Fetal webinar series
Tuesday 6/14/2022

The Single Ventricle in the Fetus

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Objectives

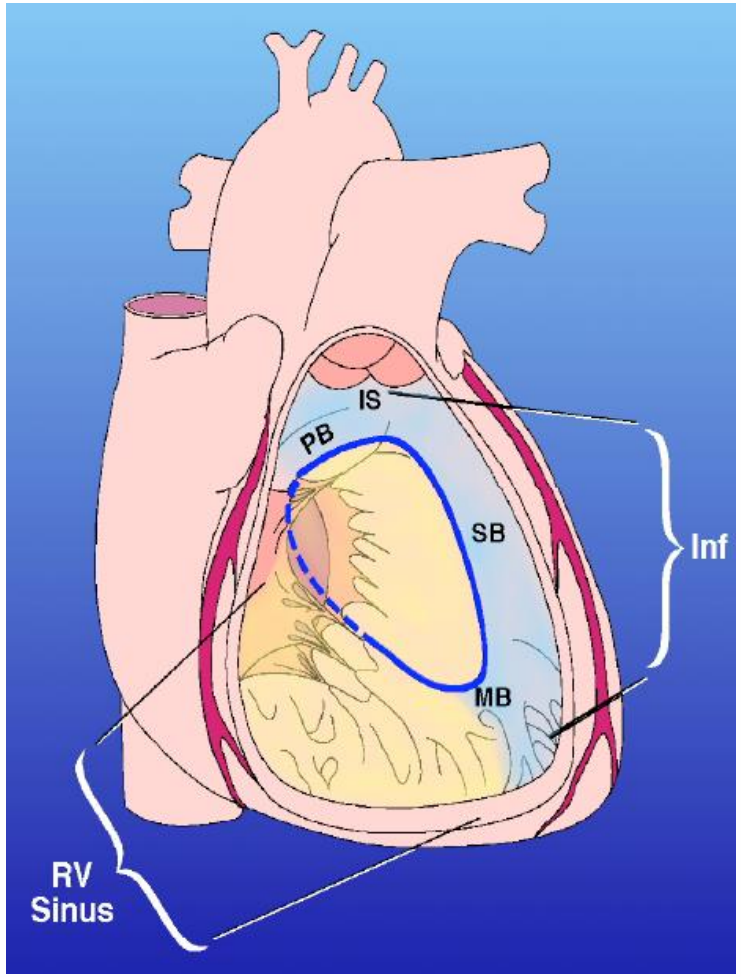
- Review the definition of single ventricle
- Review the commonly encountered anatomic SV variations
- Review the diagnostic approach to the SV
- Review appropriate counselling and management

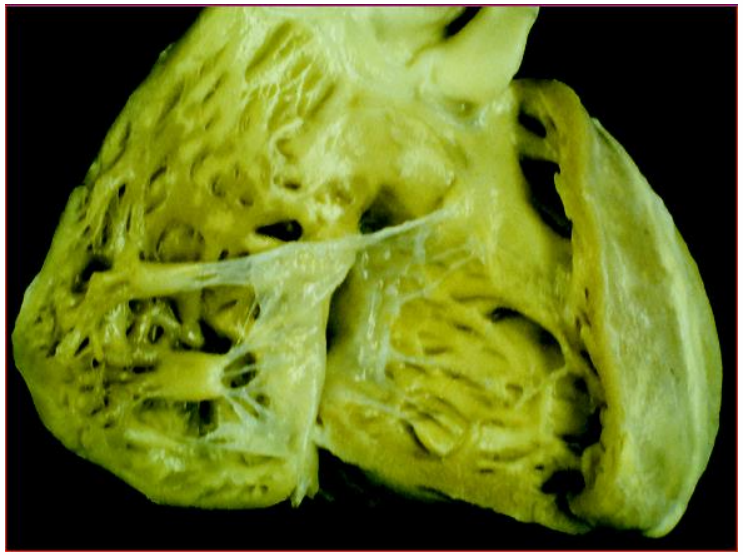
What is a Single Ventricle?

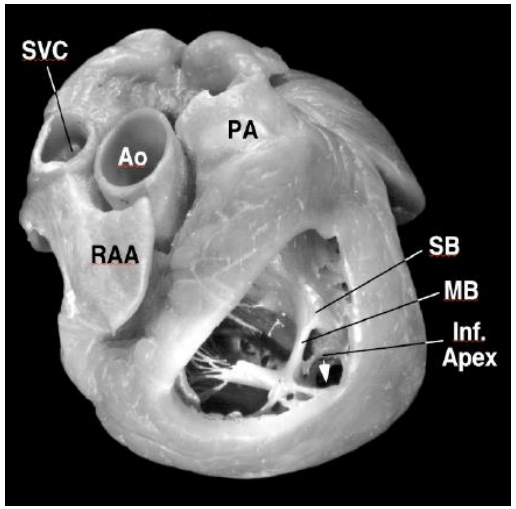
- Let's start with what is a normal ventricle?
- Normal ventricles comprise of an inlet, trabecular portion or body and of an outlet.
- Single Ventricle – doesn't exist
- Large dominant ventricle and a small rundimentary ventricle

- The terms used are Single ventricle, Univentricular heart, Common ventricle and, more recently, Single Functional ventricle.

Segmental approach to CHD







Courtesy Tal Geva, Boston Children's Hospital

Segmental approach to CHD

Fig 17. The left ventricle (LV) is elliptical as seen from posterior (left). The opened left ventricle (right) shows the smooth mid and basal septum (Sept) with fine apical trabeculations. The mitral valve (MV) has 2 free-wall papillary muscles (*).

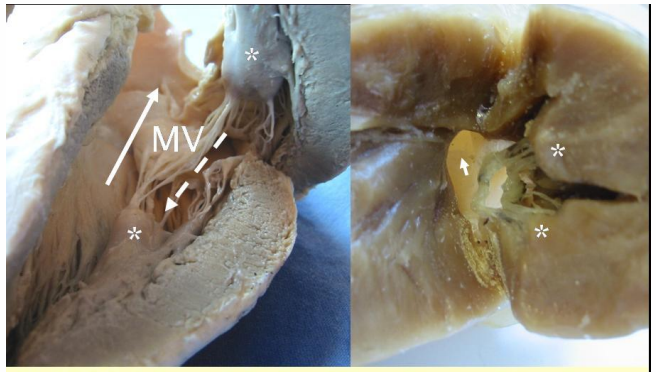
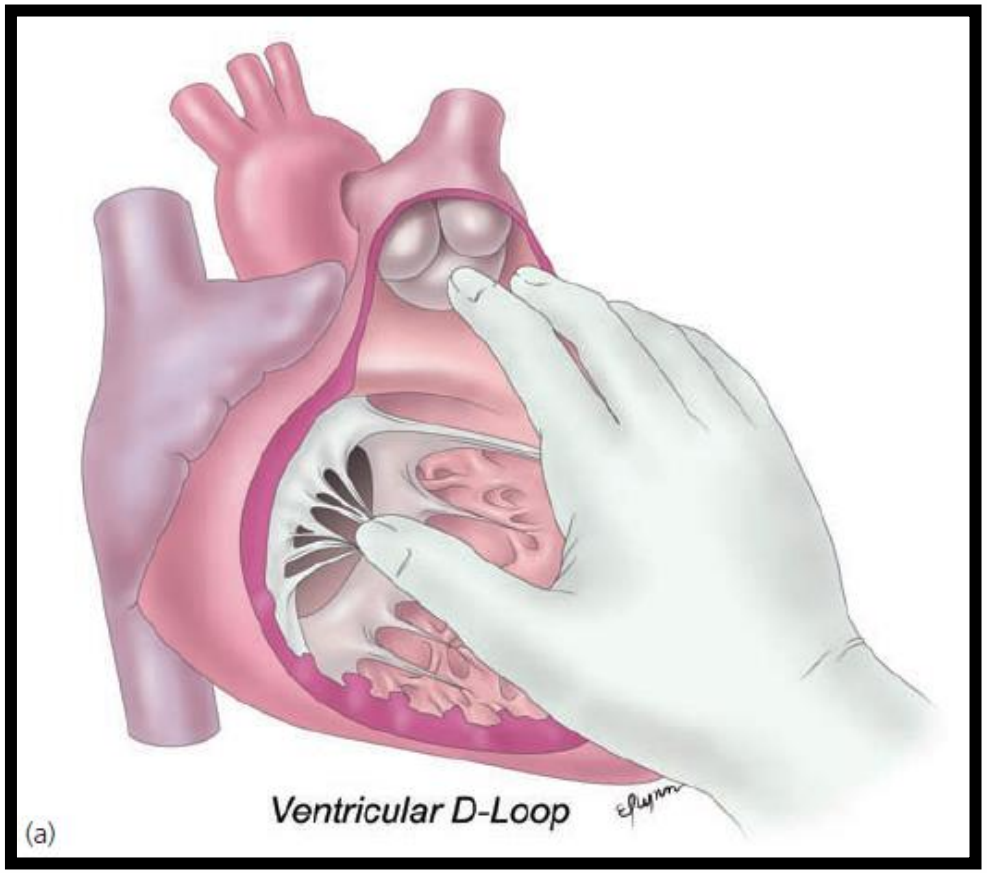
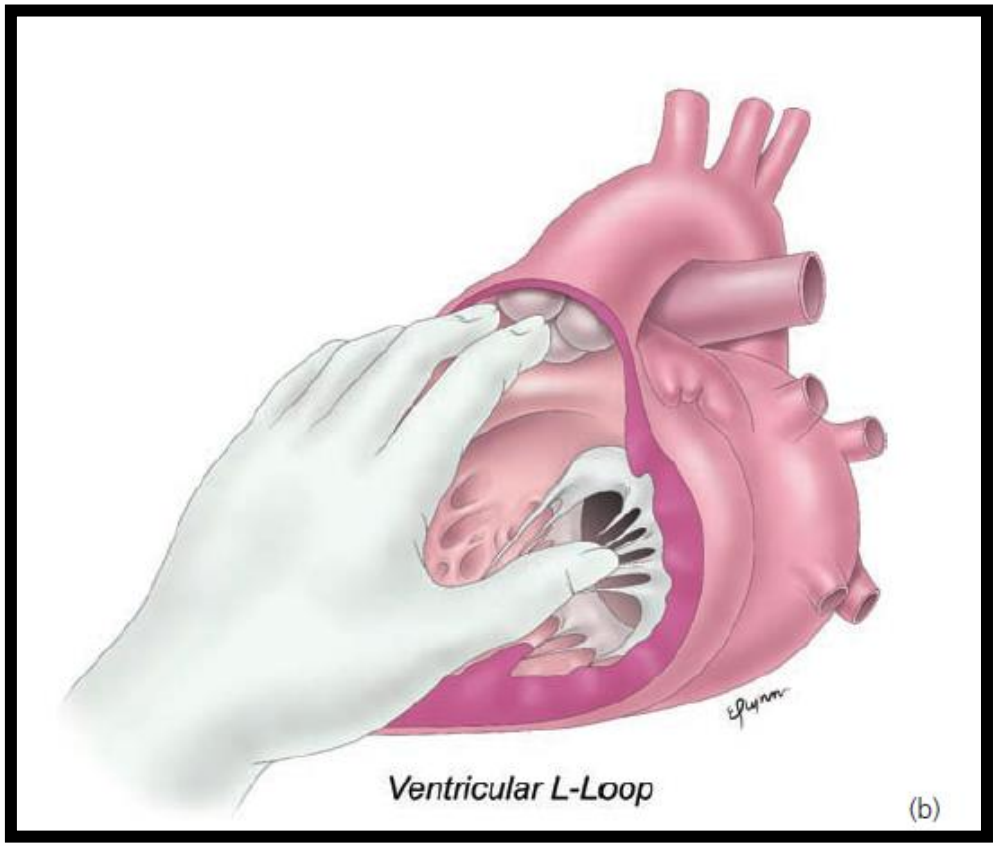


Fig 18. The left ventricle viewed from the apex shows the free wall papillary muscles (*), the adjacent and nearly parallel inflow ( ) and outflow ( ) tracts, and the outflow tract between the anterior mitral leaflet and the septum.

Segmental approach to CHD





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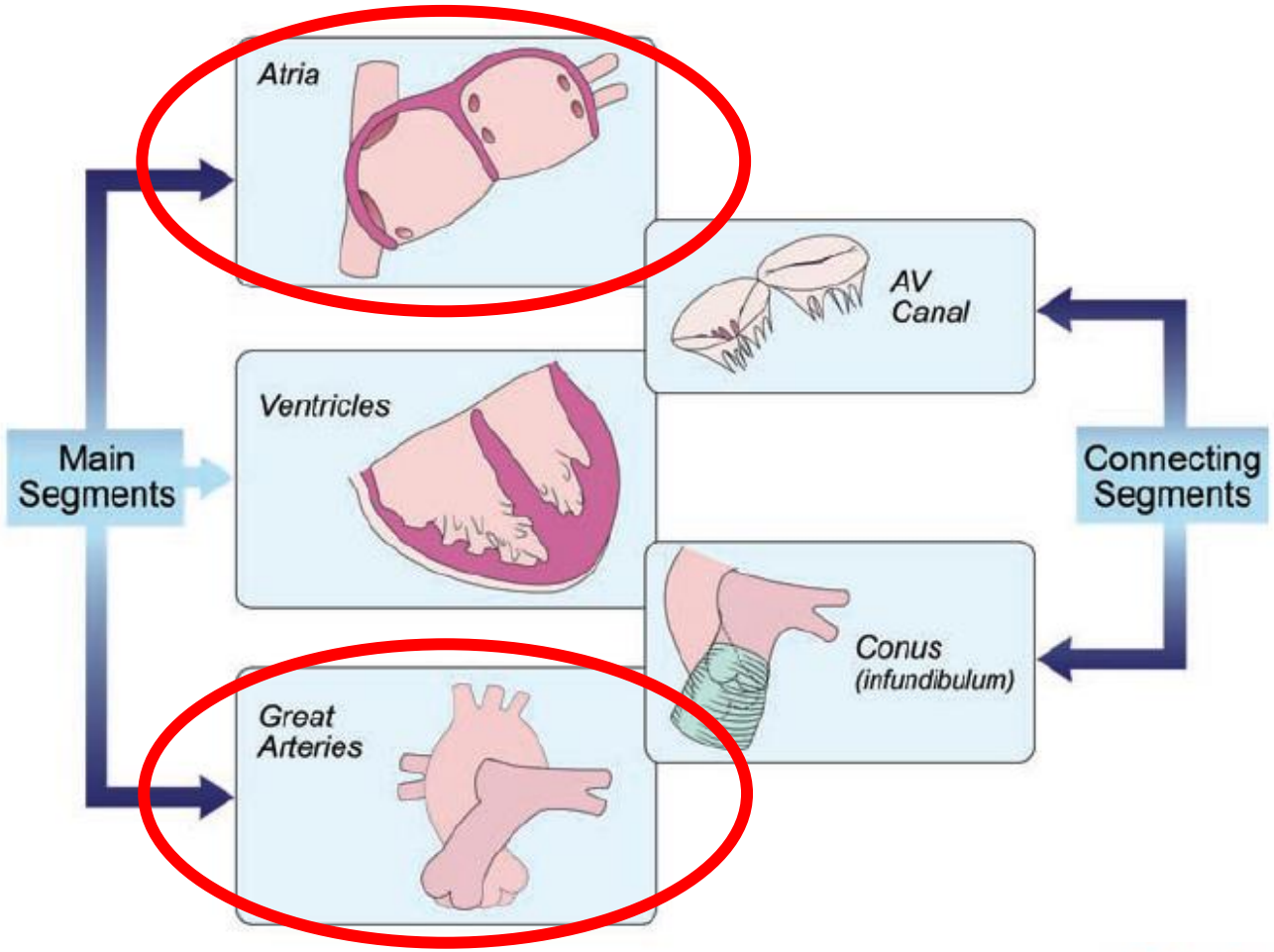
Segmental approach to CHD



{ X,X,X }

Atrial Situs

Ventricular Loop

Arteries

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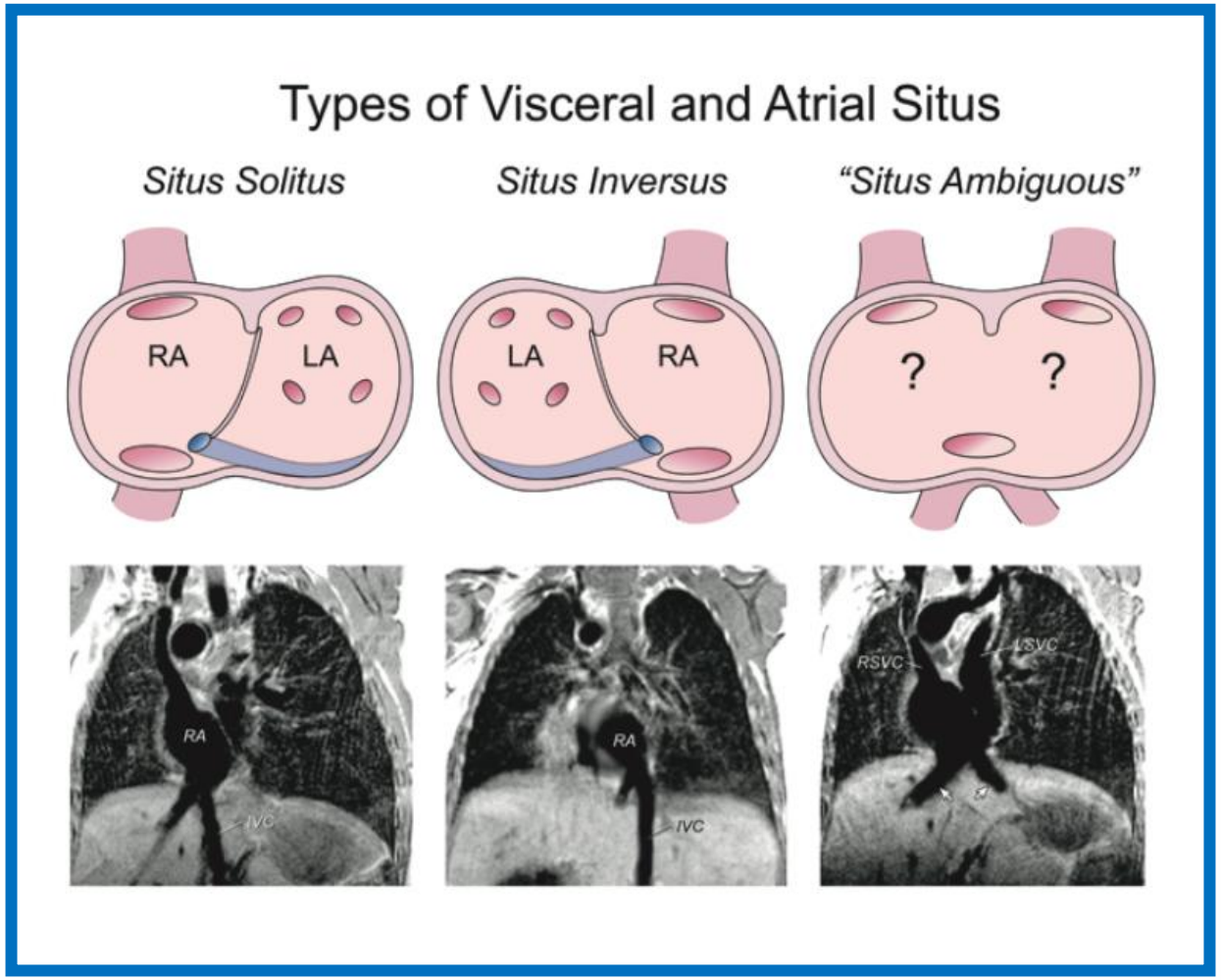
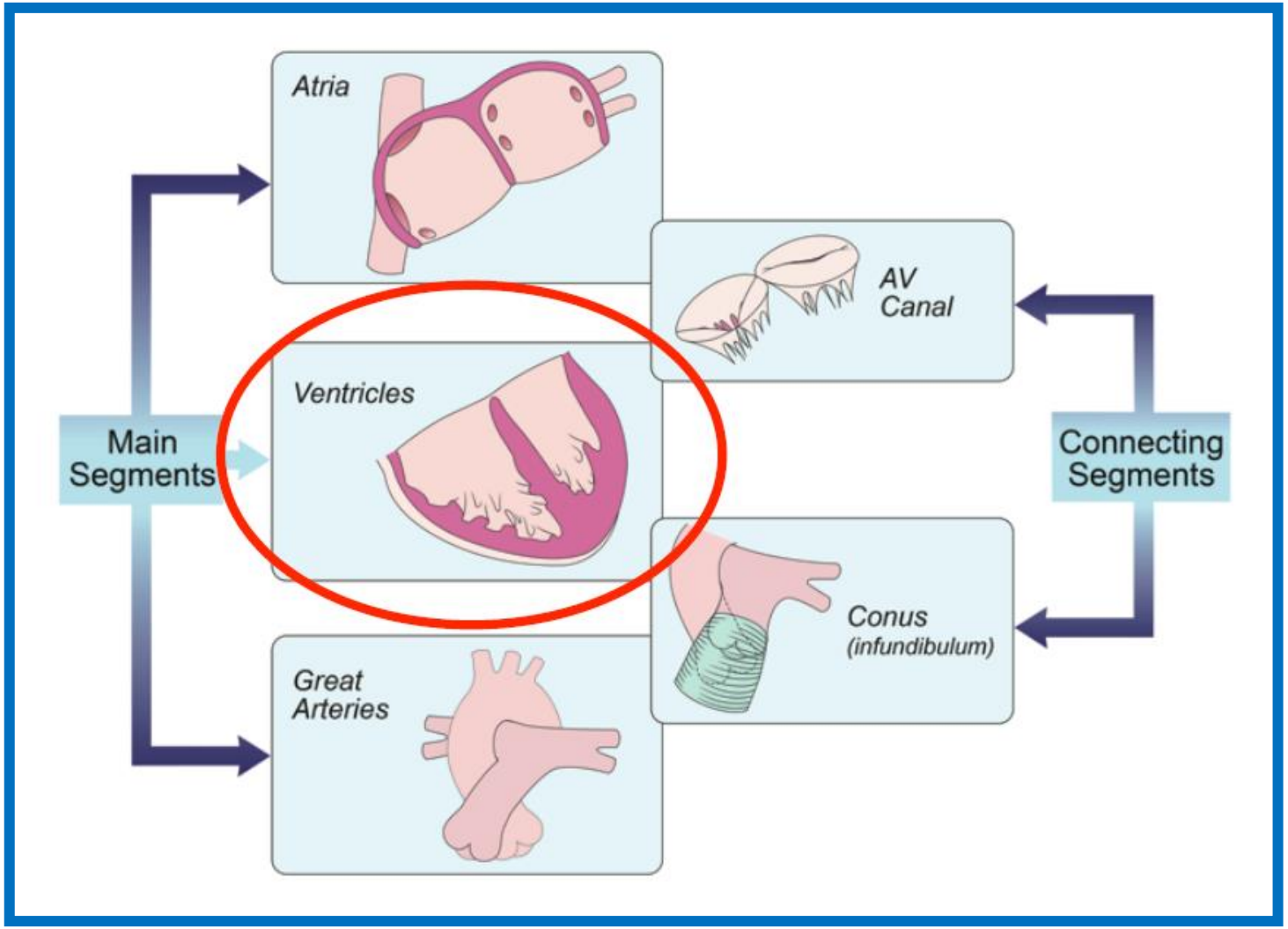
Segmental approach to CHD

1. Thoracoabdominal situs
2. Cardiac position
3. Segment-by-segment
4. Atrial situs (solitus, inversus, ambiguous)
5. Ventricular situs (D-loop, L-loop)
6. AV connection/alignment
7. Ventriculo-arterial alignment
8. Conal (infundibular) anatomy
9. Relationship between great arteries
10. Associated anomalies

Segmental approach to CHD



Segmental approach to CHD



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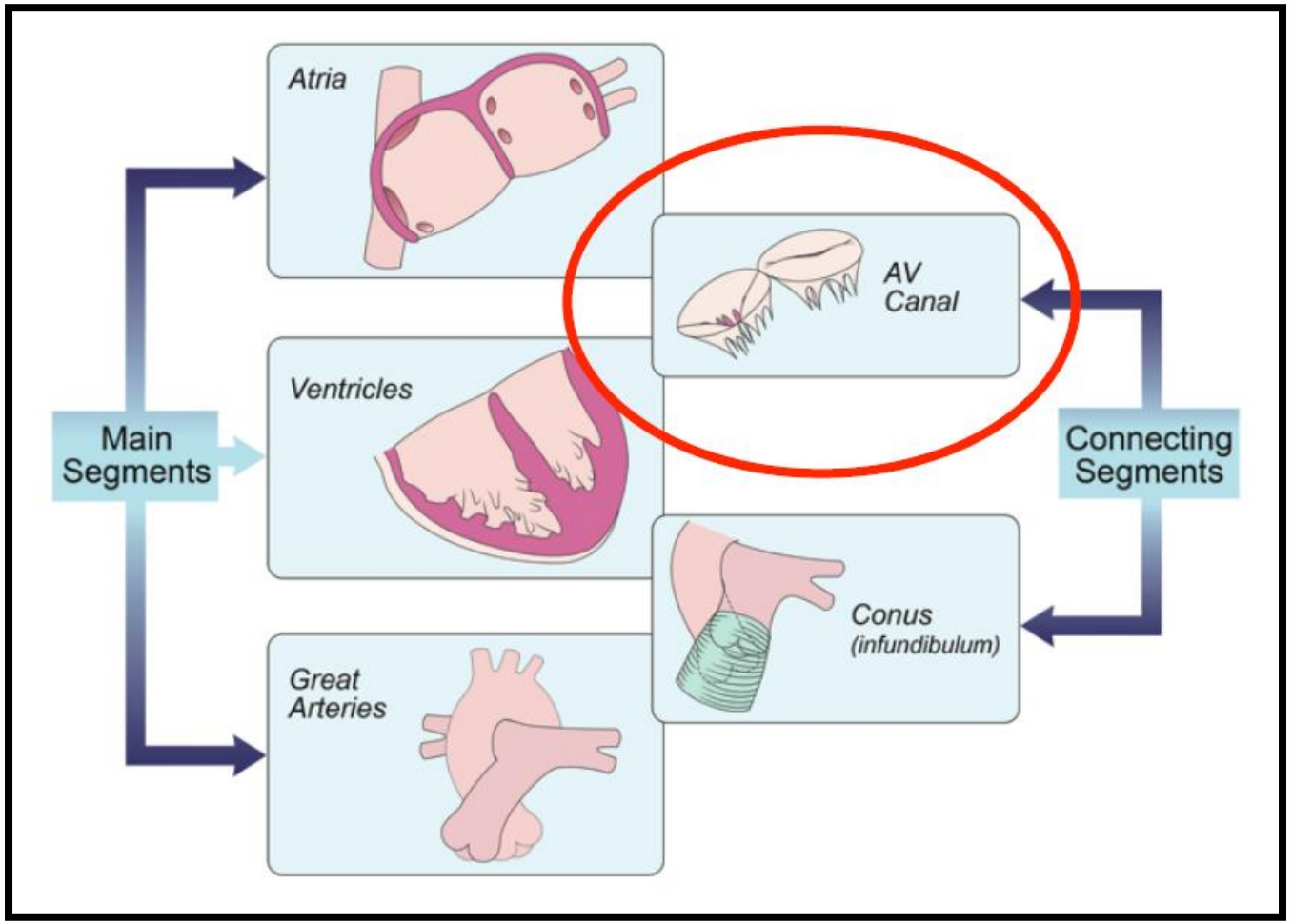

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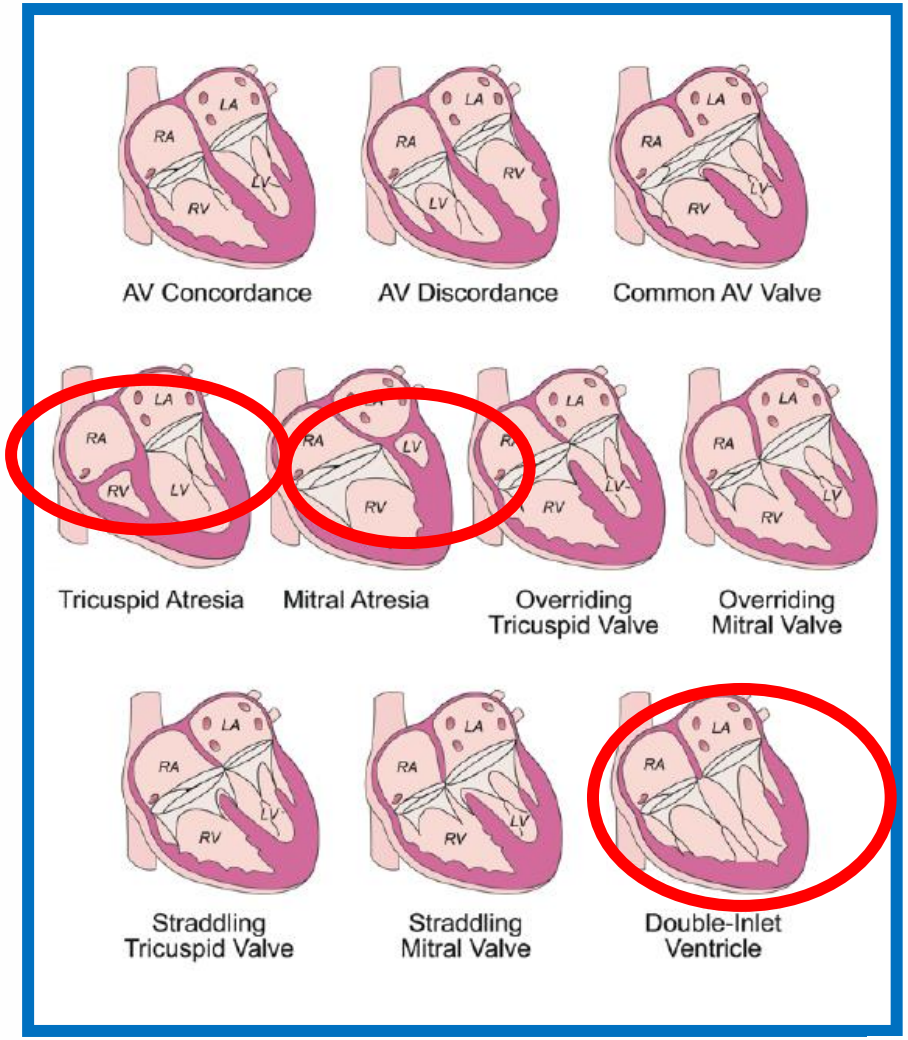

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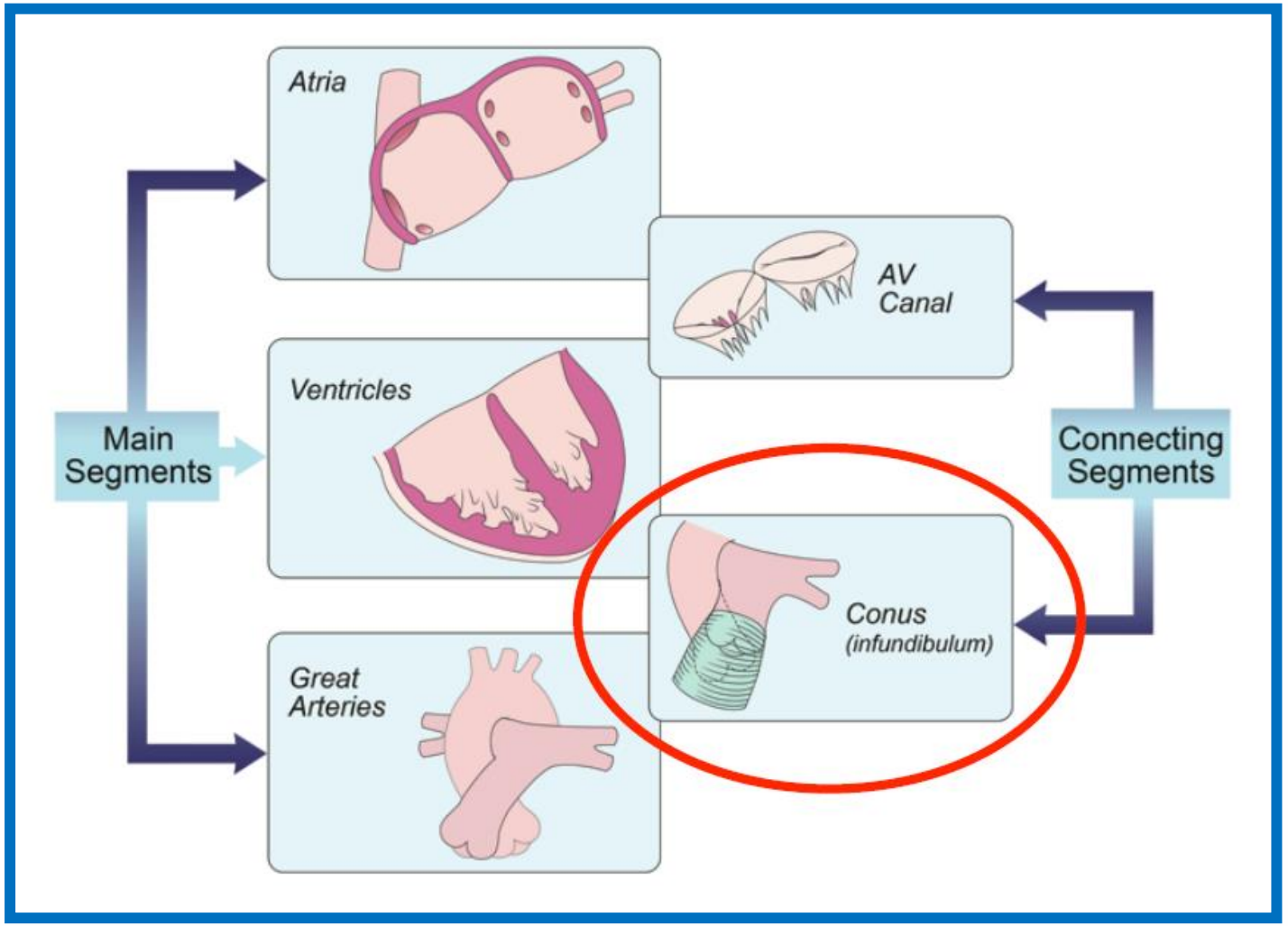

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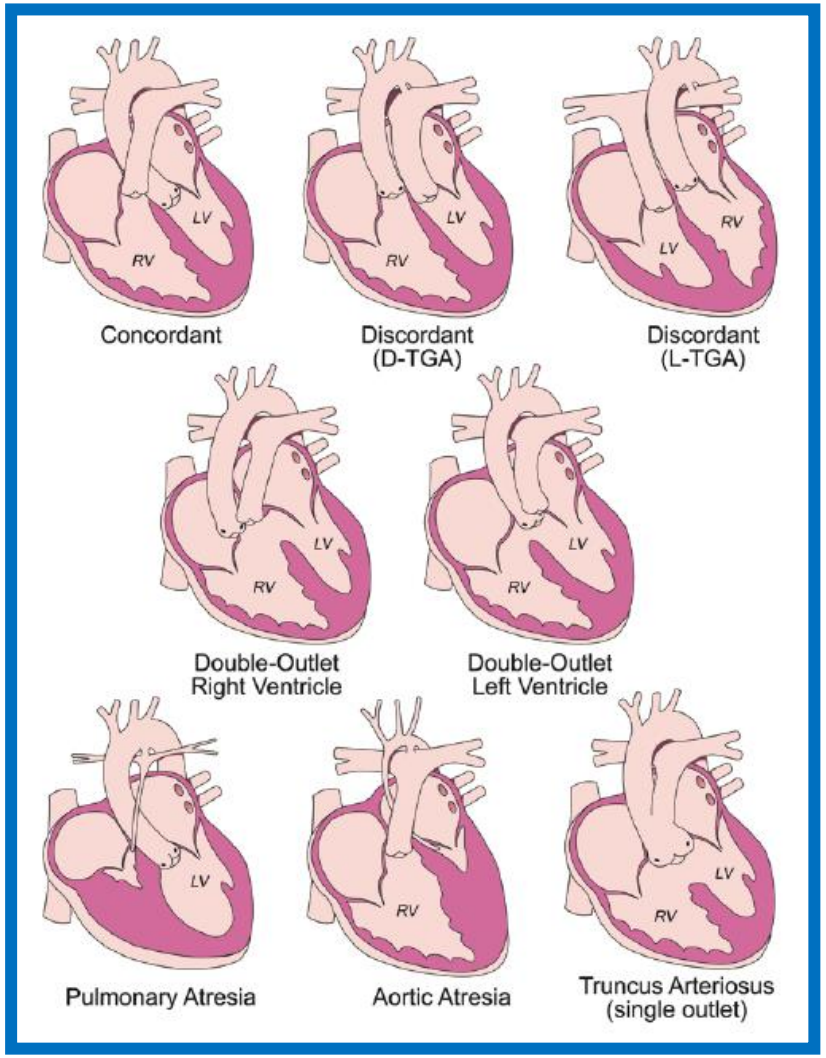

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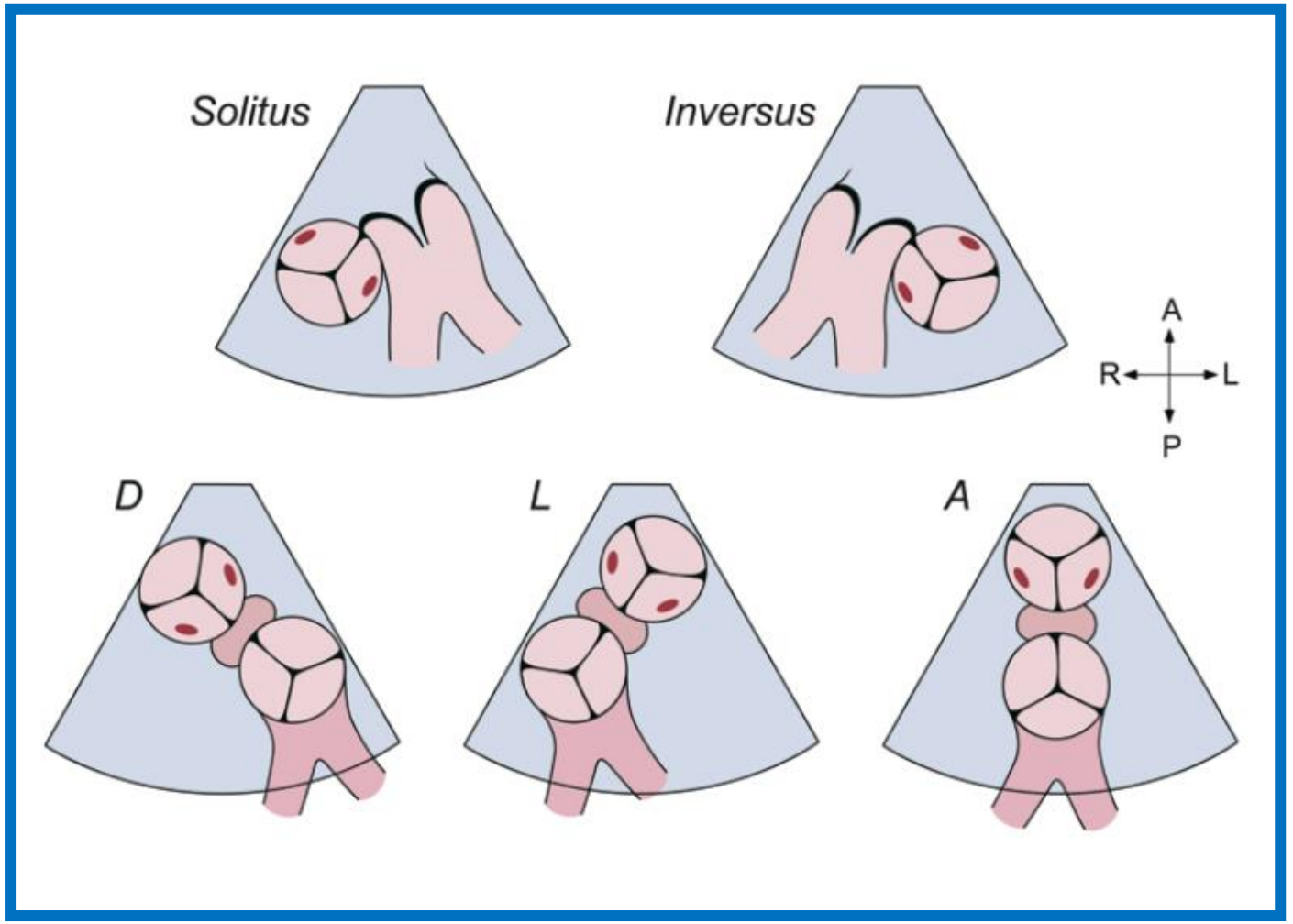

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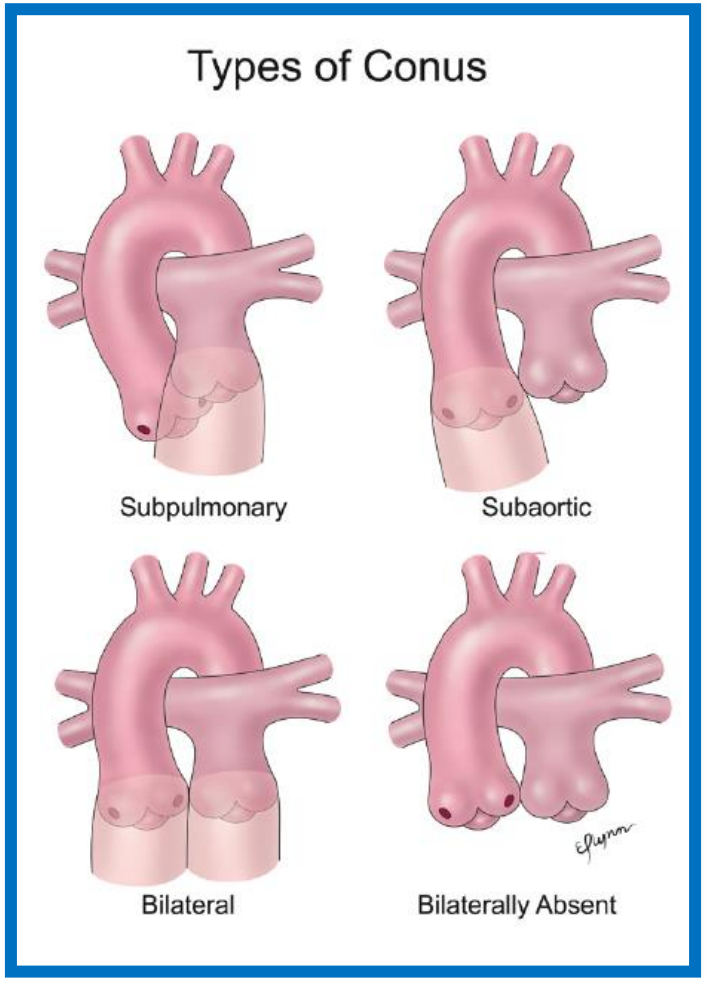

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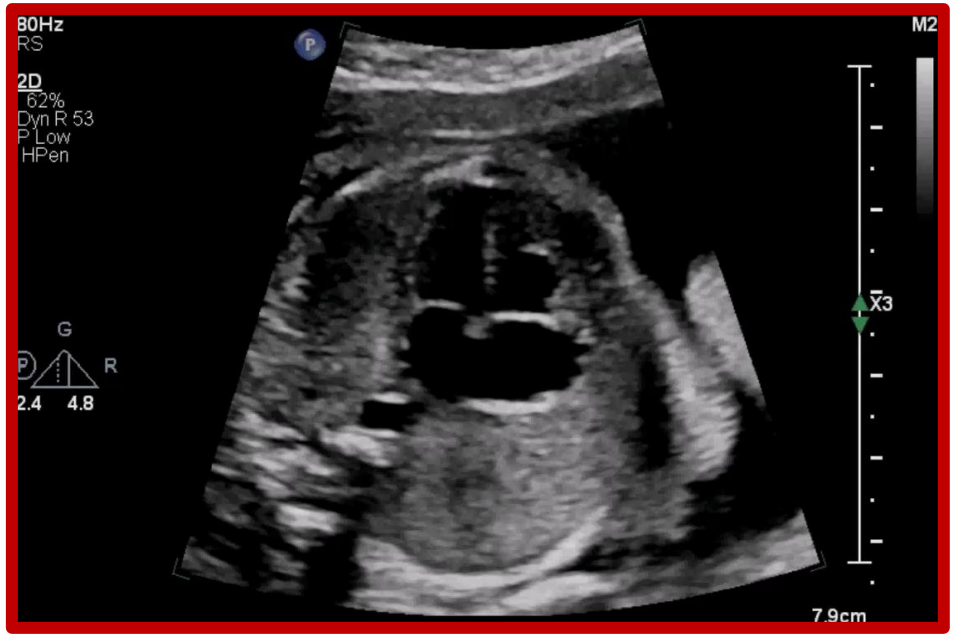
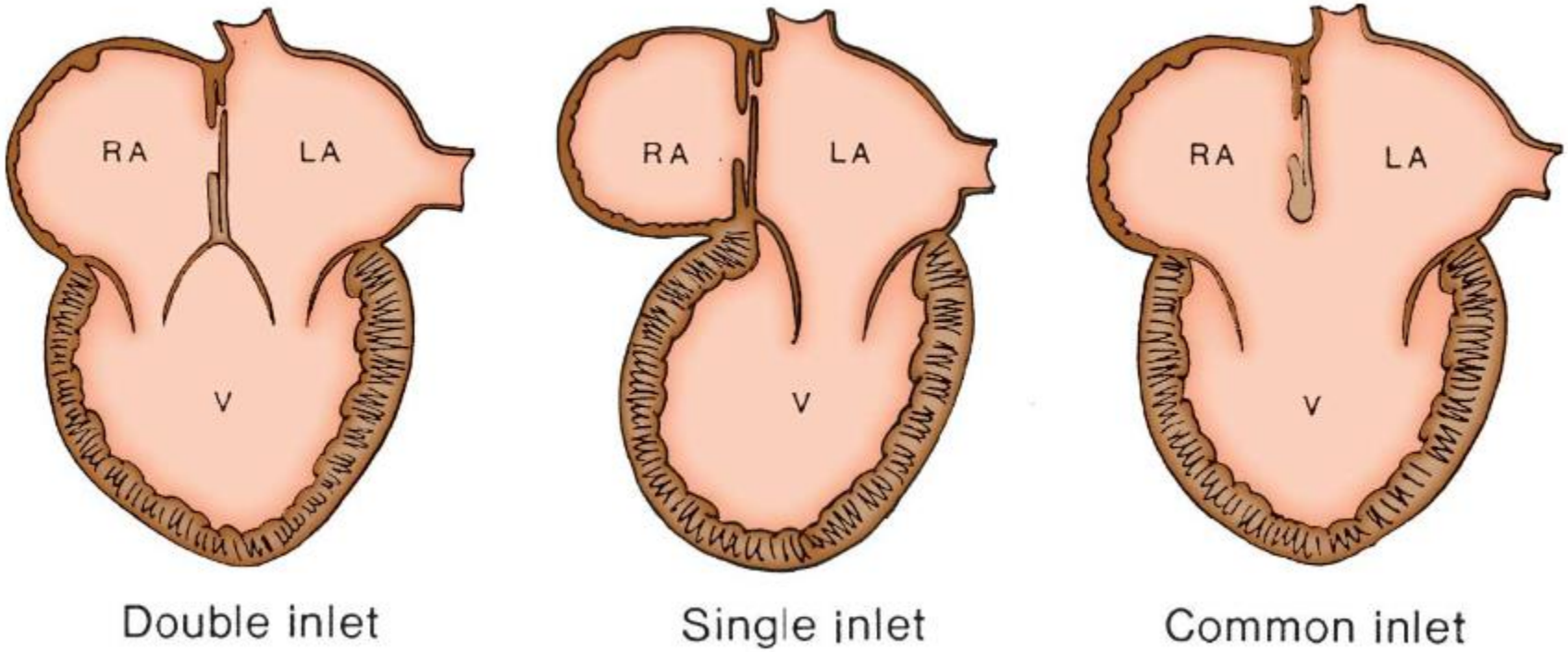
Four Chamber View



Single Ventricle



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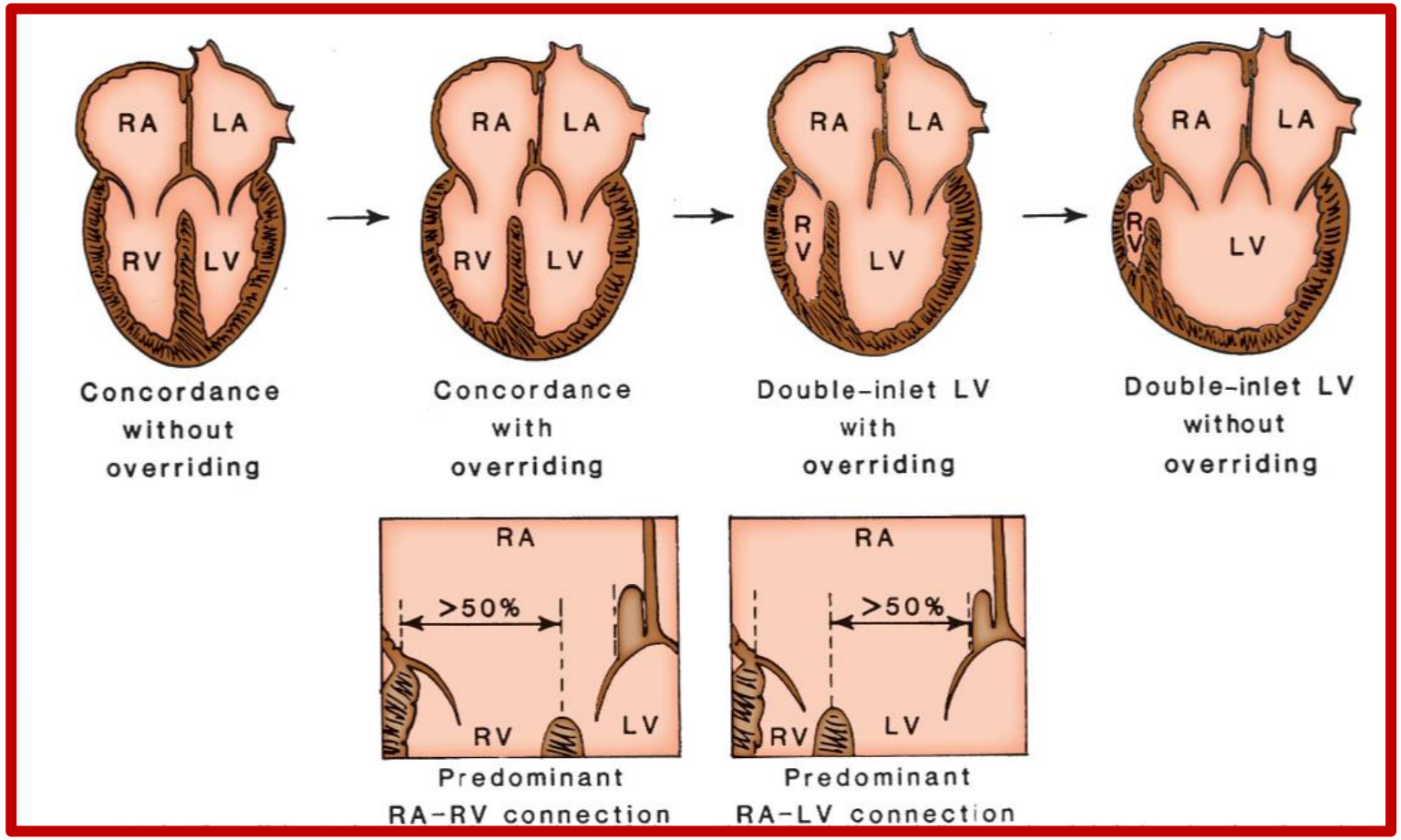

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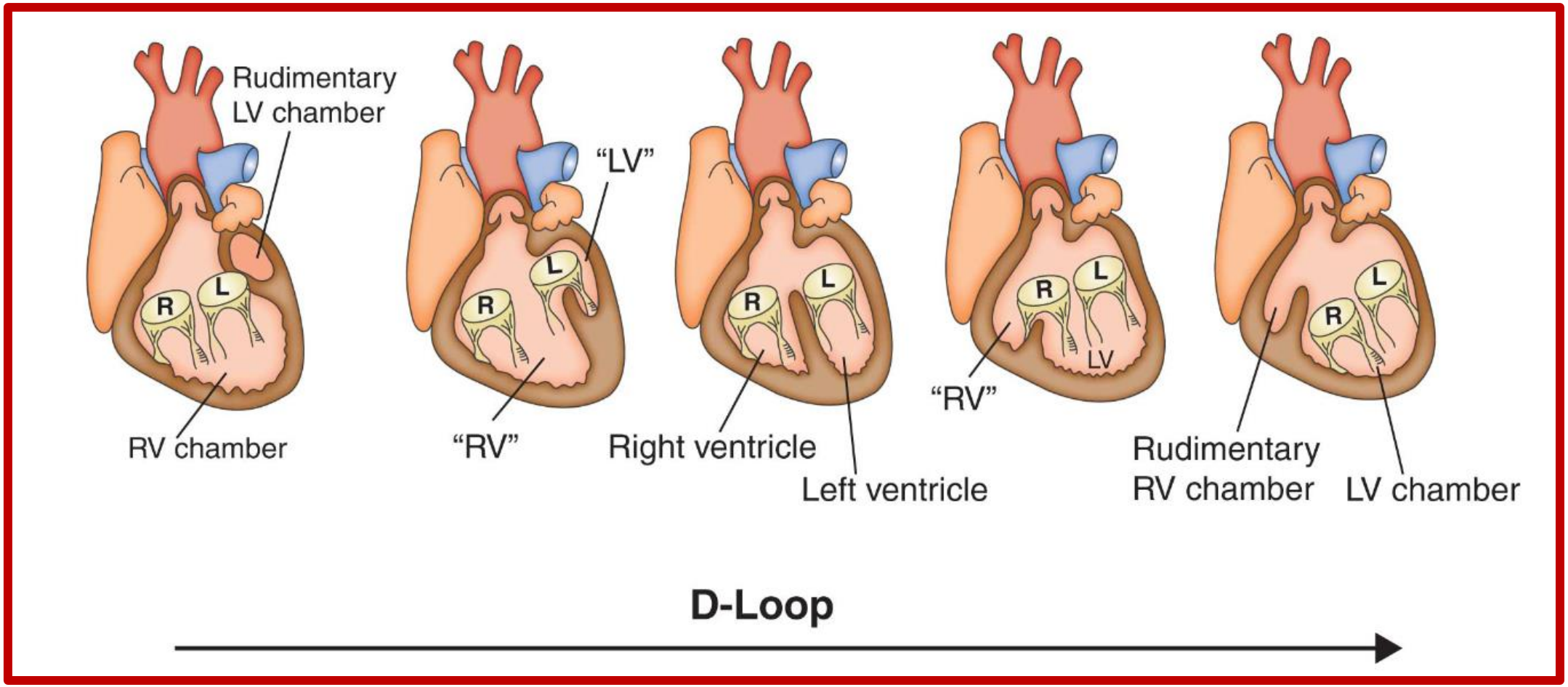
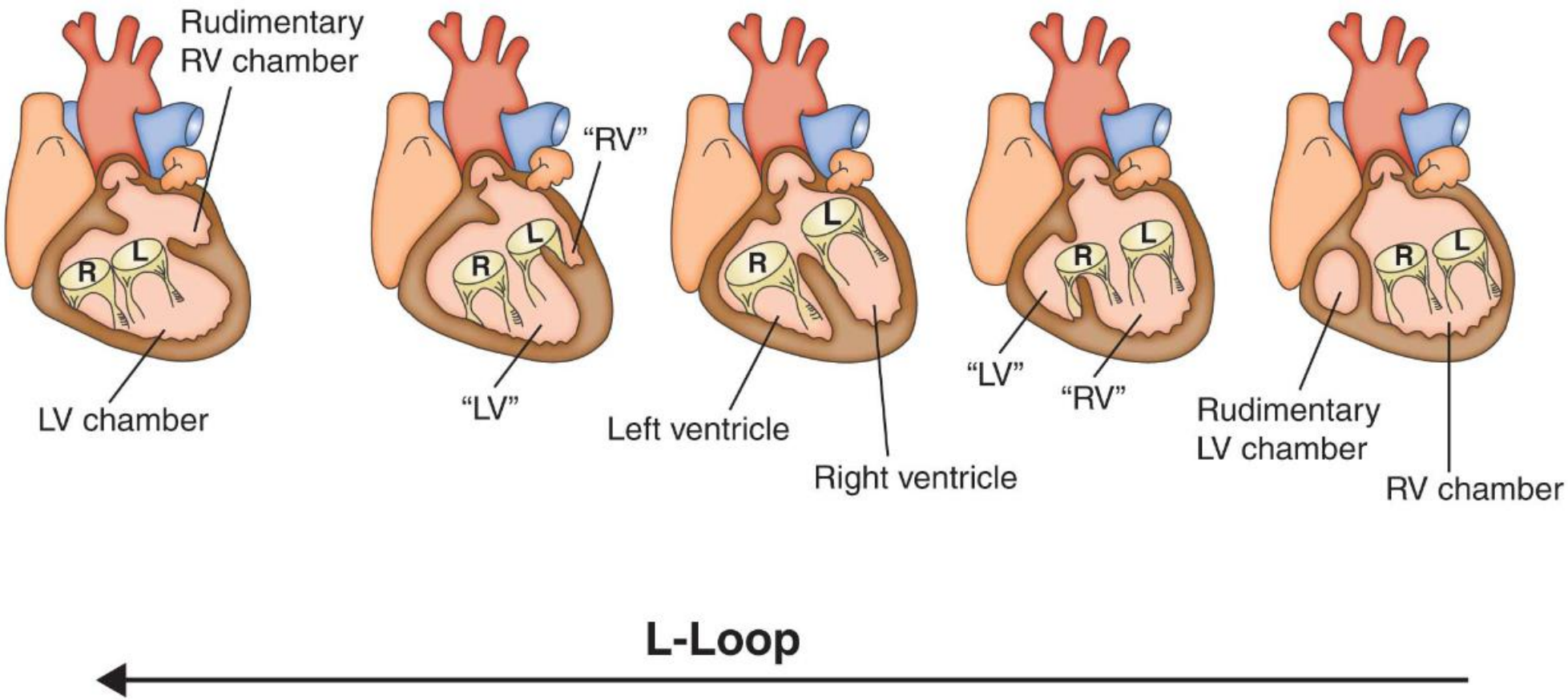


Single Ventricle



Double Inlet Left Ventricle

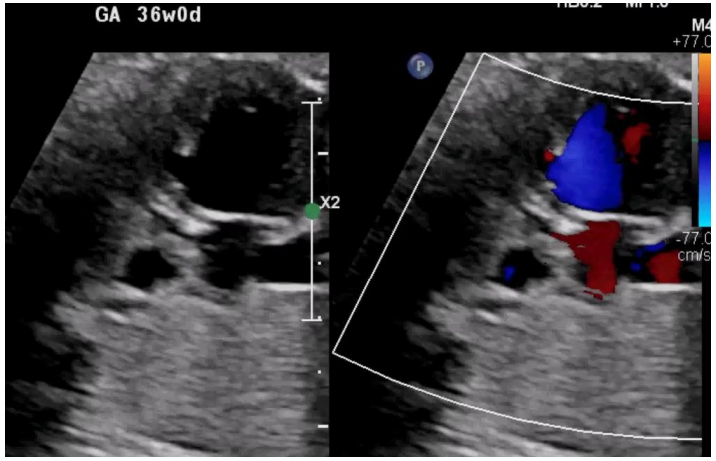
- The commonest form of Single Ventricle ~78%
- Four subgroups according to the Great Artery Relationship
  - I – Normally related great arteries
  - II – Right Anterior Aorta
  - III – Left Anterior Aorta
  - IV – Left posterior Aorta (inverted)

- Subaortic Obstruction (usually at the level of the BVF)
- Pulmonary Stenosis

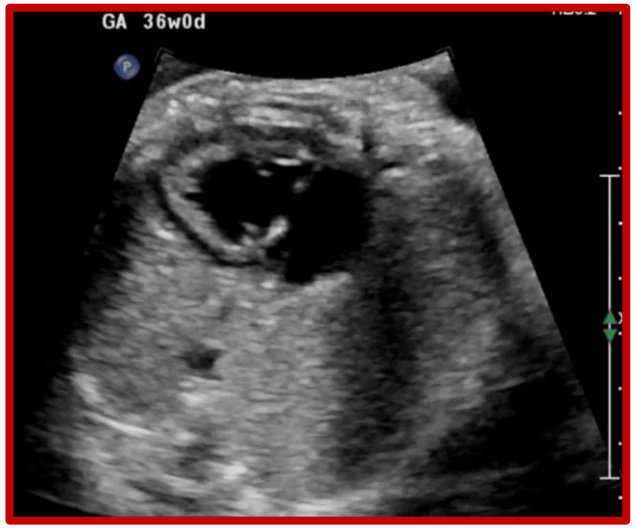
Double Inlet Left Ventricle

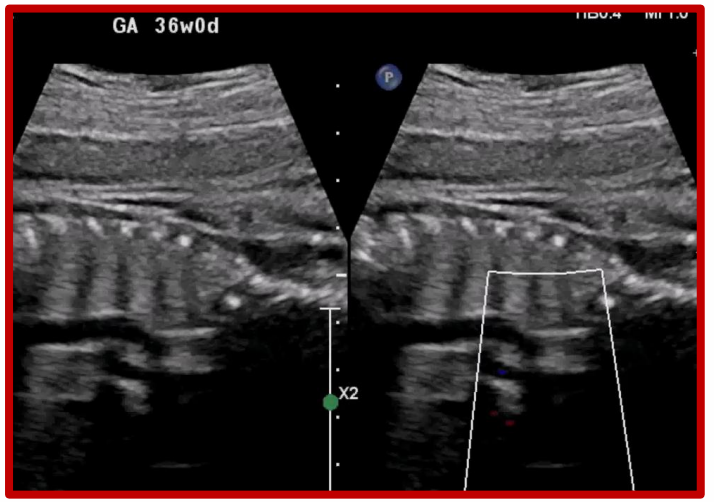
- DILV with left sided subaortic HRV and discordant VA connections
- Commonest ~38%
- Subpulmonary Obstruction

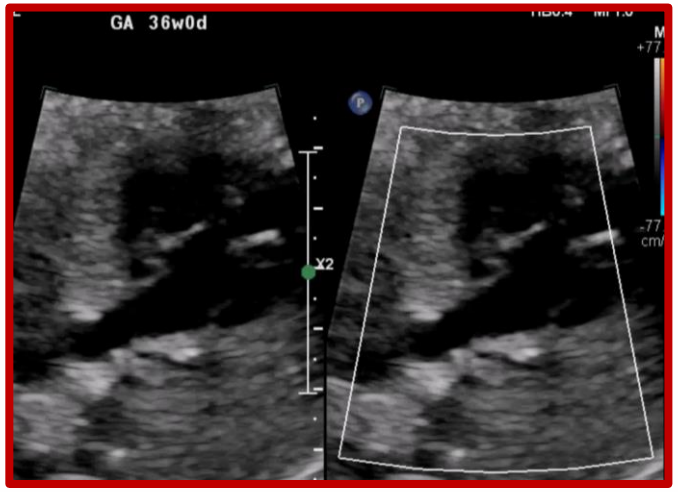
Double Inlet Left Ventricle

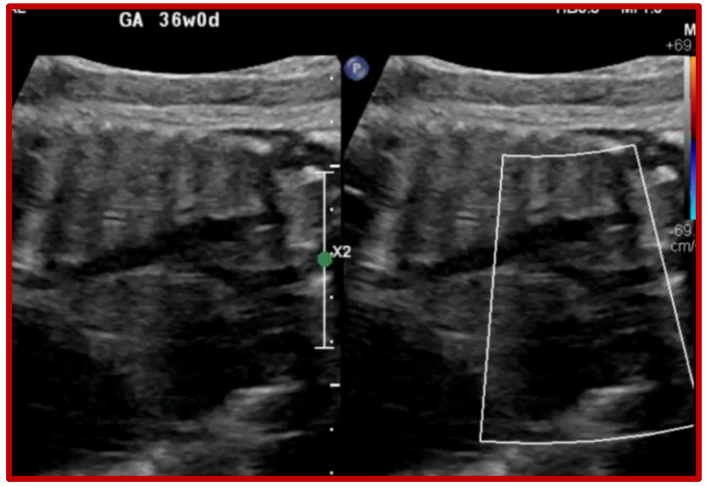


Double Inlet Left Ventricle







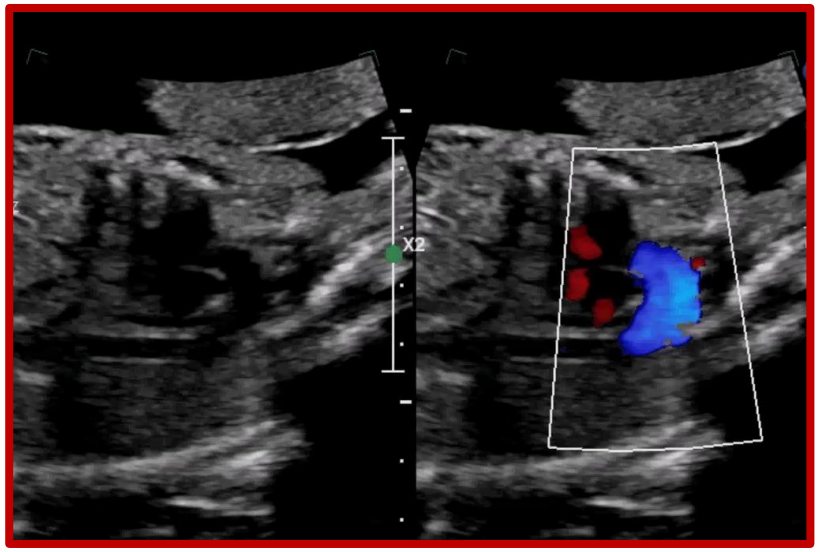


Double Inlet Left Ventricle

- DILV with Rt sided HRV
- ~ 25%
- TGA physiology

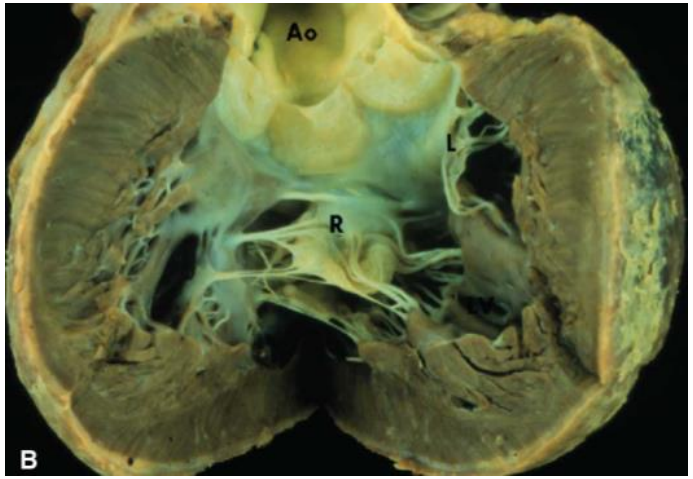
Double Inlet Left Ventricle

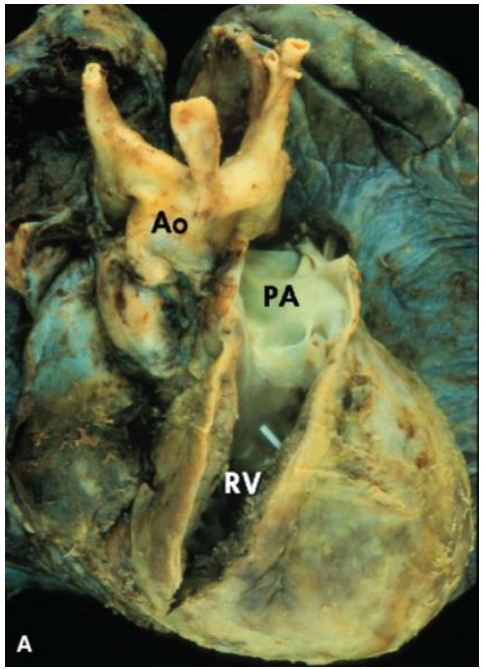
Double Inlet Left Ventricle



Double Inlet Left Ventricle

- DILV with NRGA – Holmes Heart
- Relatively rare ~15%
- Subpulmonary Obstruction





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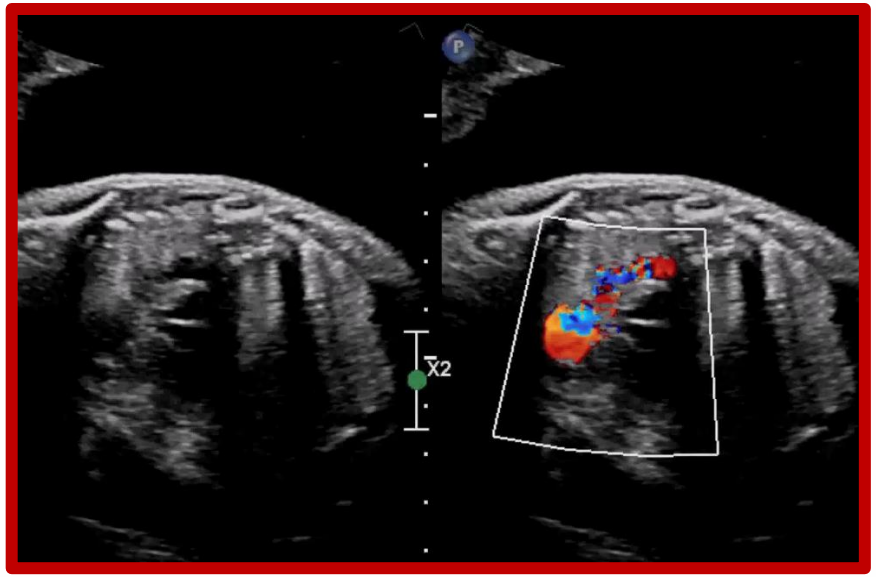

Counseling - Management

- Single ventricle by definition will go down the SV pathway
- At least 3 surgical interventions will be needed
- Prognosis depends on the individual anatomic characteristics of each fetus
- When there is hypoplasia of one great artery PGE might be needed
- Number and kind of intervention will depend on anatomy
- In {S,L,L} DILV possibility of need of a pacemaker

Conclusions

- Functional single ventricle although rare has a broad spectrum of ‘phenotypes’
- Important elements for accurate diagnosis as well as prognosis are:
  - Atrial situs
  - Atrioventricular alignments – connections
  - AV valve morphology
  - Ventricular looping and morphology
  - Communication of the dominant to the outlet (rudimentary chamber)
  - Relation and size of the great arteries
  - Heart rate and AV conduction


LOVE WILL.