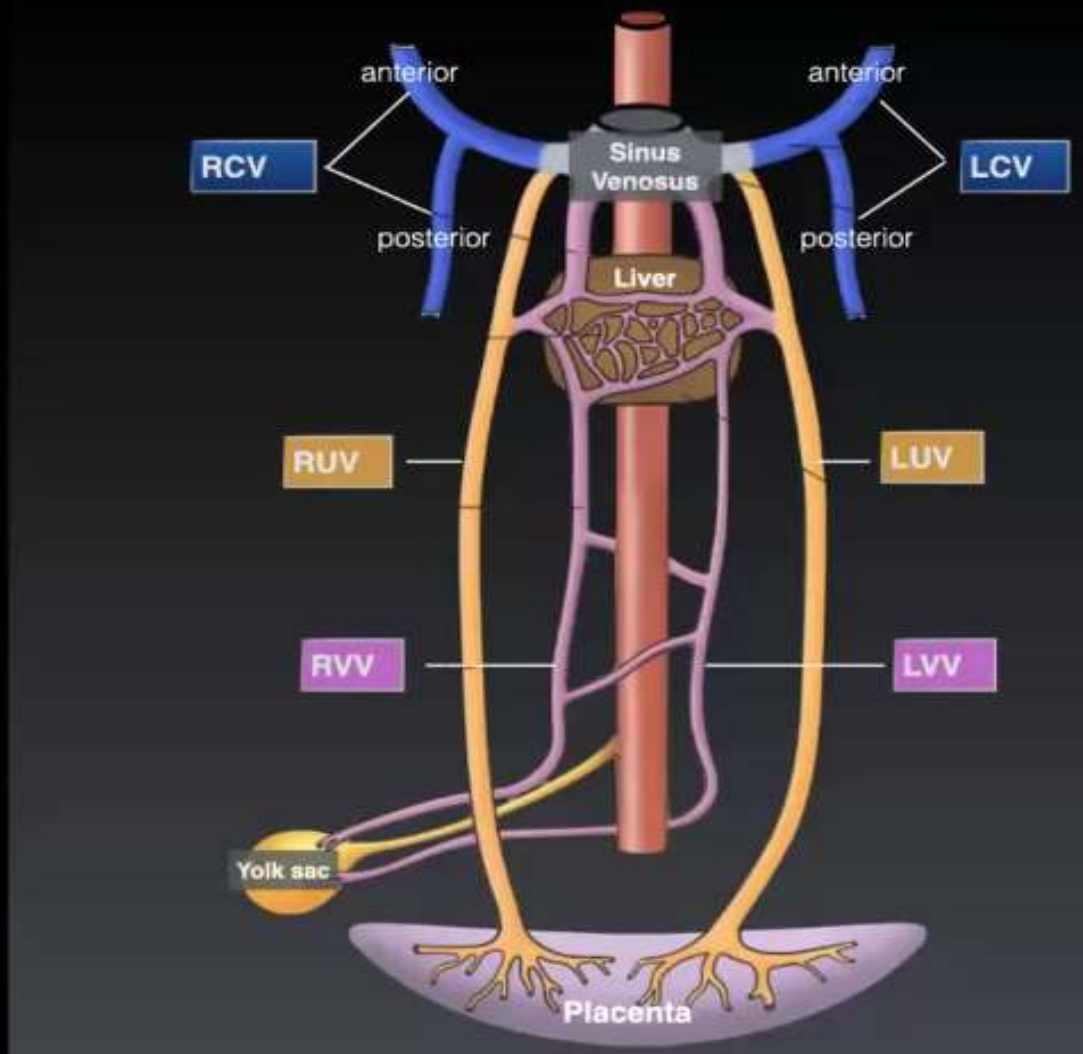
None

# Venous Embryology

**Cardinal Veins (CV)**

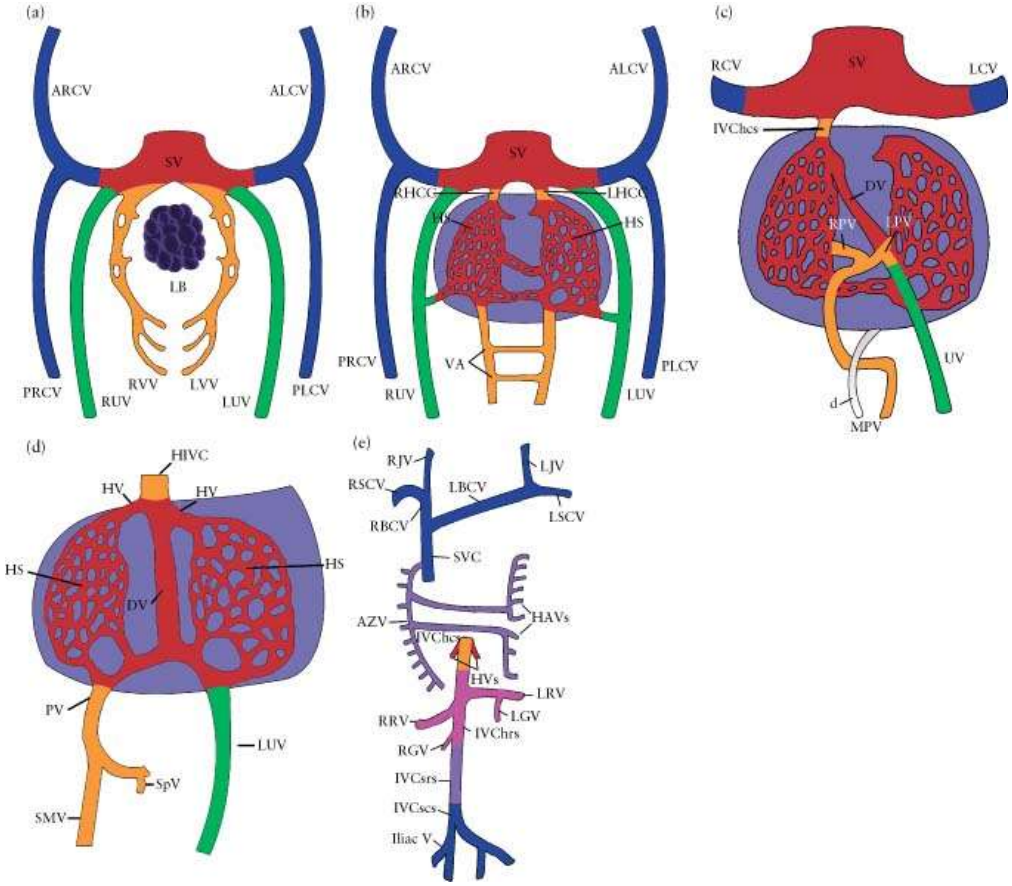
**Umbilical Veins (UV)**

**Vitelline Veins (VV)**



# Evolutionary Changes

- Liver sinusoidal connections with umbilical and vitelline veins
- Entire right umbilical vein regresses
- Proximal segment of left umbilical vein regresses
- Connection between distal left umbilical vein, ductus venosus, and proximal right vitelline vein to the right heart

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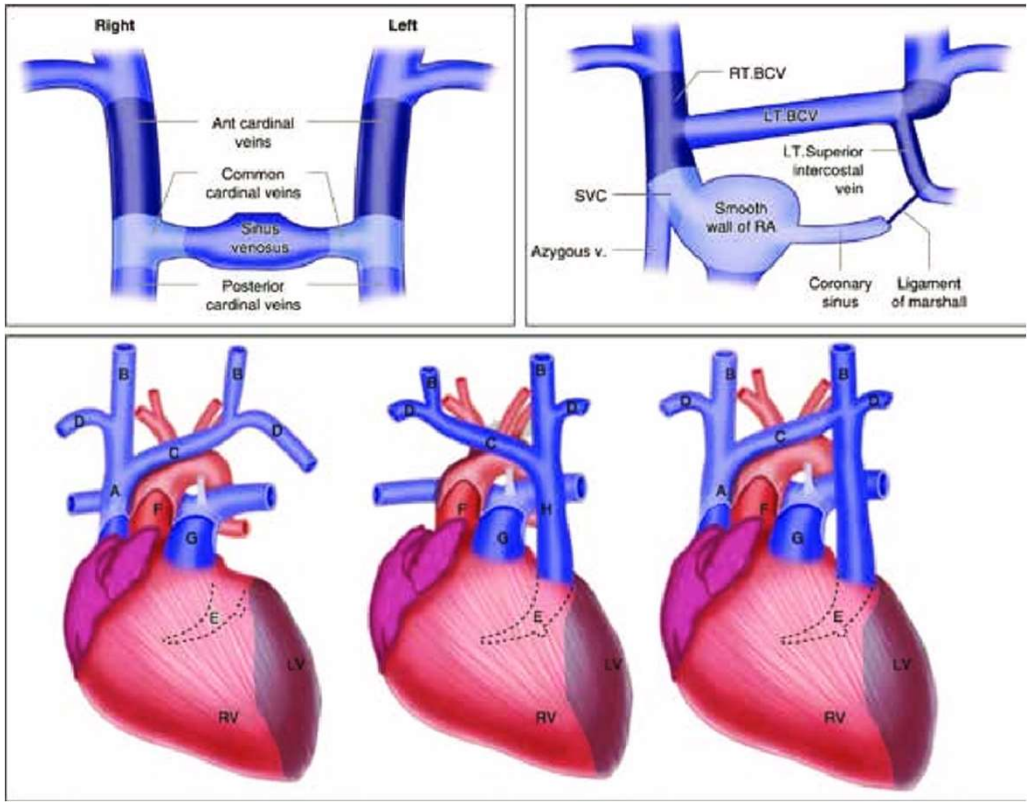
# Anterior Cardinal Veins

- Left anterior cardinal vein regresses
- Right anterior cardinal vein persists → right superior vena cava
- Left brachiocephalic vein connects your left sided venous system to the right side

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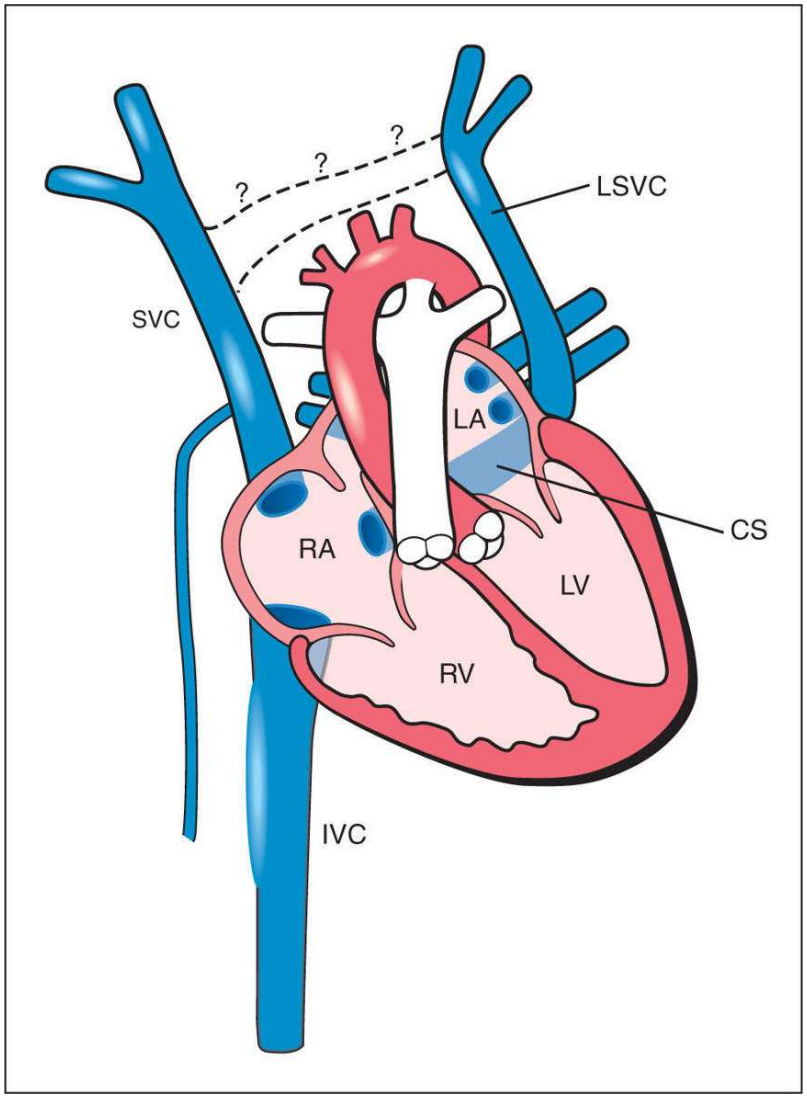
# Persistent LSVC

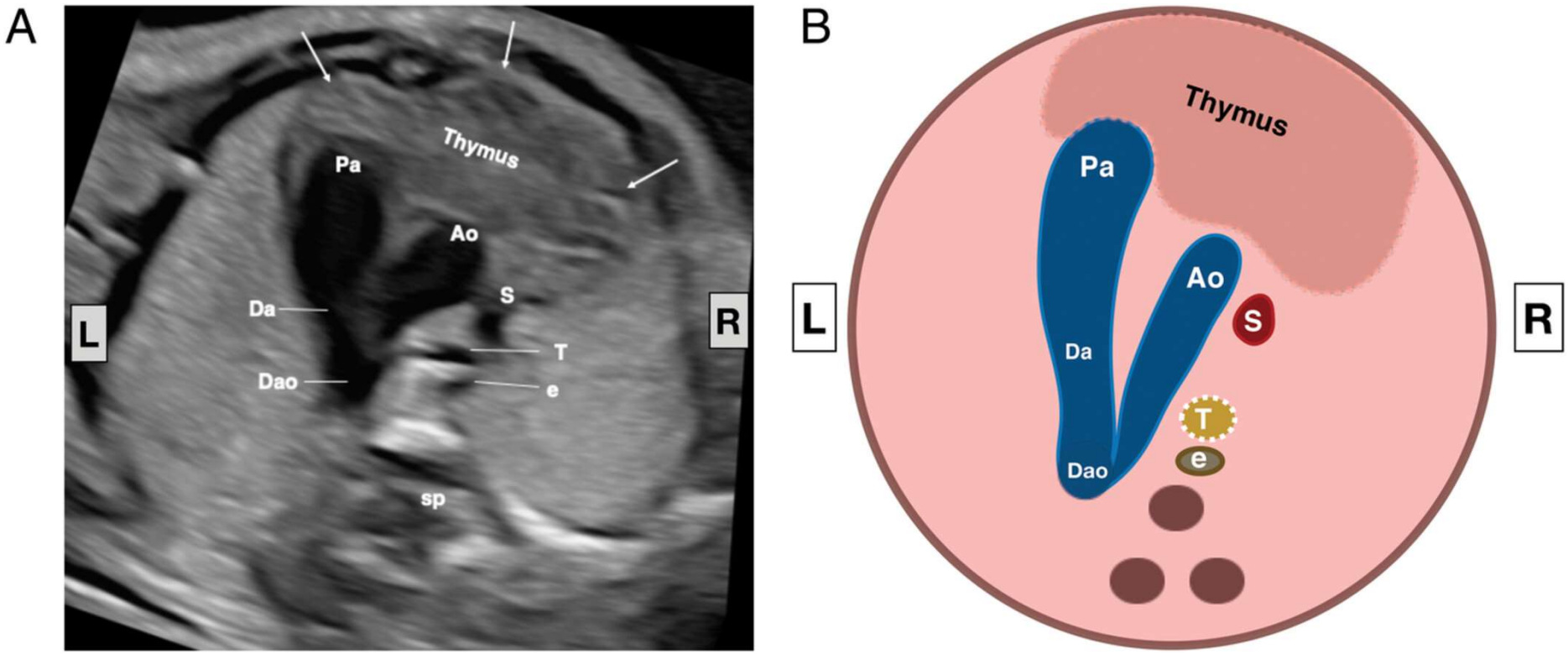
Prevalence of 0.04%-0.5%, although may be more now since the screening guidelines have recommended 3VV and 3VTV

- 80-90% of cases have an associated RSVC
- Drainage:
  - Coronary sinus
  - Partially unroofed coronary sinus
  - Directly into left atrium
- Isolated finding
- Associated with CHD
  - Heterotaxy
  - ASD
  - VSD
  - AVSD
  - Conotruncal defects (TOF)
  - Left sided obstructive lesions




https://obgynkey.com/anomalies-of-systemic-and-pulmonary-venous-connections/

# 3 Vessel Trachea View (3VTV)




https://onlinelibrary.wiley.com/doi/full/10.1002/jum.15067

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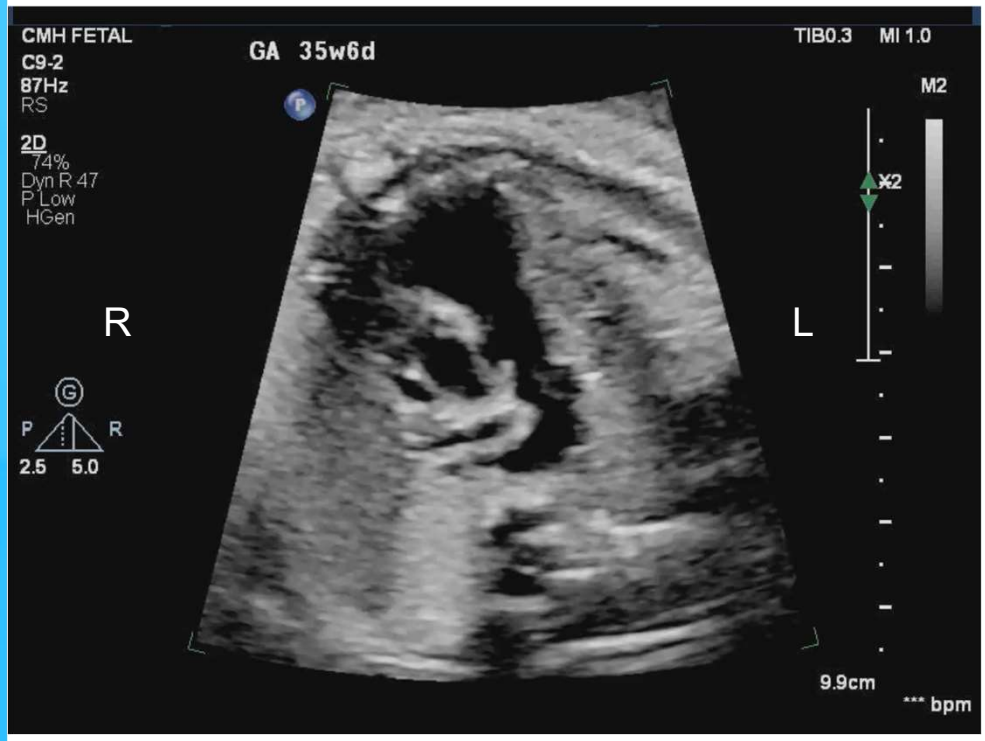
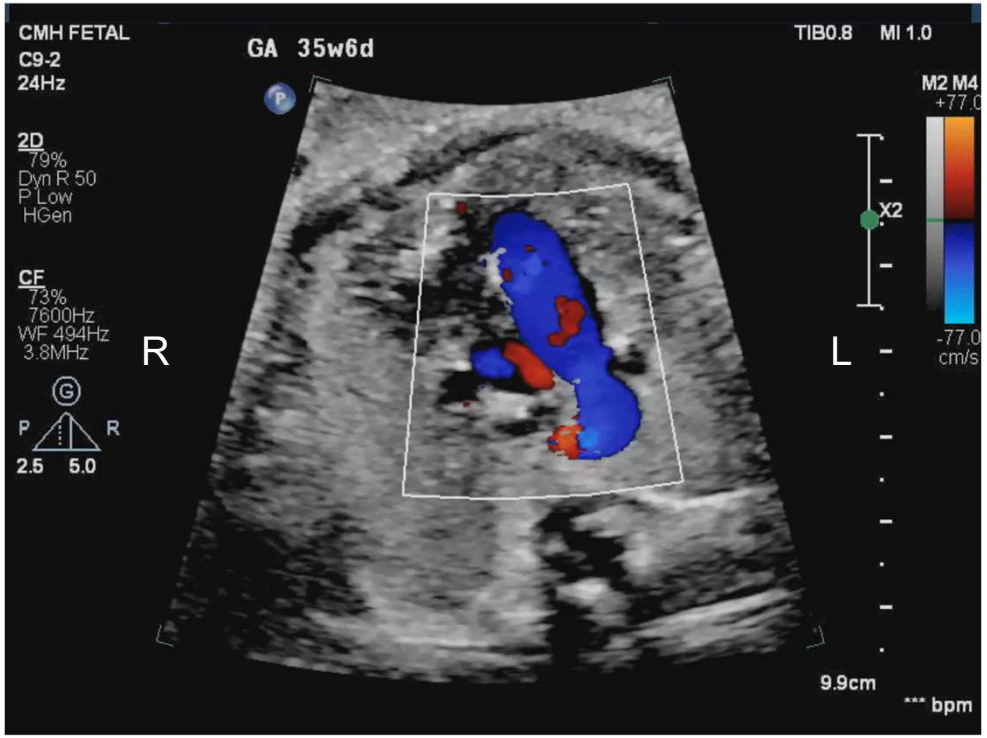
# LSVC 3VV

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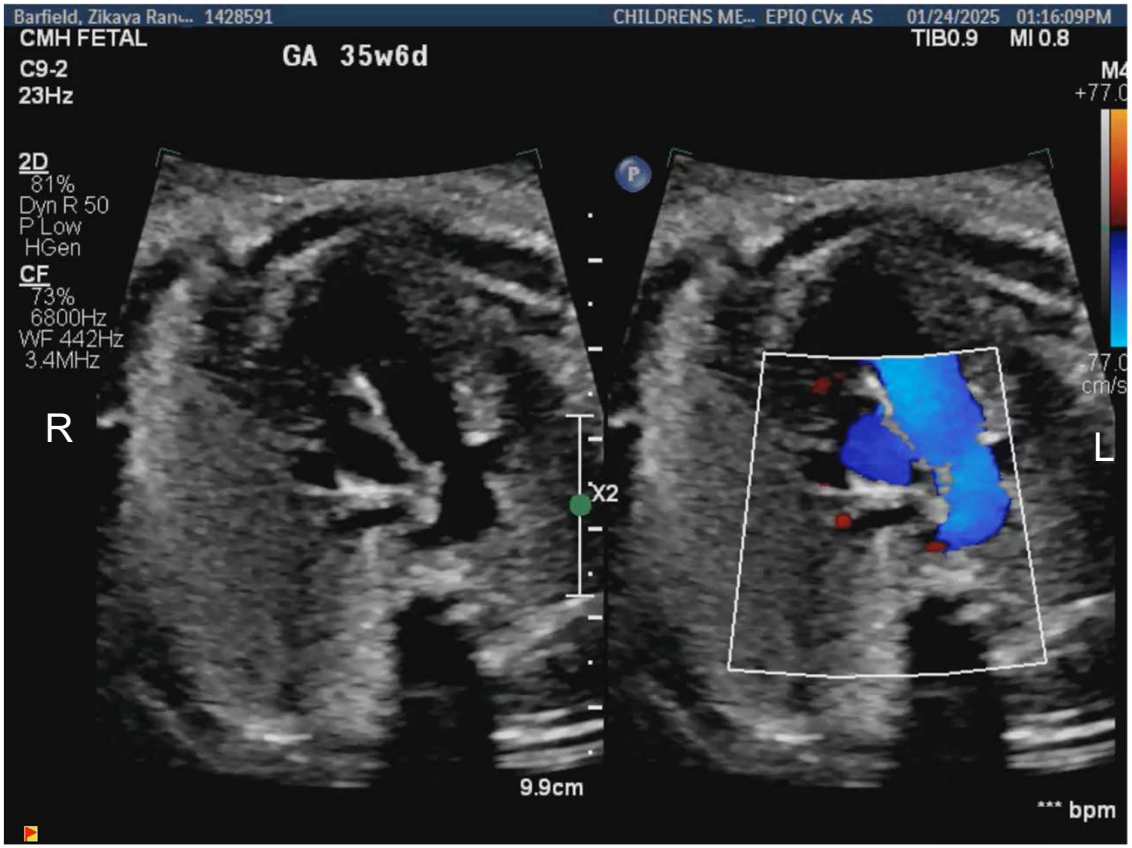
# LSVC 3VTV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01/24/2025 01:16:09PM
TIB0.9 MI 0.8
CMH FETAL
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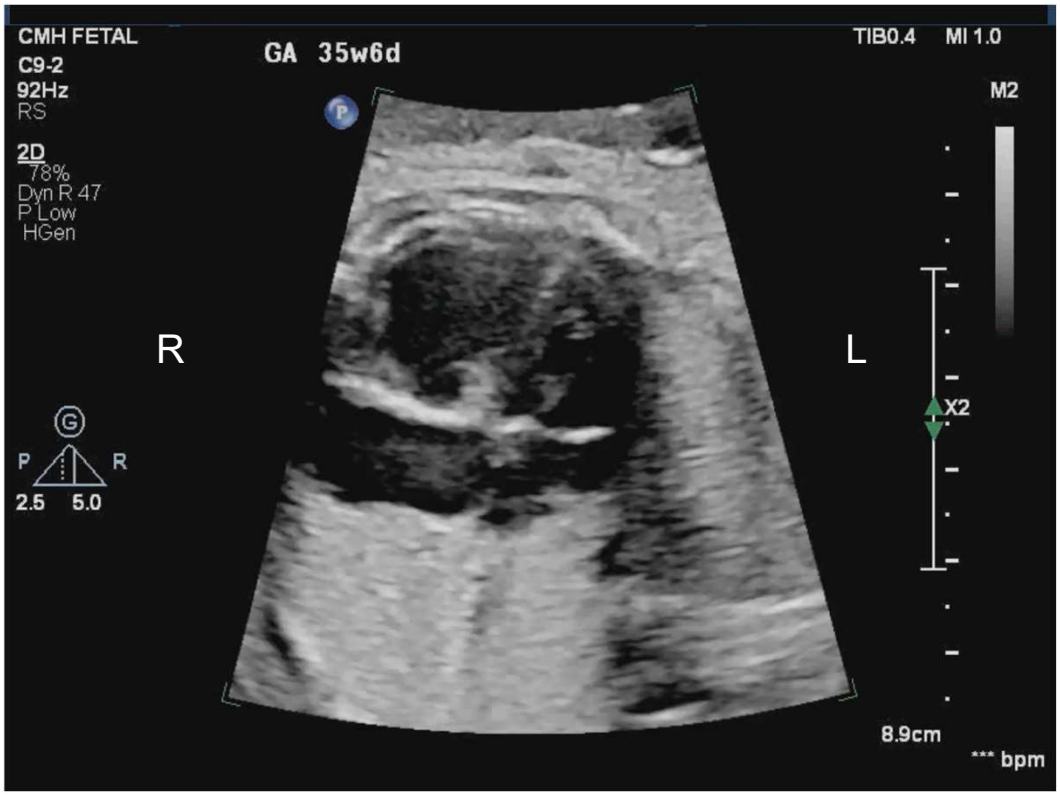
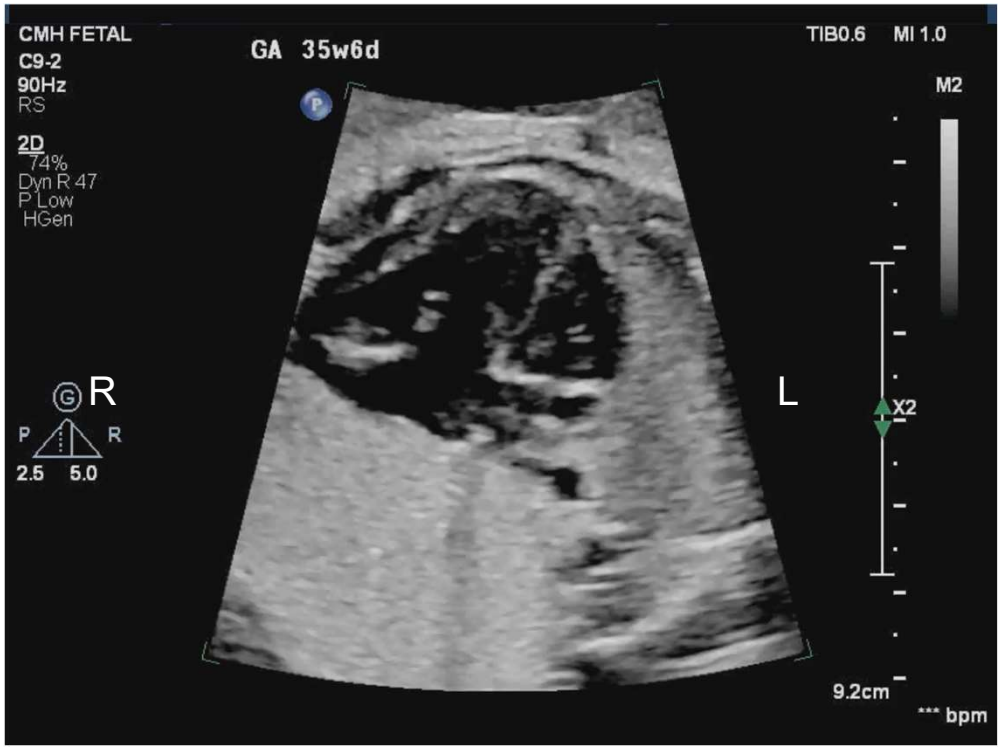
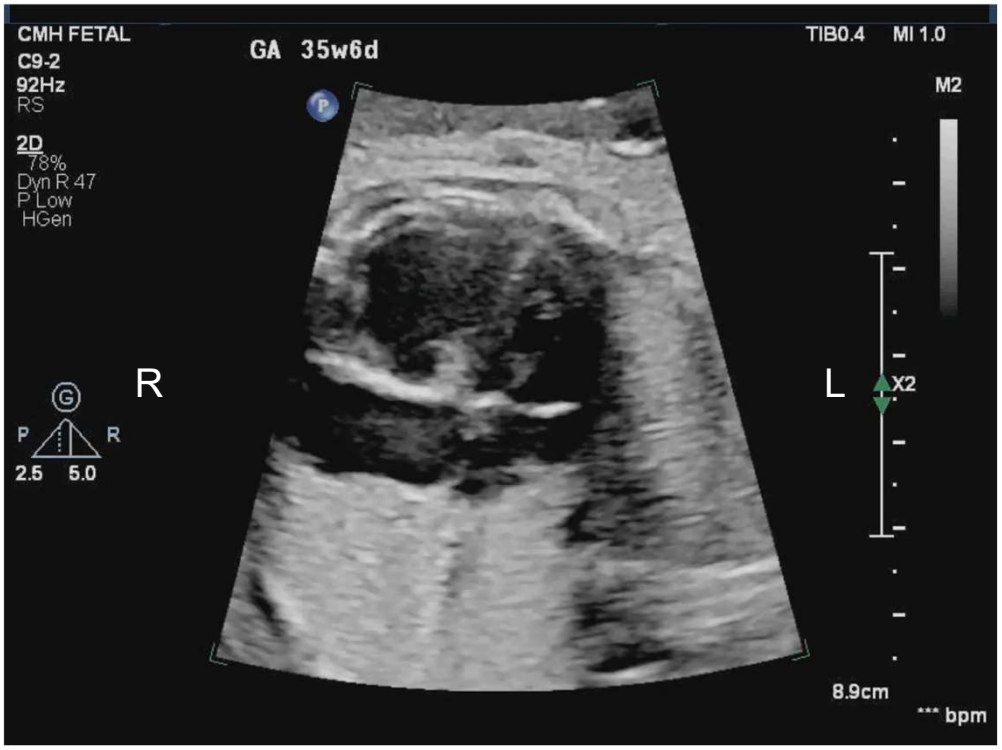
# LSVC Dilated CS



# LSVC A4C

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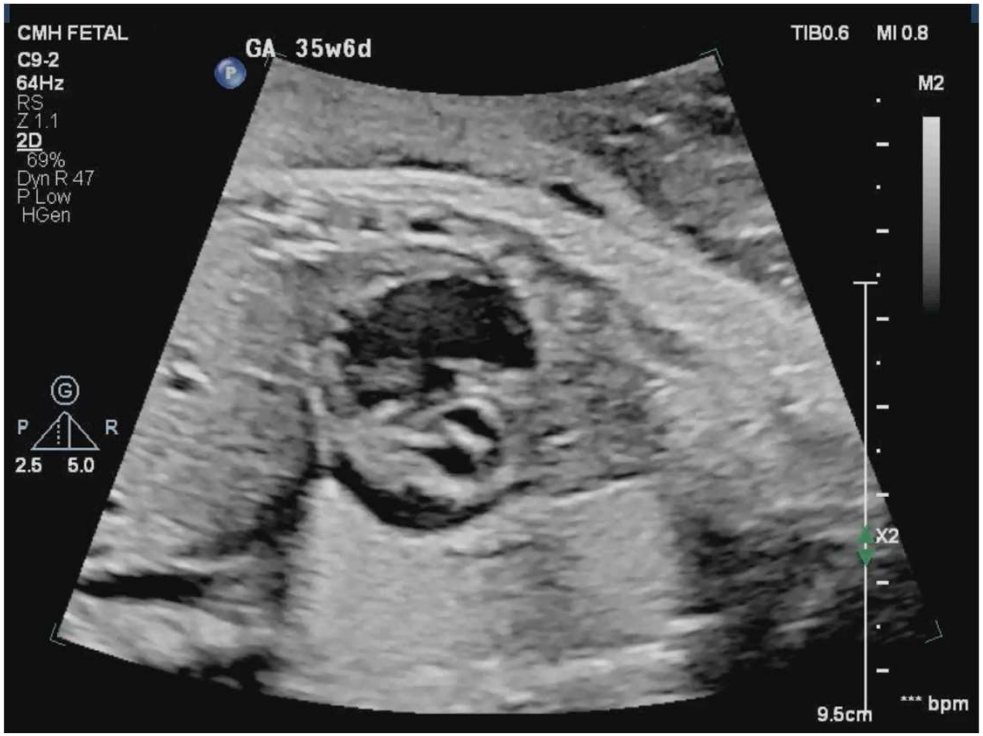
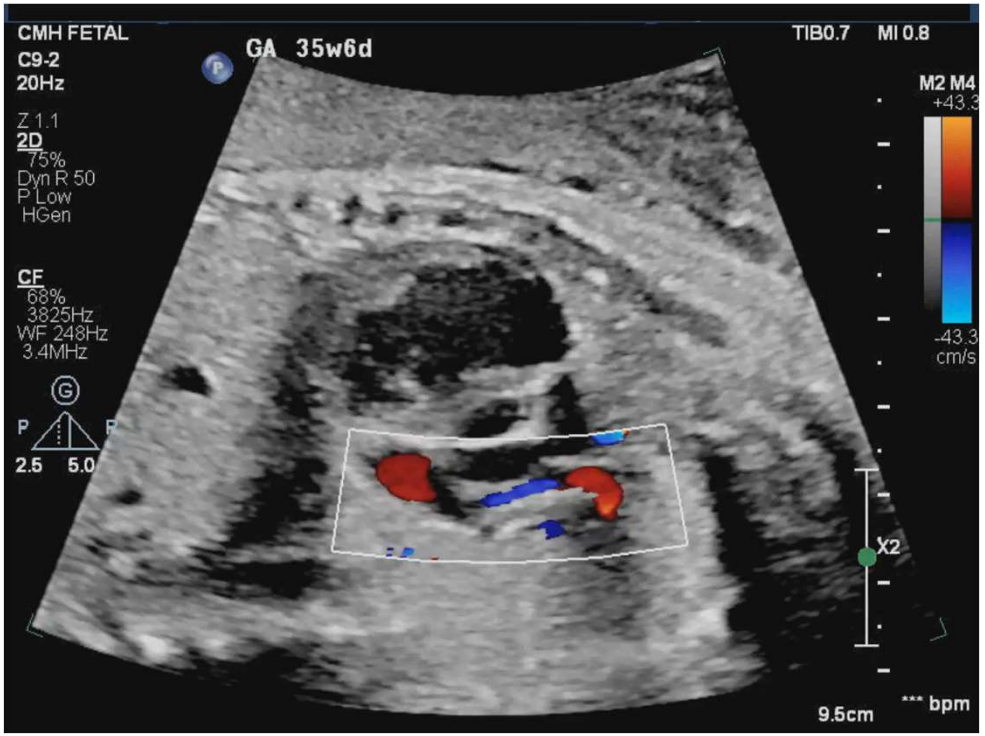
# LSVC Sagittal

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# Follow-up

Dependent on presence of associated CHD

If isolated:

- Dependent on left sided structures

Postnatal echocardiogram to confirm normal intracardiac anatomy

No long-term surveillance is currently recommended

# Potential Long-Term Sequelae

## Arrhythmias in Children Having a Single Left Superior Vena Cava and Minimal Structural Heart Disease

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From the *University of Miami School of Medicine, Jackson Memorial Hospital, Miami, Florida, USA; and †Duke University School of Medicine, Duke University Medical Center, Durham, North Carolina, USA

**Pediatric Arrhythmias with Single Left Superior Vena Cava.** ***Background:*** **The presence of a single left superior vena cava in the absence of complex congenital heart disease is uncommon, and, in the absence of hemodynamic consequences, it would not be expected to result in cardiovascular signs or symptoms. Single case reports and our anecdotal experience suggested to us that this anomaly is highly associated with cardiac arrhythmias.**

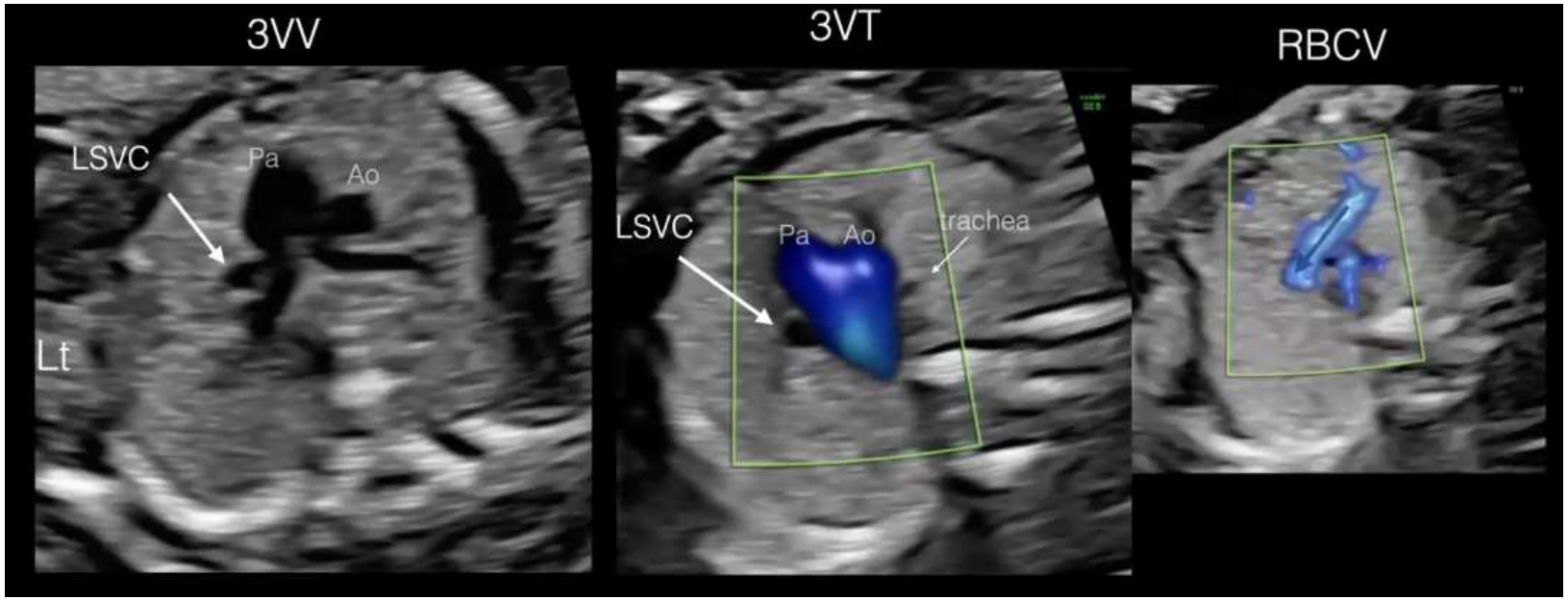
***Objective:*** **We sought to describe the clinically important arrhythmias in a population of young patients having this anomaly.**

***Methods:*** **A retrospective chart review was performed from all patients <20 years old and who were determined by echocardiography over an 11-year-period to have a single left superior vena cava and minor or no coexisting congenital heart defects. The prevalence of nonsinus pacemaker, age-corrected sinus rate percentile, and prevalence of brady- or tachyarrhythmias was compared with a control group of patients having bilateral superior vena cavae.**

***Results:*** **Eight patients having a single left and 55 patients having bilateral superior vena cava(e) were identified. The existence of this anomaly tended to be associated with a lower age-corrected sinus rate percentile (17.5% vs 75%, P = 0.09), and was associated with a higher prevalence of arrhythmias (50% vs 7%, P = 0.014) compared with the control group. In the study group, one patient each had clinically relevant sinus node dysfunction, third-degree AV block, Wolff-Parkinson-White syndrome and atrial fibrillation, and AV nodal reentrant tachycardia.**

***Conclusion:*** **Even in the absence of symptoms, patients found to have a single left superior vena cava should be monitored long-term for clinically important arrhythmias.** *(J Cardiovasc Electrophysiol, Vol. 20, pp. 182-186, February 2009)*

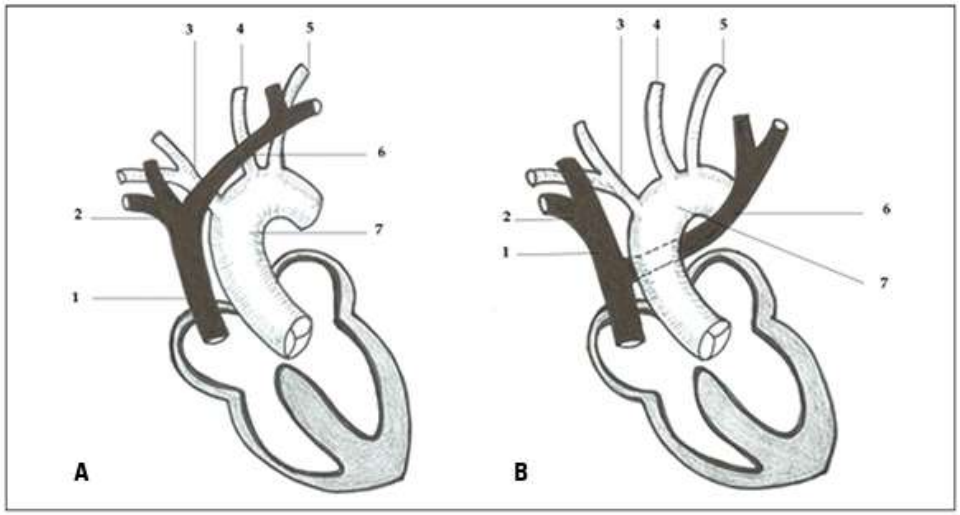
# LSVC with absent RSVC



# Retroaortic Innominate (Brachiocephalic) Vein

Rare – incidence not well known

Left brachiocephalic vein → horizontal course crossing anterior mediastinum and descending obliquely towards the right and merging with the right SVC

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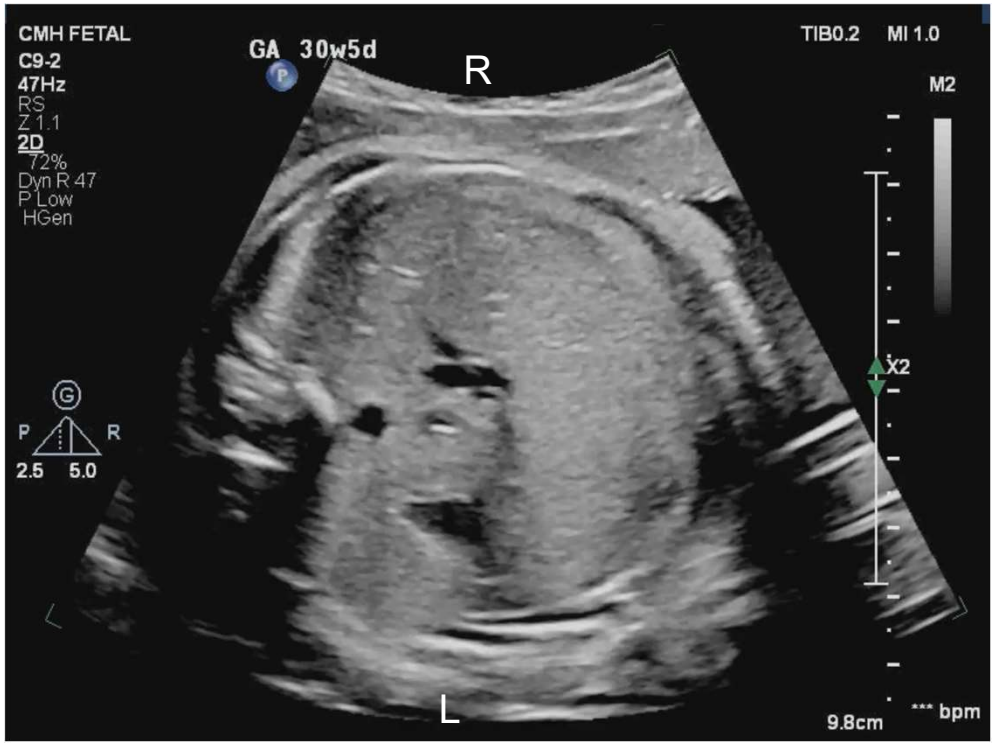
# Retroaortic Innominate Vein

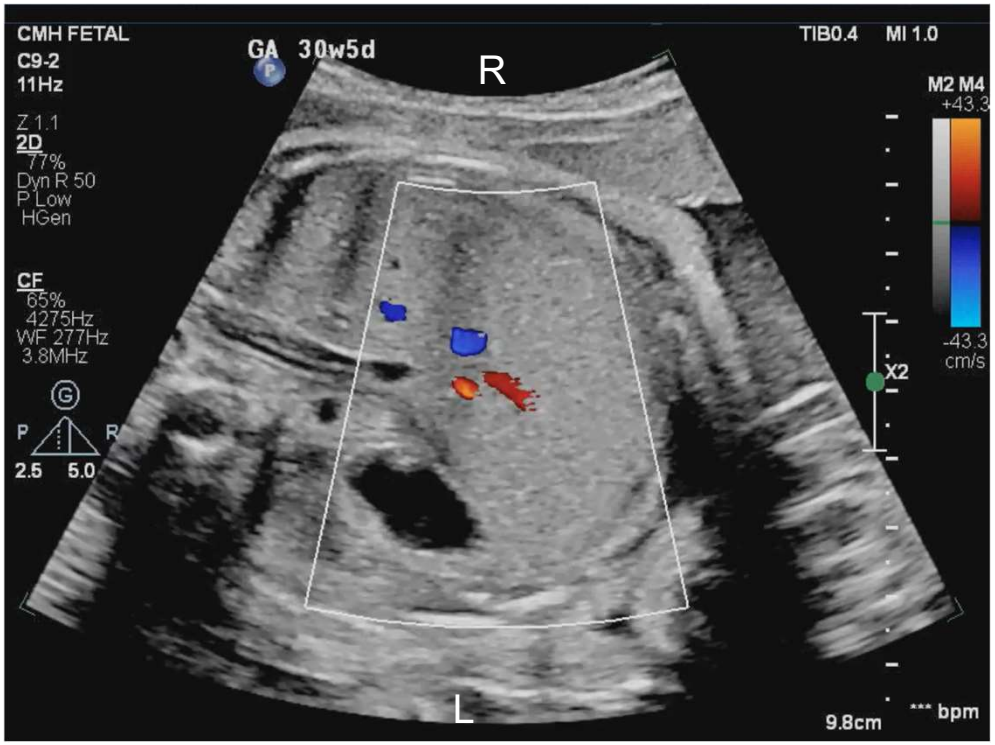
Isolated

Associated CHD:

- TOF (most common)
- ASD
- VSD
- PS
- Aortic arch anomalies

# Normal Sweep

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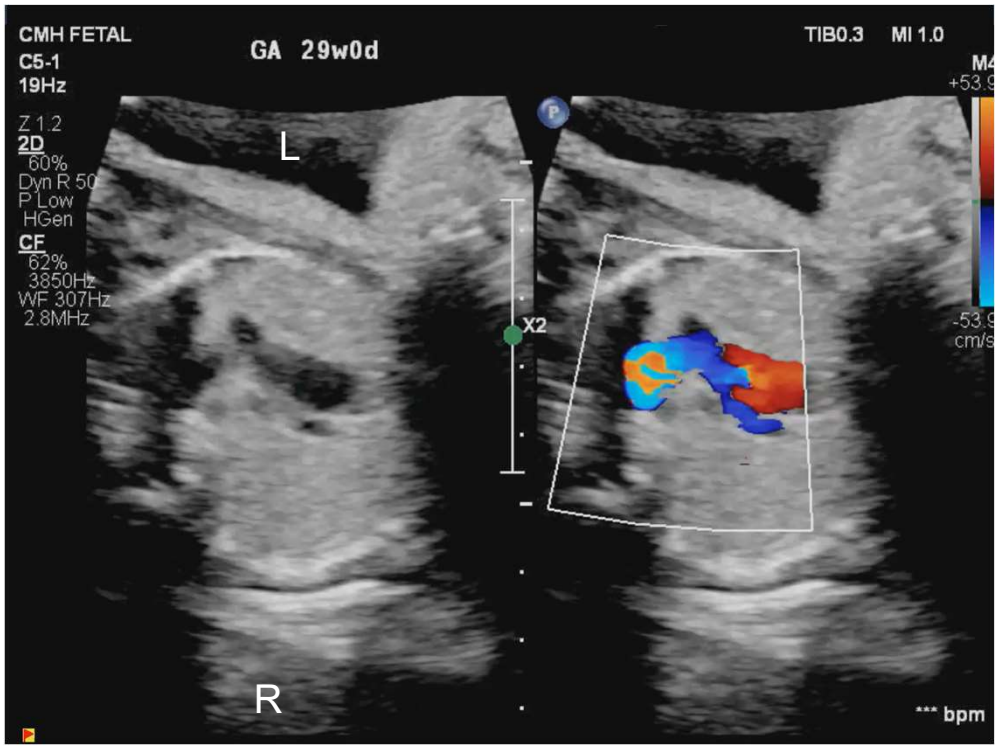
# 3VV/3VTV Retroaortic Innominate Vein



# Importance of a Retroaortic Innominate Vein

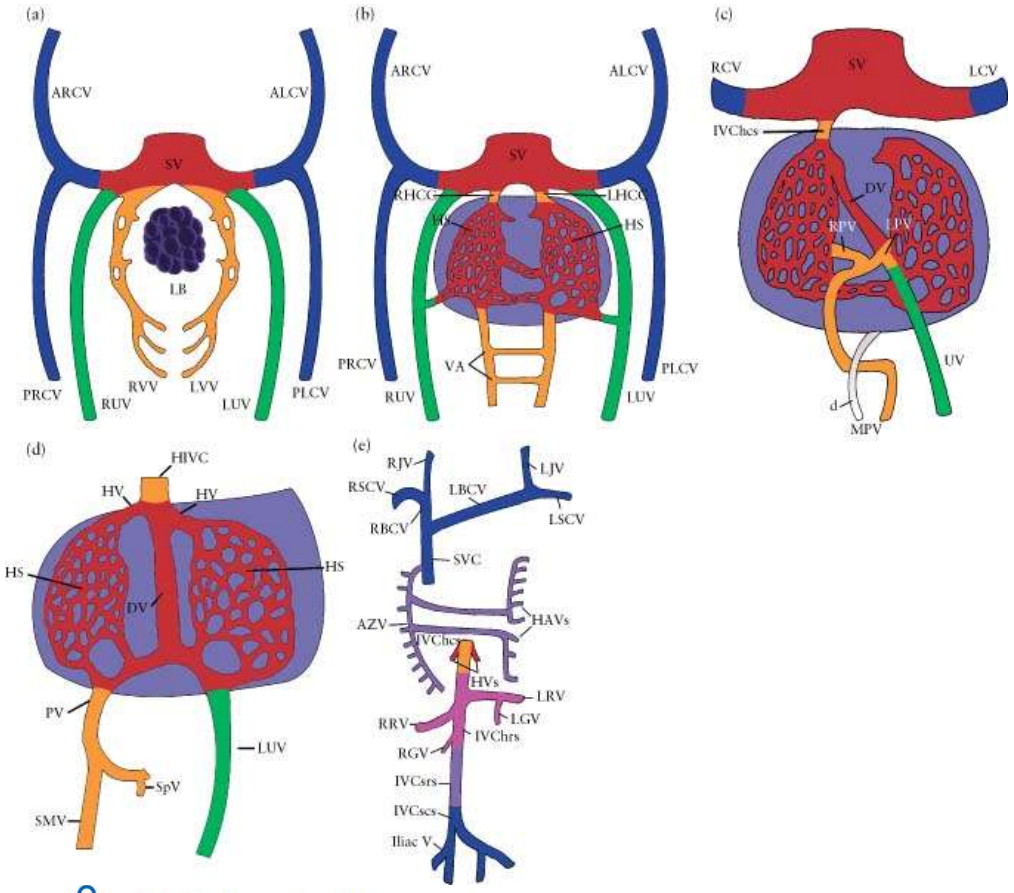
Knowledge of commonly associated CHD

Isolated finding – normal variant

Unawareness can lead to surgical complications

Difficulty with placement of left sided central lines or transvenous pacemakers

# Systemic Venous Development

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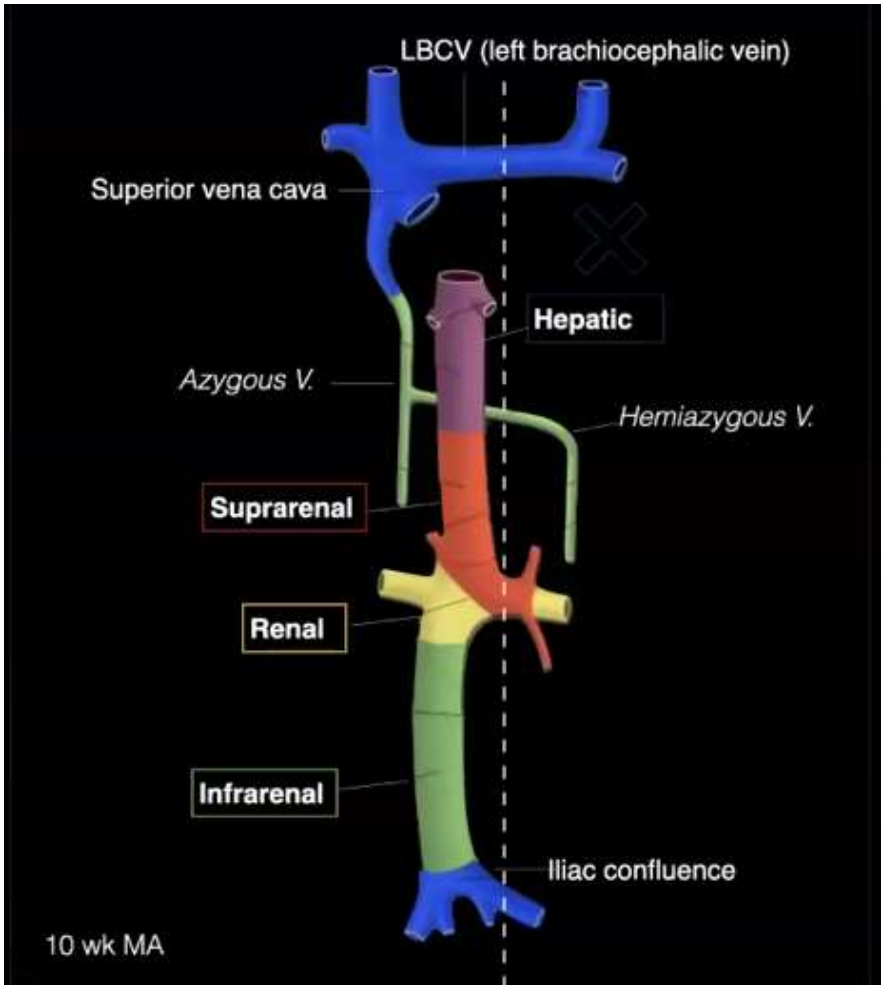

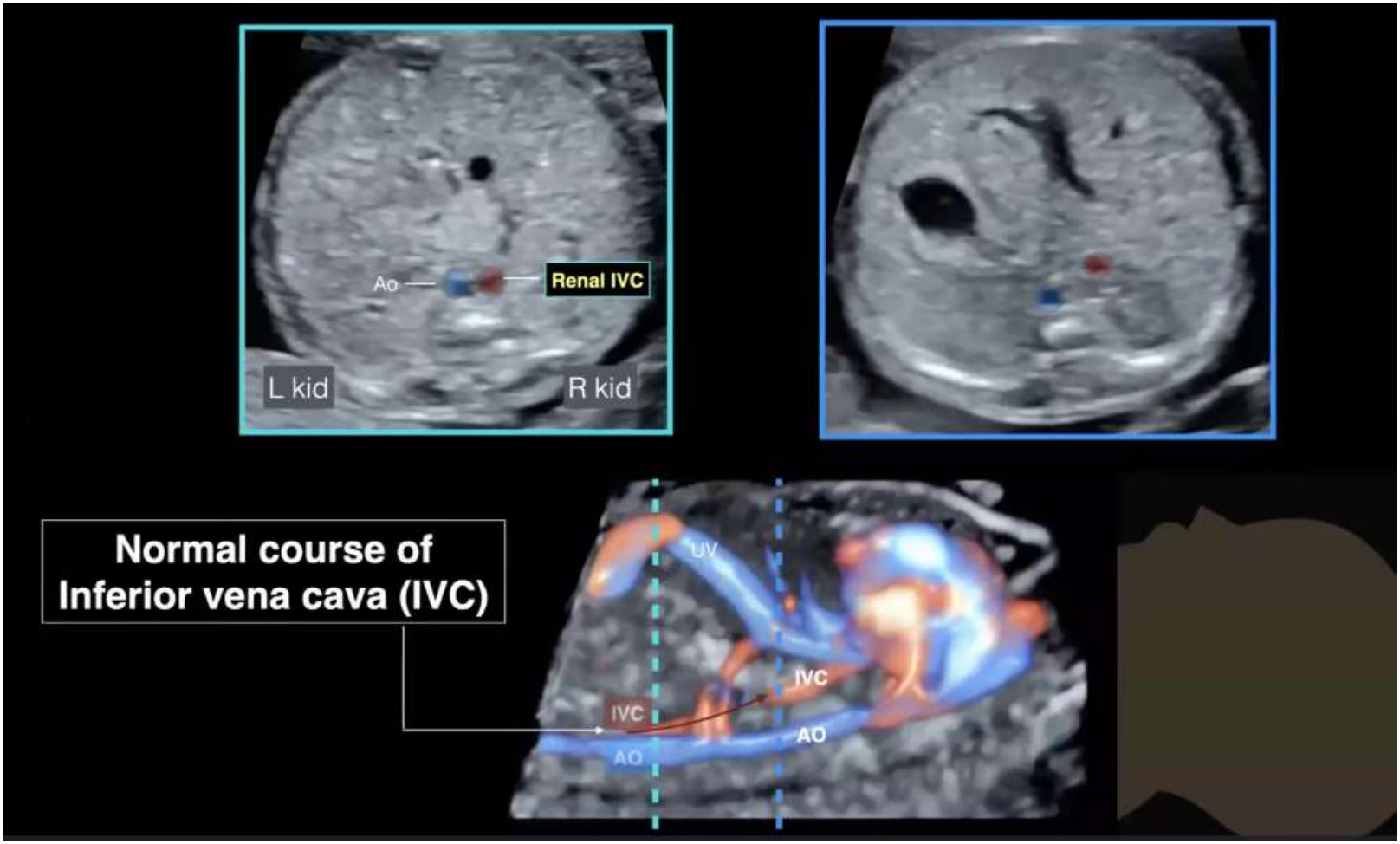


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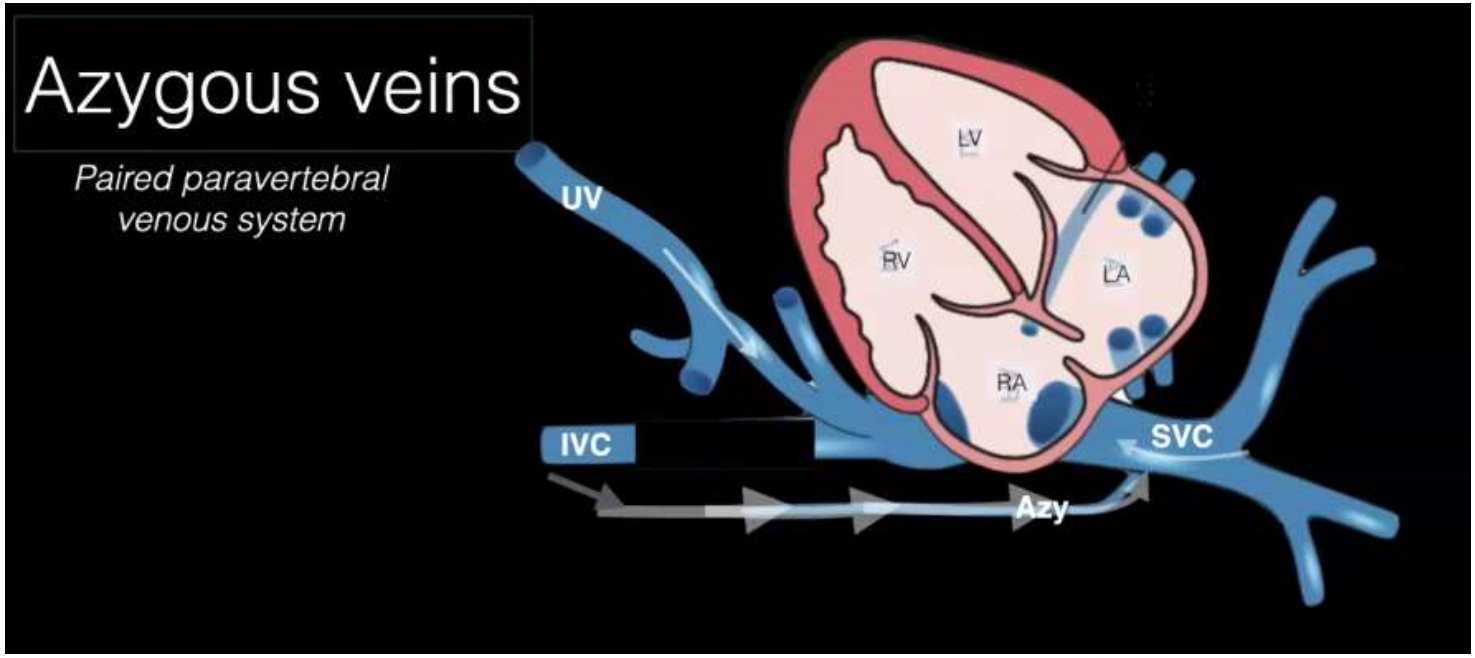
# Interrupted IVC

Occurs when there is an absent hepatic segment → infrahepatic IVC then makes connection with the azygous system



# Interrupted IVC

Prevalence of 0.2% - 3%

Isolated finding

Commonly associated with congenital heart disease:

- Heterotaxy Syndrome (Left atrial isomerism/Polysplenia Syndrome)
- Complete AV Canal Defect
- ASDs
- VSDs
- Pulmonary Stenosis/Atresia

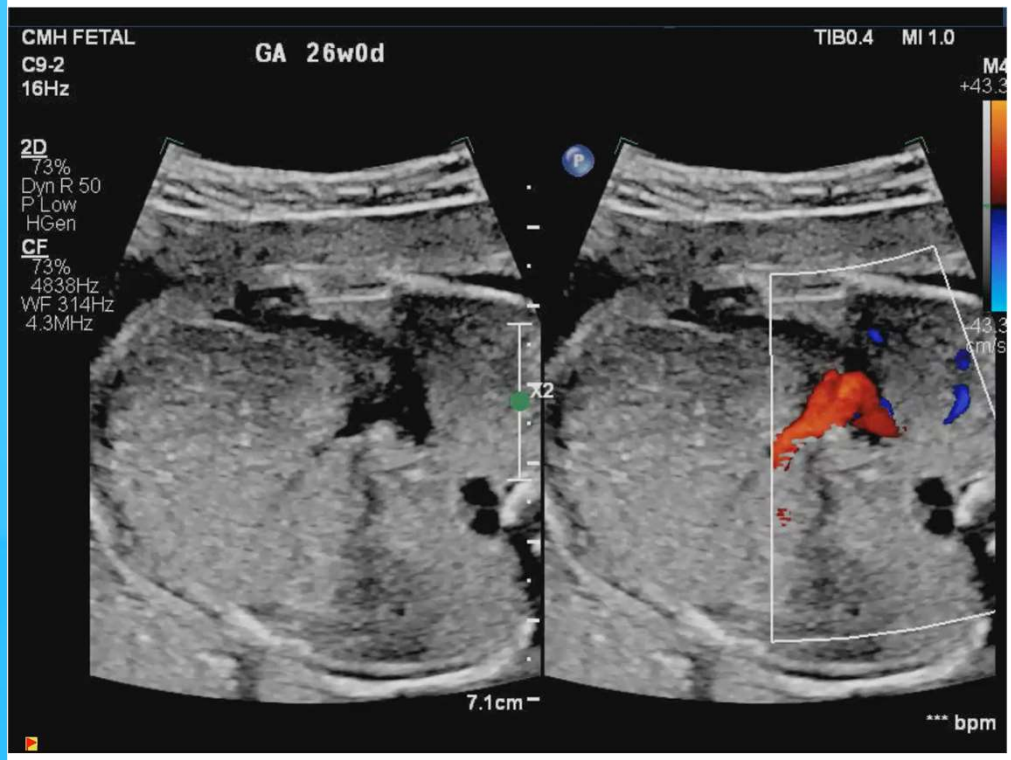
Abdominal wall defects:

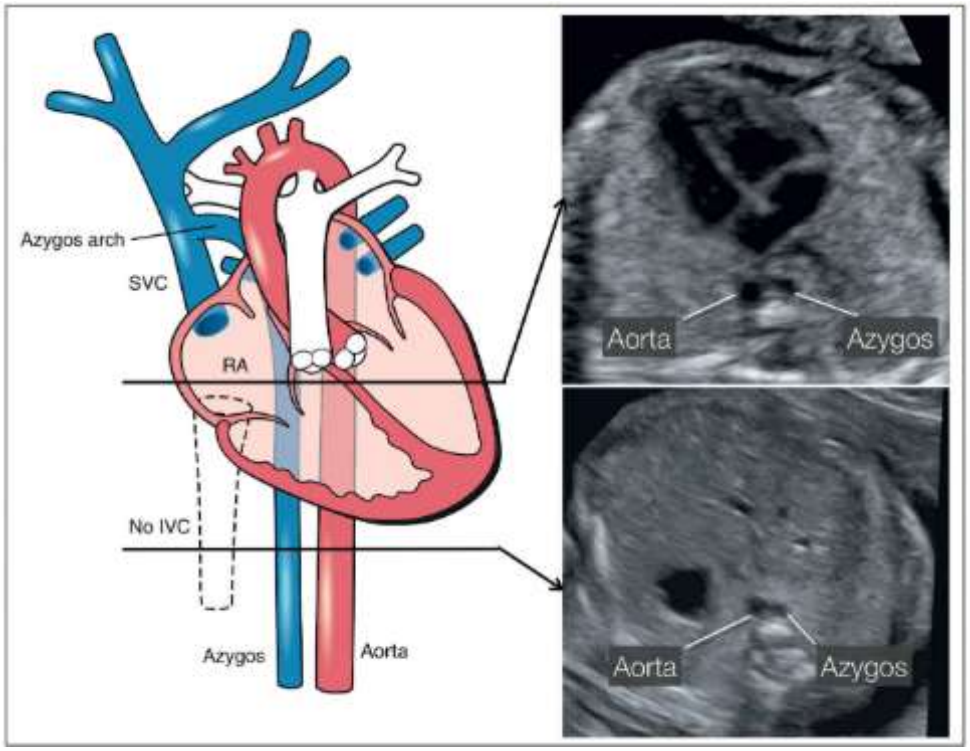
-Omphalocele

# Interrupted IVC and Heterotaxy Syndrome

| | Left isomerism (%) | Right isomerism (%) |
|---|---|---|
| Interrupted IVC | 89 | 5 |
| Left Superior Vena Cava | 28.5 | 42 |
| AVSD | 59 | 73 |
| Single Ventricle | 14 | 38 |
| RVOT obstruction | 35.5 | 67.5 |
| Conotruncal anomaly | 21 | 40 |
| TAPVC | 10 | 42 |
| Arrhythmia | 37 | 1 |

# Interrupted IVC

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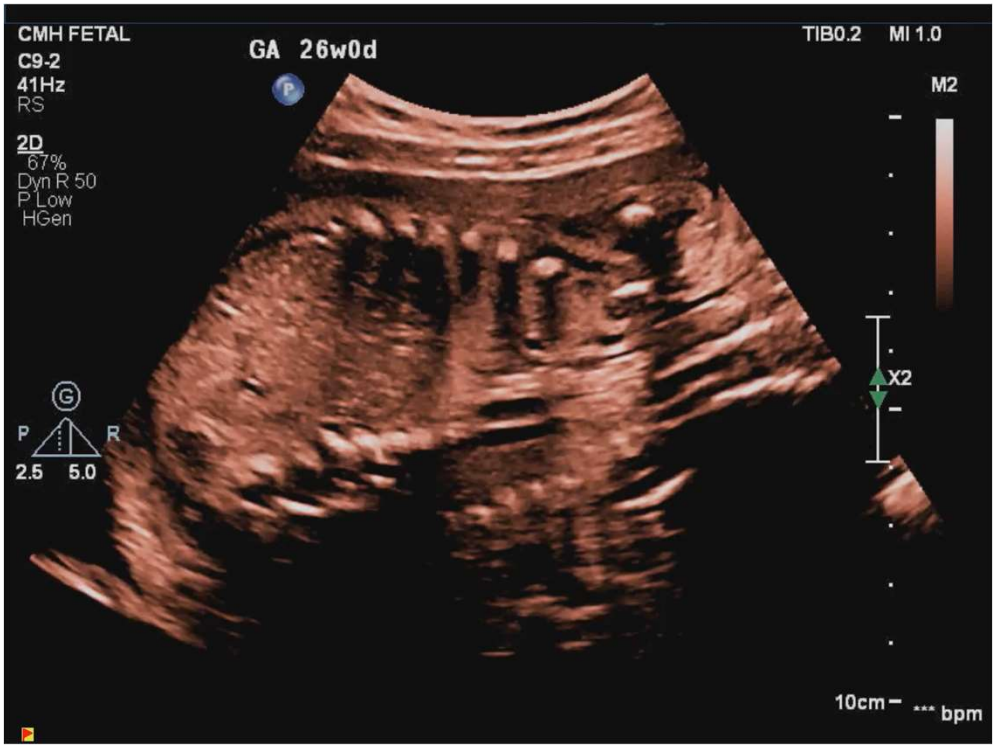
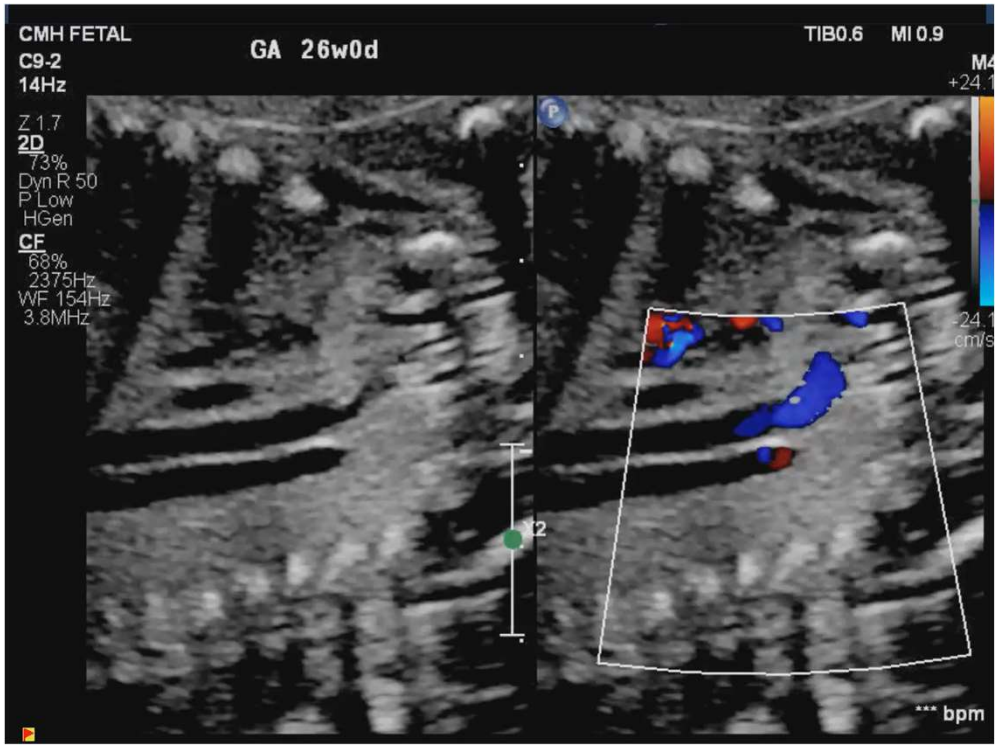
# Dilated Azygous

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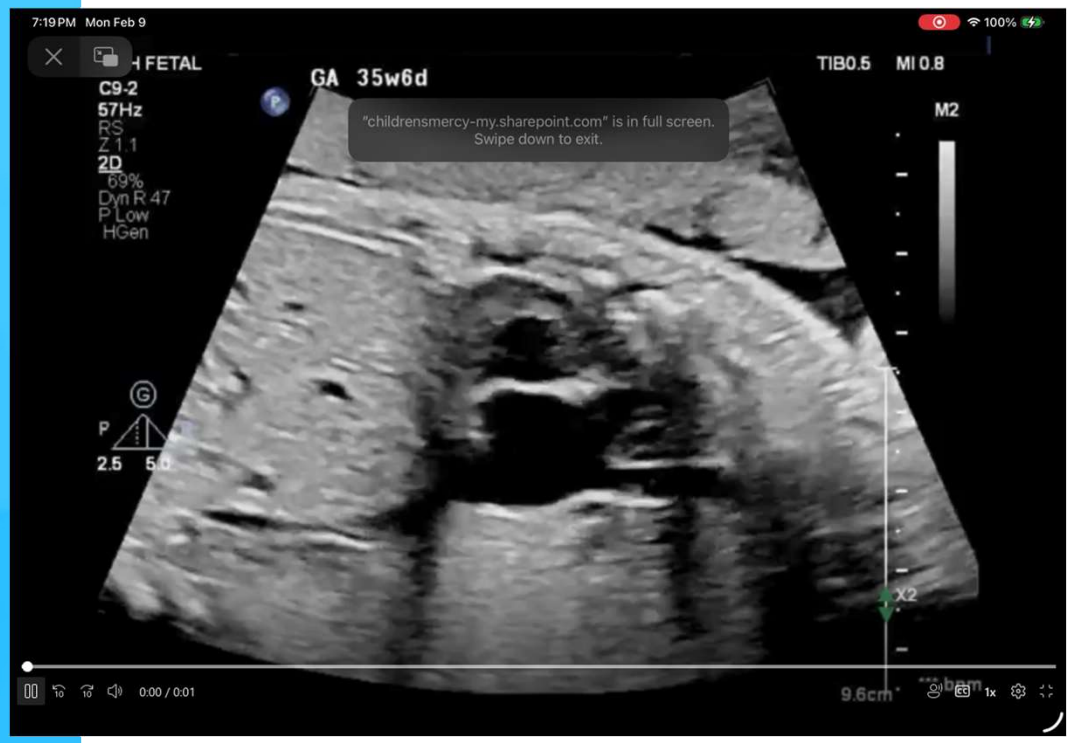
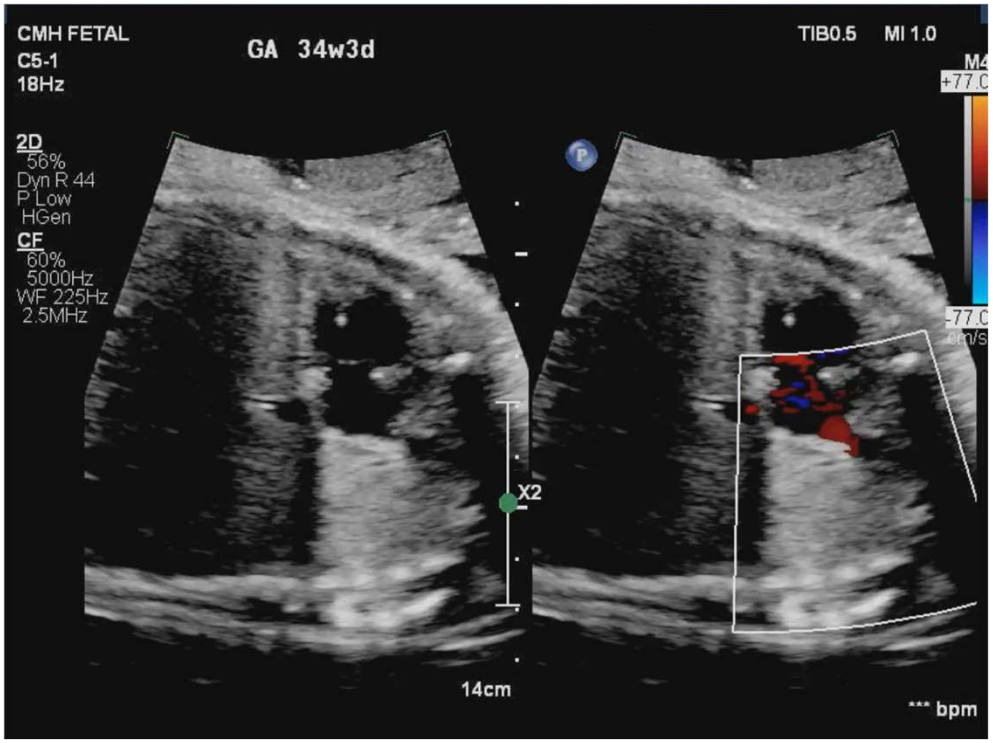
# Interrupted IVC - Bicaval

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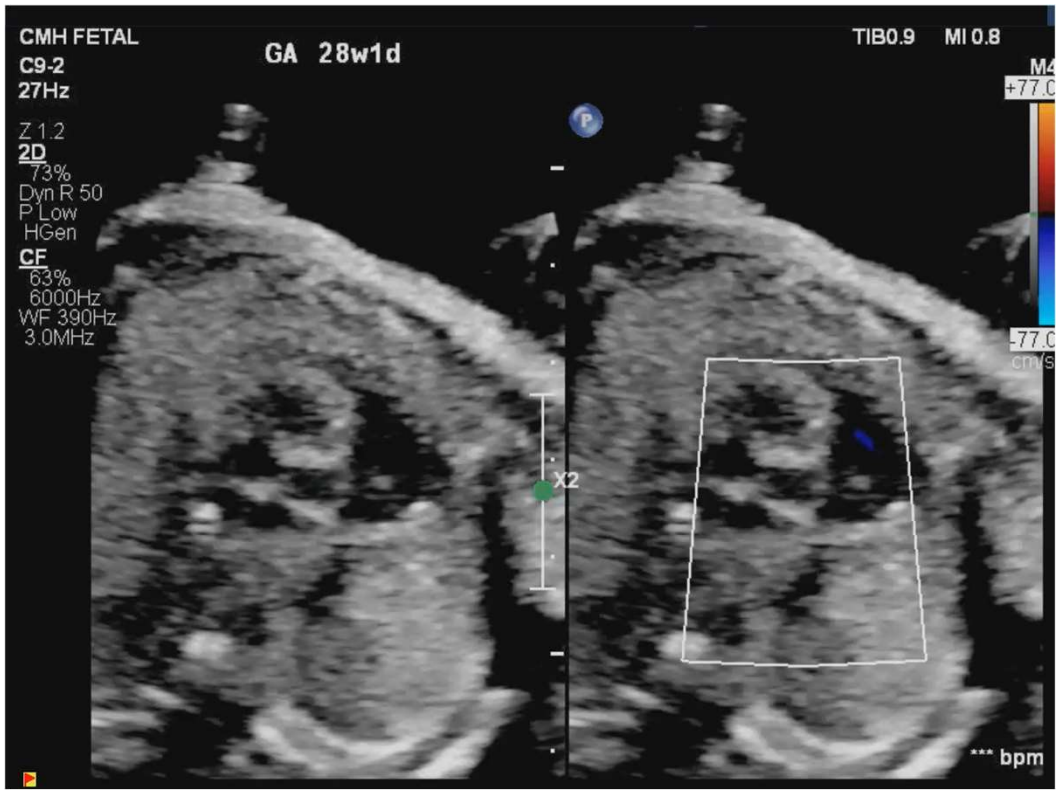
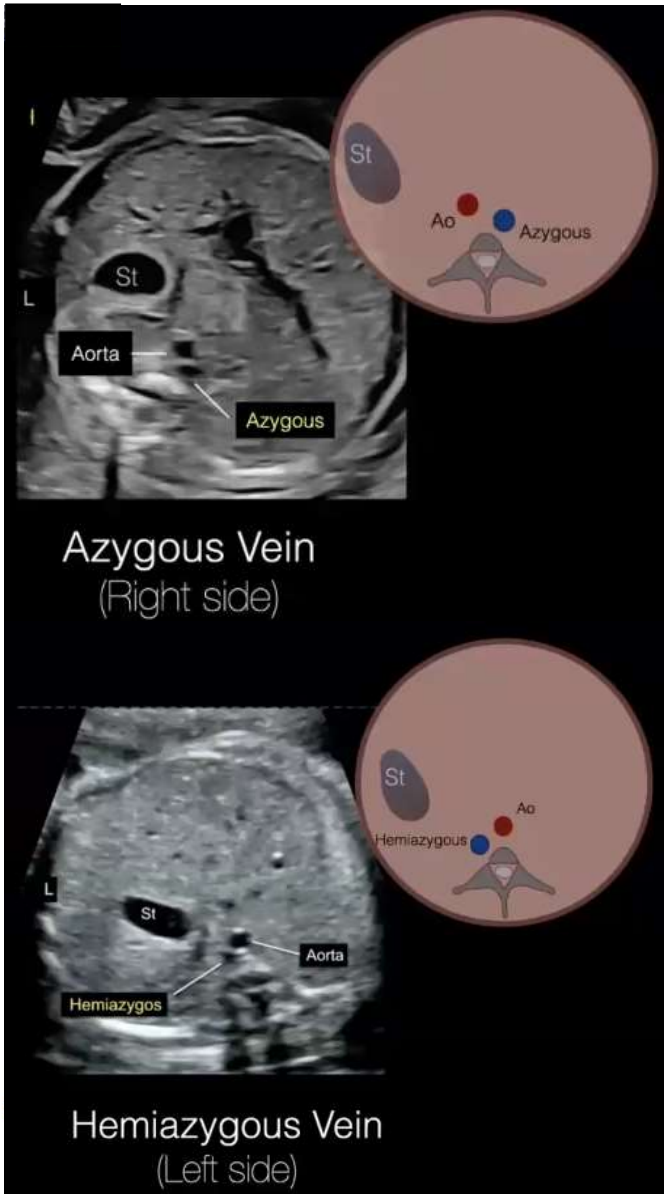
# Interrupted IVC – 3VTV

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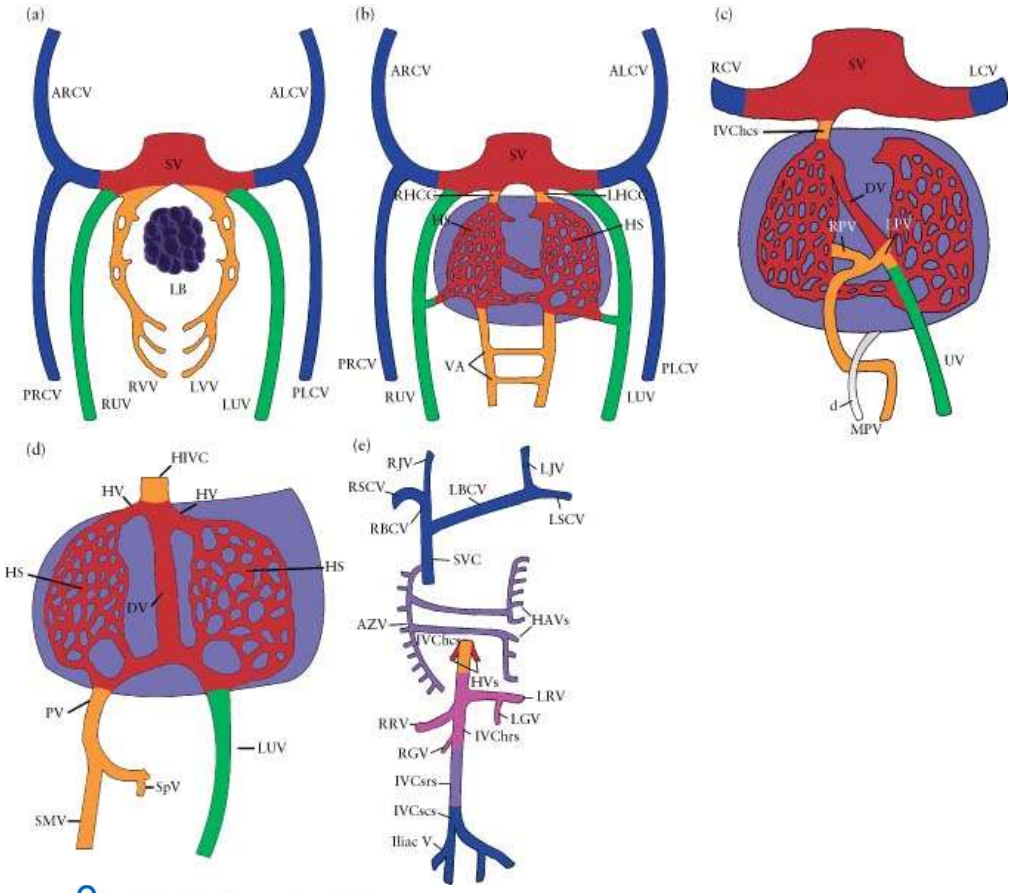
# Systemic Venous Development

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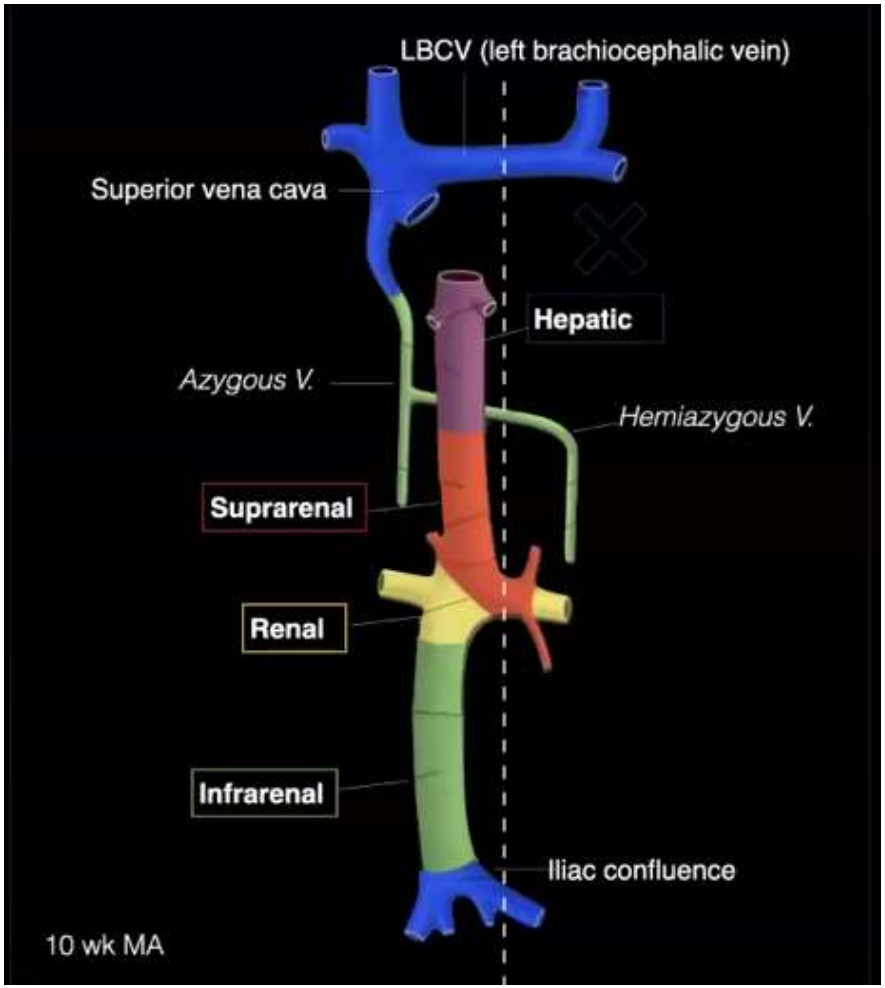
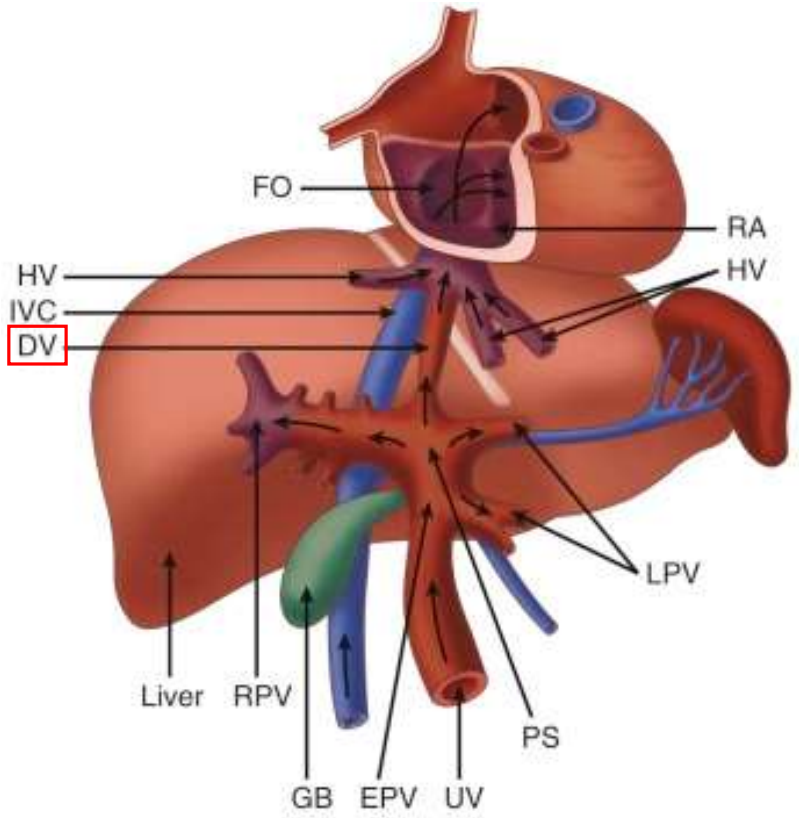


# Ductus Venosus

Ductus Venosus – Venous structure that shunts umbilical venous return to the fetal heart

Review of venous anatomy

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# Agenesis of the Ductus Venosus

- Prevalence 0.04% - 0.2%
- Umbilical venous return occurs via 1 of 2 ways:

1. Intrahepatic – umbilical vein connects directly to the portal system (usually left portal vein)
2. Extrahepatic – umbilical vein connects to a venous structure outside of the liver (femoral or iliac veins, **IVC**, directly to the coronary sinus or **right atrium**, left atrium, renal vein, SVC, right ventricle).

- Loss of preferential streaming of umbilical venous return across the foramen ovale to the left heart

# Agenesis of the Ductus Venosus

When absent, high degree of association with structural abnormalities of the heart or chromosomal/genetic syndromes

Cardiac evaluation:

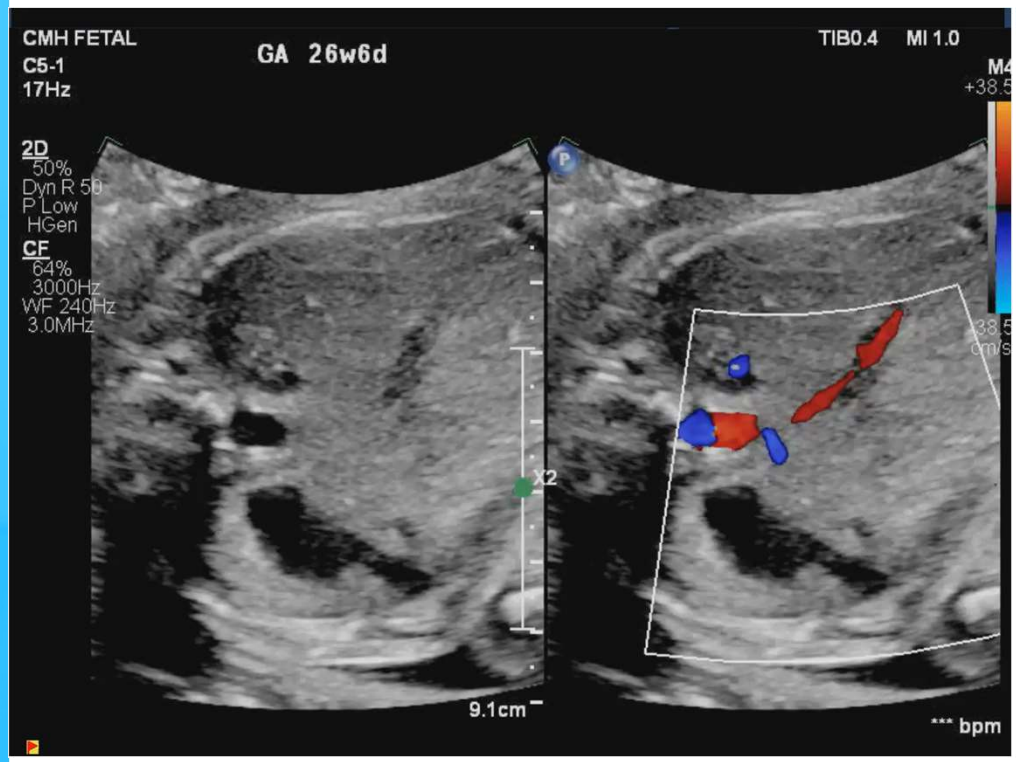
- Cardiomegaly (most common)
- VSD
- Valvar anomalies
- Pericardial effusion
- Coarctation
- DORV
- ASD

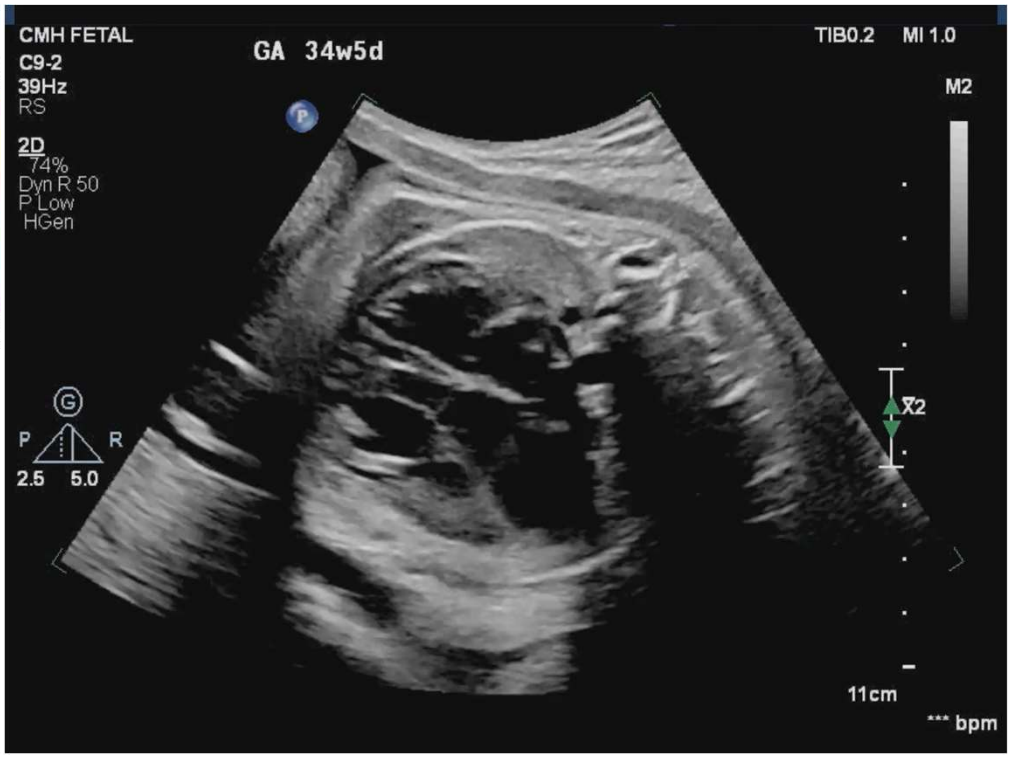
# Agenesis of the DV

Many other organ systems outside of the heart:

- MSK: Facial and limb anomalies
- GI: Diaphragmatic hernia, TE fistula
- Chromosomal: Turner Syndrome, T21, T18
- GU: Hydronephrosis, Ectopic Kidney

# Extrahepatic ADV

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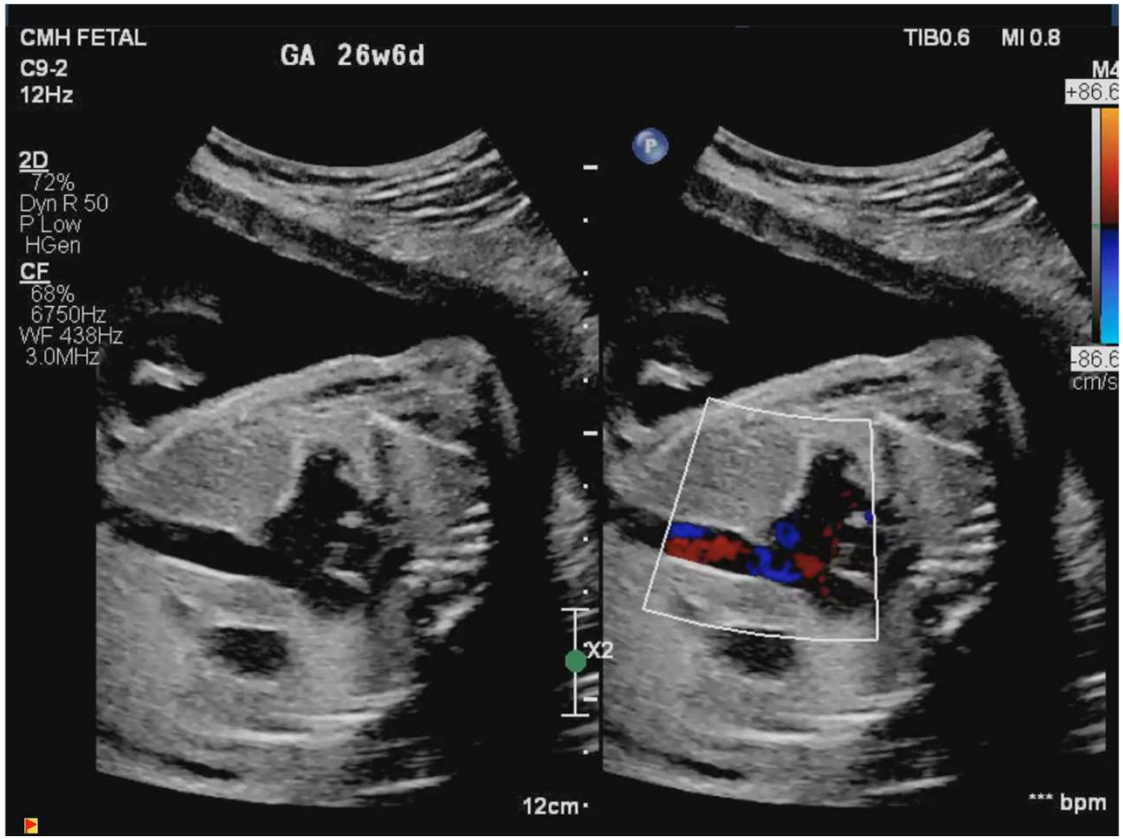
# Extrahepatic ADV



# ADV - Prognosis

Dependent on associated abnormalities and type of umbilical venous drainage

- Intrahepatic – overall good prognosis
- Extrahepatic:
  - Drainage site – direct umbilical vein drainage to the right atrium = highest risk
  - Shunt diameter - combined cardiac output (CCO) exceeding **750 to 800 mL/kg/min** is high risk for development of fetal hydrops
  - Presence or absence of portal venous system
  - Associated structural heart defects
  - Fetal Hydrops
  - Chromosomal abnormalities
- Association with portal vein stenosis, benign and malignant hepatic tumors, hepatopulmonary syndrome

# PRUV

PRUV – Left umbilical vein regresses and right remains open

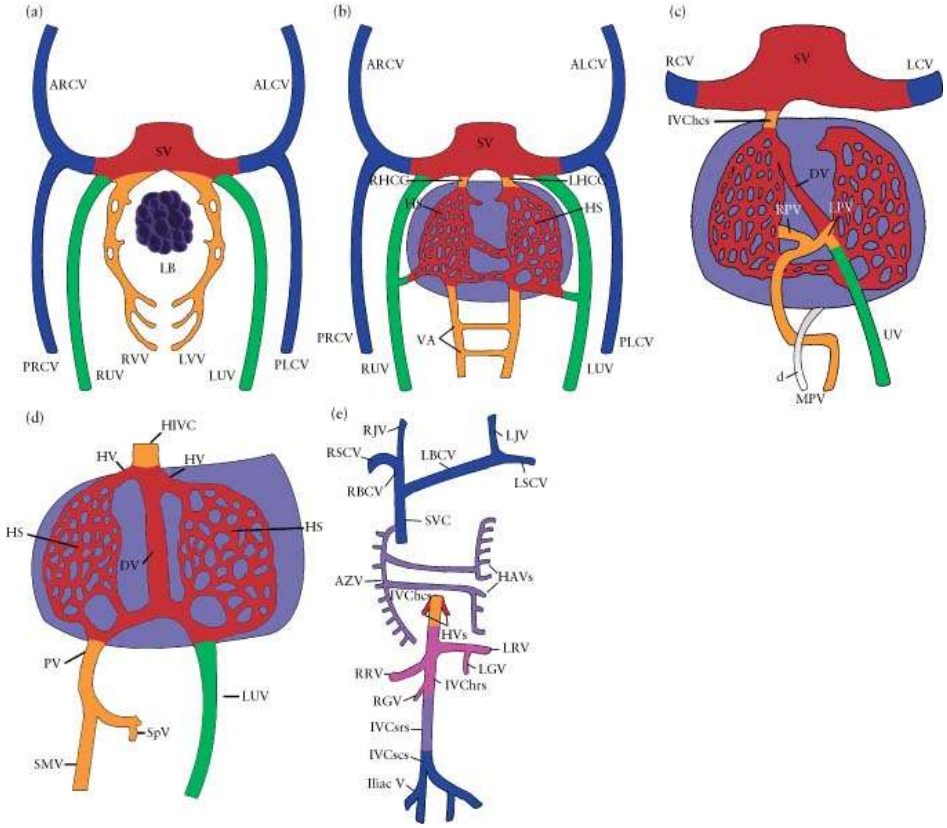
Incidence  0.08% - 0.4%

Types:

- Intrahepatic (95%) – better prognosis
- Extrahepatic – associated with absent ductus venosus

Typically an isolated anomaly

- Cardiac 8% (VSD, LSVC, single ventricle, CAVC, Truncus, TGA, TOF, R aortic arch)
- Placental/umbilical chord 7%
- GU 6%
- CNS 4%
- Chromosomal 1%.

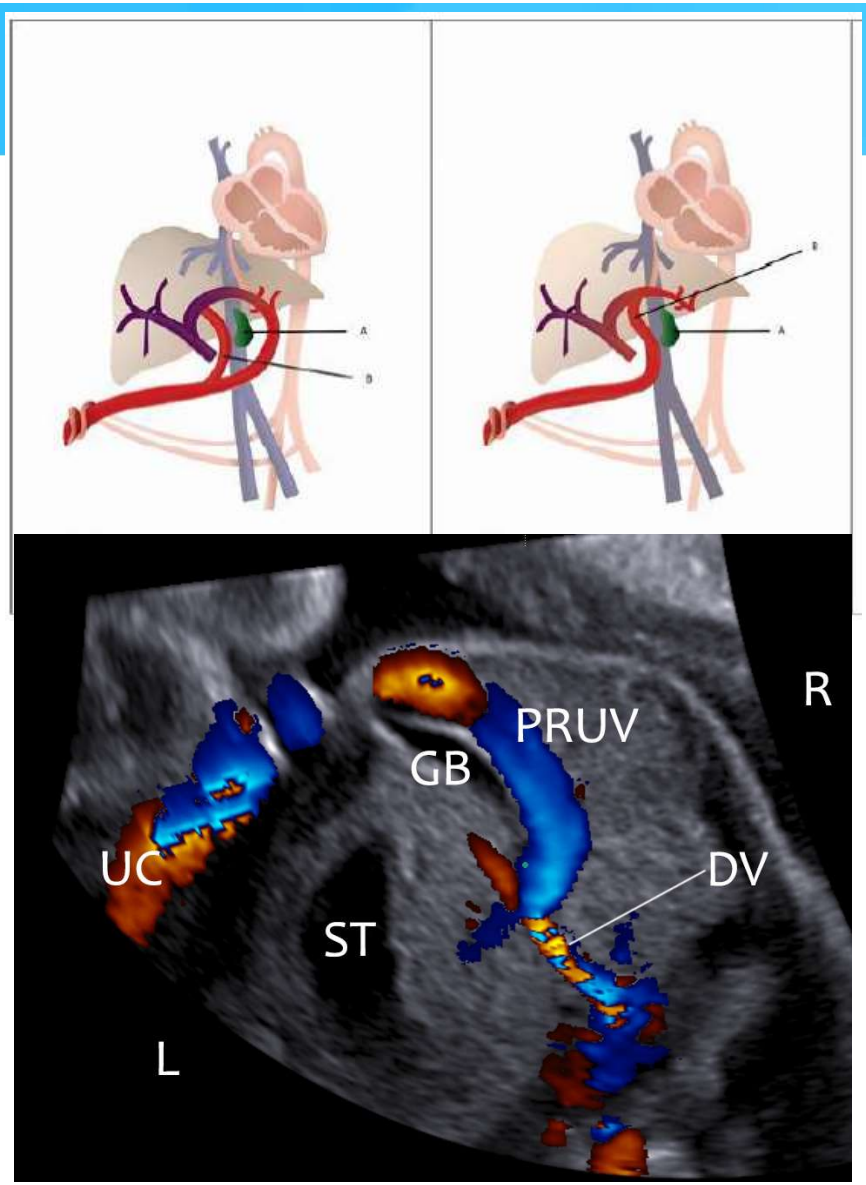


# PRUV

*Gallbladder is MEDIAL to right UV*

Usually curves towards stomach

Usually retains a connection to the portal system (95% of cases)

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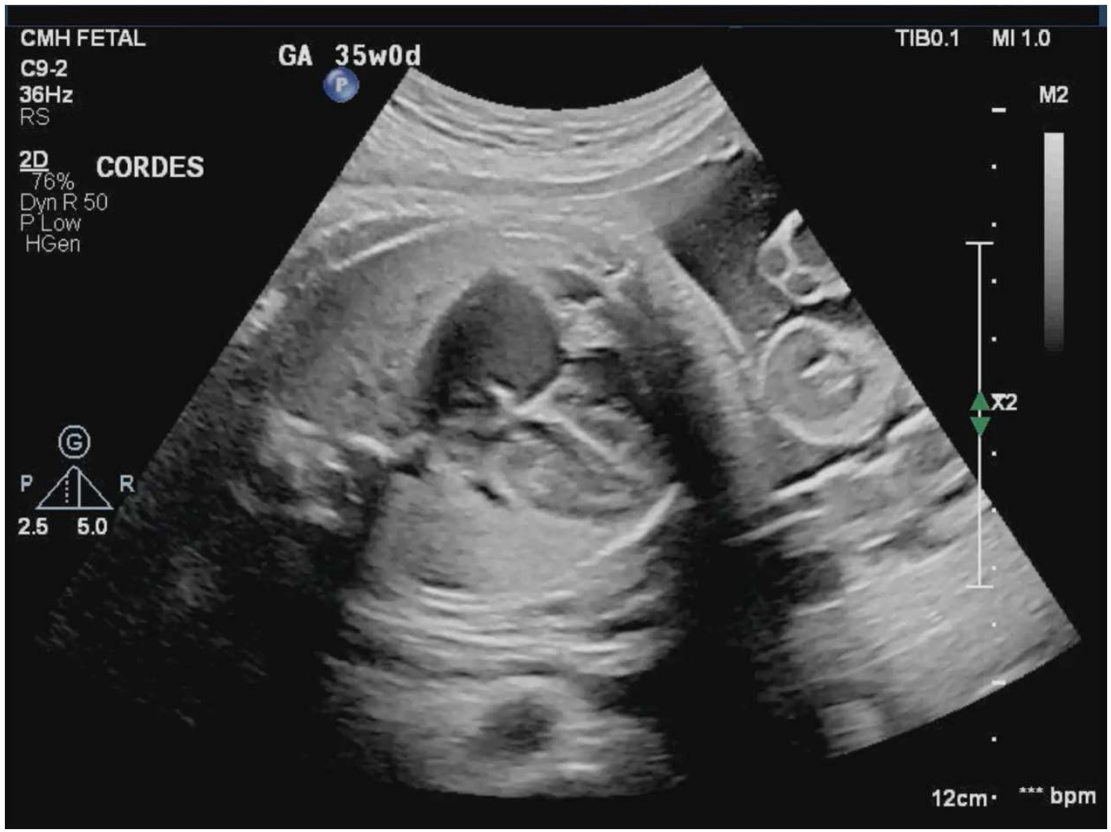
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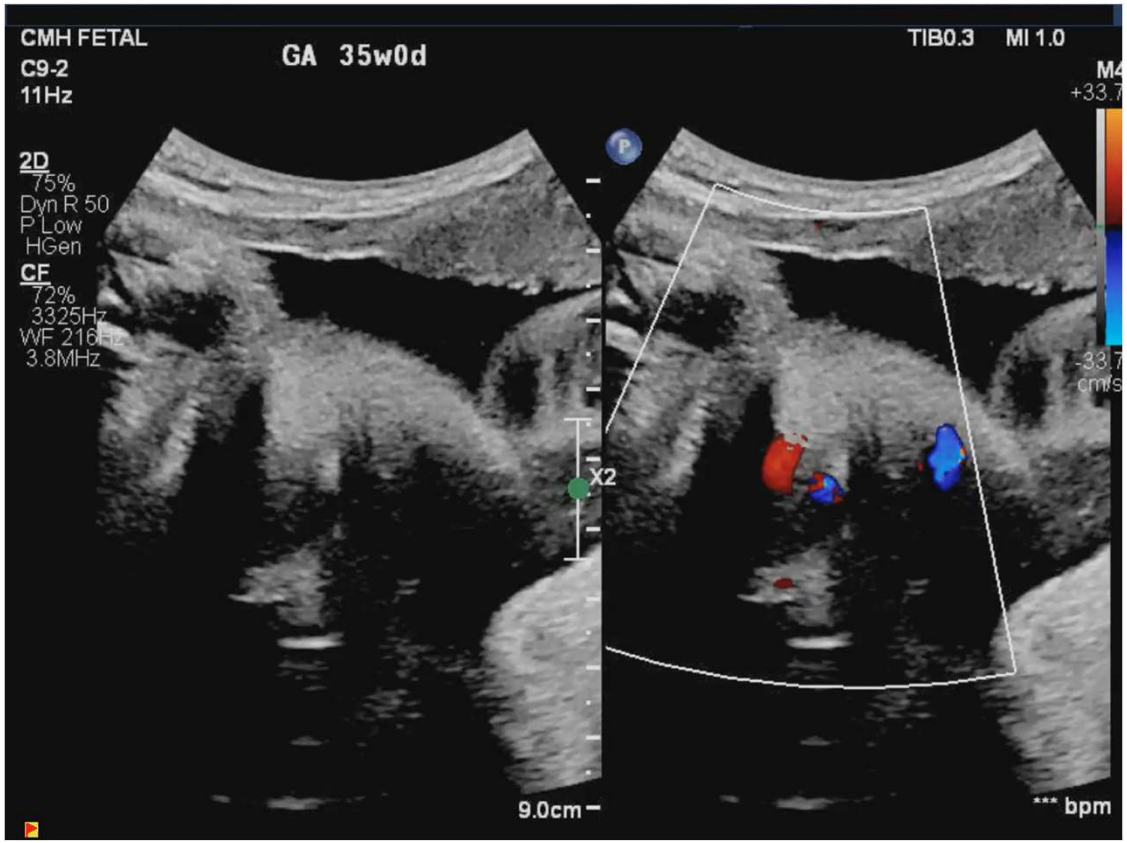
C9-2
36Hz
RS
2D
76%
Dyn R 50
P Low
HGen
CORDES
GA 35w0d
TIB0.1
MI 1.0
M2
X2
G
P
R
2.5
5.0
12cm
*** bpm


CMH FETAL
C9-2
11Hz
GA 35w0d
TIB0.3 MI 1.0
2D
75%
Dyn R 50
P Low
HGen
CF
72%
3325Hz
WF 216Hz
3.8MHz
X2
9.0cm
*** bpm

# Umbilical Vein Varix

Focal dilation of the umbilical vein from its entry in the abdominal wall to the portal system

- Extra-abdominal UV varices have also been reported

Incidence 0.04 – 0.1%

Diagnosis:

- Diameter exceeds 2 SD for the gestational age
- Diameter >9mm at term gestation
- Diameter 1.5x intrahepatic portion or >50% of non-dilated portion

Can be an isolated finding

Associated with aneuploidy (9-10% of cases)

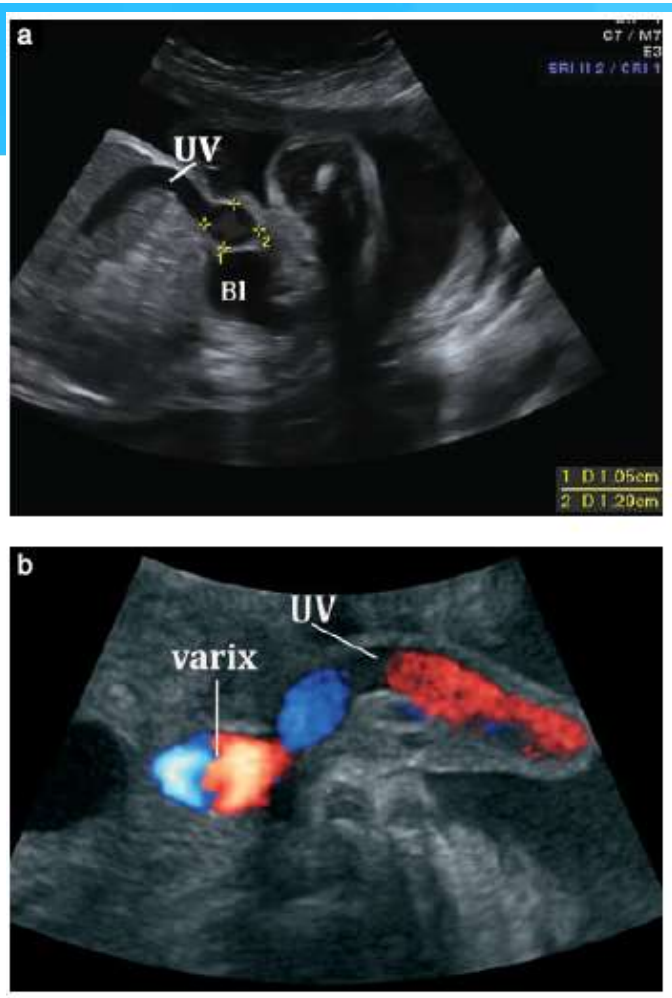


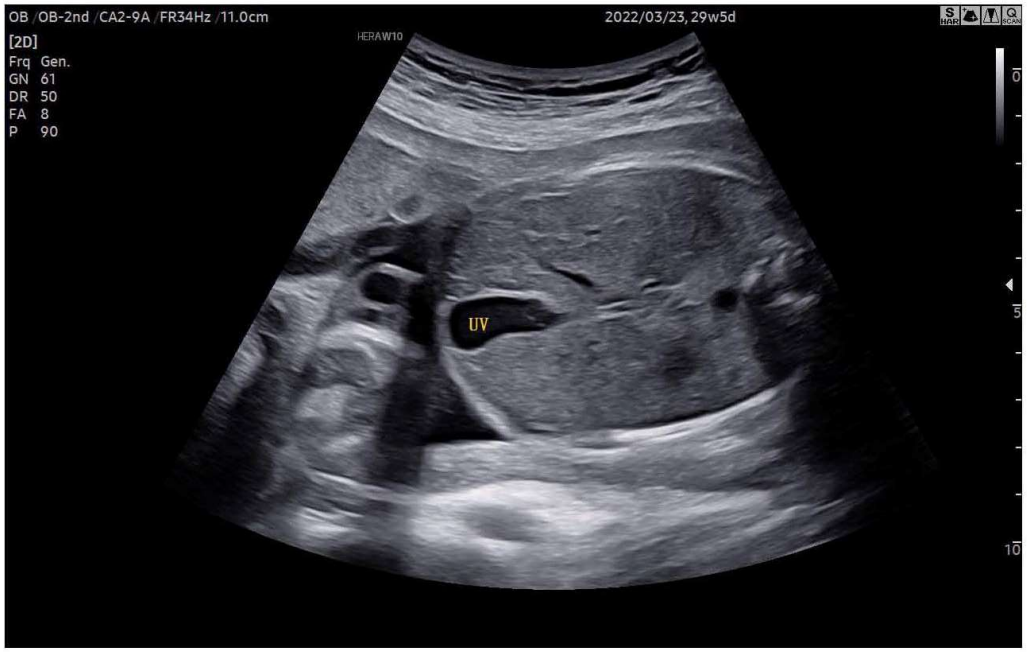

Figure 5 Fetal intra-abdominal umbilical vein (UV) varix: an anechoic cystic mass (calipers) is seen between the abdominal wall and the lower liver edge in a longitudinal section (a) and imaged with high-definition power flow Doppler (b). The varix is shown to be 1.5 times the diameter of the UV. Bl, bladder.

High association with anomalies: fetal aneuploidy, CV, GU malformations, diaphragmatic hernia, pulmonary sequestration, ventriculomegaly, echogenic bowel, **sudden intrauterine demise (5-8%)**

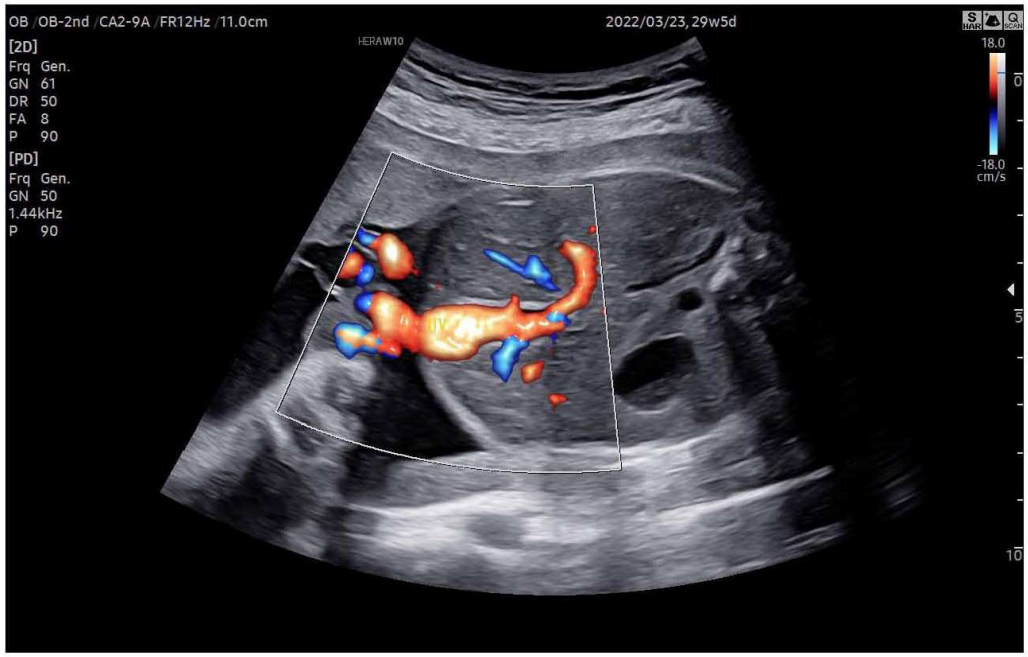
Prognosis dependent on factors:

- Diameter
- Presence of turbulent flow and thrombosis
- Associated fetal anomalies
- Growth parameters
- Isolated finding

**a**




**b**

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# References


Yagel, S., Kivilevitch, Z., Cohen, S., Valsky, D., Messing, B., Shen, O. & Achiron, R. (2010). The fetal venous system, Part I: normal embryology, anatomy, hemodynamics, ultrasound evaluation and Doppler investigation. Ultrasound in obstetrics & gynecology : the official journal of the International Society of Ultrasound in Obstetrics and Gynecology. 35. 741-50. 10.1002/uog.7618.

Yagel, S., Kivilevitch, Z., Cohen, S., Valsky, D., Messing, B., Shen, O. & Achiron, R. (2010). The fetal venous system, Part II: Ultrasound Evaluation of the Fetus with Congenital Venous System Malformation or Developing Circulatory Compromise. Ultrasound in obstetrics & gynecology : the official journal of the International Society of Ultrasound in Obstetrics and Gynecology. 36. 93-111. DOI 10.1002/uog.7622

Keleş A, Yılmaz O, Dağdeviren G, Çelik ÖY, Yücel A, Şahin D. Persistent Left Superior Vena Cava: Why is Prenatal Diagnosis Important? Fetal Pediatr Pathol. 2022 Aug;41(4):592-602. doi: 10.1080/15513815.2021.1933662. Epub 2021 Jun 9. PMID: 34106033.

Azizova, A., Onder, O., Arslan, S. *et al*. Persistent left superior vena cava: clinical importance and differential diagnoses. *Insights Imaging* **11**, 110 (2020). https://doi.org/10.1186/s13244-020-00906-2

Kahkouee, S., Sadr, M., Pedarzadeh, S., Fardin, A., Borhani, S., Gholami, G., & Amjad. (2016). Anomalous left brachiocephalic vein: important vascular anomaly concomitant with congenital anomalies and heart diseases. Folia Morphology. 76(1):51-57. doi: 10.5603/fm.A2016.0031

Karmegaraj B, Vijayakumar S. Prenatal diagnosis conventional/four-dimensional direct volume rendering fetal echocardiography pregnancy and postnatal outcomes of isolated anomalous course of left brachiocephalic vein. Pediatr Radiol. 2025 Sep;55(10):2054-2063. doi: 10.1007/s00247-025-06367-3. Epub 2025 Aug 18. PMID: 40824298.

Fetal Heart Society Webinar. Diagnosing Systemic Venous Anomalies in the Fetus: More than Meets the Eye. June, 2024.

Chen SJ, Wu MH, Wang JK. Clinical implications of congenital interruption of inferior vena cava. J Formos Med Assoc. 2022 Oct;121(10):1938-1944. doi: 10.1016/j.jfma.2022.01.021. Epub 2022 Feb 5. PMID: 35135704.

Bernard O, Franchi-Abella S, Branchereau S, Pariente D, Gauthier F, Jacquemin E. Congenital portosystemic shunts in children: recognition, evaluation, and management. Semin Liver Dis. 2012 Nov;32(4):273-87. doi: 10.1055/s-0032-1329896. Epub 2013 Feb 8. PMID: 23397528.

DiPaola F, Trout AT, Walther AE, Gupta A, Sheridan R, Campbell KM, Tiao G, Bezerra JA, Bove KE, Patel M, Nathan JD. Congenital Portosystemic Shunts in Children: Associations, Complications, and Outcomes. Dig Dis Sci. 2020 Apr;65(4):1239-1251. doi: 10.1007/s10620-019-05834-w. Epub 2019 Sep 23. PMID: 31549332; PMCID: PMC8180198.

Moaddab A, Tonni G, Grisolia G, Bonasoni MP, Araujo Júnior E, Rolo LC, Prefumo F, de la Fuente S, Sepulveda W, Ayres N, Ruano R. Predicting outcome in 259 fetuses with agenesis of ductus venosus - a multicenter experience and systematic review of the literature (.). J Matern Fetal Neonatal Med. 2016 Nov;29(22):360

Davey B, Szwast A, Rychik J. Diagnosis and management of heart failure in the fetus. Minerva Pediatrica. 2012 Oct;64(5):471-492. PMID: 22992530; PMCID: PMC4704789.6-14. doi: 10.3109/14767058.2016.1144743. Epub 2016 Mar 3. PMID: 26809266.

Sinkovskaya, E., Klassen, A, Abuhamad, A. (2013). A Novel Systematic Approach to the Evaluation of the Fetal Venous System. Seminars in Fetal & Neonatal Medicine. 18:269-278

Moradi, B., Tabatabaei, M., Atashi, S., Shahsavan, K., Kazemi, M., Rahmani, M., Radmard, A., Davari-Tanha, F. (2024). Ultrasound of Fetal Venous System: Normal anatomy, Variation, and Anomalies: A Review Article. Journal of Obstetrics, Gynecology, and Cancer Research 9(6):599-612. doi 10.30699/jogcr.9.6.599

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