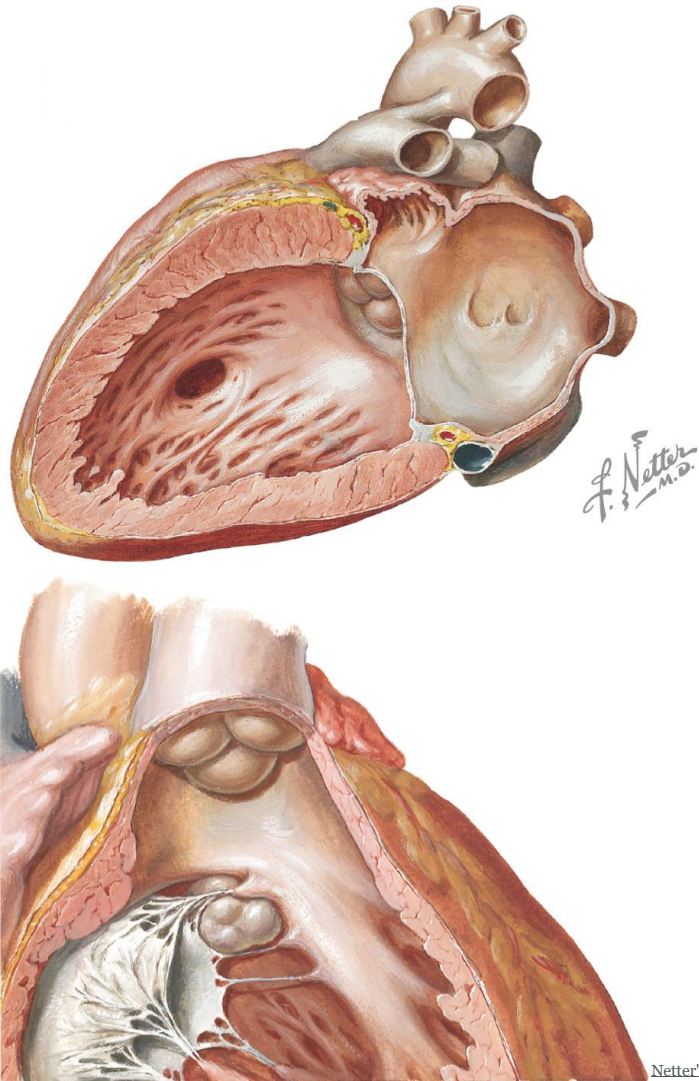


6TH PEDIATRIC ECHOCARDIOGRAPHY SYMPOSIUM  
SEPTEMBER 7<sup>TH</sup> 2024

# Ventricular Septal Defects: Anatomy and Physiology

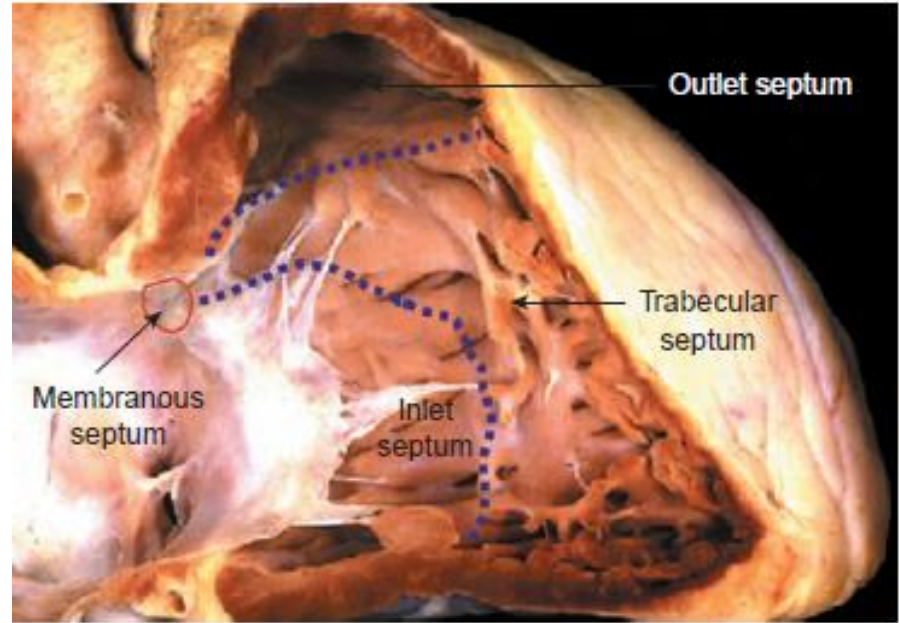
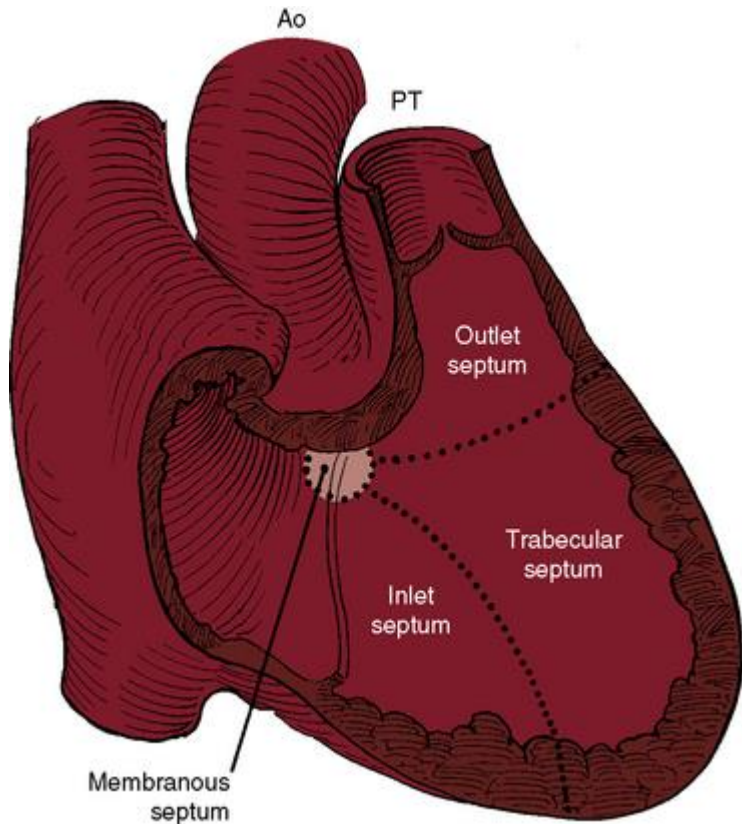
Charlie Simpkin, DO  
Advanced Cardiac Imaging Fellow  
Pulmonary Hypertension Fellow





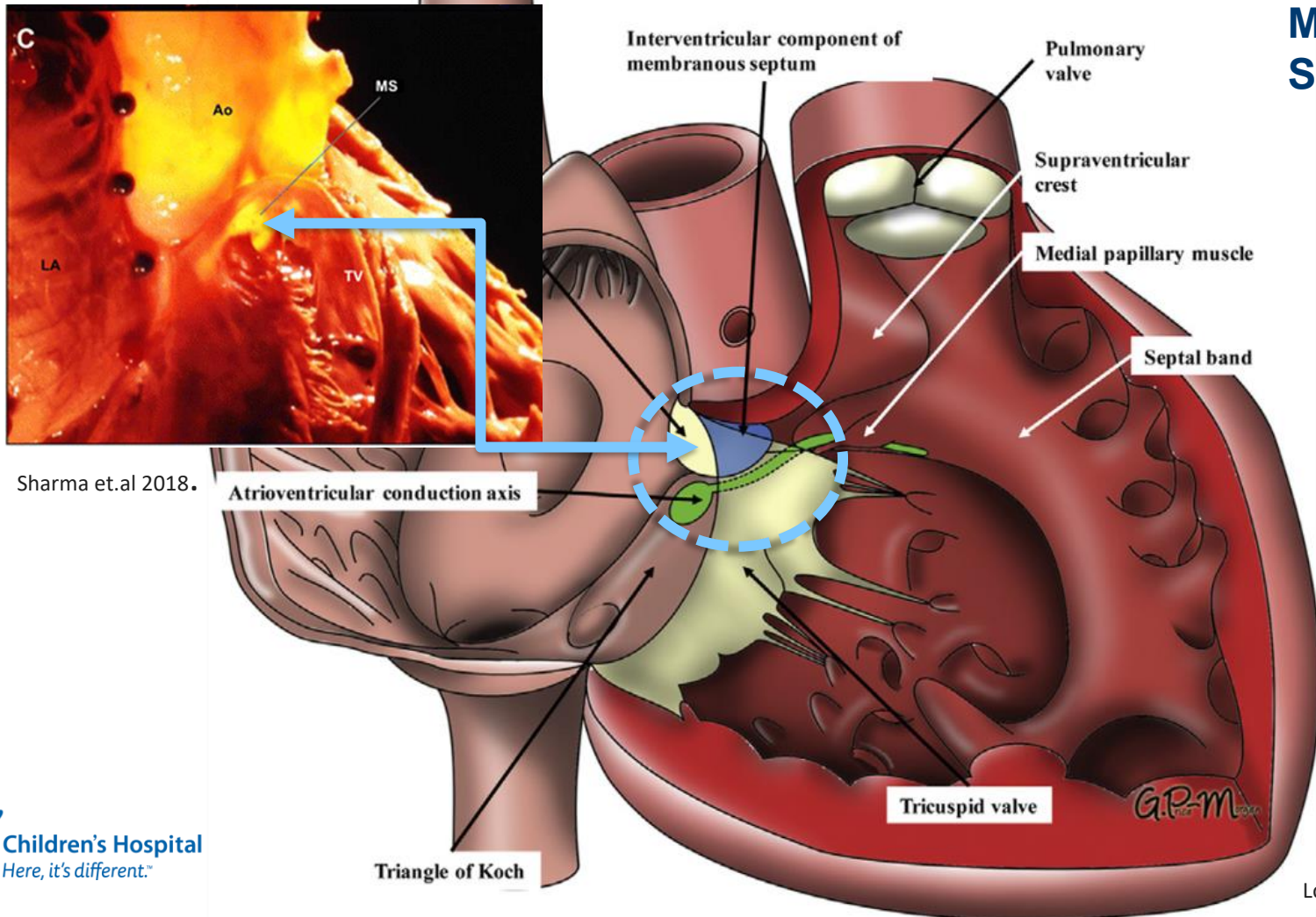
# Disclosures:

- **None**



(Modified from Anderson RH, Becker AE, Lucchese FE, Meier MA, Rigby ML, Soto B. Forbus and Shirali 2009  
 Morphology of congenital heart disease. Baltimore: University Park Press; 1983)

# Membranous Septum



Sharma et.al 2018.

Atrioventricular conduction axis

Interventricular component of membranous septum

Pulmonary valve

Supraventricular crest

Medial papillary muscle

Septal band

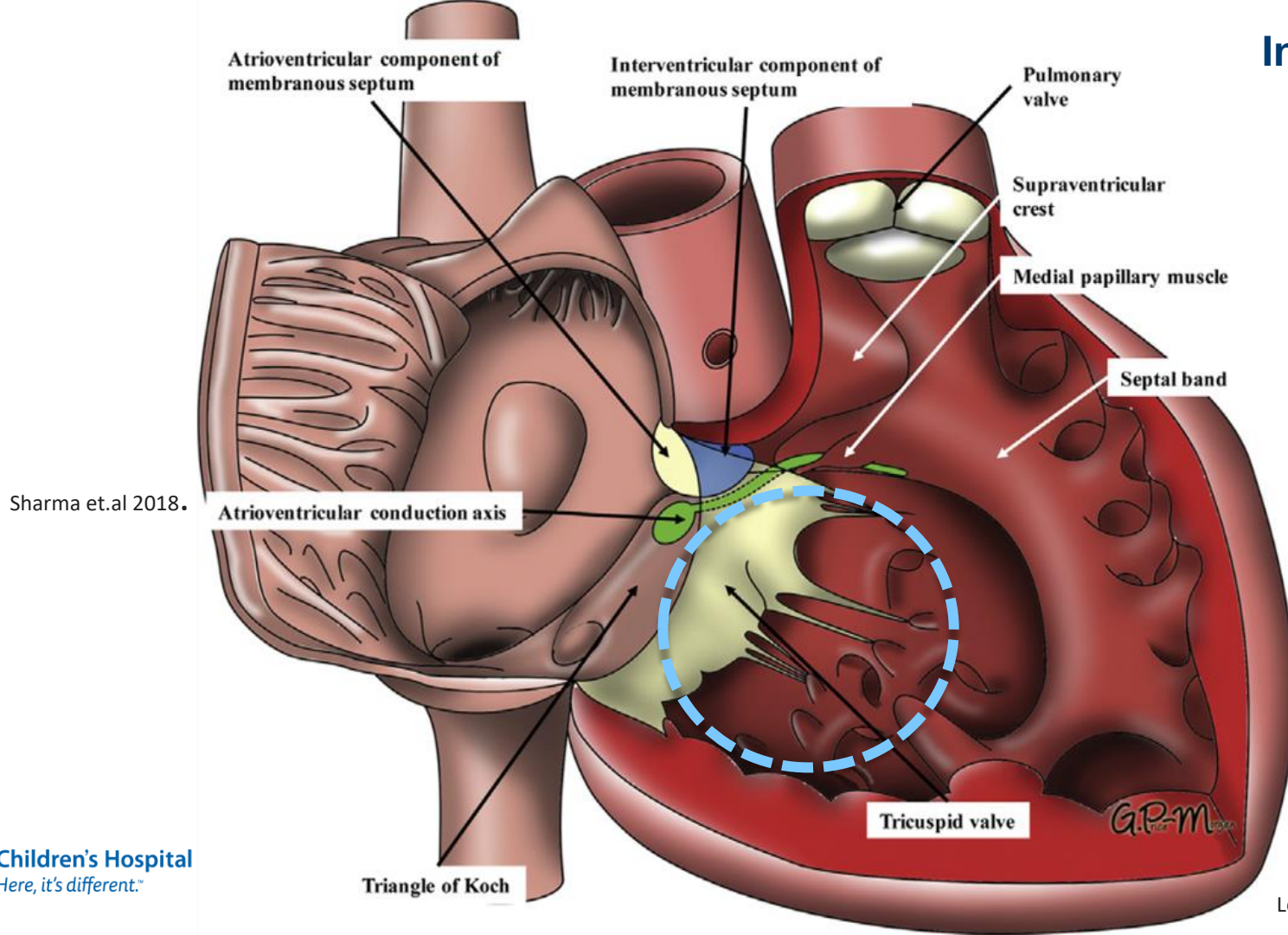
Tricuspid valve

Triangle of Koch

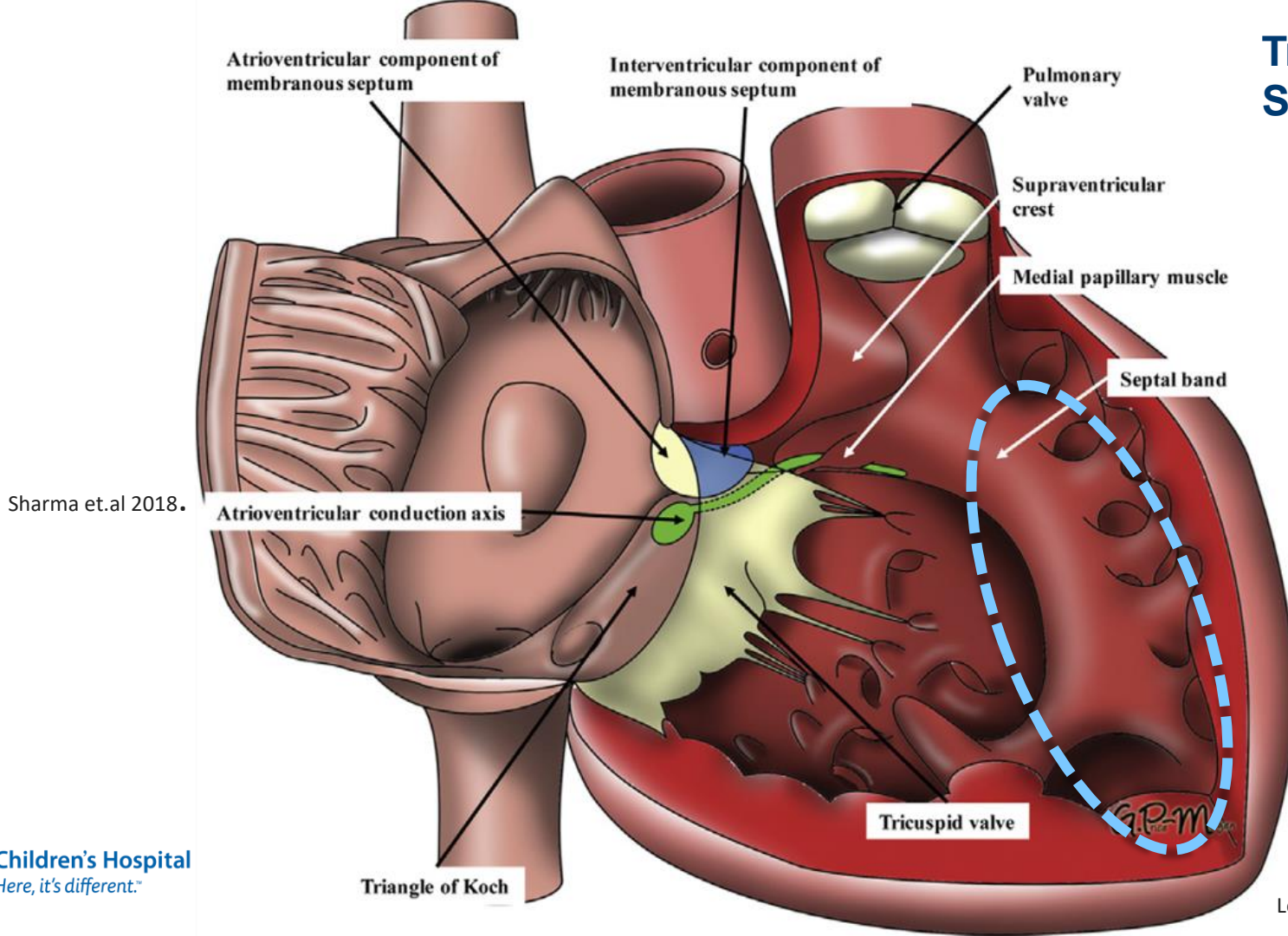
G.P.M. 2008

Lopez L, et.al 2018.

# Inlet Septum

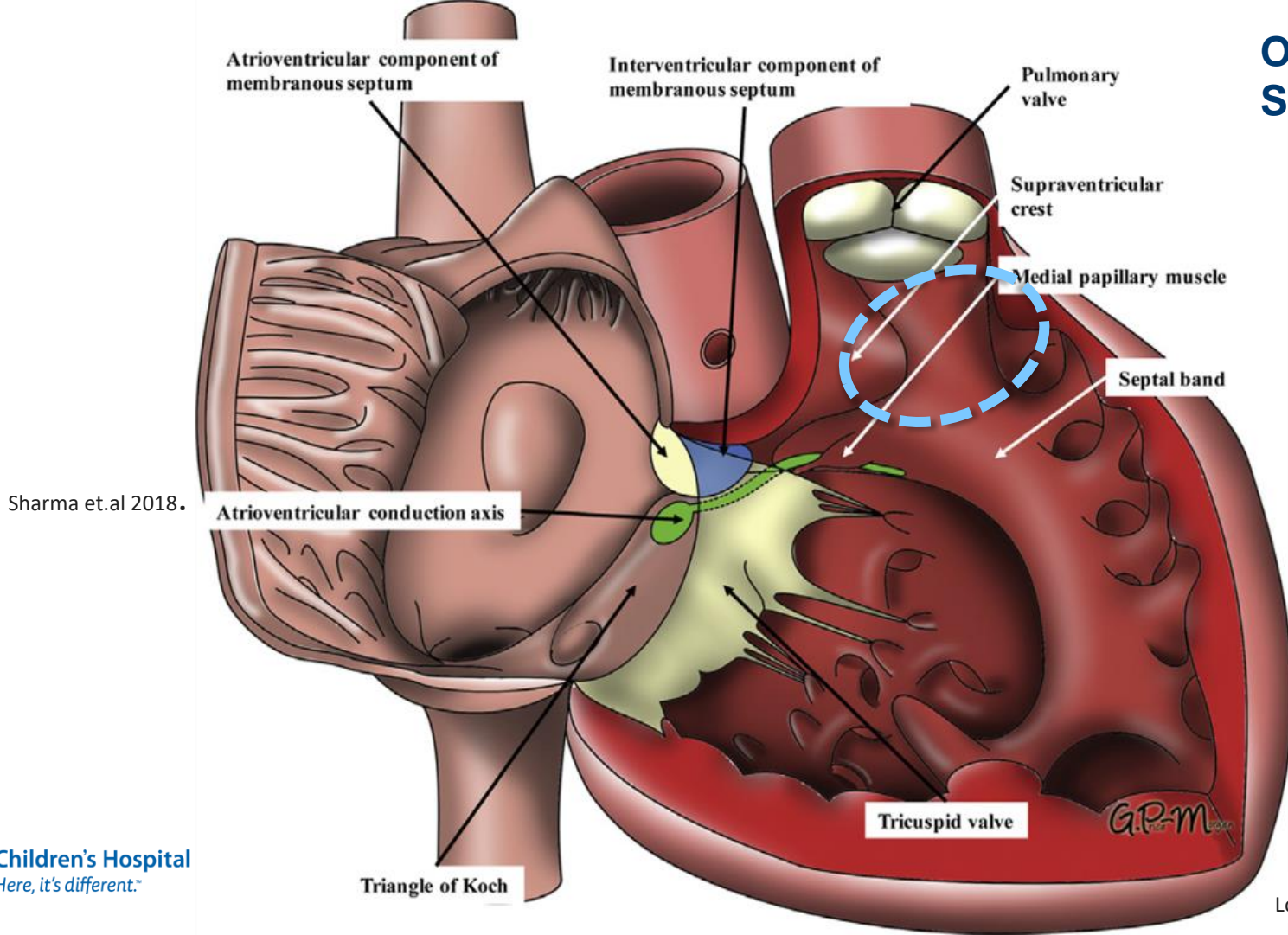


# Trabecular Septum



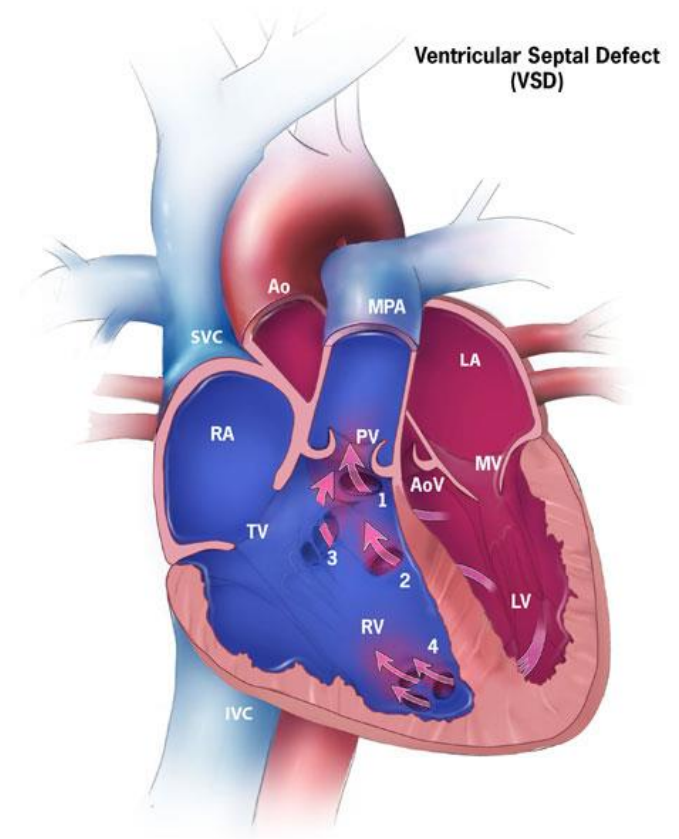
Sharma et.al 2018.

# Outlet Septum



# VSD Physiology

## Blood Flows Down The Path Of Least Resistance



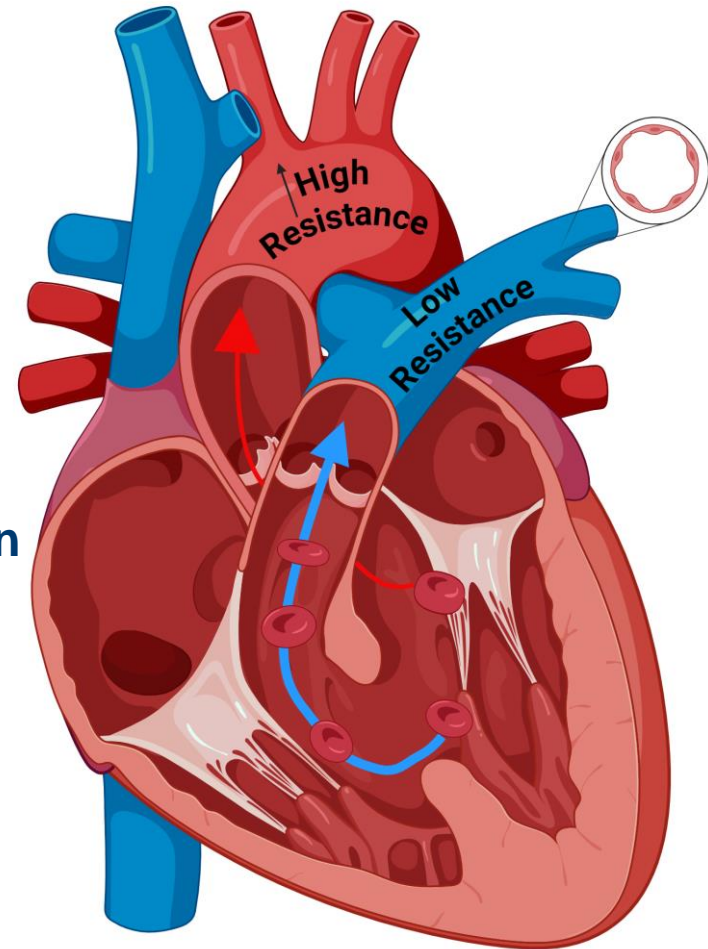
- |                     |                            |                     |                                |
|---------------------|----------------------------|---------------------|--------------------------------|
| RA. Right Atrium    | SVC. Superior Vena Cava    | TV. Tricuspid Valve | 1. Conoventricular, malaligned |
| RV. Right Ventricle | IVC. Inferior Vena Cava    | MV. Mitral Valve    | 2. perimembranous              |
| LA. Left Atrium     | MPA. Main Pulmonary Artery | PV. Pulmonary Valve | 3. inlet                       |
| LV. Left Ventricle  | Ao. Aorta                  | AoV. Aortic Valve   | 4. muscular                    |



# VSD Physiology

## Typical Course:

- As PVR falls: L → R Shunting
- ↑ Pulmonary blood flow
- Volume loads LA and LV
- Stimulates catecholamine response to maintain cardiac output to systemic circulation
- Congestive heart failure develops ~3-4 months of life

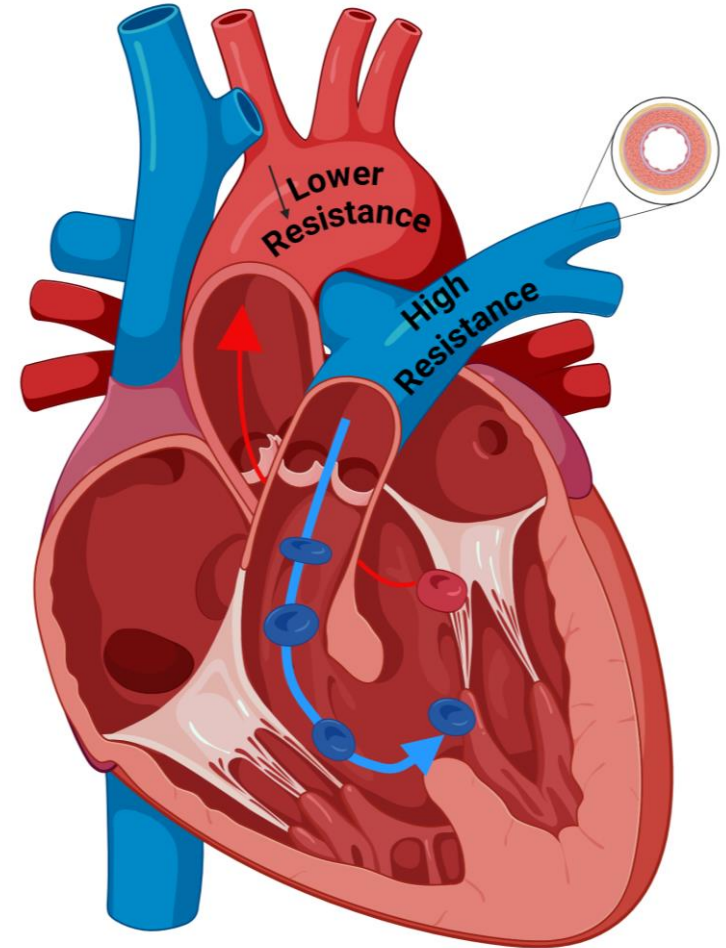


# VSD Physiology

Left untreated:

- Pulmonary vascular remodeling
- Reversal of shunt R → L
- “Eisenmenger Physiology”

Depending on the child, ideal age for repair is  
4-6mo of life





# Defects

# Classification of Ventricular Septal Defects for the Eleventh Iteration of the International Classification of Diseases—Striving for Consensus: A Report From the International Society for Nomenclature of Paediatric and Congenital Heart Disease

Presented at the Seventh World Congress of Paediatric Cardiology and Cardiac Surgery, Barcelona, Spain, July 16–21, 2017.

[Leo Lopez](#), MD<sup>a,\*,†</sup>, [Lucile Houyel](#), MD<sup>b</sup>, [Steven D. Colan](#), MD<sup>c</sup>, [Robert H. Anderson](#), MD, PhD (Hon)<sup>d</sup>, [Marie J. Béland](#), MDCM<sup>e</sup>, [Vera D. Aiello](#), MD, PhD<sup>f</sup>, [Frederique Bailliard](#), MD, MS<sup>g</sup>, [Meryl S. Cohen](#), MD<sup>h</sup>, [Jeffrey P. Jacobs](#), MD<sup>i</sup>, [Hiromi Kurosawa](#), MD<sup>j</sup>, [Stephen P. Sanders](#), MD<sup>c</sup>, [Henry L. Walters III](#), MD<sup>k</sup>, [Paul M. Weinberg](#), MD<sup>h</sup>, [Jeffrey R. Boris](#), MD<sup>h</sup>, [Andrew C. Cook](#), PhD<sup>l</sup>, [Adrian Crucean](#), MD, PhD<sup>m</sup>, [Allen D. Everett](#), MD<sup>n</sup>, [J. William Gaynor](#), MD<sup>n</sup>, [Jorge Giroud](#), MD<sup>i</sup>, [Kristine J. Guleserian](#), MD<sup>a</sup>, [Marina L. Hughes](#), DPhil, FRACP<sup>o</sup>, [Amy L. Juraszek](#), MD<sup>p</sup>, [Otto N. Krogmann](#), MD<sup>q</sup>, [Bohdan J. Maruszewski](#), MD, PhD<sup>r</sup>, [James D. St. Louis](#), MD<sup>s</sup>, [Stephen P. Seslar](#), MD, PhD<sup>t</sup>, [Diane E. Spicer](#), BS, PA<sup>u</sup>, [Shubhika Srivastava](#), MBBS<sup>v</sup>, [Giovanni Stellin](#), MD<sup>w</sup>, [Christo I. Tchervenkov](#), MD<sup>x</sup>, [Lianyi Wang](#), MD<sup>y</sup>, [Rodney C.G. Franklin](#), MD<sup>z</sup>



Children's Hospital Colorado  
Here, it's different.™



Department of Pediatrics  
SCHOOL OF MEDICINE  
UNIVERSITY OF COLORADO ANSCHUTZ MEDICAL CAMPUS

Table 1. *International Society for Nomenclature of Paediatric and Congenital Heart Disease Classification Scheme for Ventricular Septal Defect (International Paediatric and Congenital Cardiac Code 07.10.00) as Incorporated in the International Classification of Diseases-11th Iteration*

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1. Perimembranous central VSD (07.10.01)
2. Inlet VSD without a common AV junction (07.14.05)<sup>†</sup>
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  - b. Inlet perimembranous VSD with AV septal malalignment and without a common AV junction (07.14.06)
  - c. Inlet muscular VSD (07.11.02)
3. Trabecular muscular VSD (07.11.01)
  - a. Trabecular muscular VSD: midseptal (07.11.04)
  - b. Trabecular muscular VSD: apical (07.11.03)
  - c. Trabecular muscular VSD: postero-inferior (07.11.12)
  - d. Trabecular muscular VSD: anterosuperior (07.11.07)
  - e. Trabecular muscular VSD: multiple ("Swiss cheese" septum) (07.11.05)
4. Outlet VSD (07.12.00)
  - a. Outlet VSD without malalignment (07.12.09)
    - i. Outlet muscular VSD without malalignment (07.11.06)
    - ii. Doubly committed juxta-arterial VSD without malalignment (07.12.01)
      1. Doubly committed juxta-arterial VSD without malalignment and with a muscular postero-inferior rim (07.12.02)
      2. Doubly committed juxta-arterial VSD without malalignment and with a fibrous postero-inferior rim (perimembranous extension) (07.12.03)
  - b. Outlet VSD with anteriorly malaligned outlet septum (07.10.17)
    - i. Outlet muscular VSD with anteriorly malaligned outlet septum (07.11.15)
    - ii. Outlet perimembranous VSD with anteriorly malaligned outlet septum (07.10.04)
  - iii. Doubly committed juxta-arterial VSD with anteriorly malaligned fibrous outlet septum (07.12.12)
    1. Doubly committed juxta-arterial VSD with anteriorly malaligned fibrous outlet septum and a muscular postero-inferior rim (07.12.07)
    2. Doubly committed juxta-arterial VSD with anteriorly malaligned fibrous outlet septum and a fibrous postero-inferior rim (perimembranous extension) (07.12.05)
  - c. Outlet VSD with posteriorly malaligned outlet septum (07.10.18)
    - i. Outlet muscular VSD with posteriorly malaligned outlet septum (07.11.16)
    - ii. Outlet perimembranous VSD with posteriorly malaligned outlet septum (07.10.19)
  - iii. Doubly committed juxta-arterial VSD with posteriorly malaligned fibrous outlet septum (07.12.13)
    1. Doubly committed juxta-arterial VSD with posteriorly malaligned fibrous outlet septum and a muscular postero-inferior rim (07.12.08)
    2. Doubly committed juxta-arterial VSD with posteriorly malaligned fibrous outlet septum and a fibrous postero-inferior rim (perimembranous extension) (07.12.06)

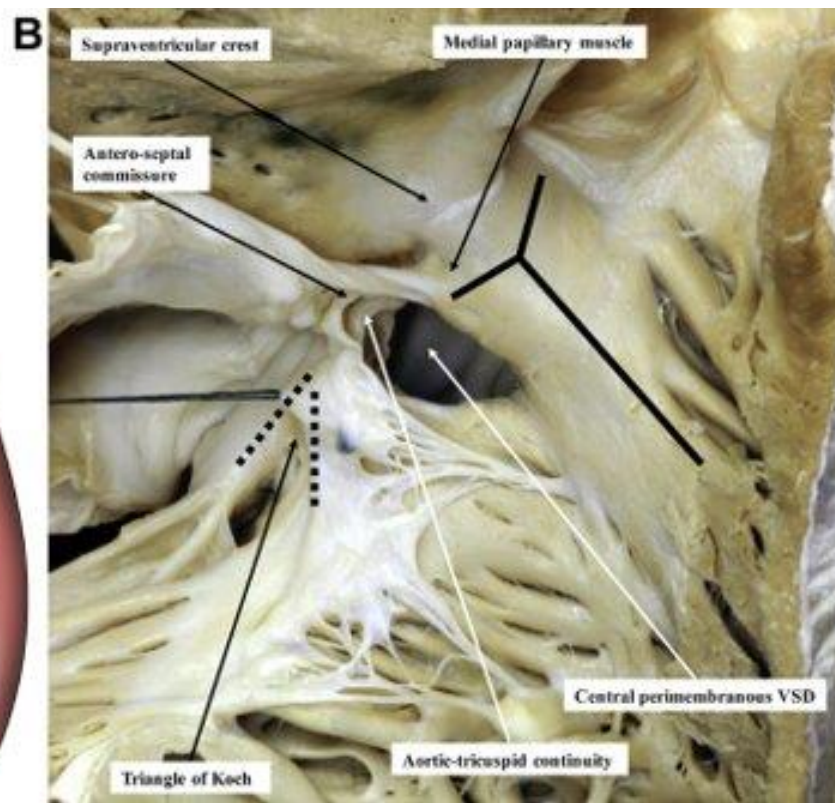
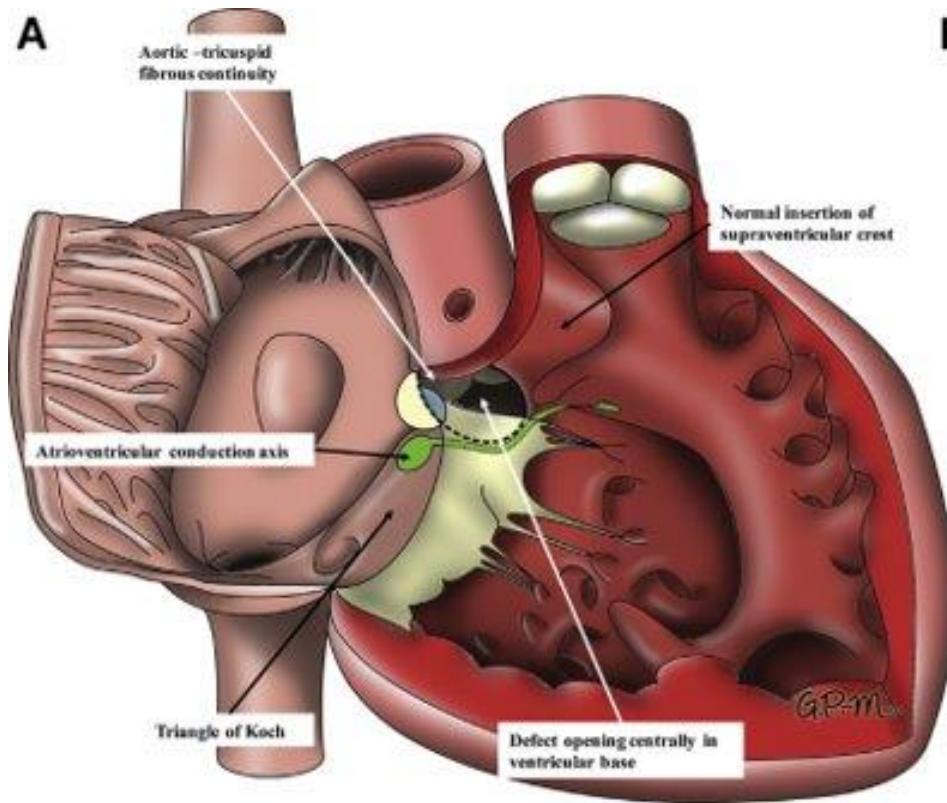
Defects are named based on:

**Their geographic location**  
**And**  
**Their margins (borders)**

# Central Perimembranous

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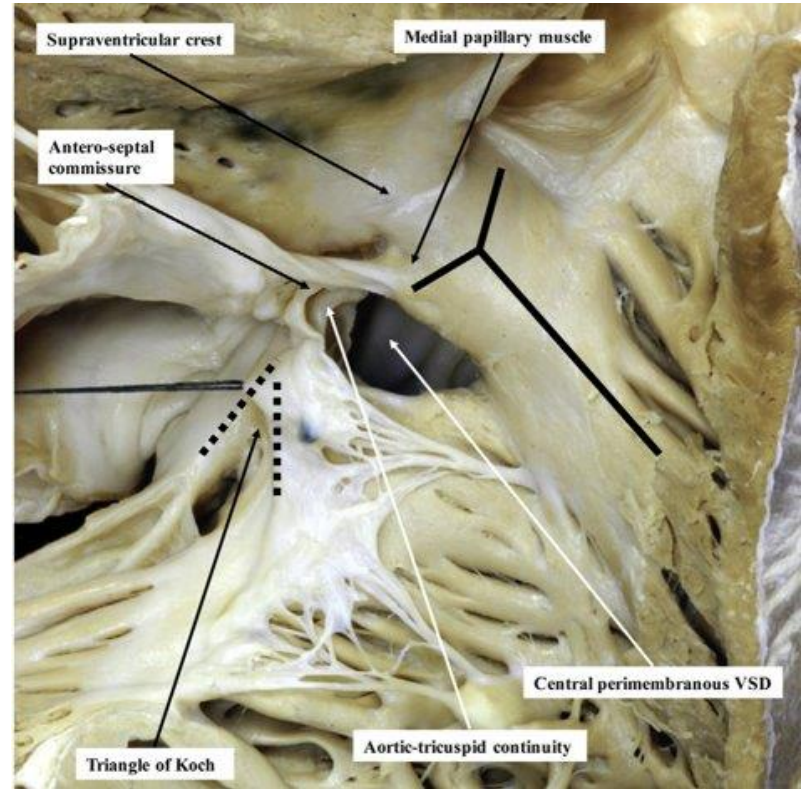
Lopez L, et.al 2018.



- Location

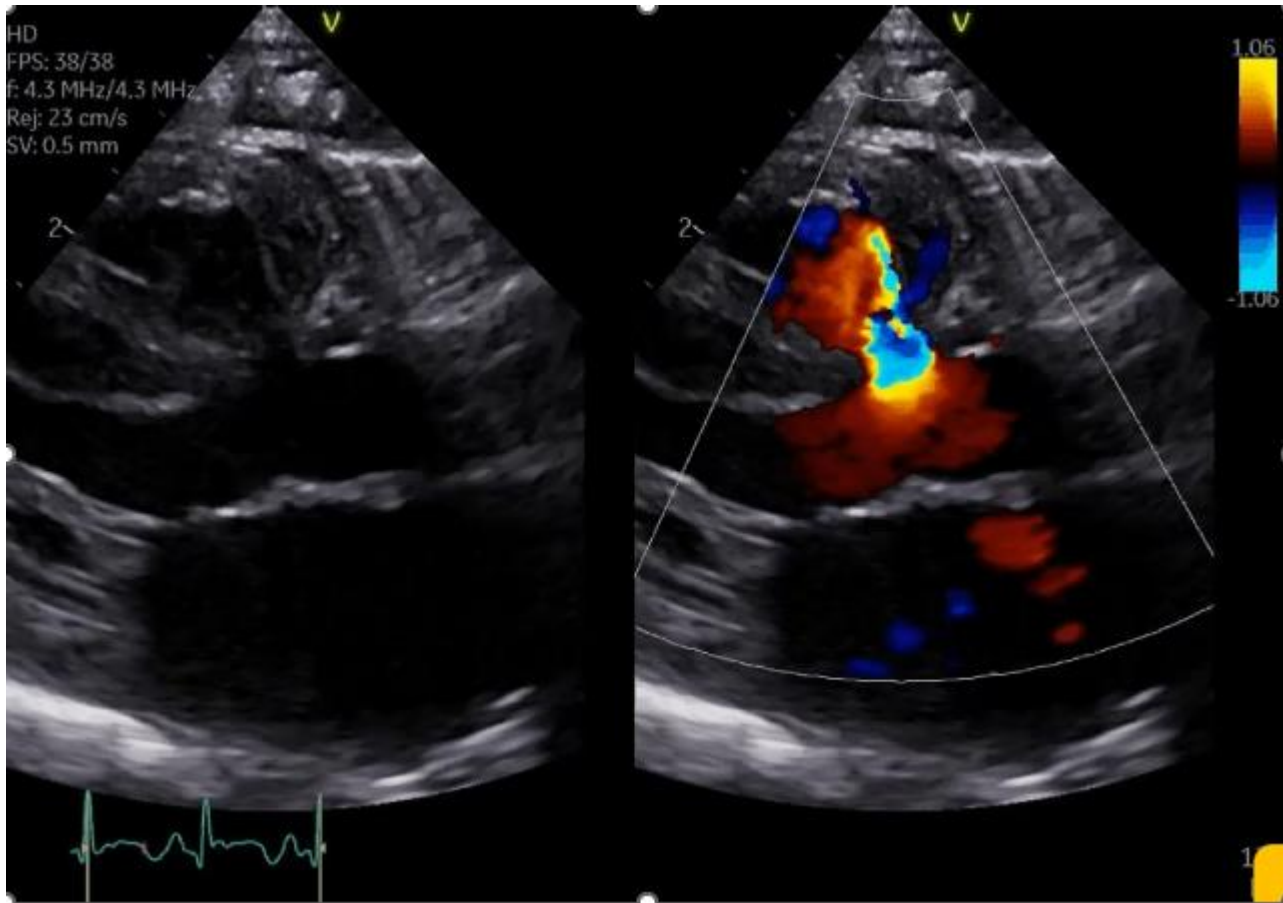
- Opens in Center of interventricular part of membranous septum
- Behind the septal leaflet of tricuspid valve
- Below right/non coronary cusps of aortic valve
- Below postero-inferior limb of septal band (Y) and medial papillary muscle

- Fibrous Continuity between TV and AoV



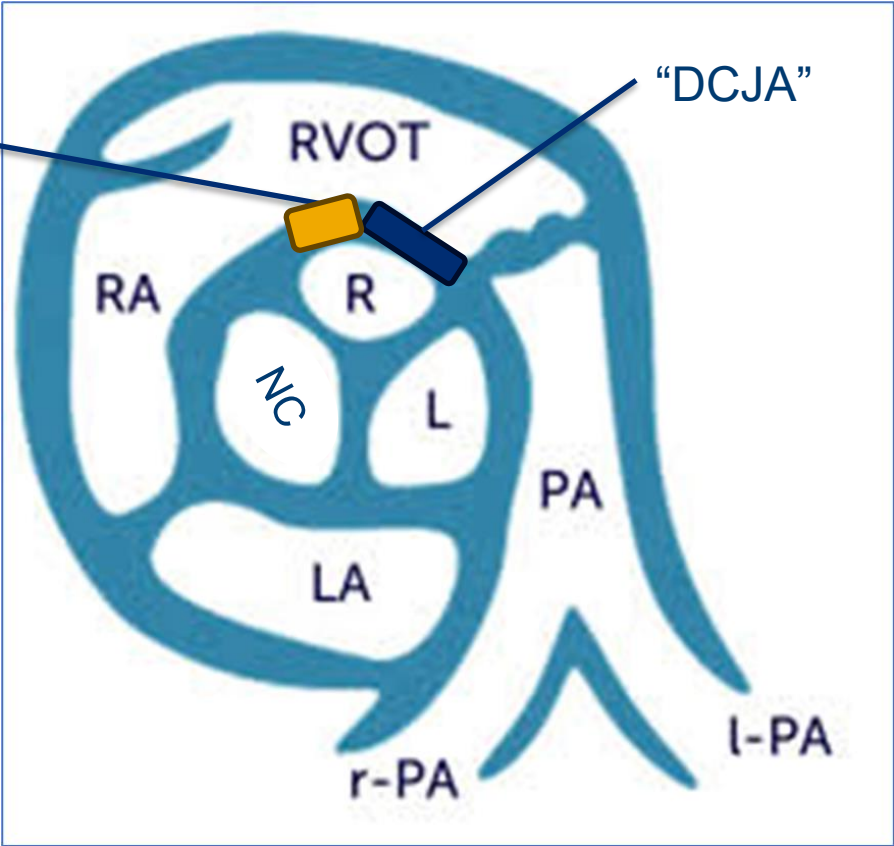
Lopez L, et.al 2018.

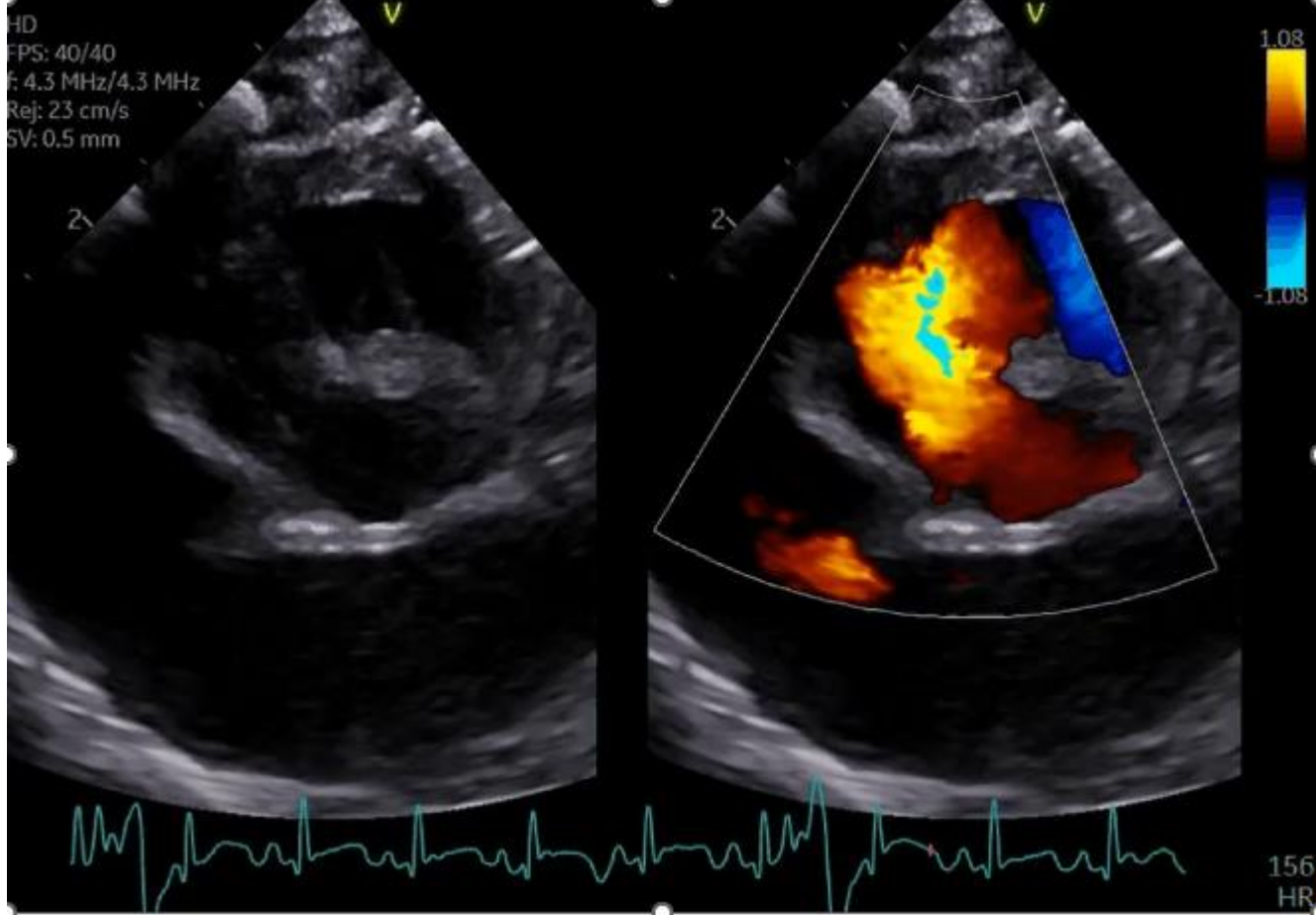
Central  
Perimembranous



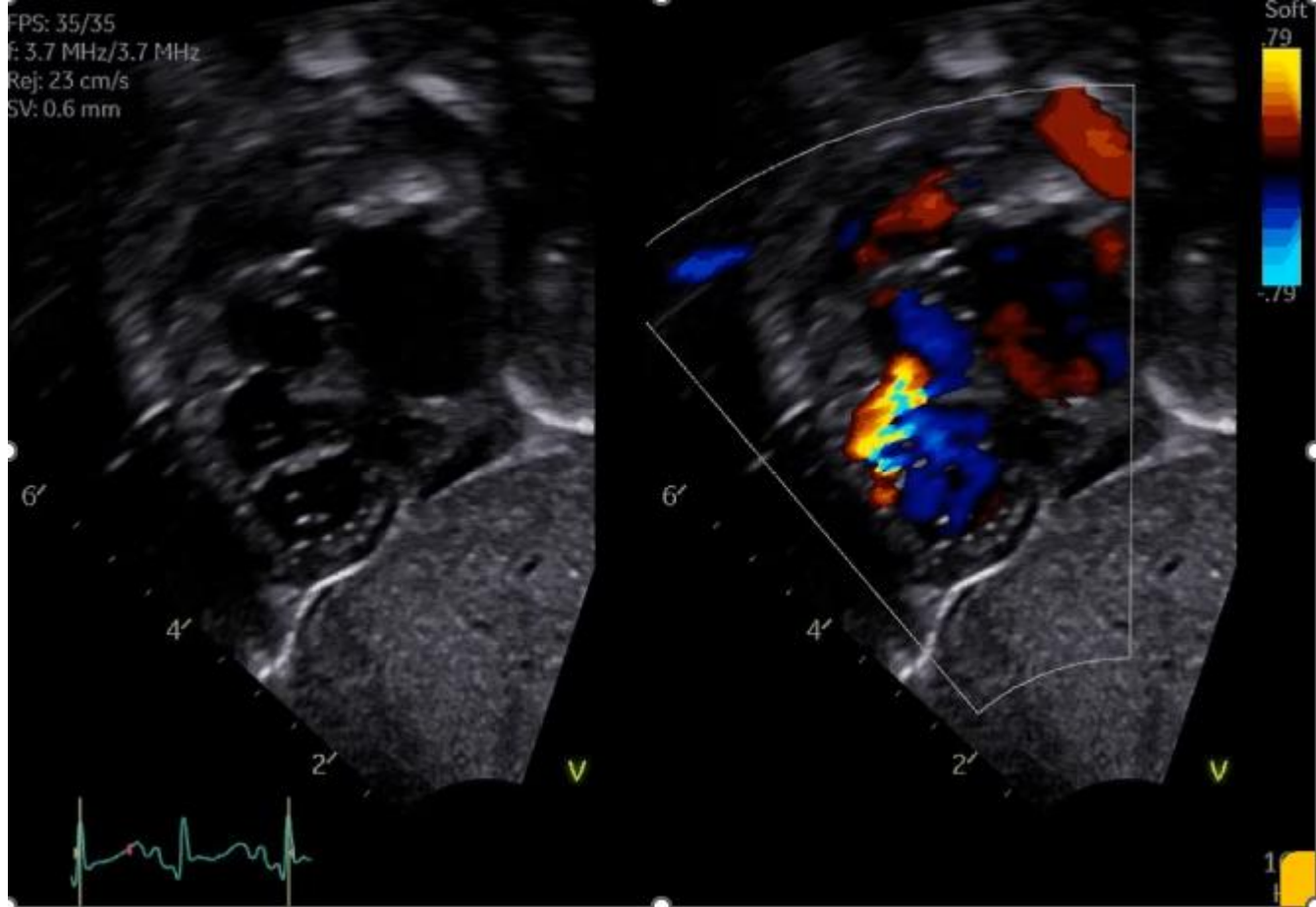
Central  
Perimembranous

Central  
Perimembranous





Central  
Perimembranous



Central  
Perimembranous

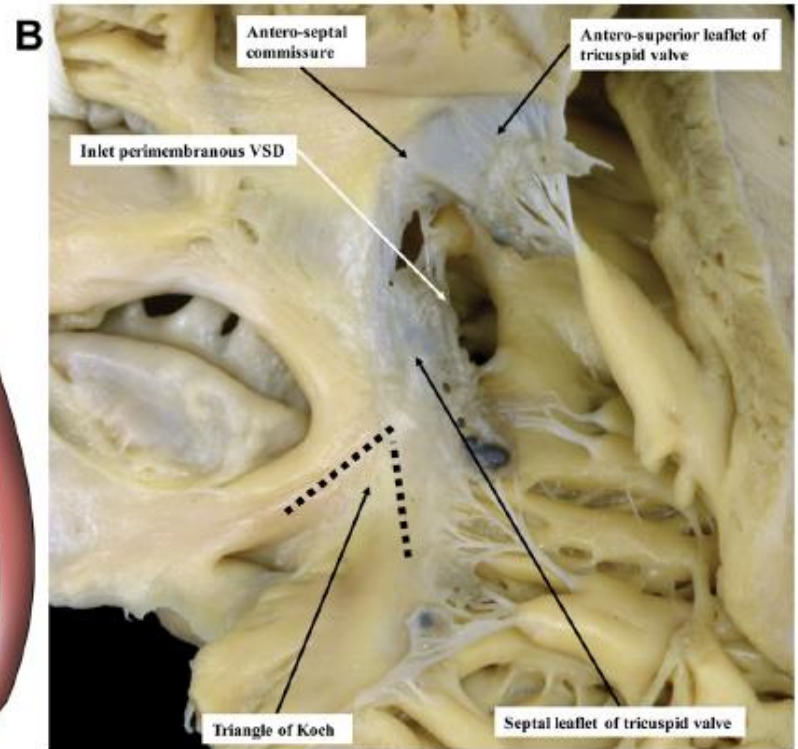
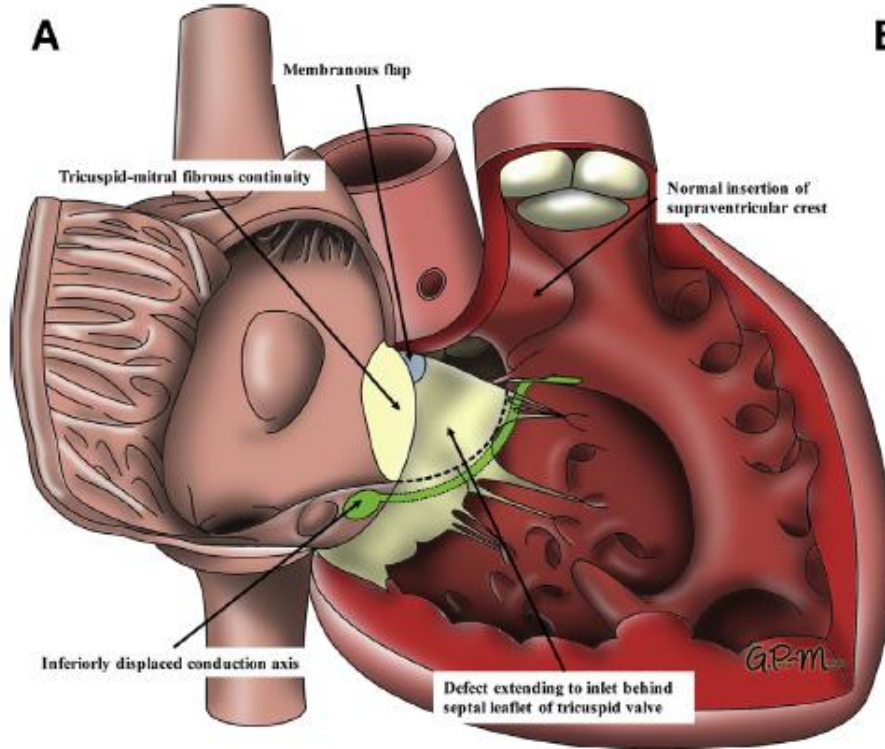
# Inlet

= AVSD\*

\*There is a SUPER rare occurrence where an Inlet VSD is present with straddling TV but NOT in association with an AVSD

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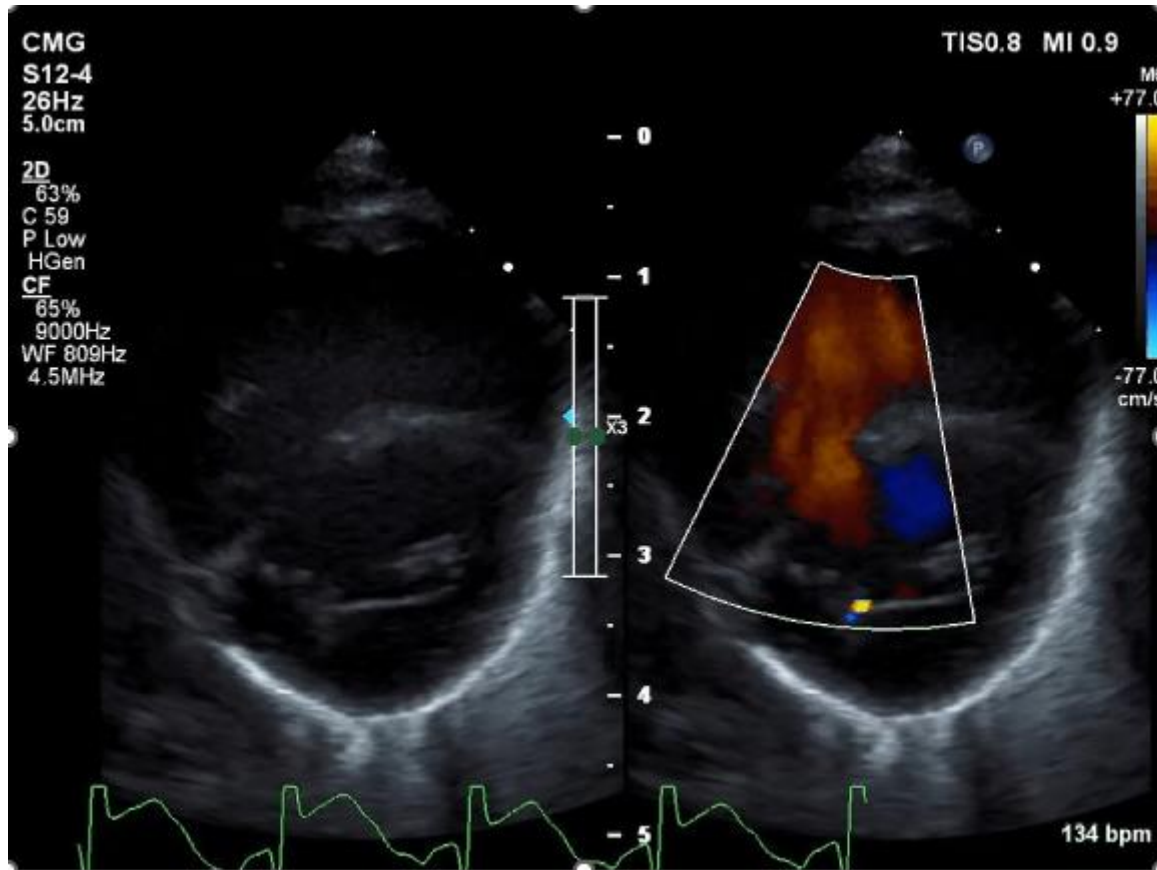
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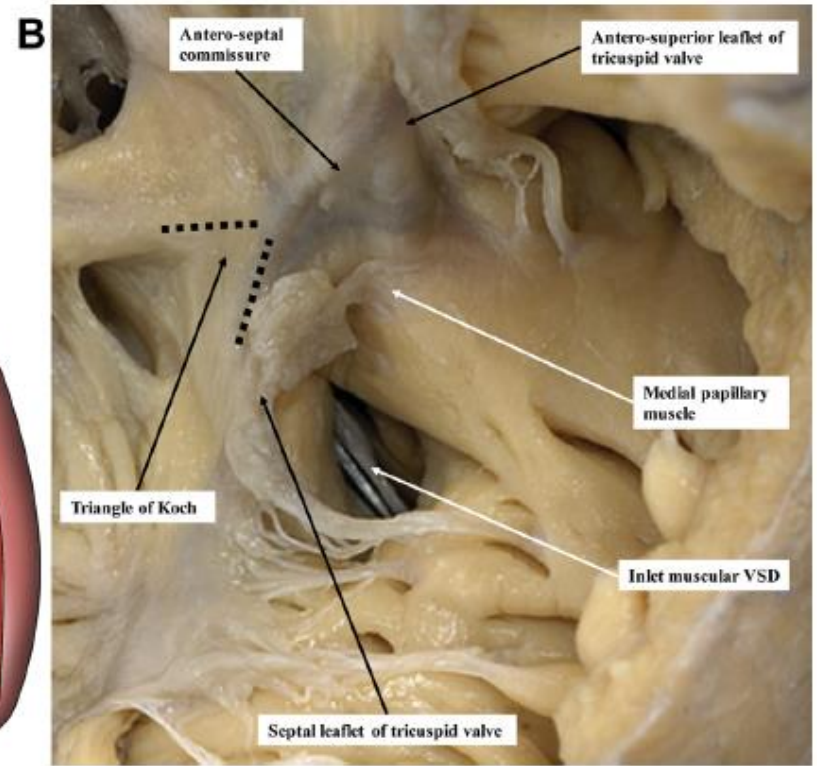
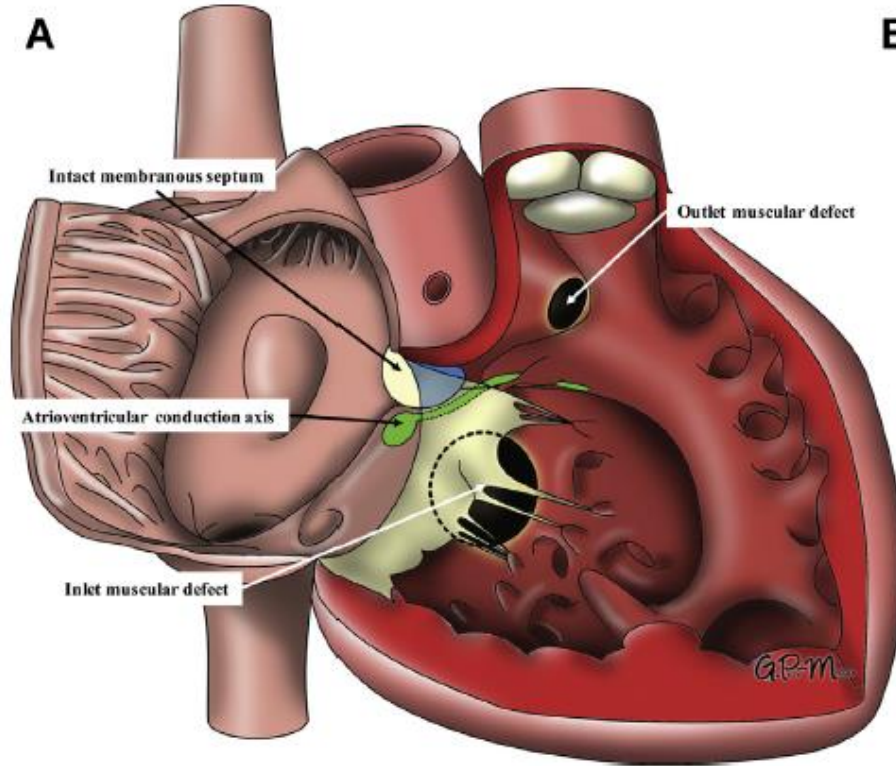


Inlet  
Perimembranous

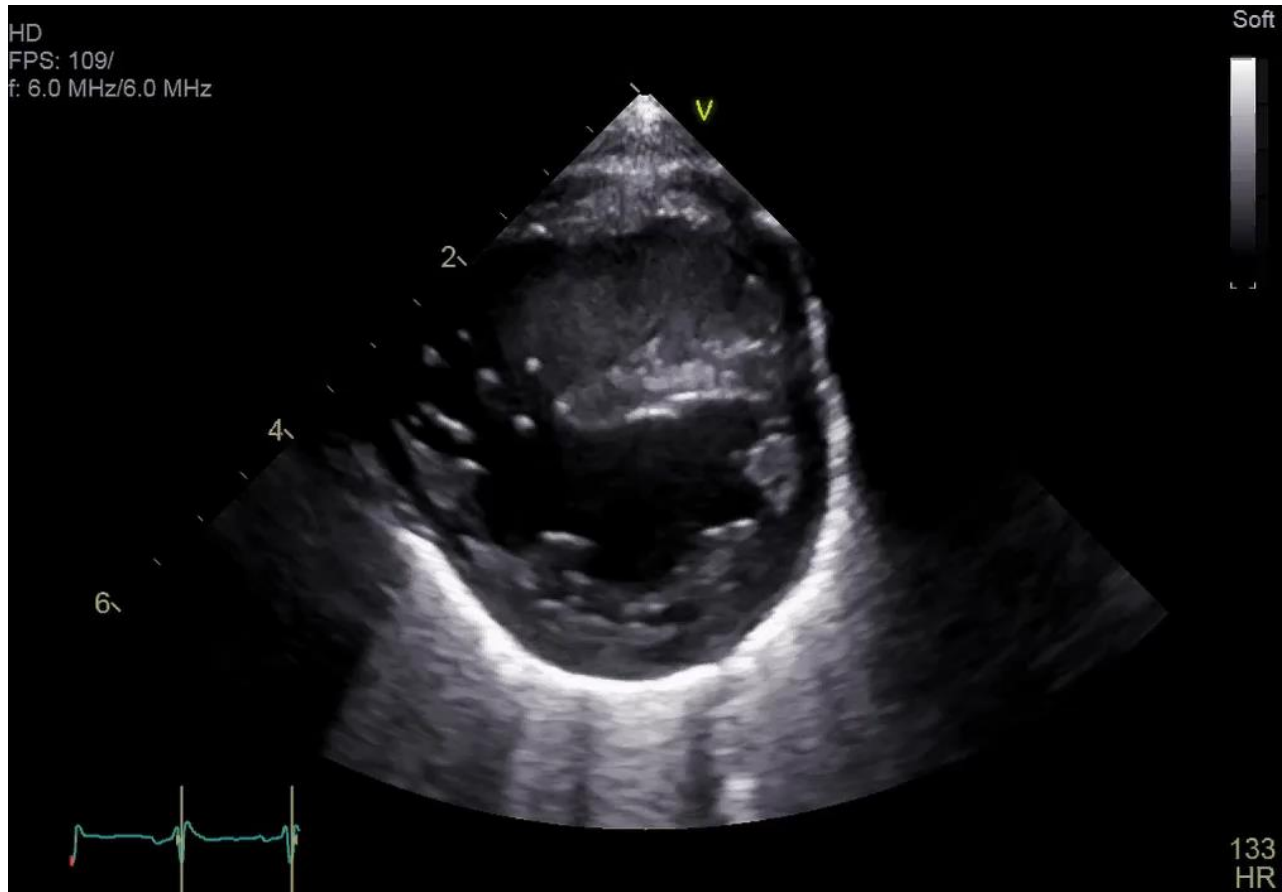




Inlet  
 Perimembranous



Lopez L, et.al 2018.



# Trabecular Muscular

Mid-septal

Apical

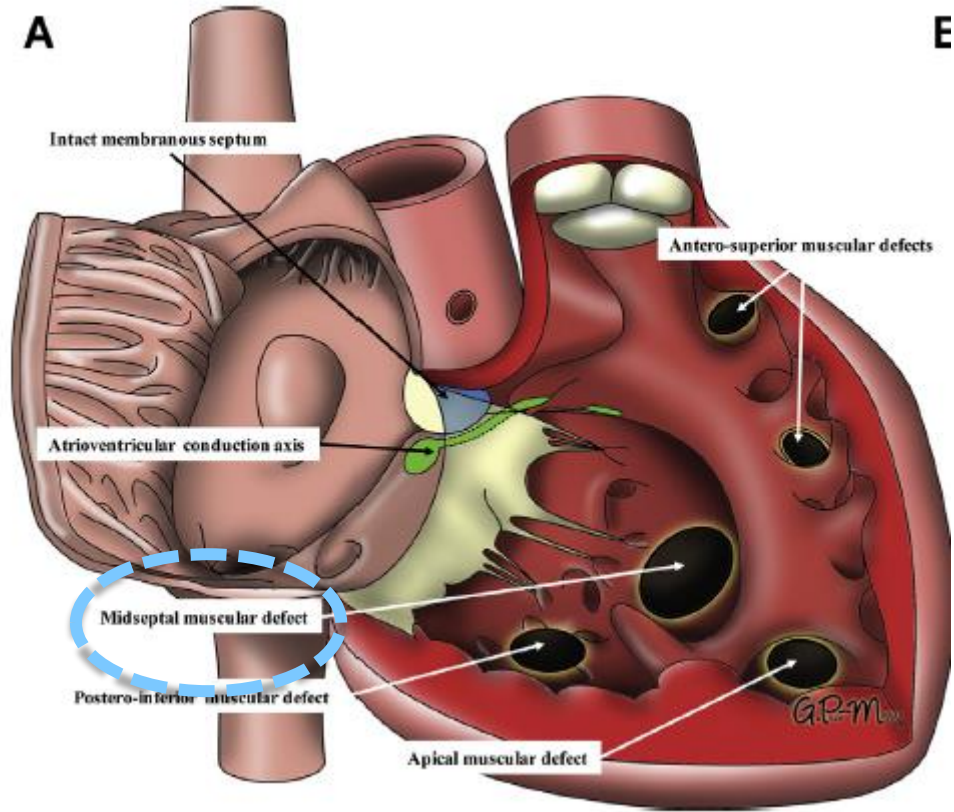
Postero-inferior

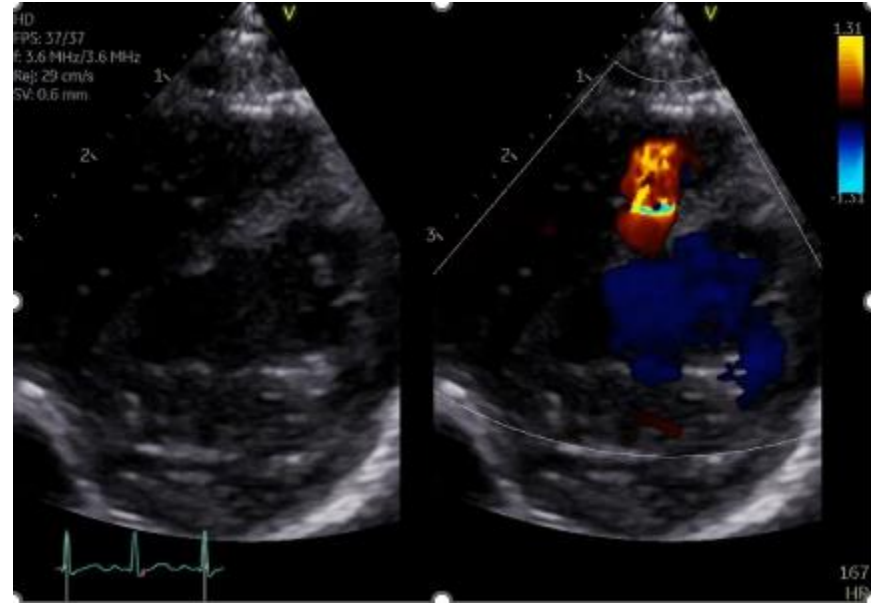
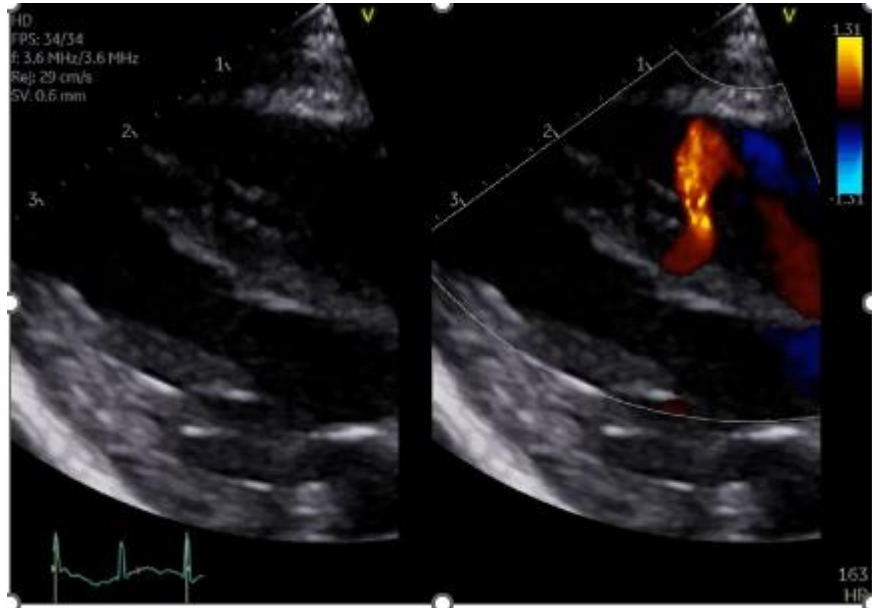
Anterior

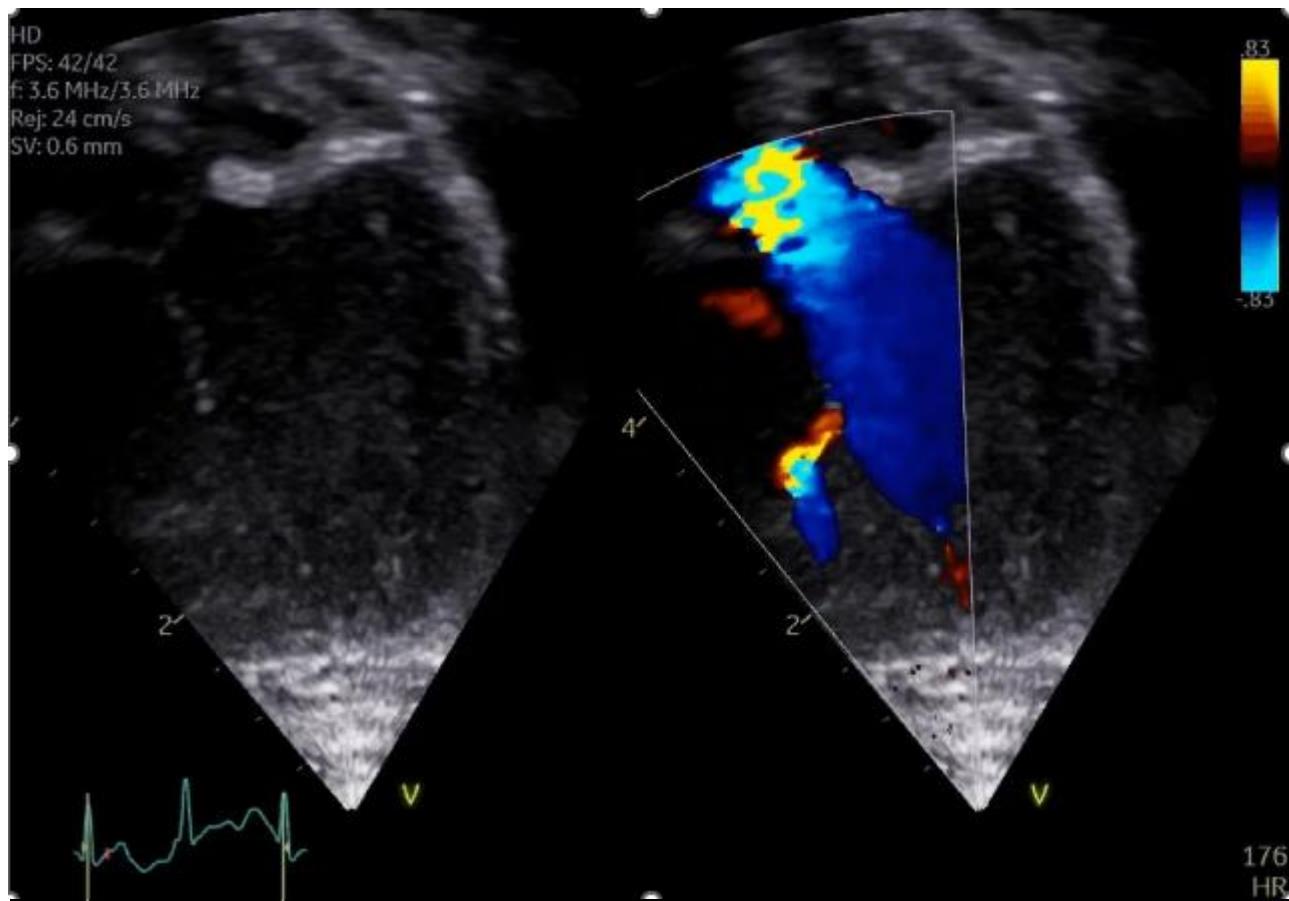
Multiple

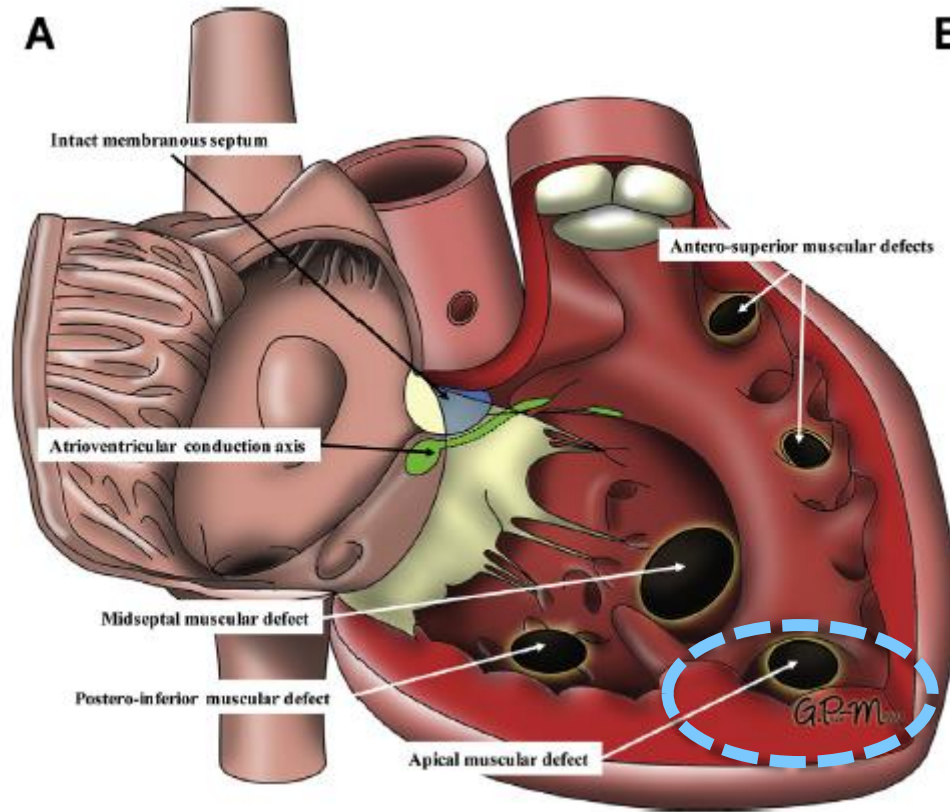
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3. Trabecular muscular VSD (07.11.01) <ul style="list-style-type: none"> <li>a. Trabecular muscular VSD: midseptal (07.11.04)</li> <li>b. Trabecular muscular VSD: apical (07.11.03)</li> <li>c. Trabecular muscular VSD: postero-inferior (07.11.12)</li> <li>d. Trabecular muscular VSD: anterosuperior (07.11.07)</li> <li>e. Trabecular muscular VSD: multiple ("Swiss cheese" septum) (07.11.05)</li> </ul>
4. Outlet VSD (07.12.00) <ul style="list-style-type: none"> <li>a. Outlet VSD without malalignment (07.12.09)               <ul style="list-style-type: none"> <li>i. Outlet muscular VSD without malalignment (07.11.06)</li> <li>ii. Doubly committed juxta-arterial VSD without malalignment (07.12.01)                   <ul style="list-style-type: none"> <li>1. Doubly committed juxta-arterial VSD without malalignment and with a muscular postero-inferior rim (07.12.02)</li> <li>2. Doubly committed juxta-arterial VSD without malalignment and with a fibrous postero-inferior rim (perimembranous extension) (07.12.03)</li> </ul> </li> </ul> </li> <li>b. Outlet VSD with anteriorly malaligned outlet septum (07.10.17)               <ul style="list-style-type: none"> <li>i. Outlet muscular VSD with anteriorly malaligned outlet septum (07.11.15)</li> <li>ii. Outlet perimembranous VSD with anteriorly malaligned outlet septum (07.10.04)</li> </ul> </li> <li>iii. Doubly committed juxta-arterial VSD with anteriorly malaligned fibrous outlet septum (07.12.12)               <ul style="list-style-type: none"> <li>1. Doubly committed juxta-arterial VSD with anteriorly malaligned fibrous outlet septum and a muscular postero-inferior rim (07.12.07)</li> <li>2. Doubly committed juxta-arterial VSD with anteriorly malaligned fibrous outlet septum and a fibrous postero-inferior rim (perimembranous extension) (07.12.05)</li> </ul> </li> <li>c. Outlet VSD with posteriorly malaligned outlet septum (07.10.18)               <ul style="list-style-type: none"> <li>i. Outlet muscular VSD with posteriorly malaligned outlet septum (07.11.16)</li> <li>ii. Outlet perimembranous VSD with posteriorly malaligned outlet septum (07.10.19)</li> </ul> </li> <li>iii. Doubly committed juxta-arterial VSD with posteriorly malaligned fibrous outlet septum (07.12.13)               <ul style="list-style-type: none"> <li>1. Doubly committed juxta-arterial VSD with posteriorly malaligned fibrous outlet septum and a muscular postero-inferior rim (07.12.08)</li> <li>2. Doubly committed juxta-arterial VSD with posteriorly malaligned fibrous outlet septum and a fibrous postero-inferior rim (perimembranous extension) (07.12.06)</li> </ul> </li> </ul>



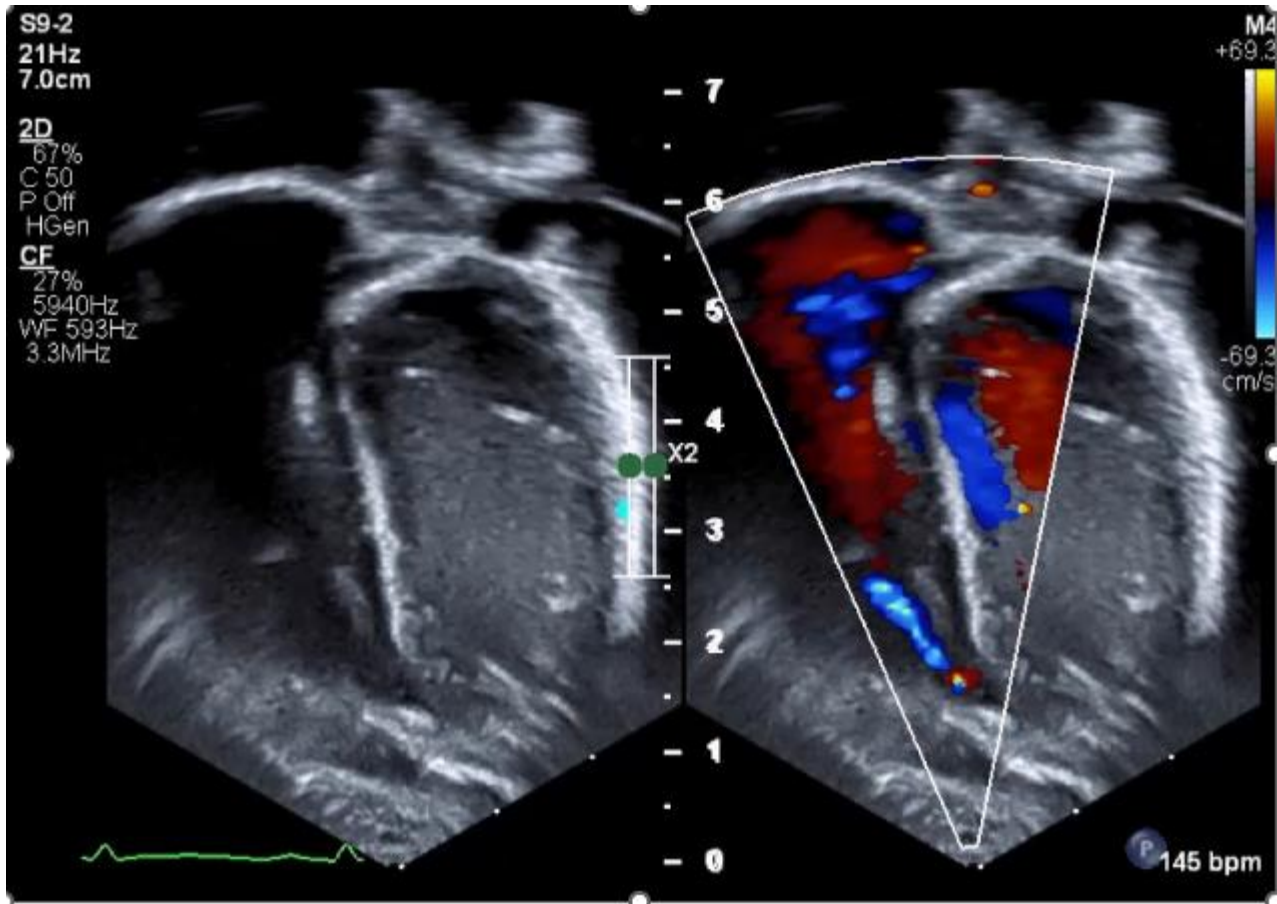






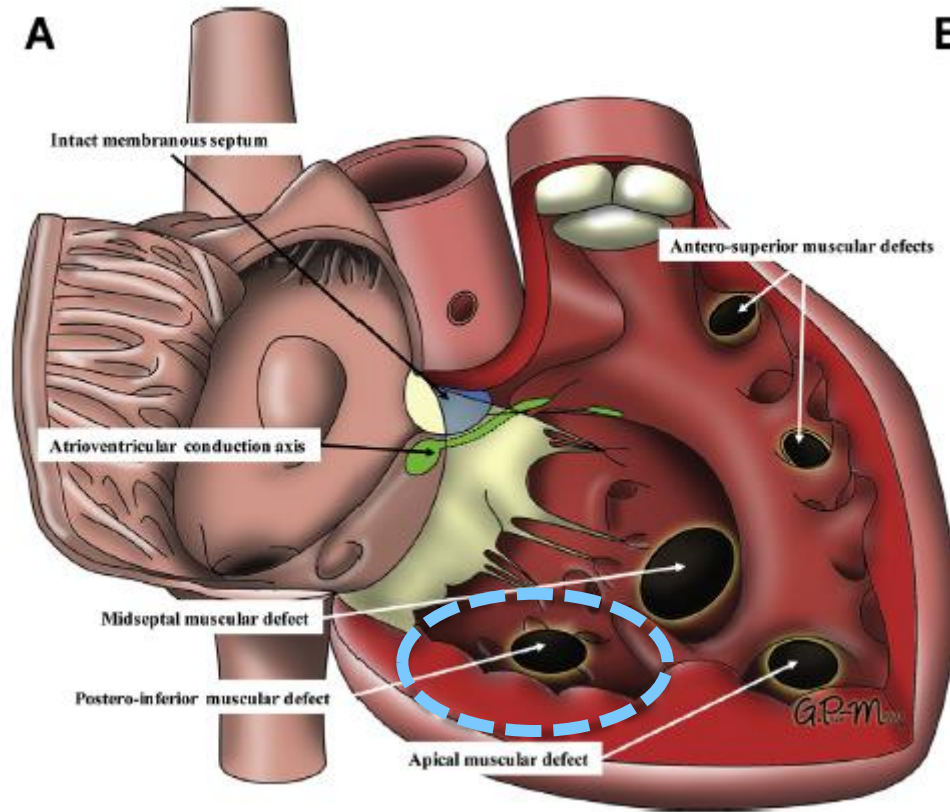
Lopez L, et.al 2018.



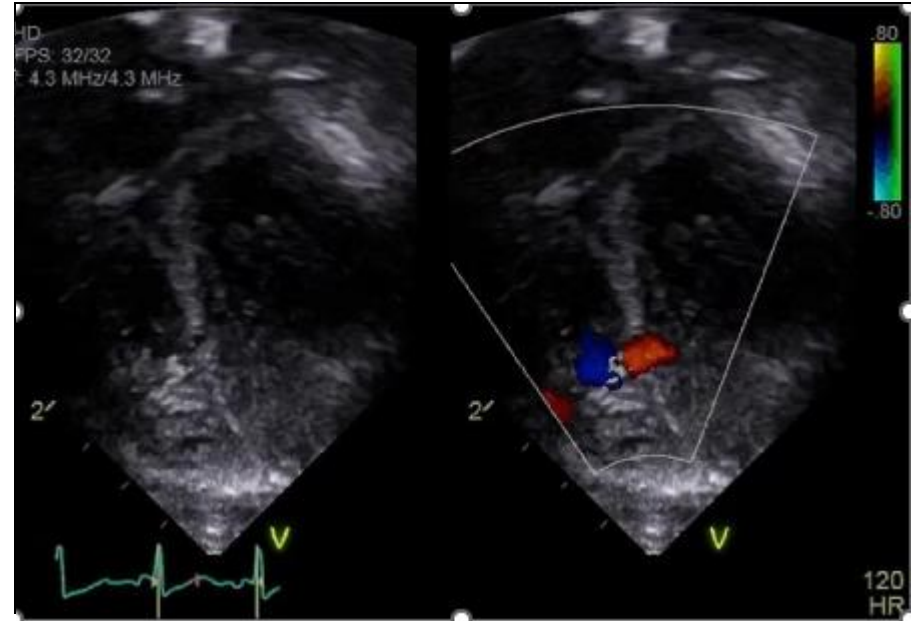
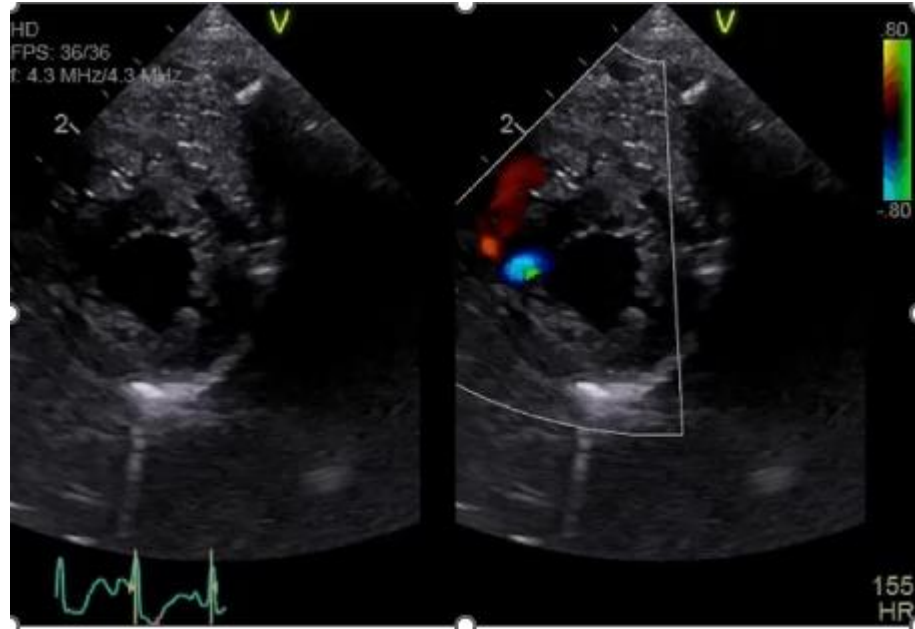


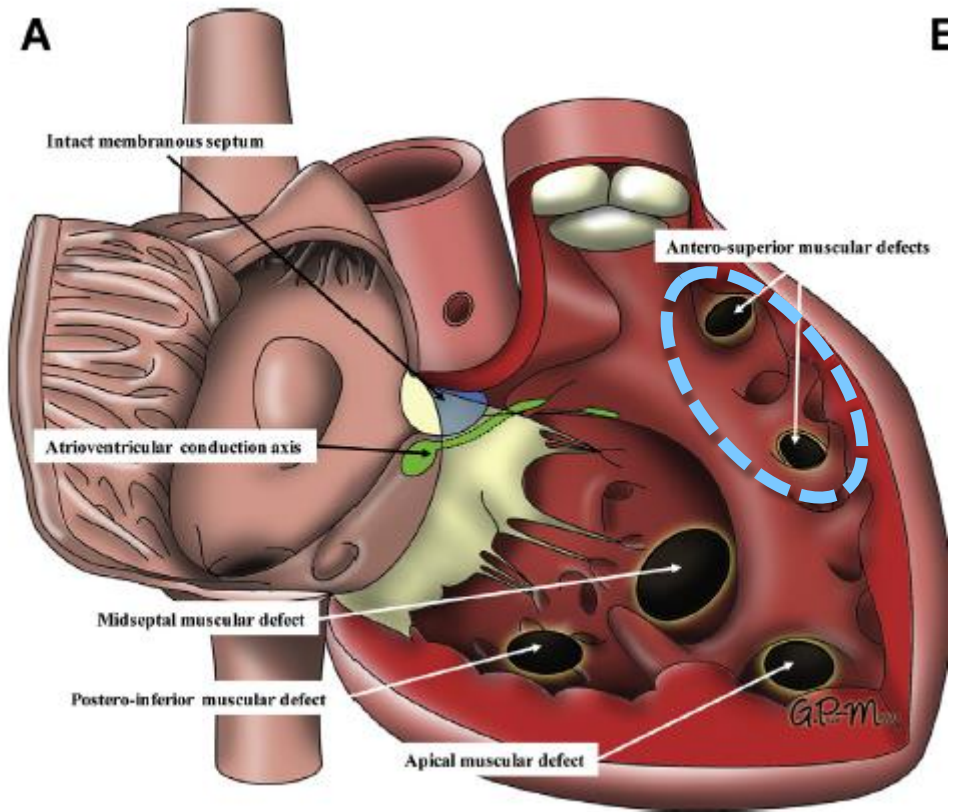
Trabecular  
Muscular

Apical

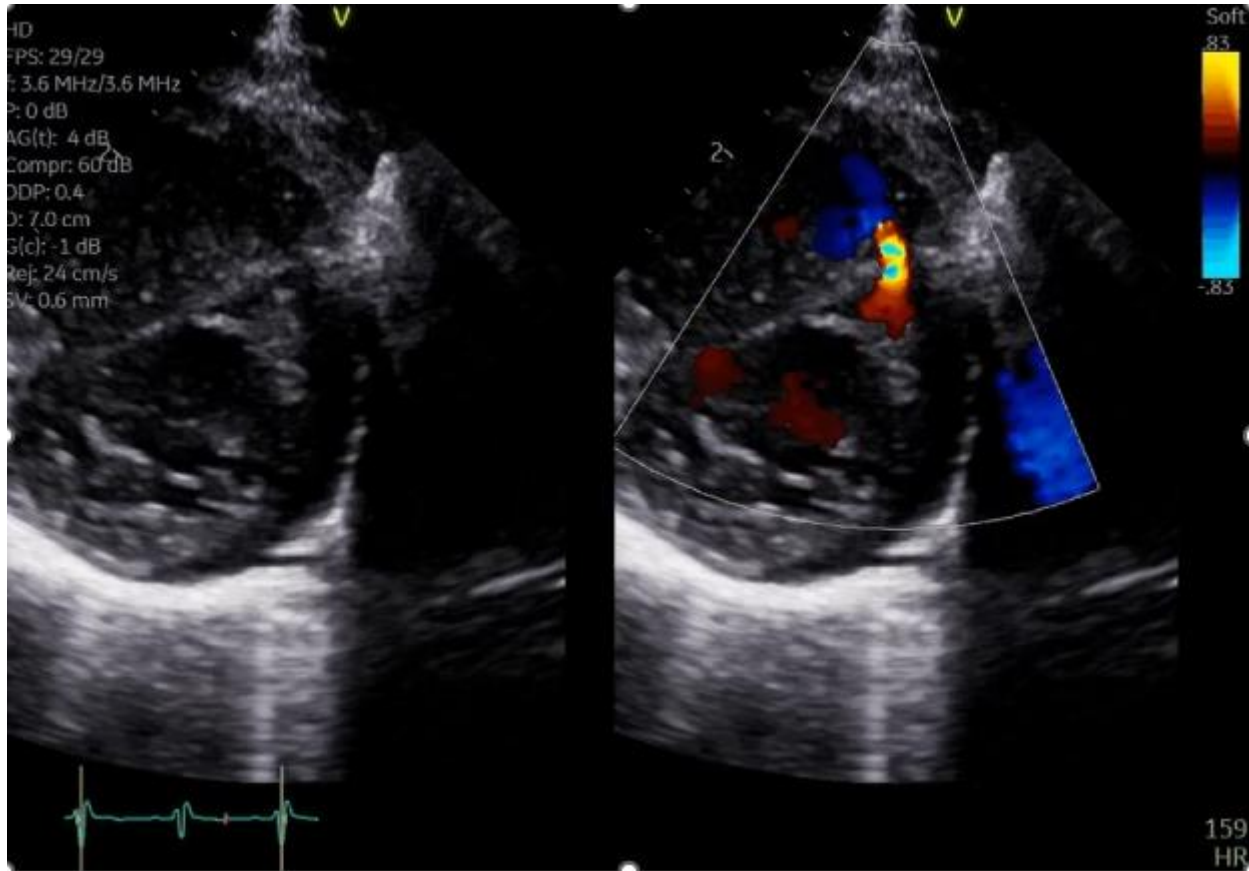


Lopez L, et.al 2018.



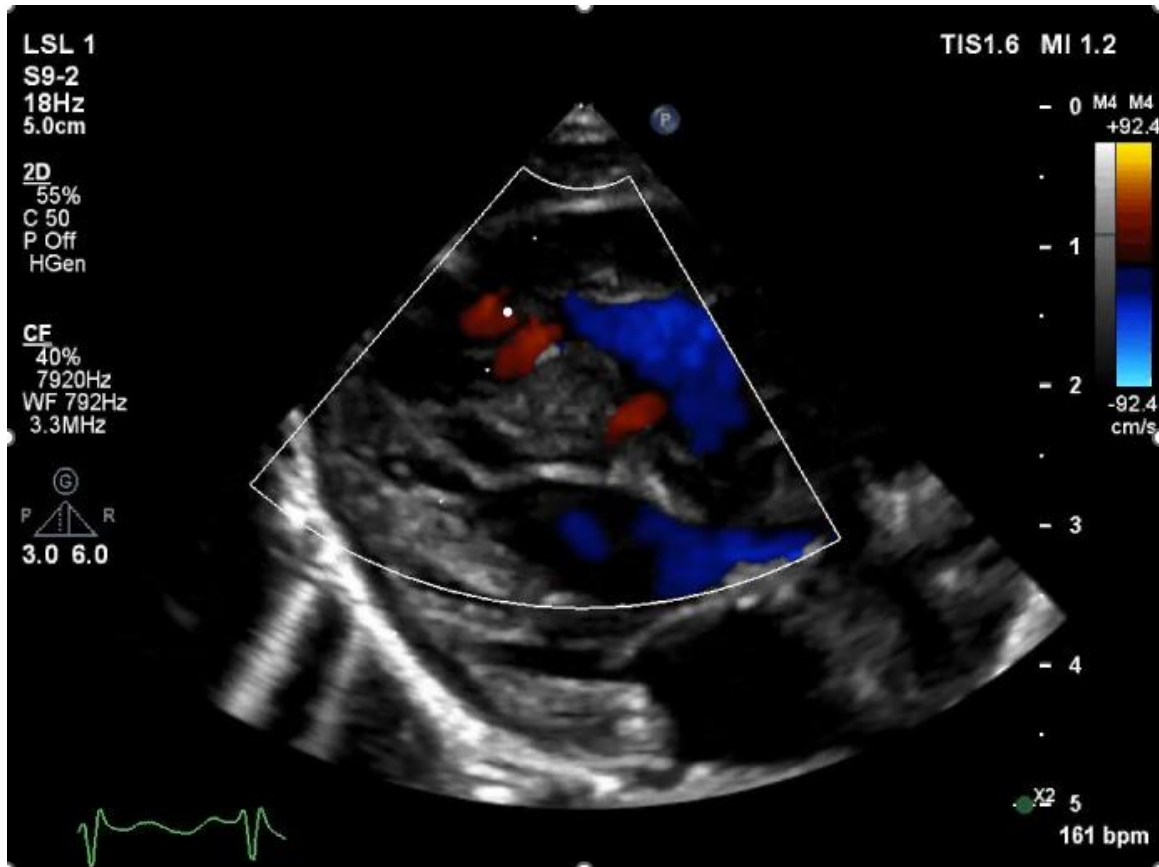


Lopez L, et.al 2018.



Trabecular  
Muscular

Anterior

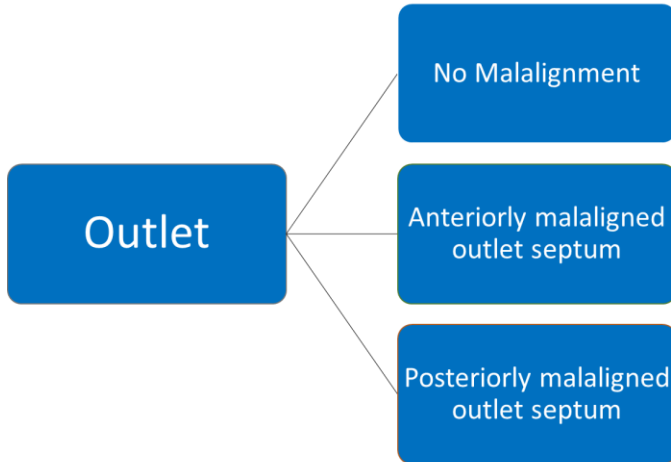


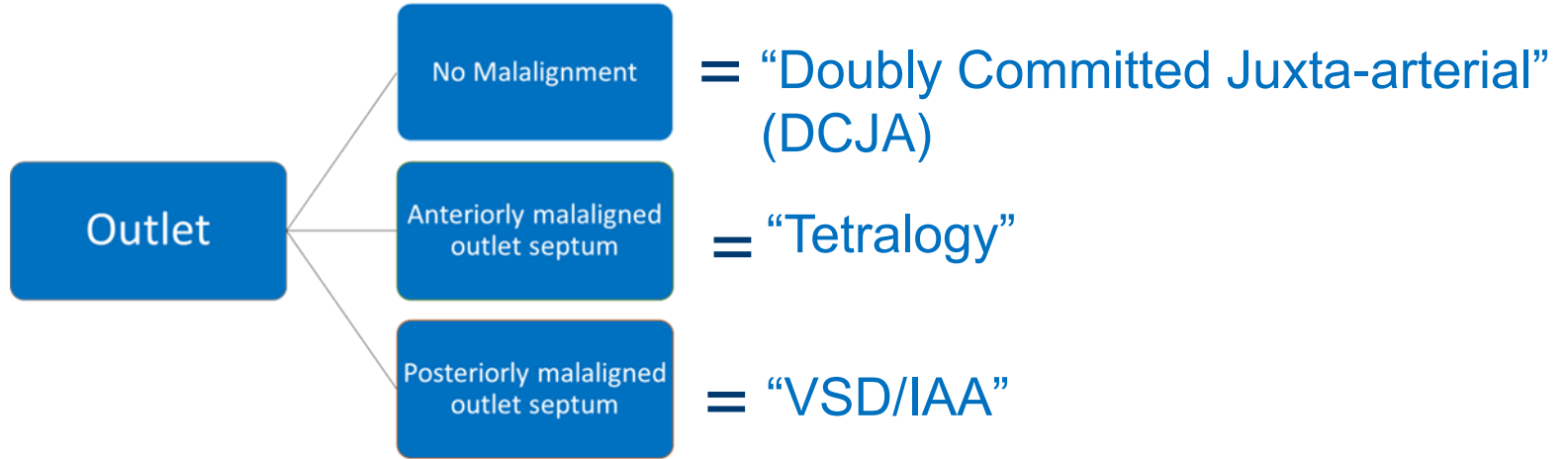
Trabecular  
Muscular

Multiple

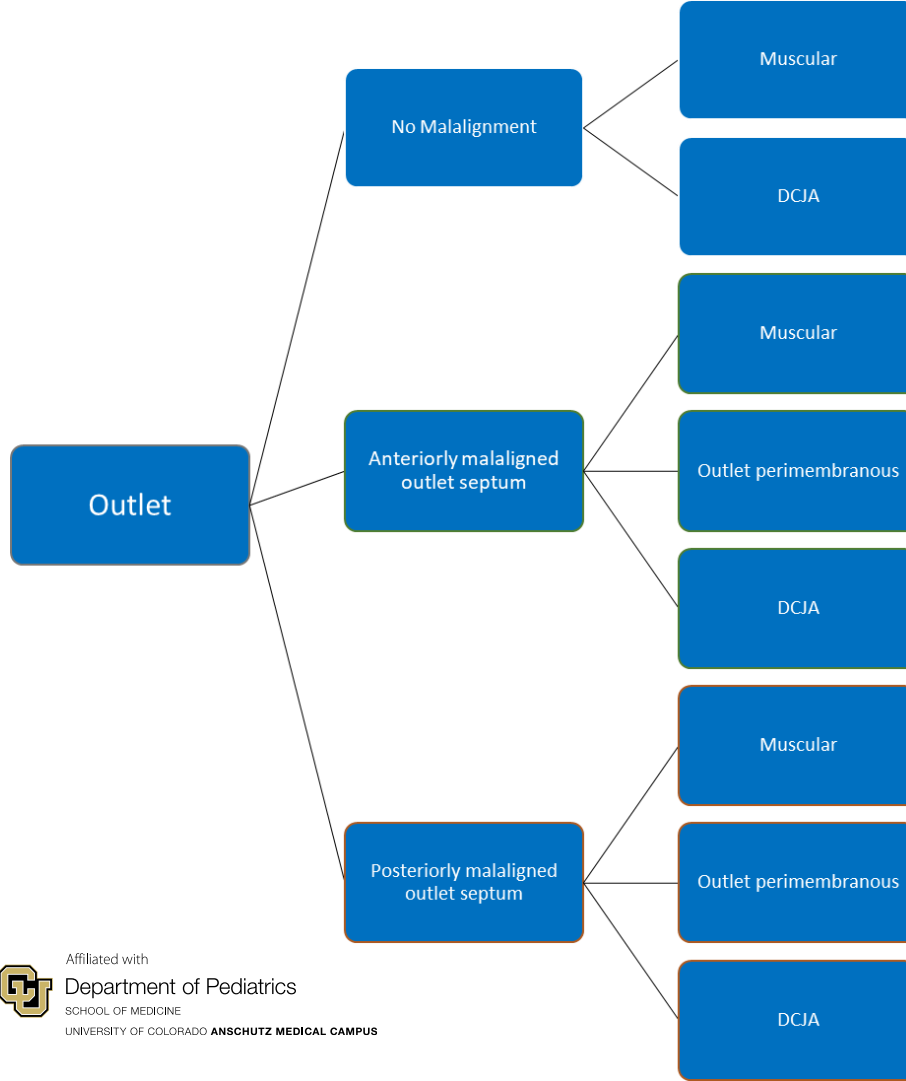
**Table 1. International Society for Nomenclature of Paediatric and Congenital Heart Disease Classification Scheme for Ventricular Septal Defect (International Paediatric and Congenital Cardiac Code 07.10.00) as Incorporated in the International Classification of Diseases-11th Iteration**

1. Perimembranous central VSD (07.10.01)
2. Inlet VSD without a common AV junction (07.14.05) <sup>a</sup> <ul style="list-style-type: none"> <li>a. Inlet perimembranous VSD without AV septal malalignment and without a common AV junction (07.10.02)</li> <li>b. Inlet perimembranous VSD with AV septal malalignment and without a common AV junction (07.14.06)</li> <li>c. Inlet muscular VSD (07.11.02)</li> </ul>
3. Trabecular muscular VSD (07.11.01) <ul style="list-style-type: none"> <li>a. Trabecular muscular VSD: midseptal (07.11.04)</li> <li>b. Trabecular muscular VSD: apical (07.11.03)</li> <li>c. Trabecular muscular VSD: postero-inferior (07.11.12)</li> <li>d. Trabecular muscular VSD: anterosuperior (07.11.07)</li> <li>e. Trabecular muscular VSD: multiple ("Swiss cheese" septum) (07.11.05)</li> </ul>
4. Outlet VSD (07.12.00) <ul style="list-style-type: none"> <li>a. Outlet VSD without malalignment (07.12.09) <ul style="list-style-type: none"> <li>i. Outlet muscular VSD without malalignment (07.11.06)</li> <li>ii. Doubly committed juxta-arterial VSD without malalignment (07.12.01) <ul style="list-style-type: none"> <li>1. Doubly committed juxta-arterial VSD without malalignment and with a muscular postero-inferior rim (07.12.02)</li> <li>2. Doubly committed juxta-arterial VSD without malalignment and with a fibrous postero-inferior rim (perimembranous extension) (07.12.03)</li> </ul> </li> </ul> </li> <li>b. Outlet VSD with anteriorly malaligned outlet septum (07.10.17) <ul style="list-style-type: none"> <li>i. Outlet muscular VSD with anteriorly malaligned outlet septum (07.11.15)</li> <li>ii. Outlet perimembranous VSD with anteriorly malaligned outlet septum (07.10.04) <ul style="list-style-type: none"> <li>iii. Doubly committed juxta-arterial VSD with anteriorly malaligned fibrous outlet septum (07.12.12) <ul style="list-style-type: none"> <li>1. Doubly committed juxta-arterial VSD with anteriorly malaligned fibrous outlet septum and a muscular postero-inferior rim (07.12.07)</li> <li>2. Doubly committed juxta-arterial VSD with anteriorly malaligned fibrous outlet septum and a fibrous postero-inferior rim (perimembranous extension) (07.12.05)</li> </ul> </li> </ul> </li> </ul> </li> <li>c. Outlet VSD with posteriorly malaligned outlet septum (07.10.18) <ul style="list-style-type: none"> <li>i. Outlet muscular VSD with posteriorly malaligned outlet septum (07.11.16)</li> <li>ii. Outlet perimembranous VSD with posteriorly malaligned outlet septum (07.10.19) <ul style="list-style-type: none"> <li>iii. Doubly committed juxta-arterial VSD with posteriorly malaligned fibrous outlet septum (07.12.13) <ul style="list-style-type: none"> <li>1. Doubly committed juxta-arterial VSD with posteriorly malaligned fibrous outlet septum and a muscular postero-inferior rim (07.12.08)</li> <li>2. Doubly committed juxta-arterial VSD with posteriorly malaligned fibrous outlet septum and a fibrous postero-inferior rim (perimembranous extension) (07.12.06)</li> </ul> </li> </ul> </li> </ul> </li> </ul>



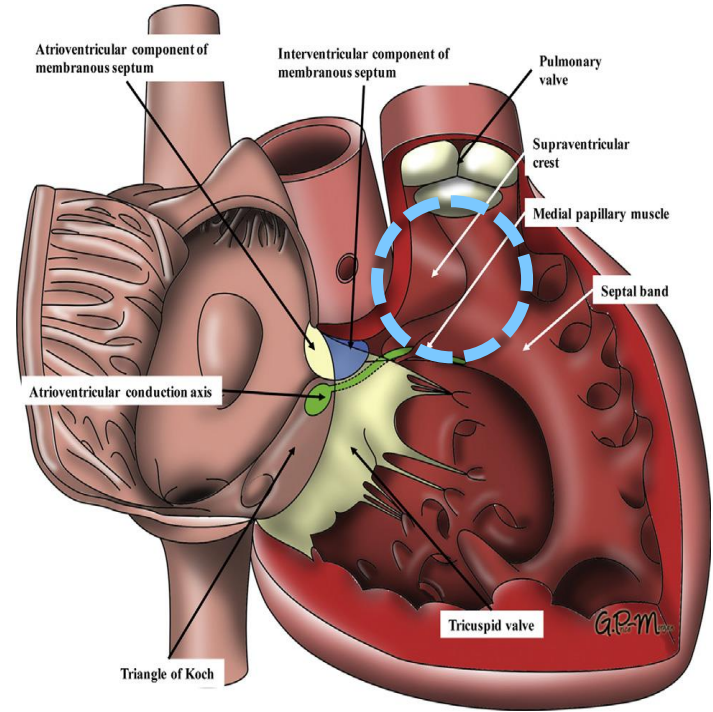






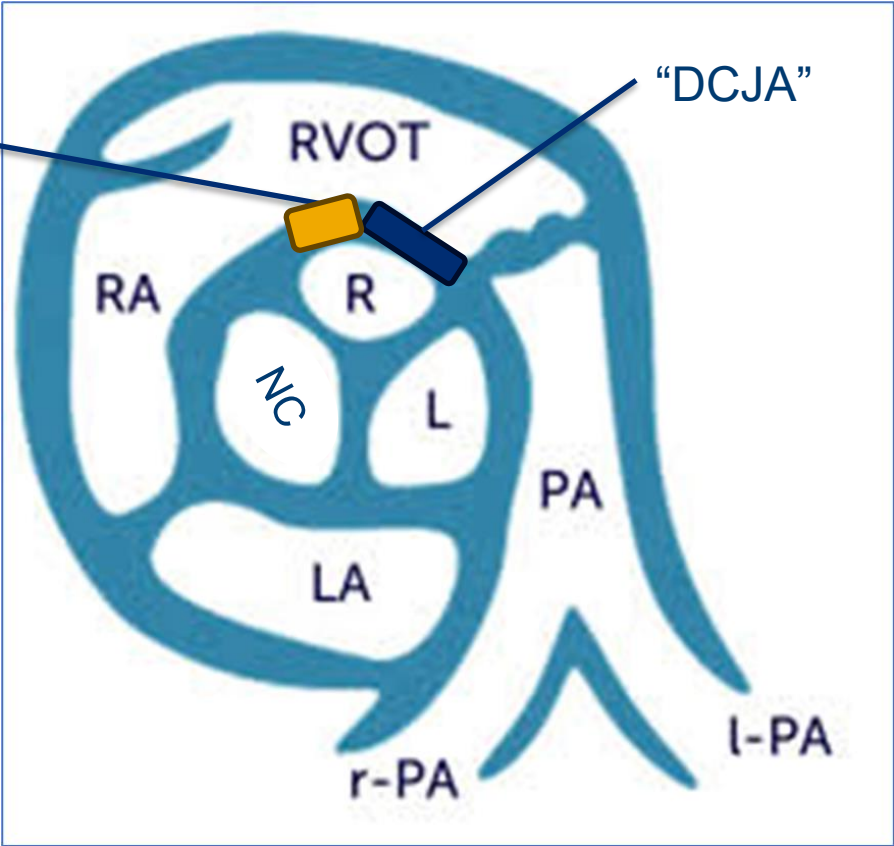
- Location

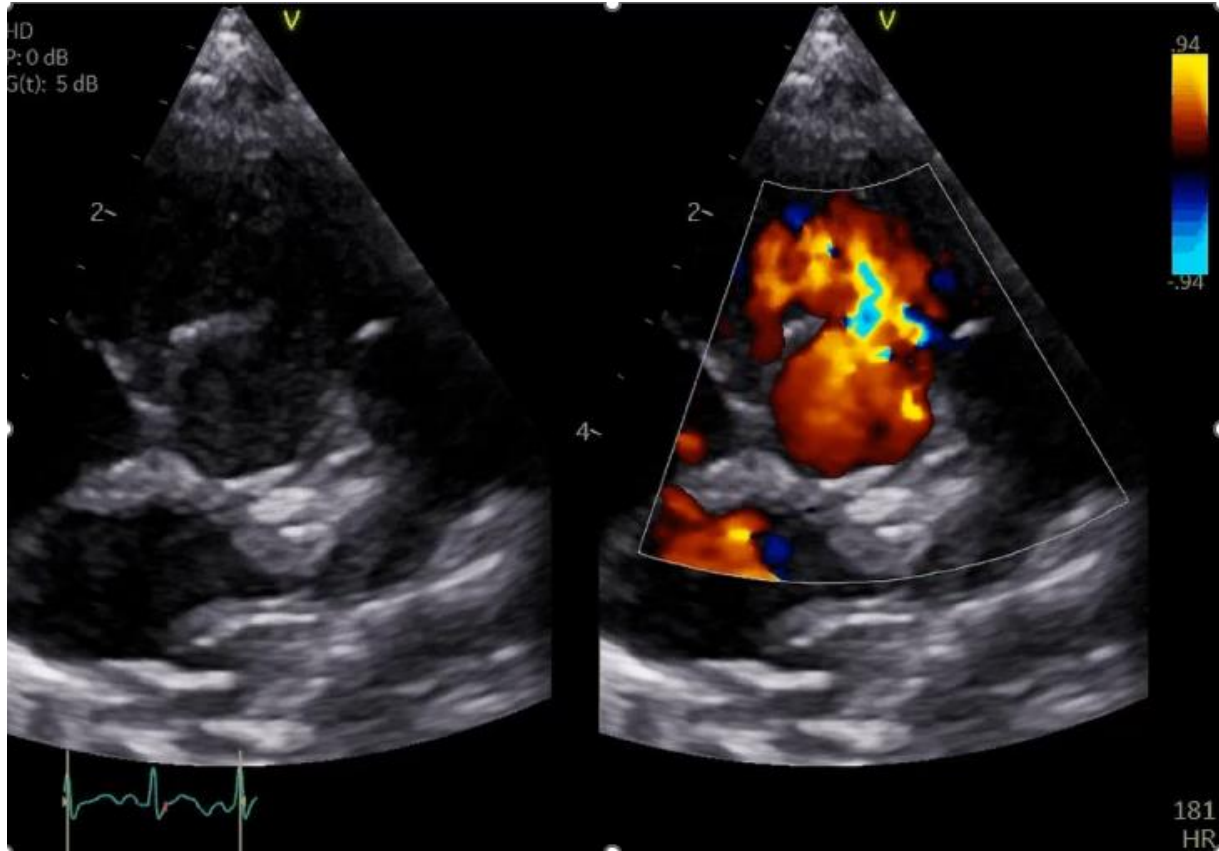
- Opens to outlet portion of RV
- Between limbs (Y) of septal band
- Behind/near anterior leaflet of TV
- Above antero-septal commissure of TV
- With/without malalignment of outlet septum



Lopez L, et.al 2018.

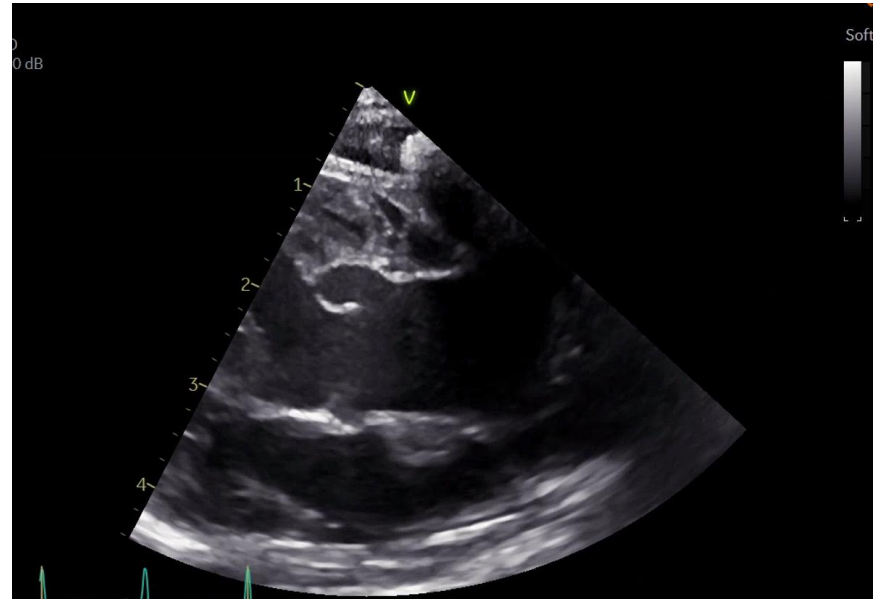
Central  
Perimembranous





Outlet

“Doubly Committed  
Juxta-arteria”



Outlet

Anteriorly malaligned  
outlet septum

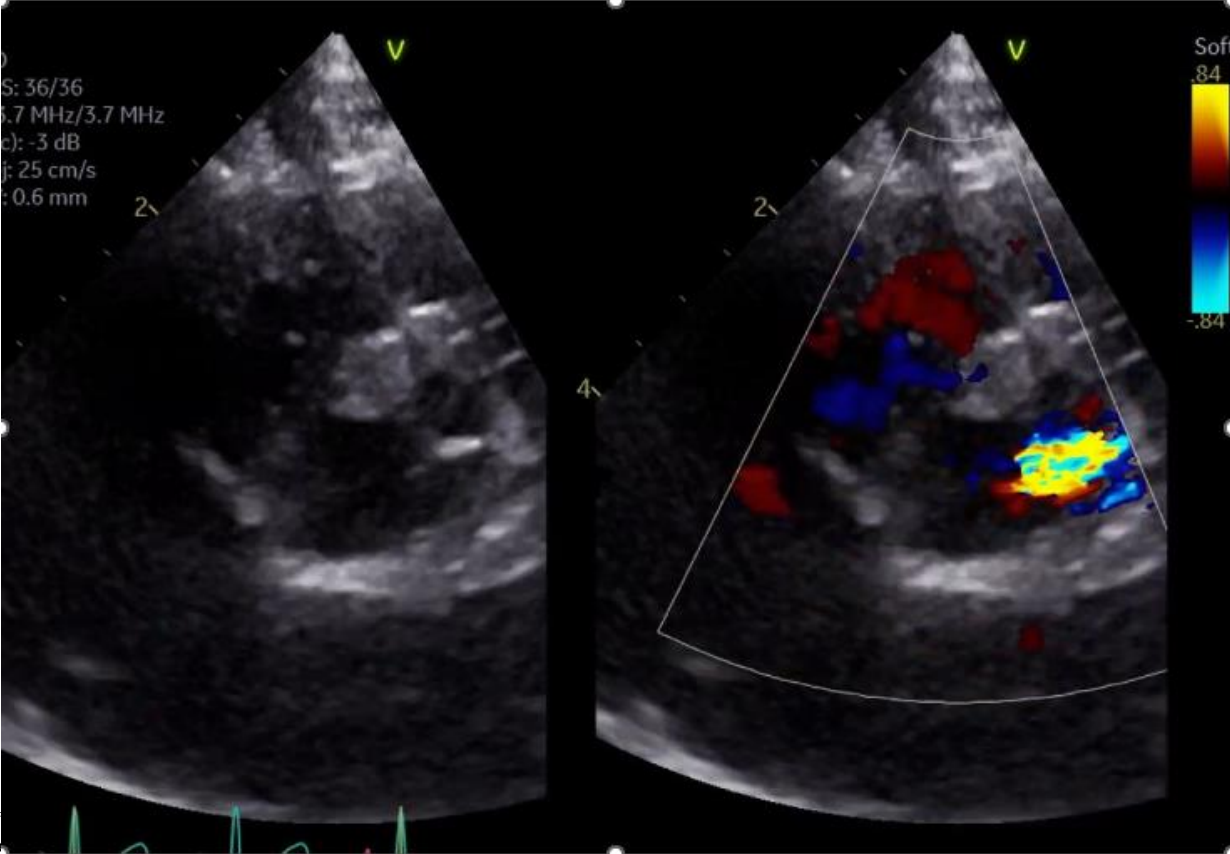
# Other Things To Look For

Left-sided  
dilation



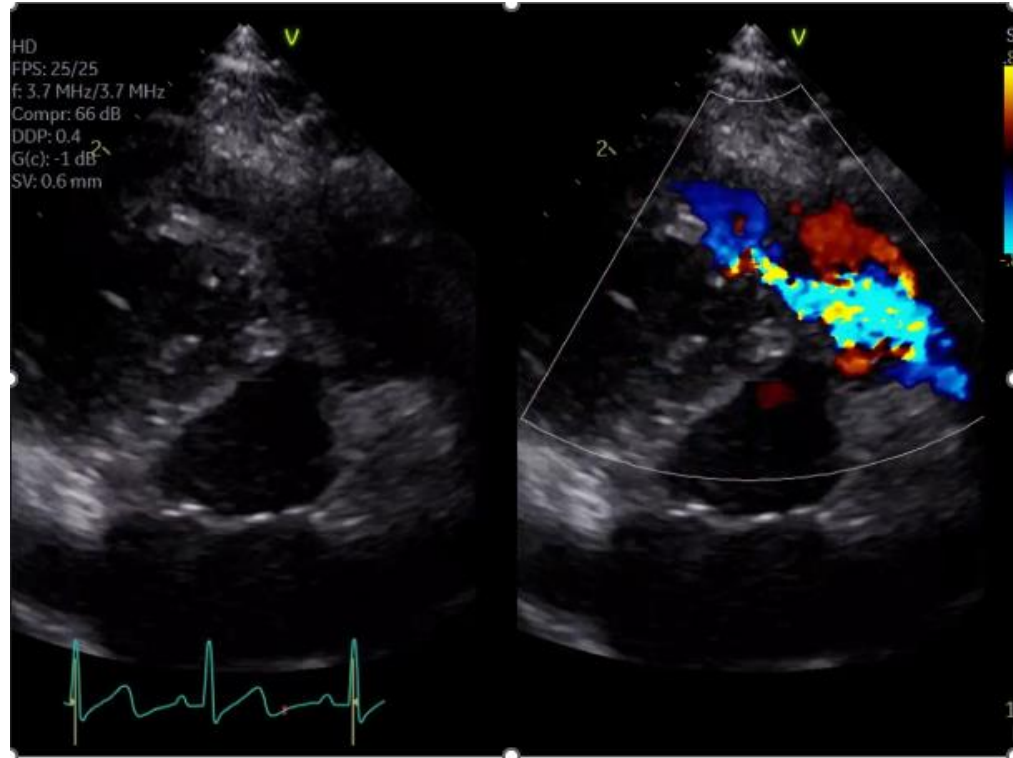
# Other Things To Look For

## Aortic Valve Insufficiency



# Other Things To Look For

## Double Chamber RV







# If All Else Fails – Describe What You See



# Thank You!



# References

- **Special Thanks to Dr. Richard Friesen** – many of the slides and echo images are borrowed from his prior presentation on VSD Nomenclature Mapping
- Lopez L, et.al. Classification of Ventricular Septal Defects for the Eleventh Iteration of the International Classification of Diseases-Striving for Consensus: A Report From the International Society for Nomenclature of Paediatric and Congenital Heart Disease. *Ann Thorac Surg.* 2018 Nov;106(5):1578-1589. doi: 10.1016/j.athoracsur.2018.06.020. Epub 2018 Jul 19. PMID: 30031844.
- Sharma PS, Naperkowski A, Bauch TD, Chan JYS, Arnold AD, Whinnett ZI, Ellenbogen KA, Vijayaraman P. Permanent His Bundle Pacing for Cardiac Resynchronization Therapy in Patients With Heart Failure and Right Bundle Branch Block. *Circ Arrhythm Electrophysiol.* 2018 Sep;11(9):e006613. doi: 10.1161/CIRCEP.118.006613. PMID: 30354292.
- Forbus GA, Shirali GS. Anomalies of the Ventricular Septum. *Echocardiography in Pediatric and Congenital Heart Disease*; 2009:175-87.