

# Discordant Atrioventricular Connections

## Morphology-Imaging-Surgical Correlation

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Department of Diagnostic Imaging

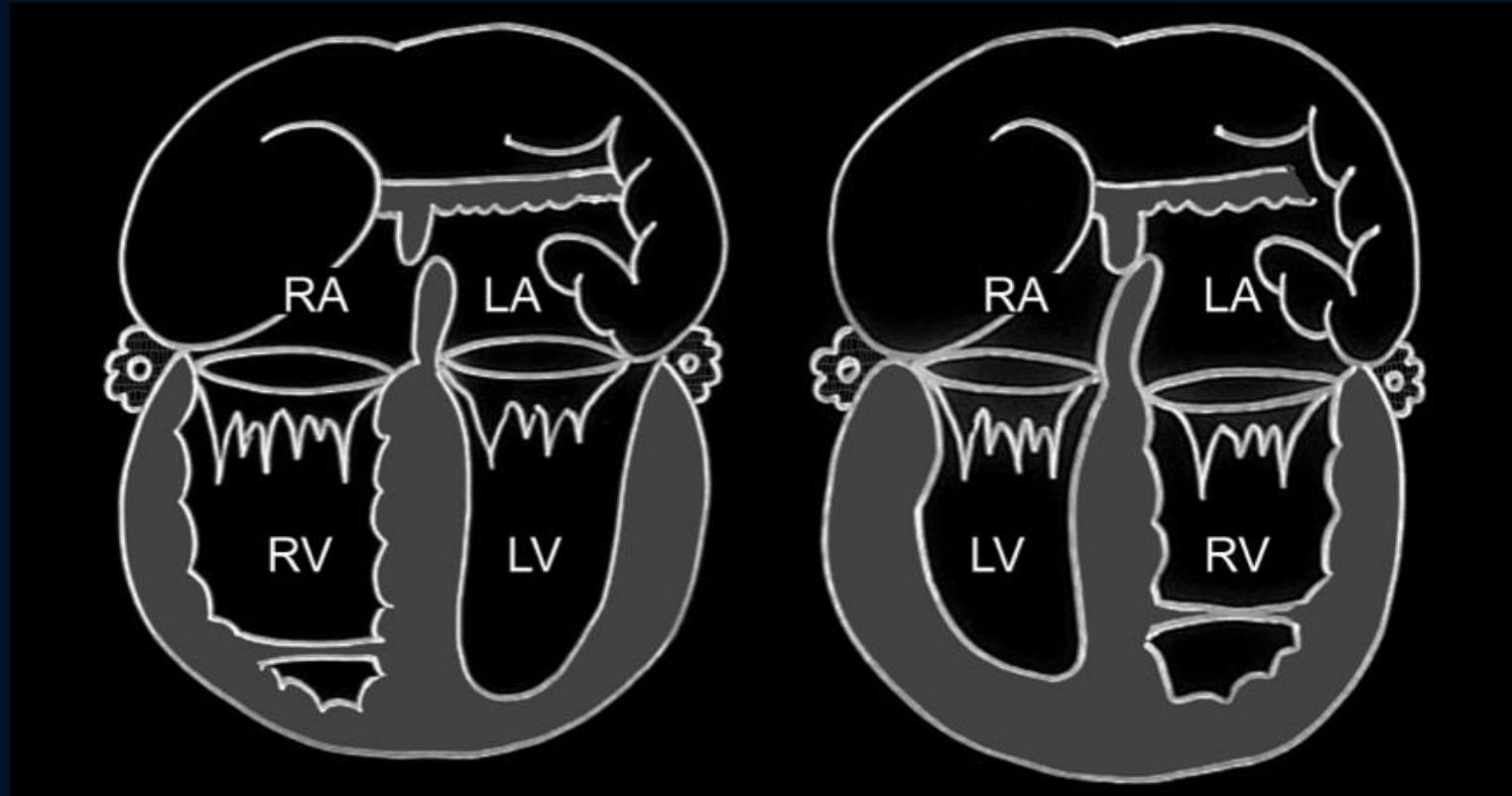
The Hospital for Sick Children and University of Toronto, Canada

No disclosures

# Outline

- Introduction to Discordant AV Connections
- Congenitally Corrected Transposition of the Great Arteries (ccTGA)
  - Typical Morphologic Features
  - Conduction Axis
  - Associated Anomalies
    - VSD
    - Tricuspid Valve Lesions
    - mLVOTO/Pulmonary Stenosis
    - Coronary Anomalies
  - Atrial Situs
  - Ventricular Relationship
  - Surgical Strategy
- Other forms of Discordant AV Connections

# Atrioventricular Connections

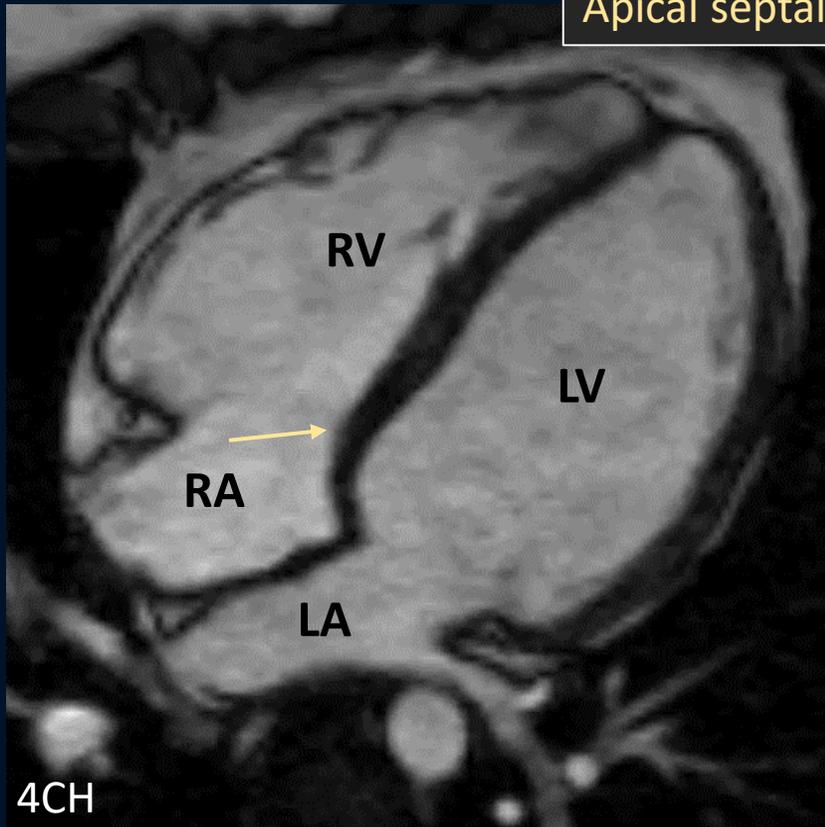


Concordant AV Connections

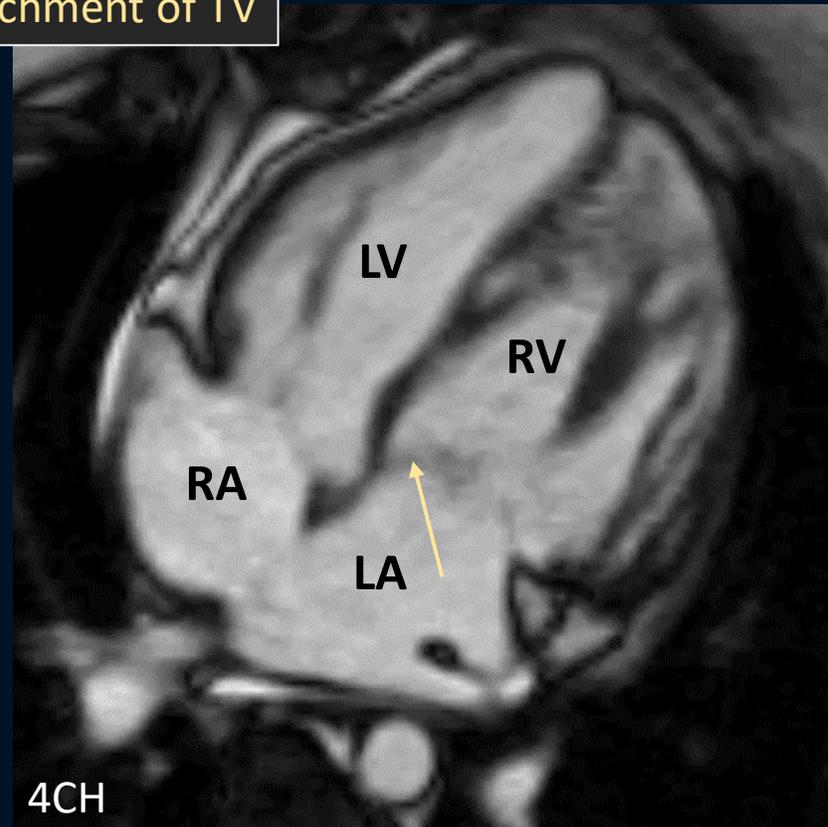
Discordant AV Connections

# Atrioventricular Connections

Apical septal attachment of TV



Concordant AV Connections



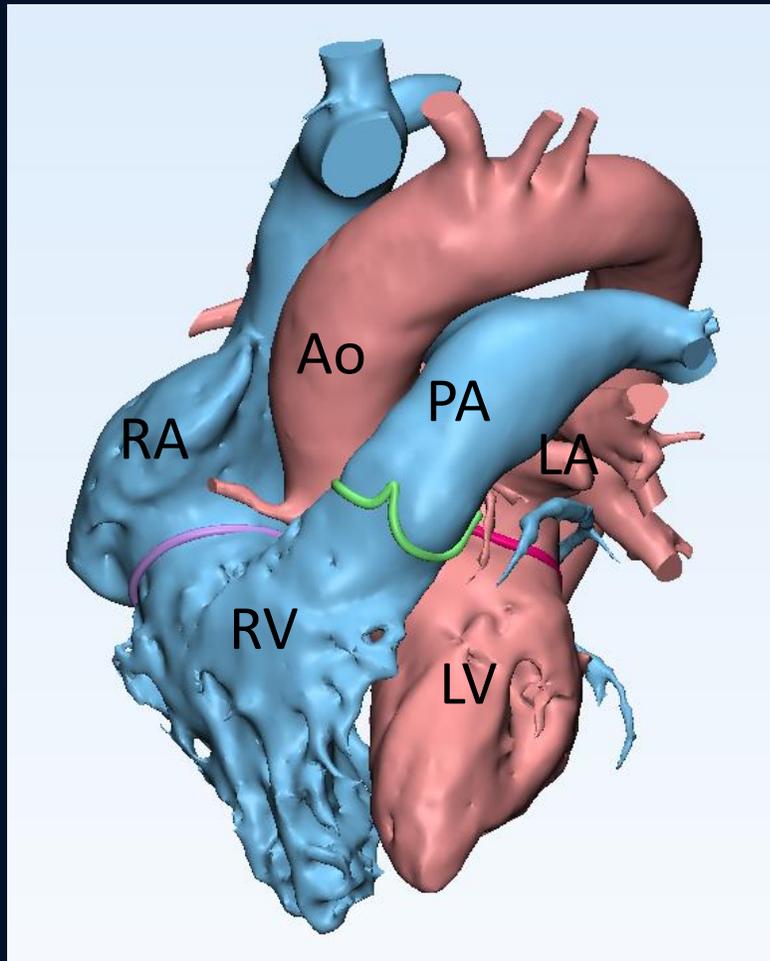
Discordant AV Connections

**Discordant AV connections = anatomic alignment, not a specific disease**

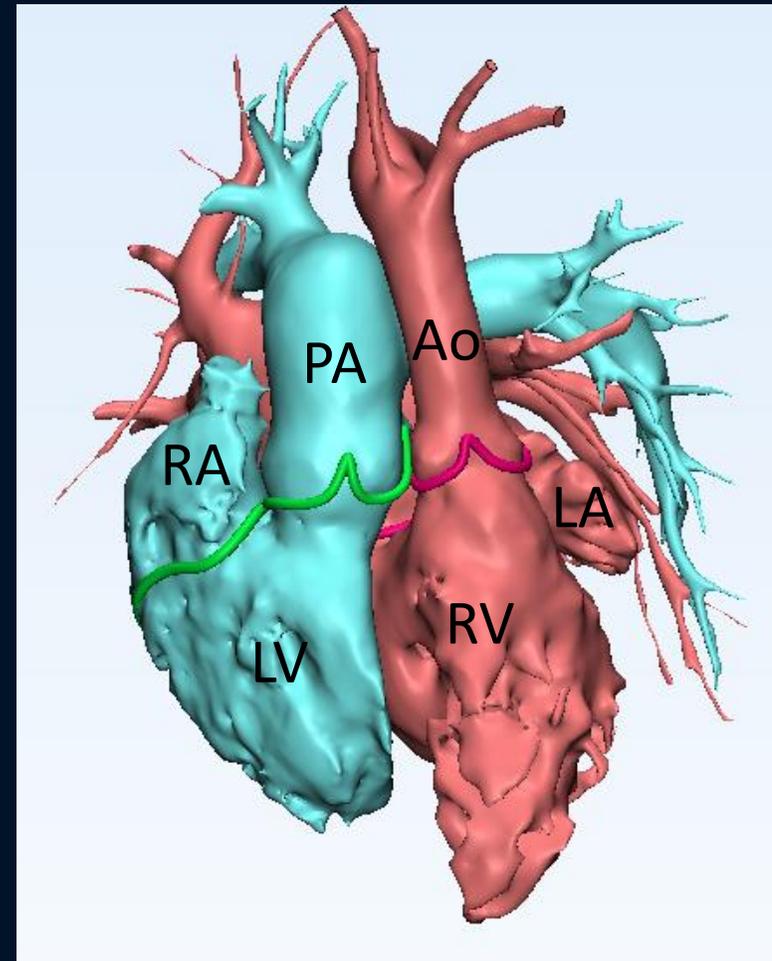
# ccTGA

❖ *Most discordant AV connections will be ccTGA*

# ccTGA = Discordant AV and VA Connections

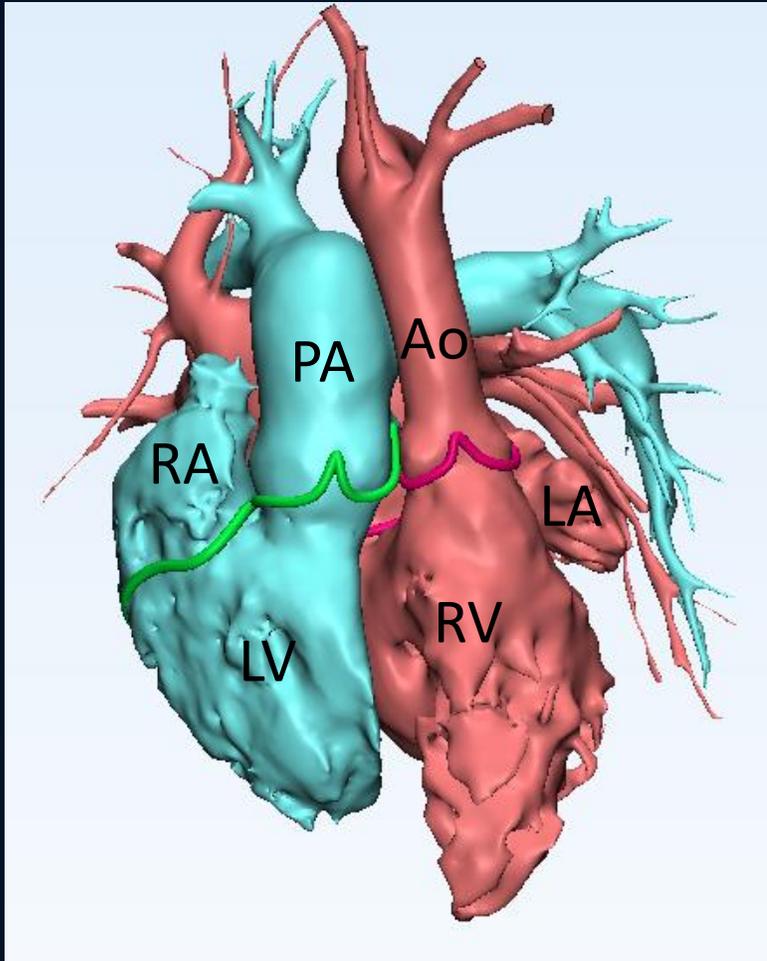


Normal



ccTGA

# ccTGA



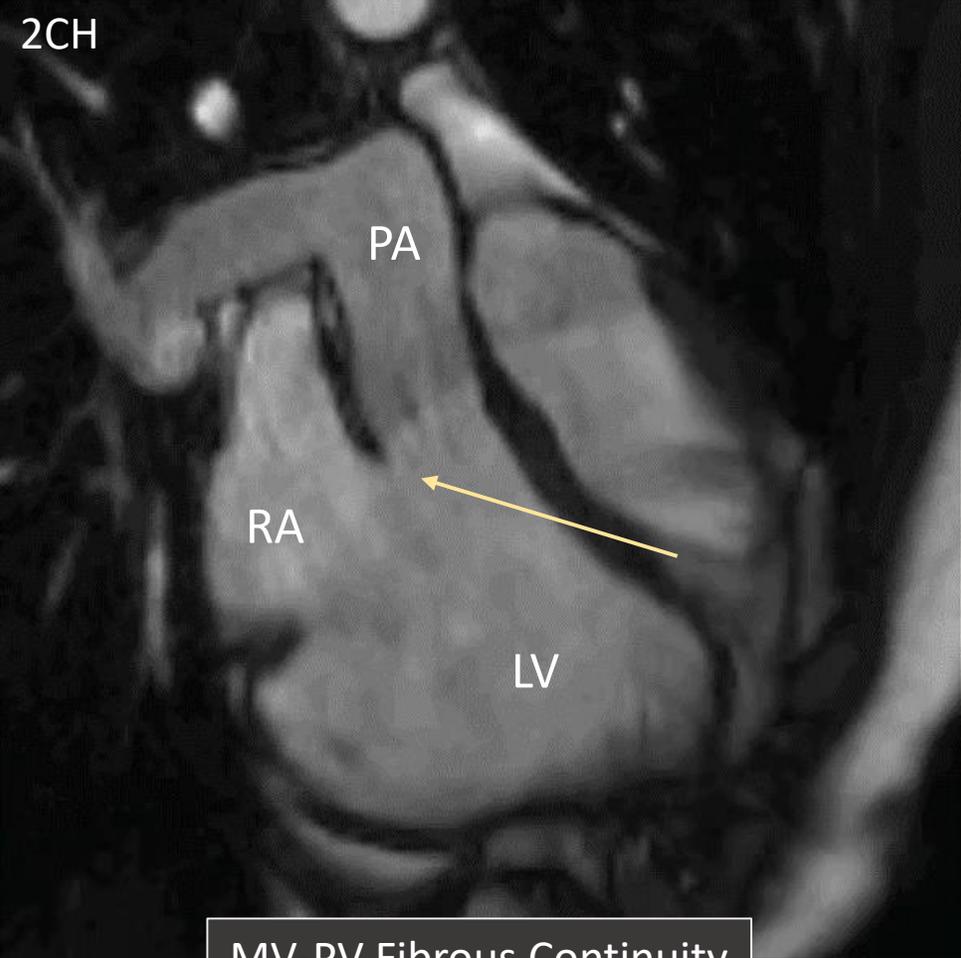
## Typical Features (Not Always)

- Mitral and pulmonary valve fibrous continuity
- Subaortic infundibulum
  - No subpulmonary infundibulum
- Parallel outflows/arterial trunks, aorta anterior & left of PA (L-TGA)
- Larger membranous septum than normal
  - Especially when ventricular septum is intact
- Abnormal AV node position

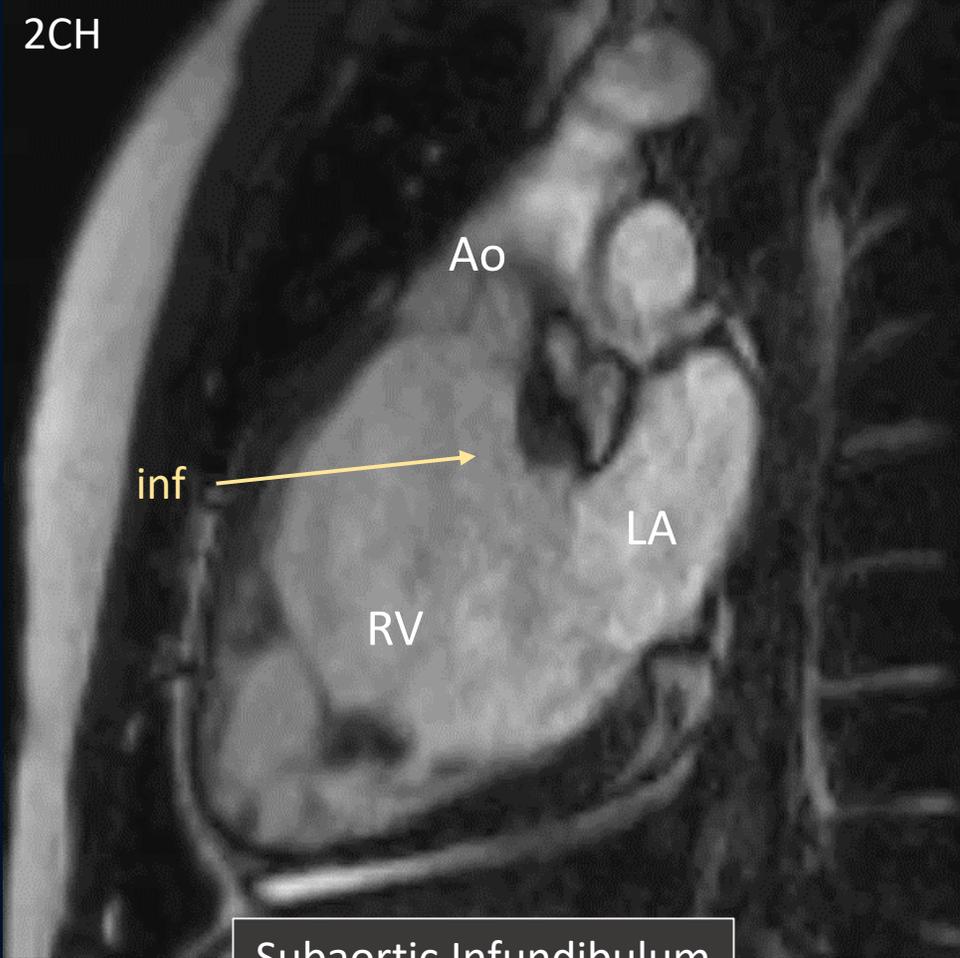
# ccTGA in Short Axis



# MV-PV Continuity / Subaortic Infundibulum

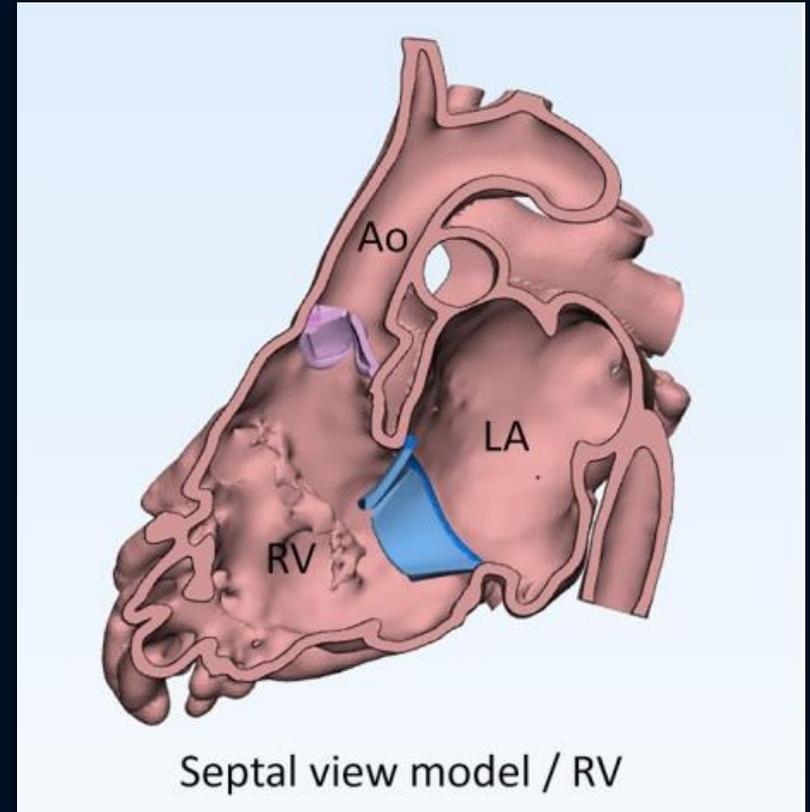
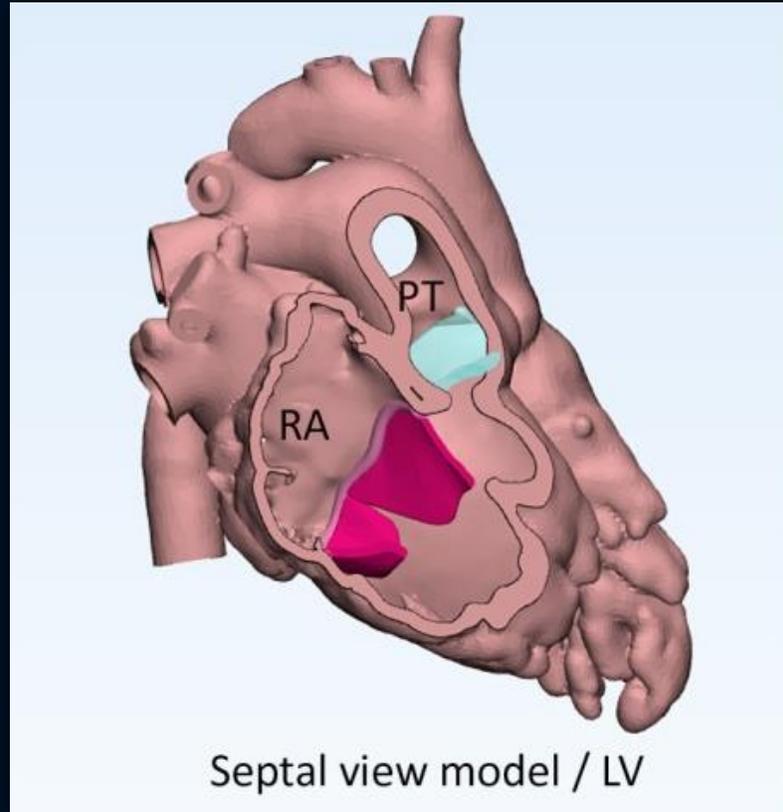
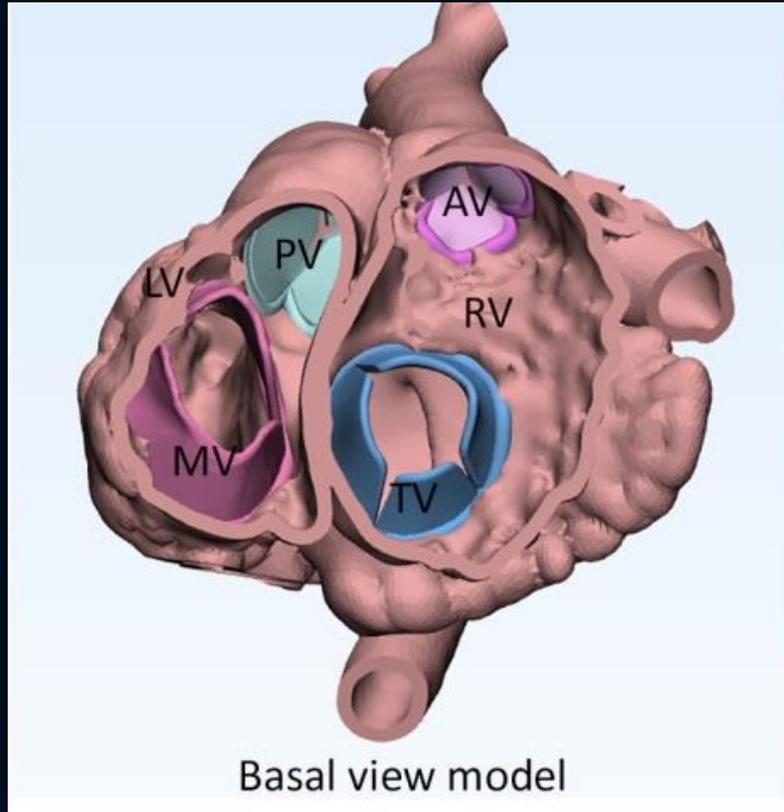


MV-PV Fibrous Continuity

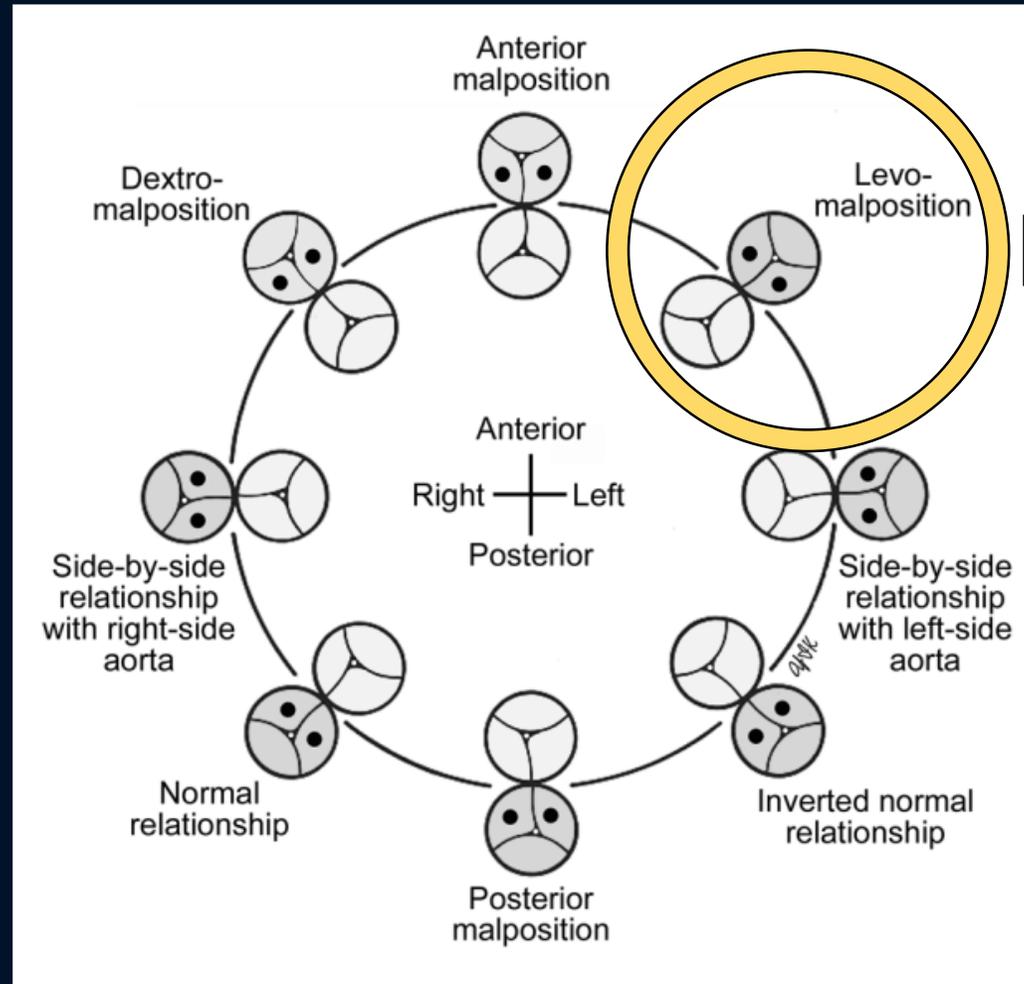


Subaortic Infundibulum

# MV-PV Continuity / Subaortic Infundibulum

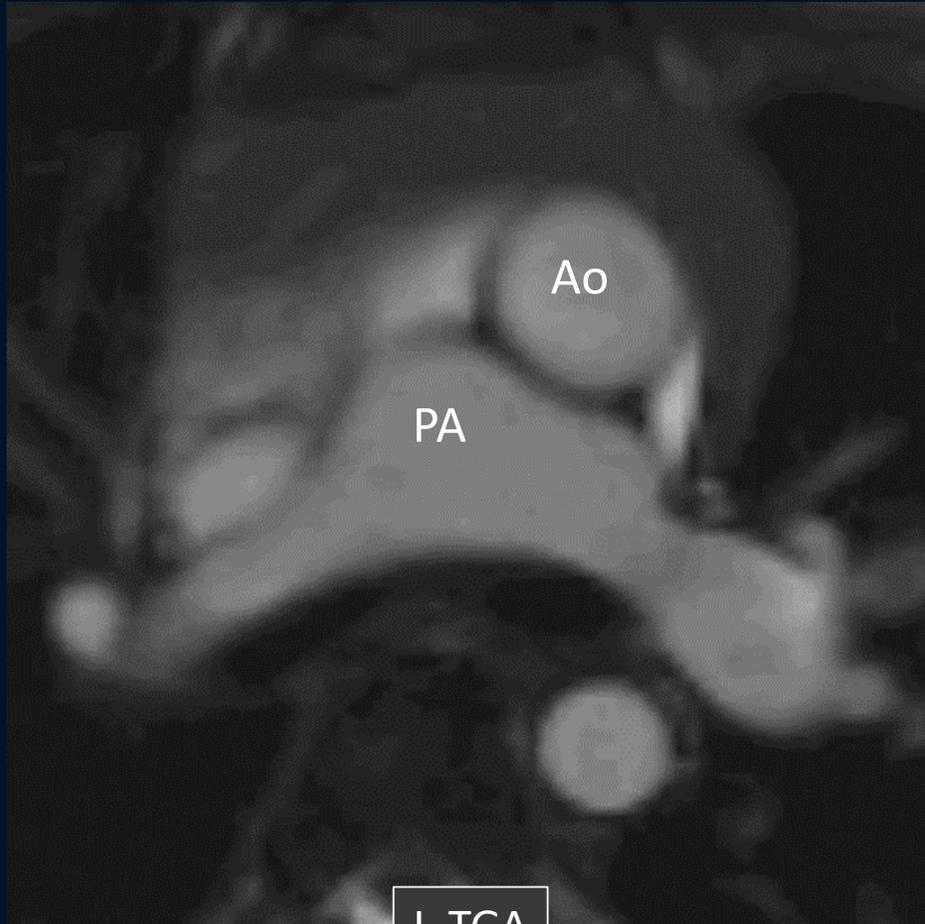


# Great Arterial Relationship

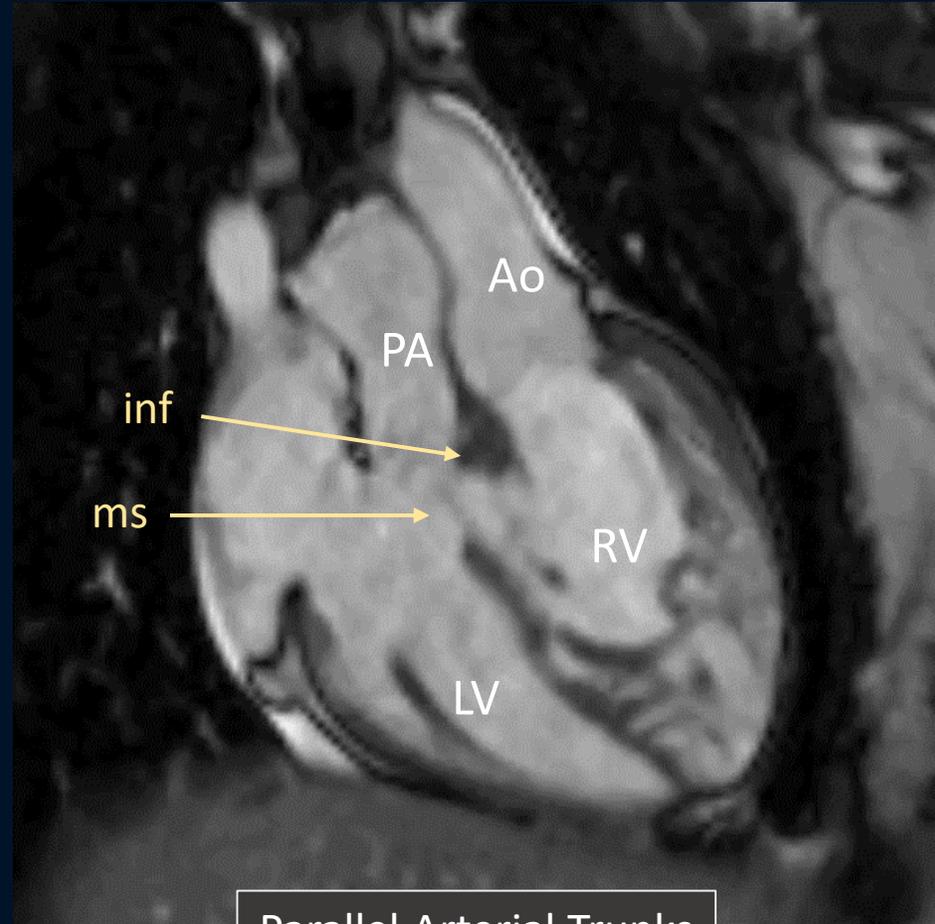


Typical orientation in ccTGA (not always)

# L-TGA / Parallel Arterial Trunks



L-TGA

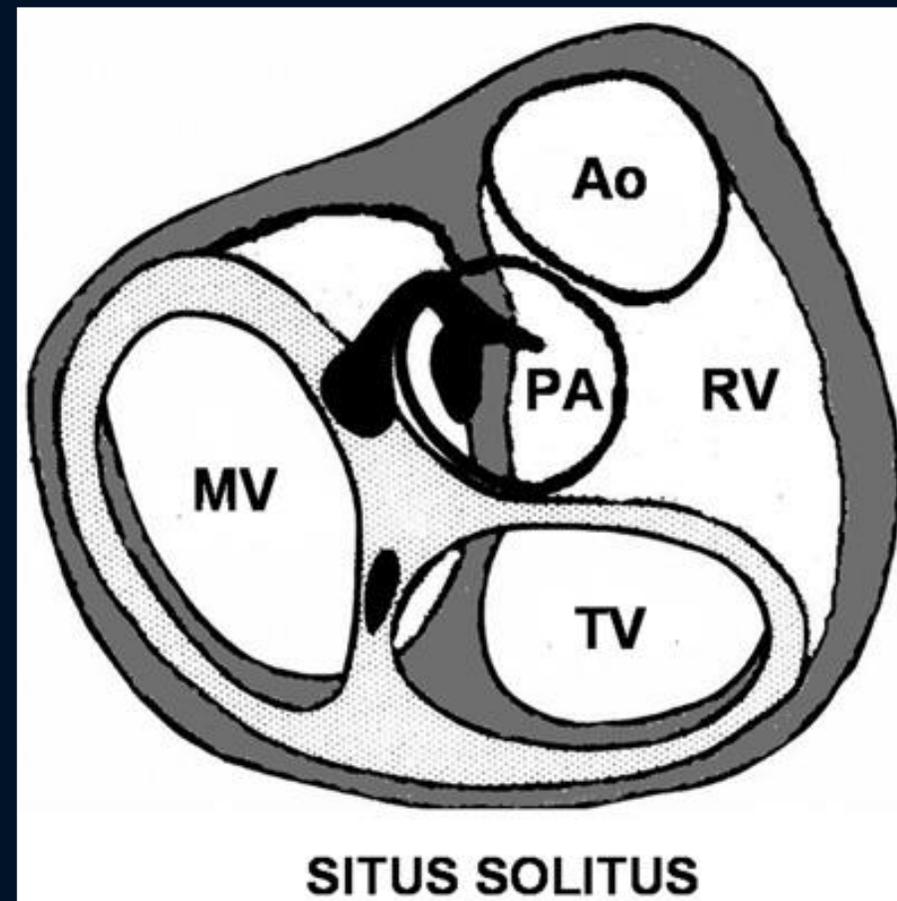


Parallel Arterial Trunks

# ccTGA: Conduction Axis

# What is the typical anatomy of the conduction axis in ccTGA?

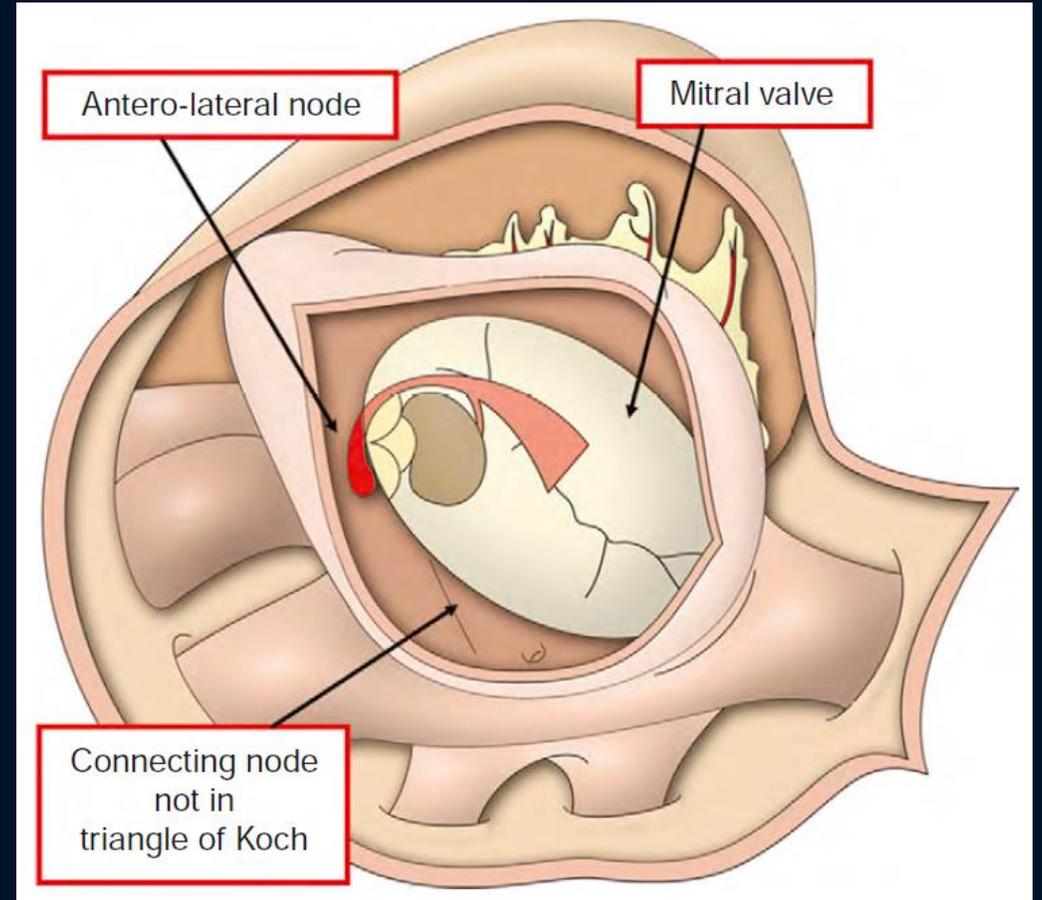
- Wedged PA leads to atrioventricular septal malalignment gap
  - Filled with large membranous septum or pmVSD
- Functional AV node in anterolateral position
  - (regular node hypoplastic at apex of triangle of Koch)
- **Conduction axis courses underneath PV annulus, anterosuperior to membranous septum**
- **Risk of AV block**
  - Congenital, post-op, or progressive (fibrosis)
  - 1.3 – 2.0% annual risk of complete heart block



Oliver JM et al. Am J Cardiol. 2012;110:1687-1691.

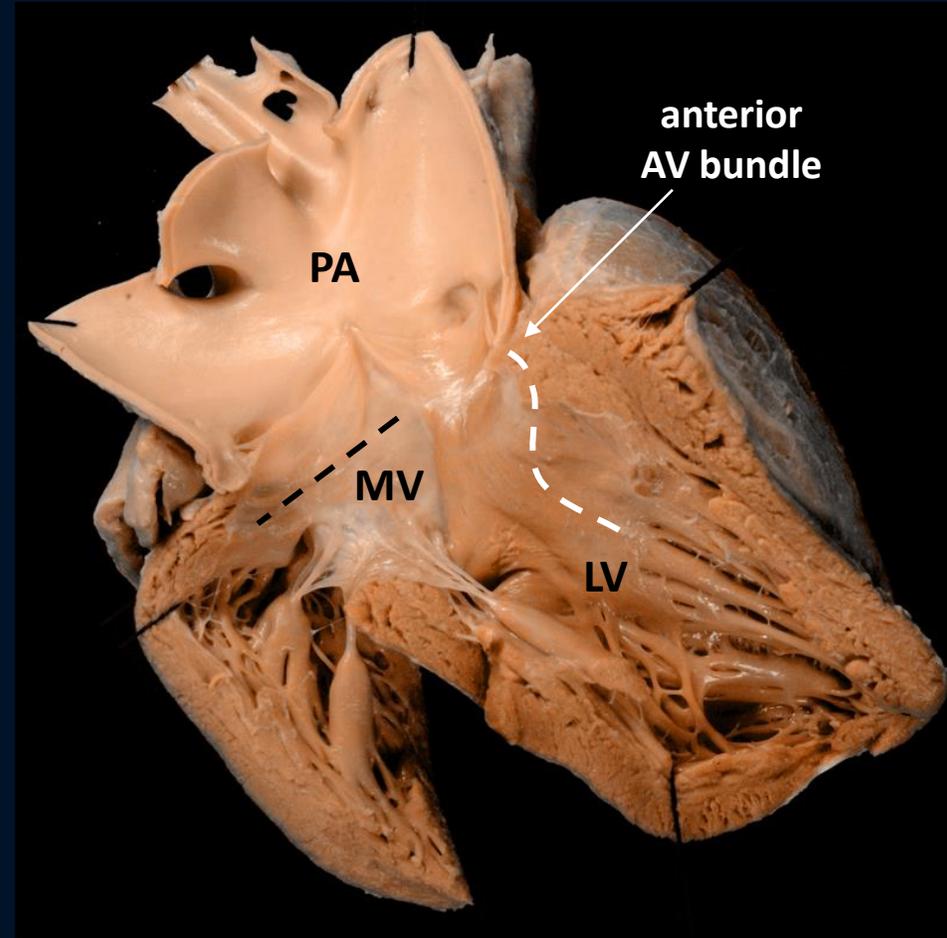
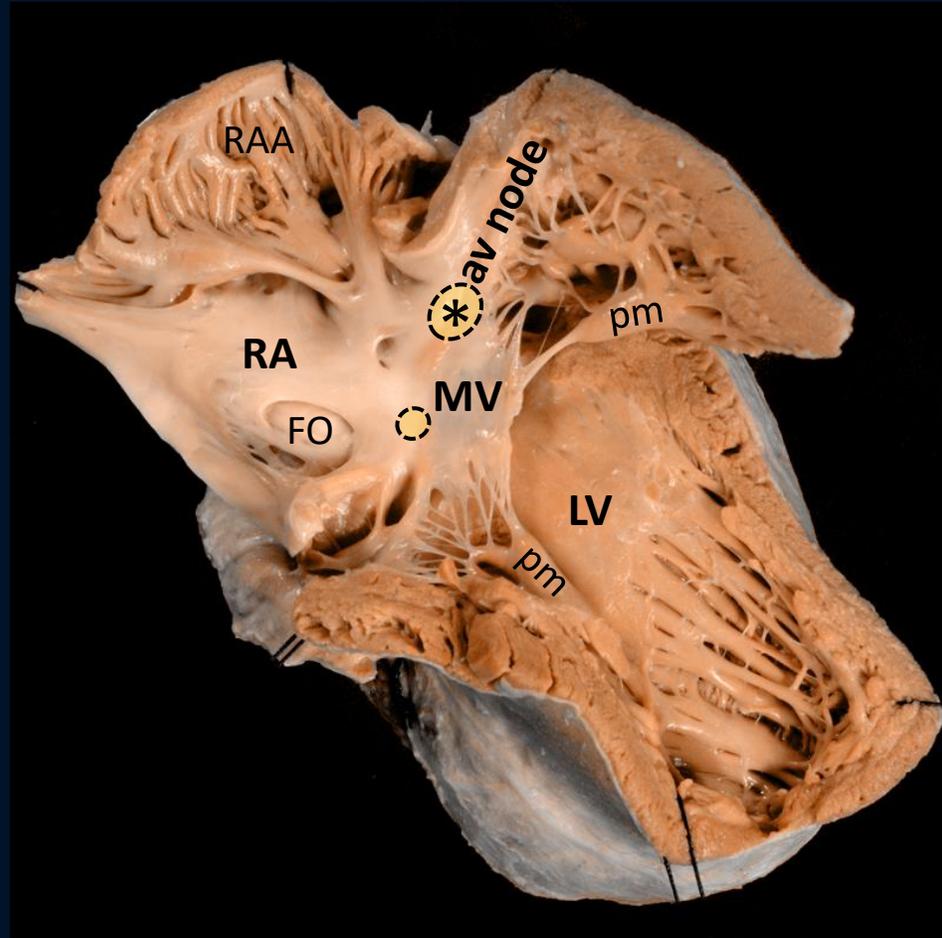
# What is the typical anatomy of the conduction axis in ccTGA?

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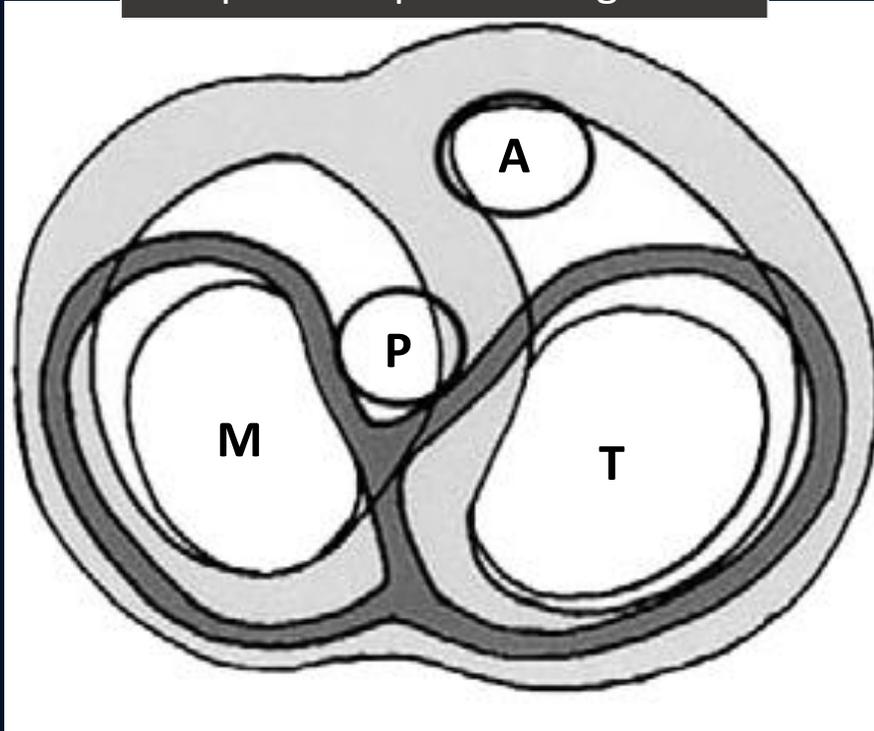
Anderson RH et al. Pediatric Cardiology. 2010. 3<sup>rd</sup> Ed.

# What is the typical anatomy of the conduction axis in ccTGA?

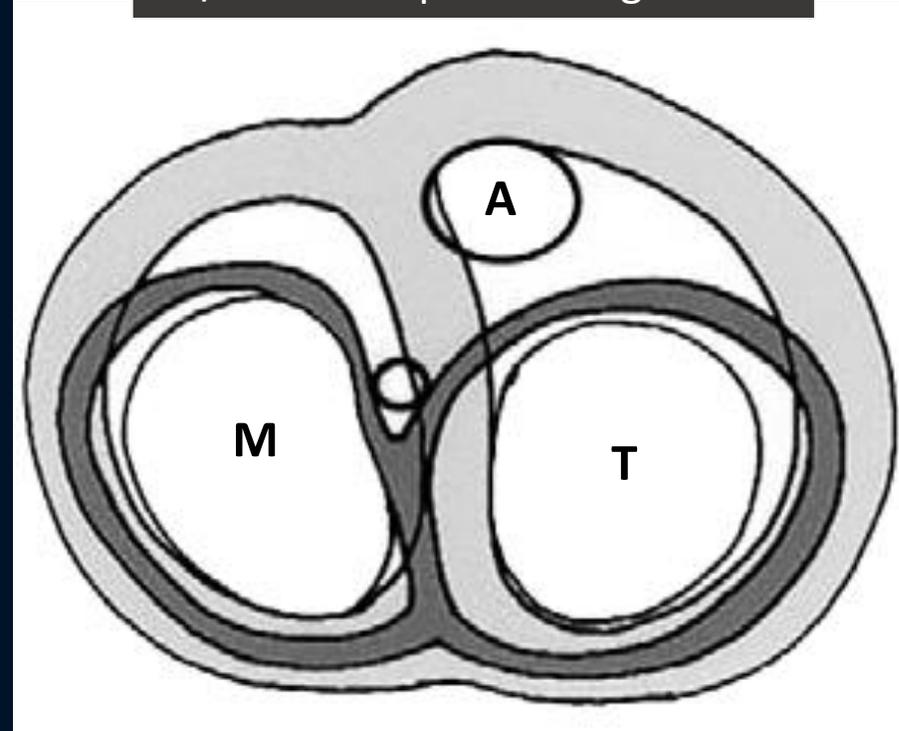


# Are there other arrangements of the conduction axis in ccTGA?

Deep PA = septal malalignment



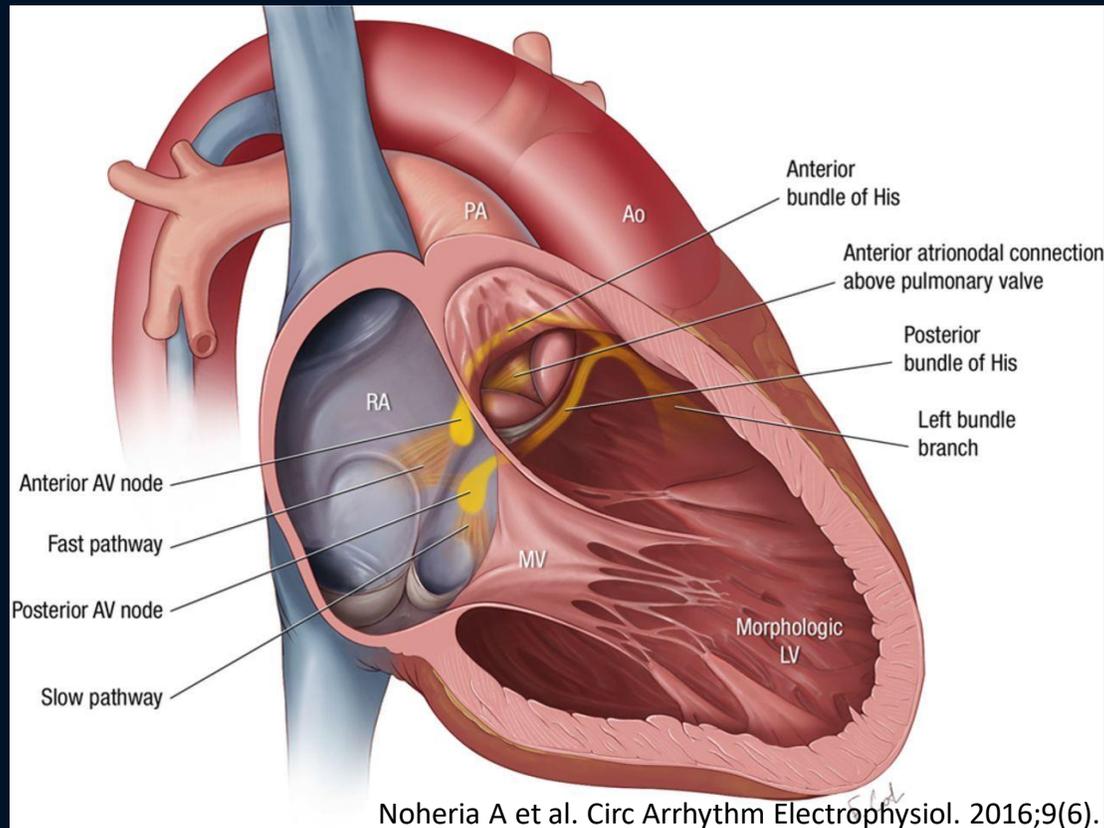
PS/PA = no septal malalignment?



Hosseinpour AR et al. Ann Thorac Surg. 2004;77:2163-6.

**Hypothesis: In PS/PA, AV node & conduction axis can be normal**

# Are there other arrangements of the conduction axis in ccTGA?



- Separate AV nodes can connect individually to anterior & posterior conduction bundles of His
- Can form so-called Monckeberg sling of conduction tissue around VSD

**Exceptions & rare variations exist, which can be mediated by underlying morphology**

# ccTGA: Associated Anomalies

- ❖ *VSD, Tricuspid Valve Lesions, mLVOTO/PS, Coronary Anomalies*
- ❖ *Combination of associated anomalies dictate clinical presentation*

# How frequent are associated anomalies in ccTGA?

Associated Anomaly	Frequency
➤ VSD	➤ 50-90%
➤ mLVOTO / Pulmonary Stenosis	➤ 33-66%
➤ Tricuspid Valve Lesions	➤ 90% at autopsy (33-50% significant)
➤ Anomalous Coronaries	➤ 20-25%

# What is the typical type of VSD in ccTGA?

## ➤ Typically perimembranous VSD

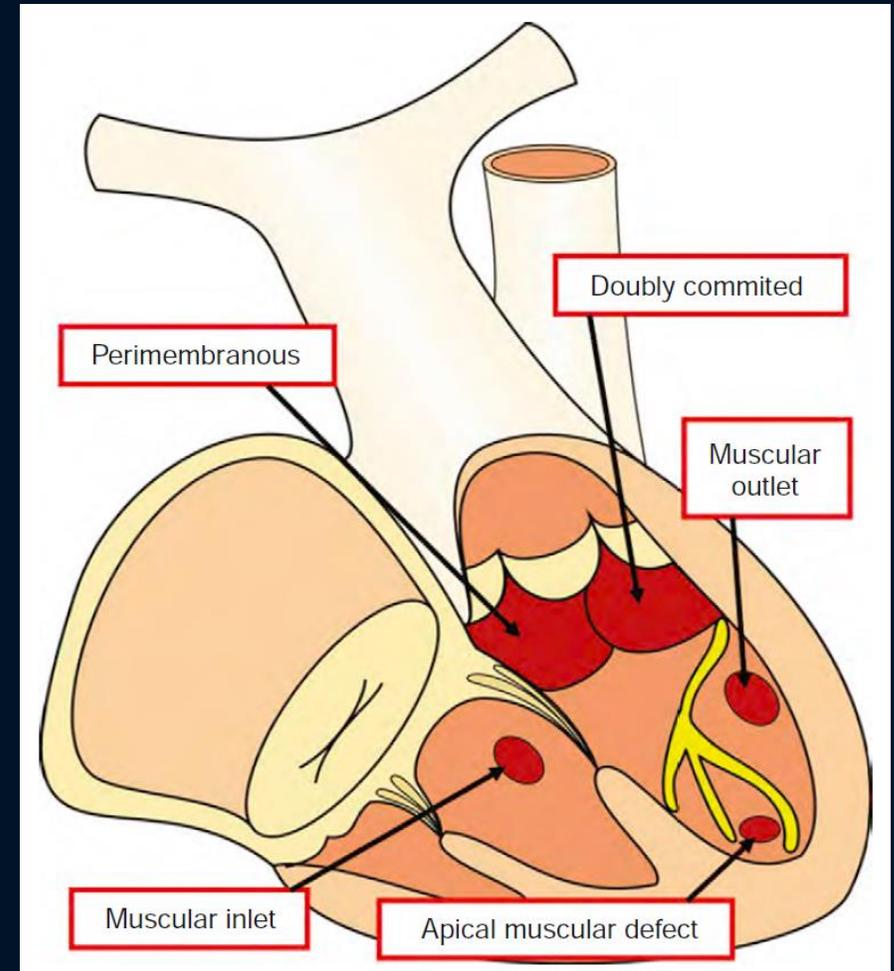
- Subpulmonic
- Often extends to mLV inlet
- *Conduction axis courses anterosuperior to VSD*

## ➤ Muscular VSD

- If subpulmonic, conduction axis also courses anterosuperior to VSD
- If between outlets, conduction axis may course posteroinferiorly (as usual)

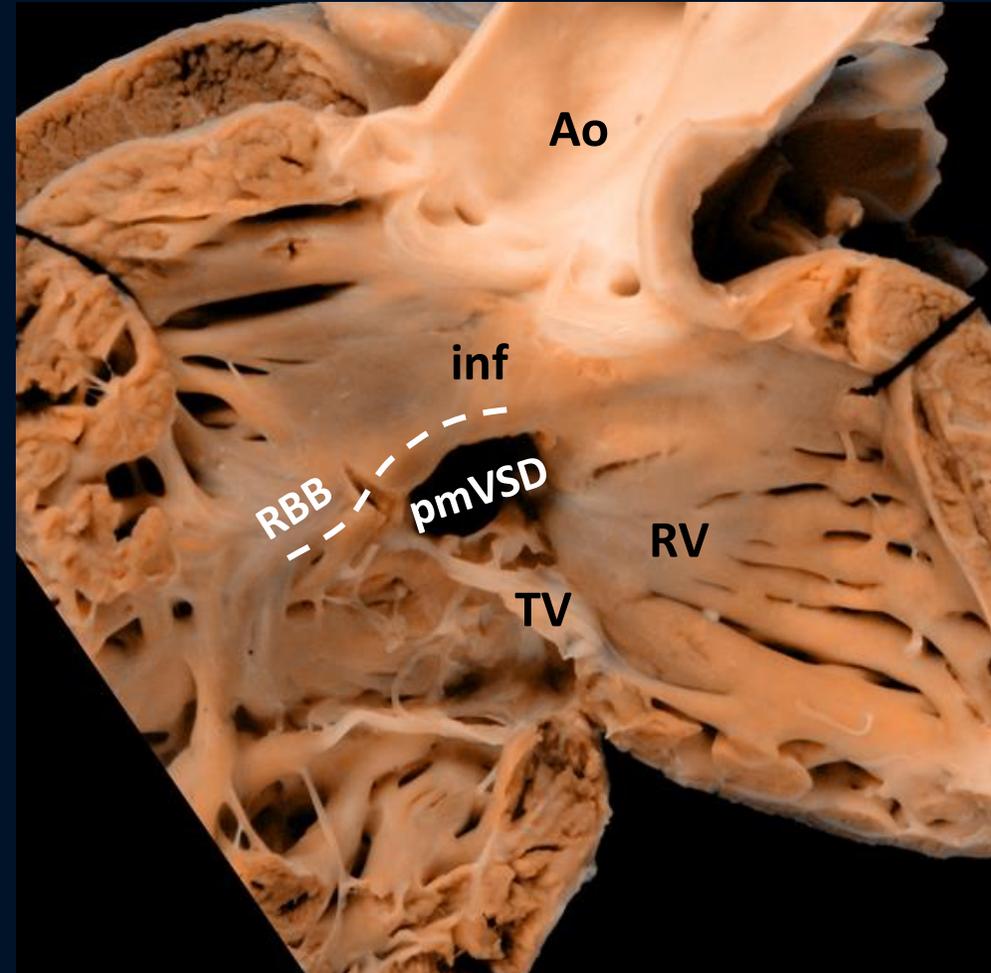
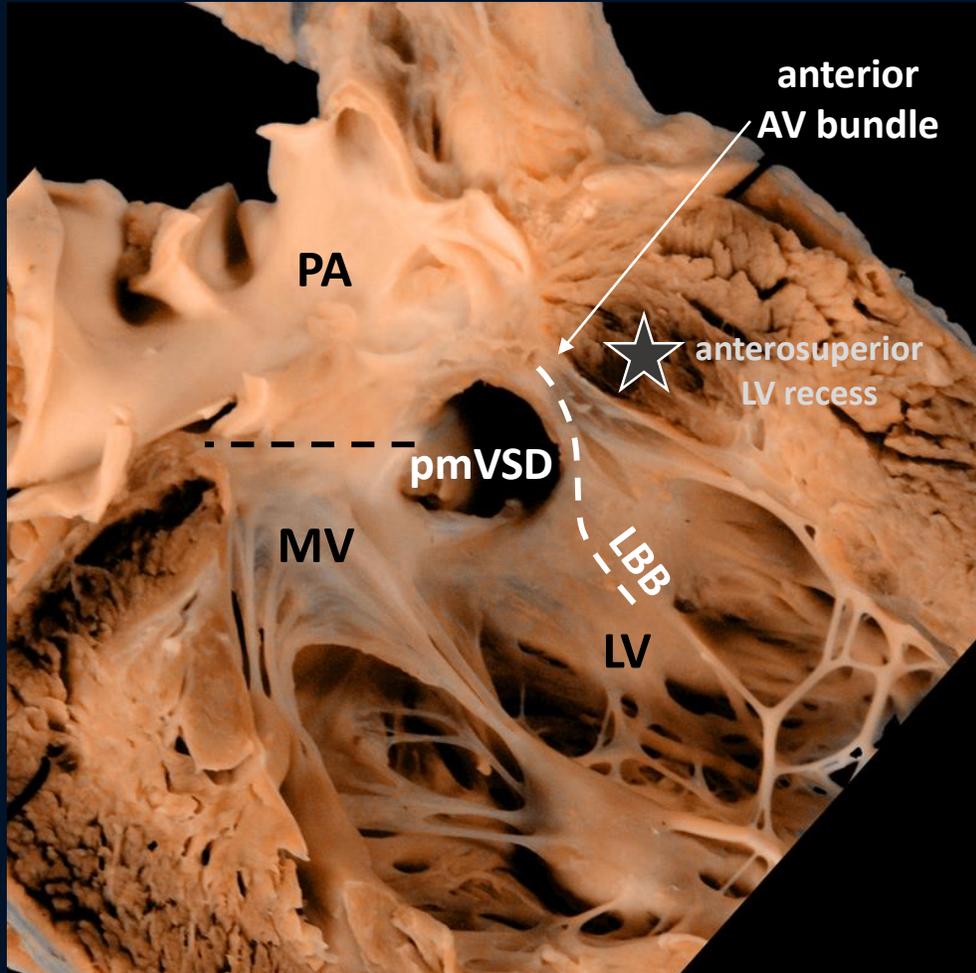
## ➤ Doubly committed VSD

- Common in Asian populations

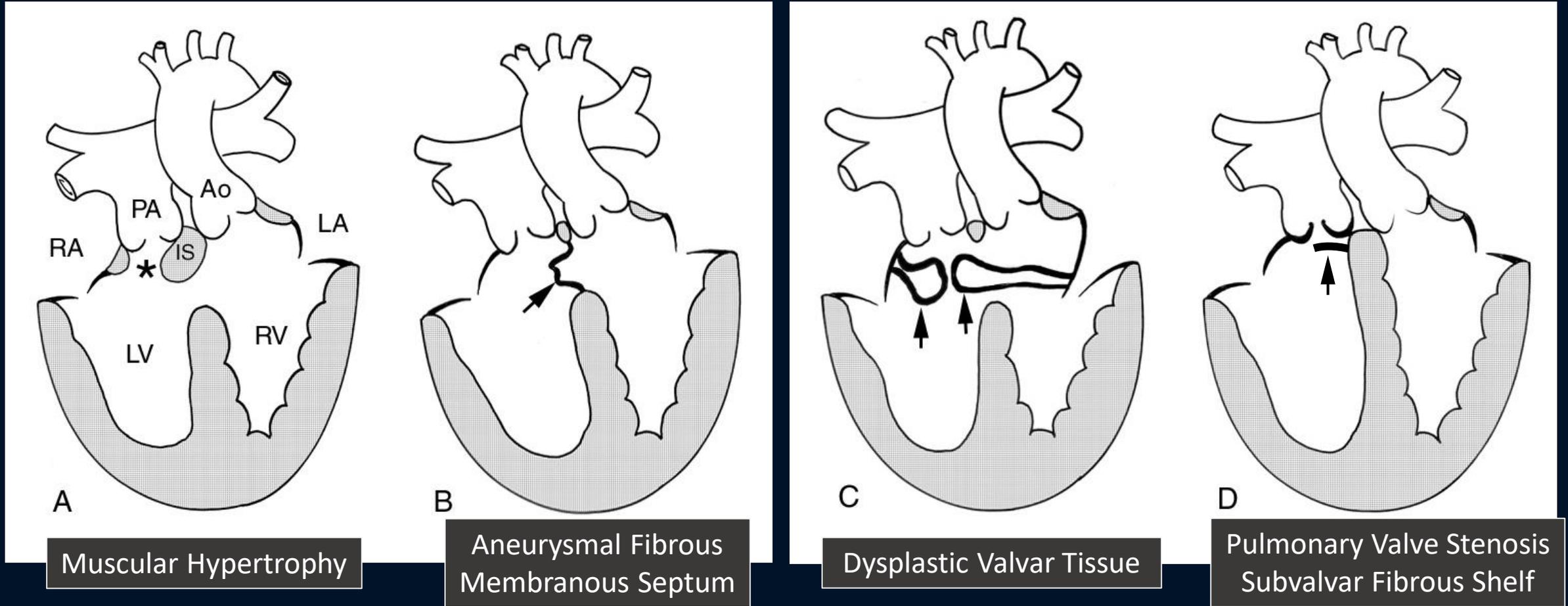


Anderson RH et al. Pediatric Cardiology. 2010. 3<sup>rd</sup> Ed.

# Perimembranous VSD in ccTGA

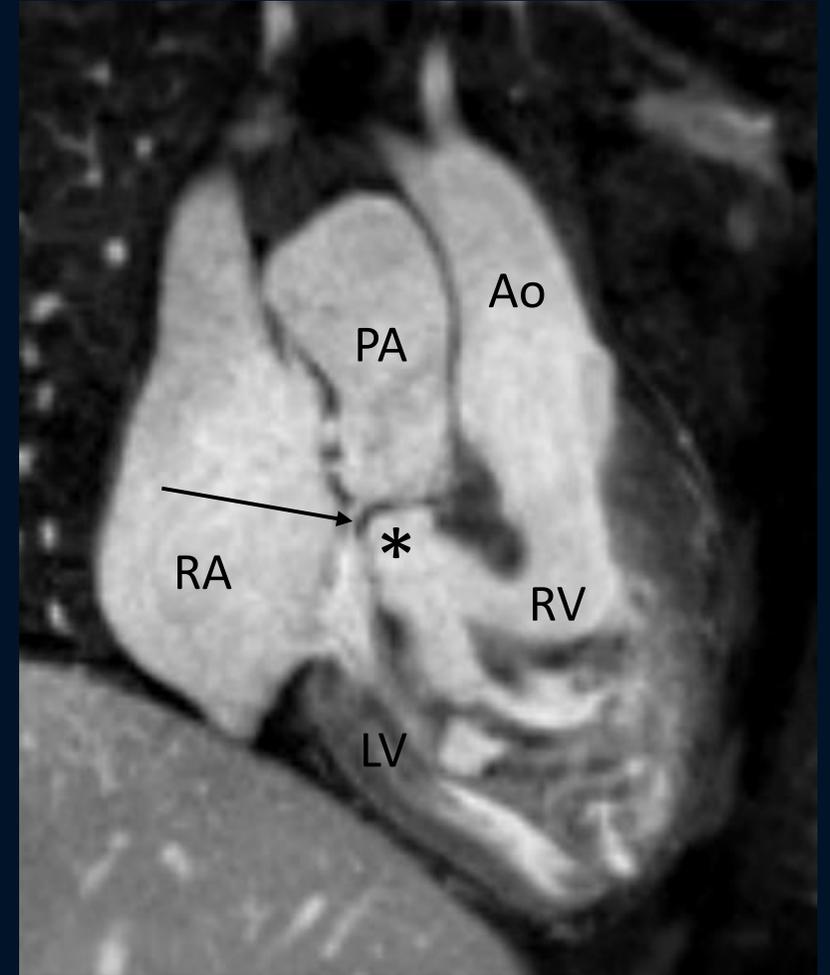
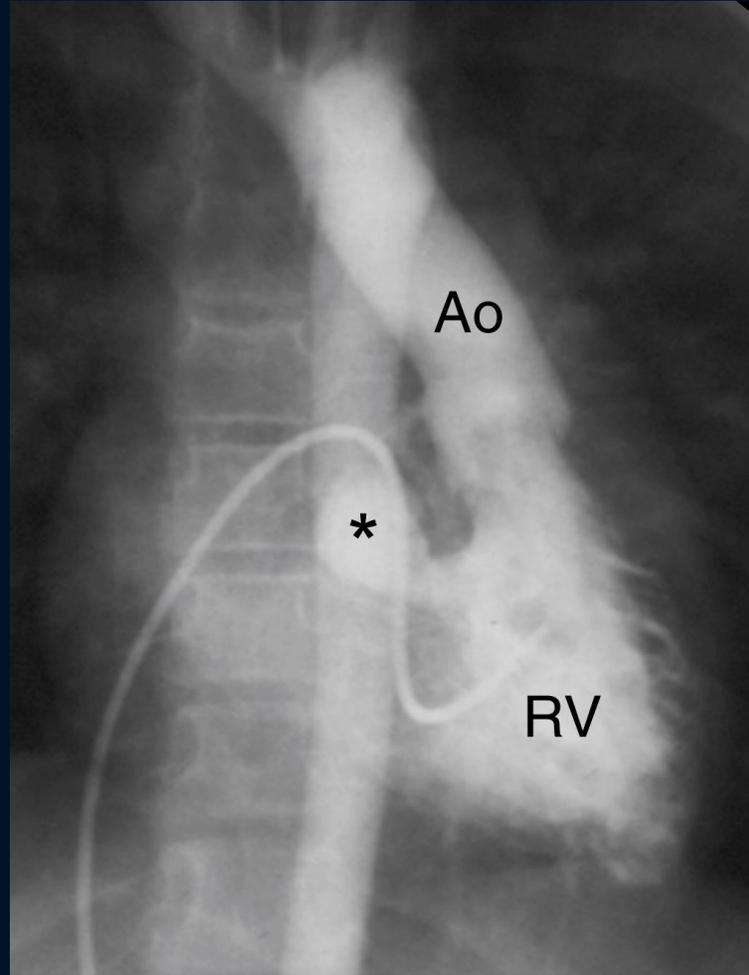
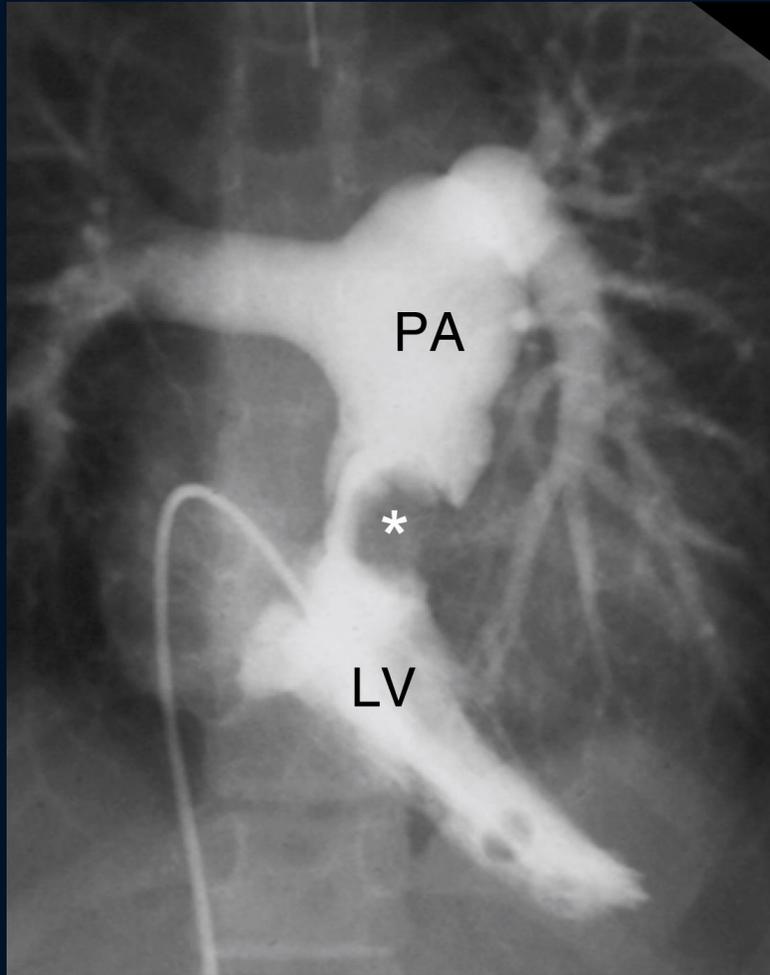


# PS / mLVOTO in ccTGA



**Conduction axis intimately associated with mLVOTO → at risk during resection**

# mLVOTO from aneurysmal fibrous intervententricular membranous septum



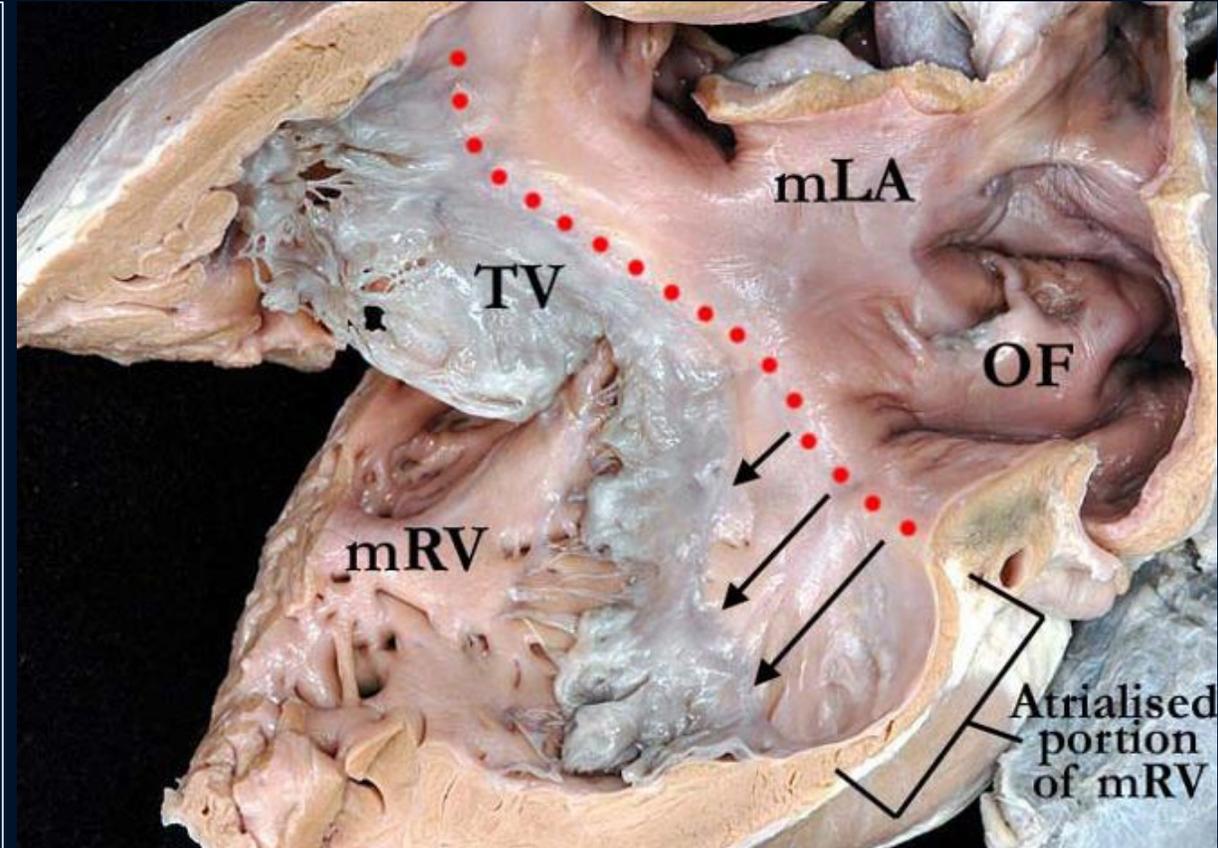
# Tricuspid Valve Lesions

- Tricuspid Valve Dysplasia
- Ebstein Malformation
- Straddling and Overriding

**Risk of progressive TR that contributes to systemic RV dysfunction**

# Ebstein Malformation of TV in ccTGA

- Associated VSD in 75%
- Can be associated with aortic arch obstruction with severe TR
- Atrialized RV usually smaller & not as thinned compared with isolated Ebstein
- Anterior leaflet not sail-like in most cases

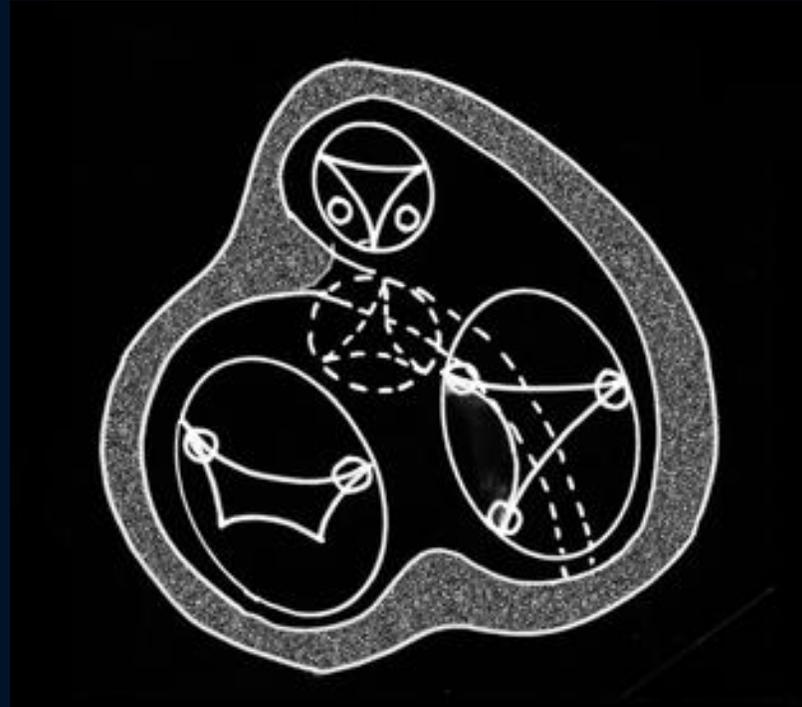


Wallis GA et al. Orphanet Journal of Rare Diseases. 2011; 6:2.

# ccTGA – Overriding / Straddling TV – DILV Spectrum



ccTGA



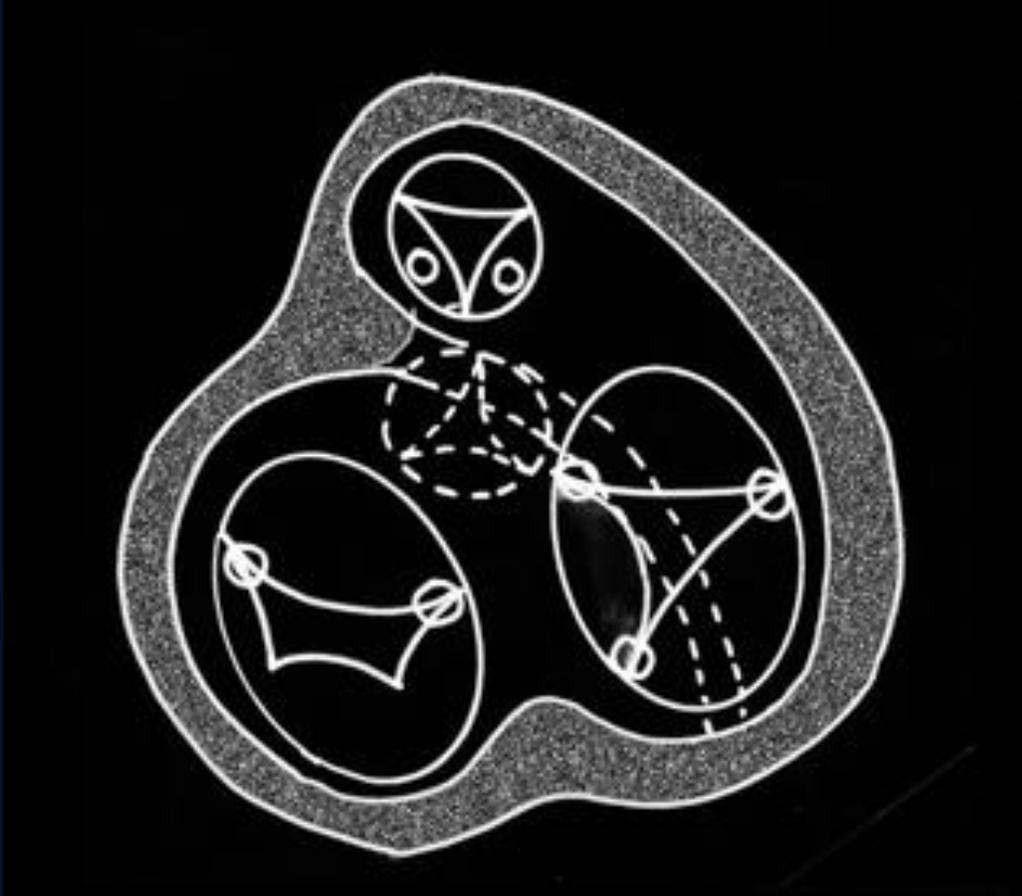
Overriding / Straddling TV



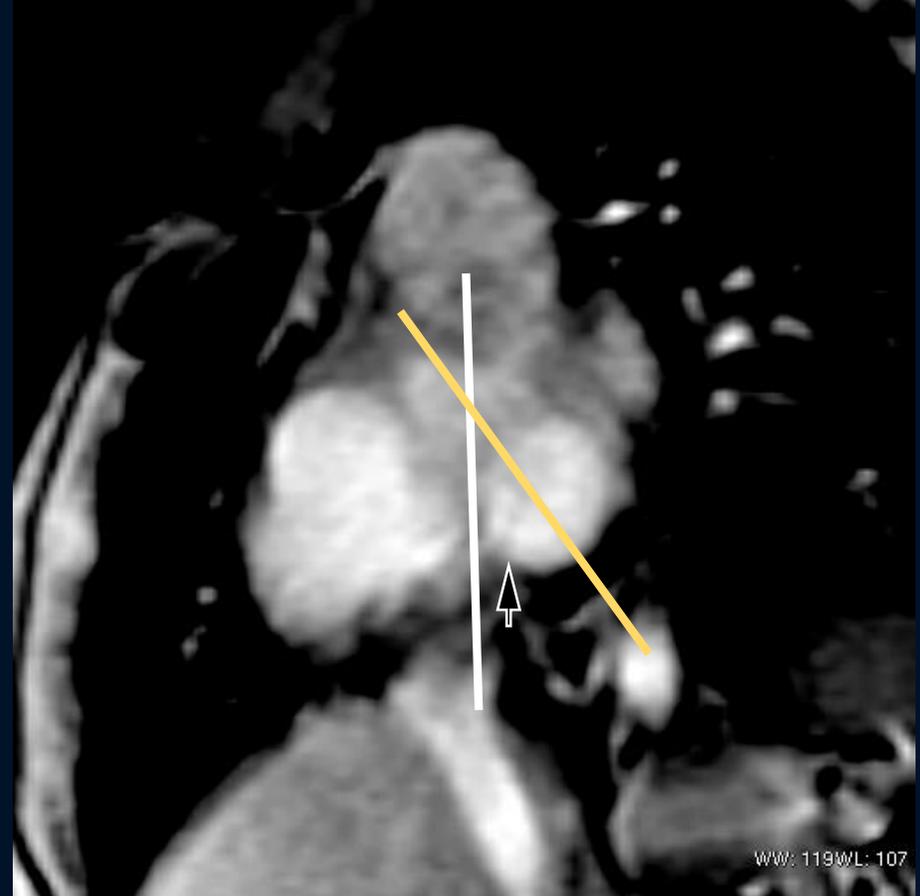
Double Inlet Left Ventricle



# ccTGA – Overriding / Straddling TV – DILV Spectrum



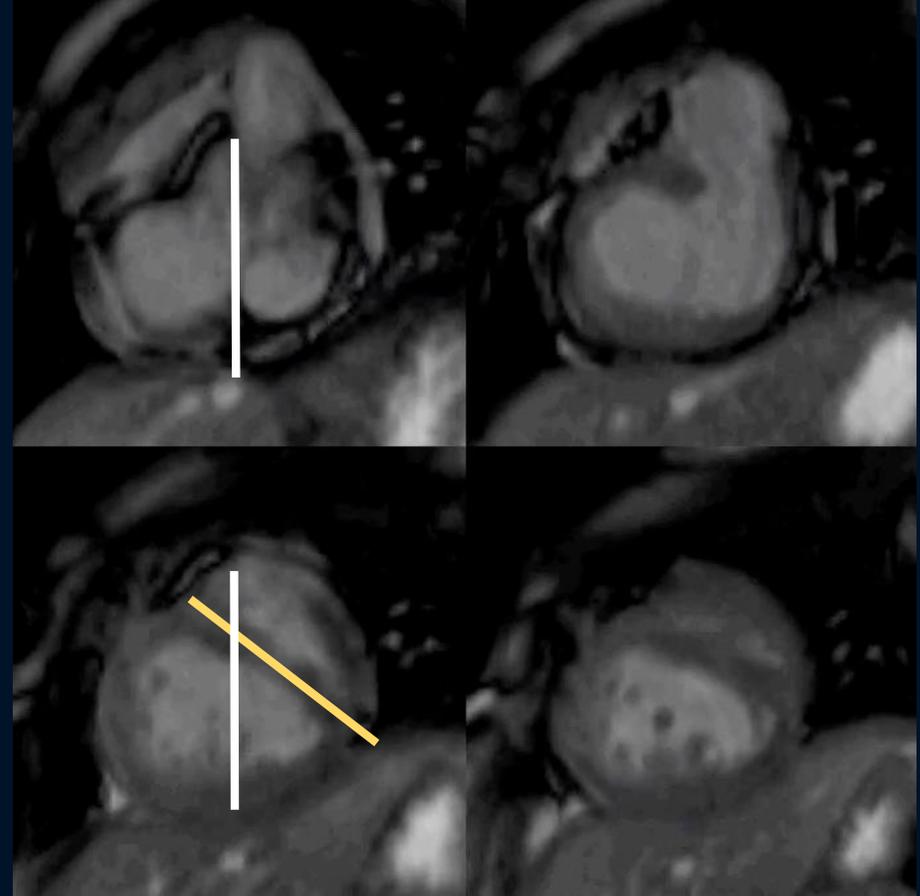
Overriding / Straddling TV



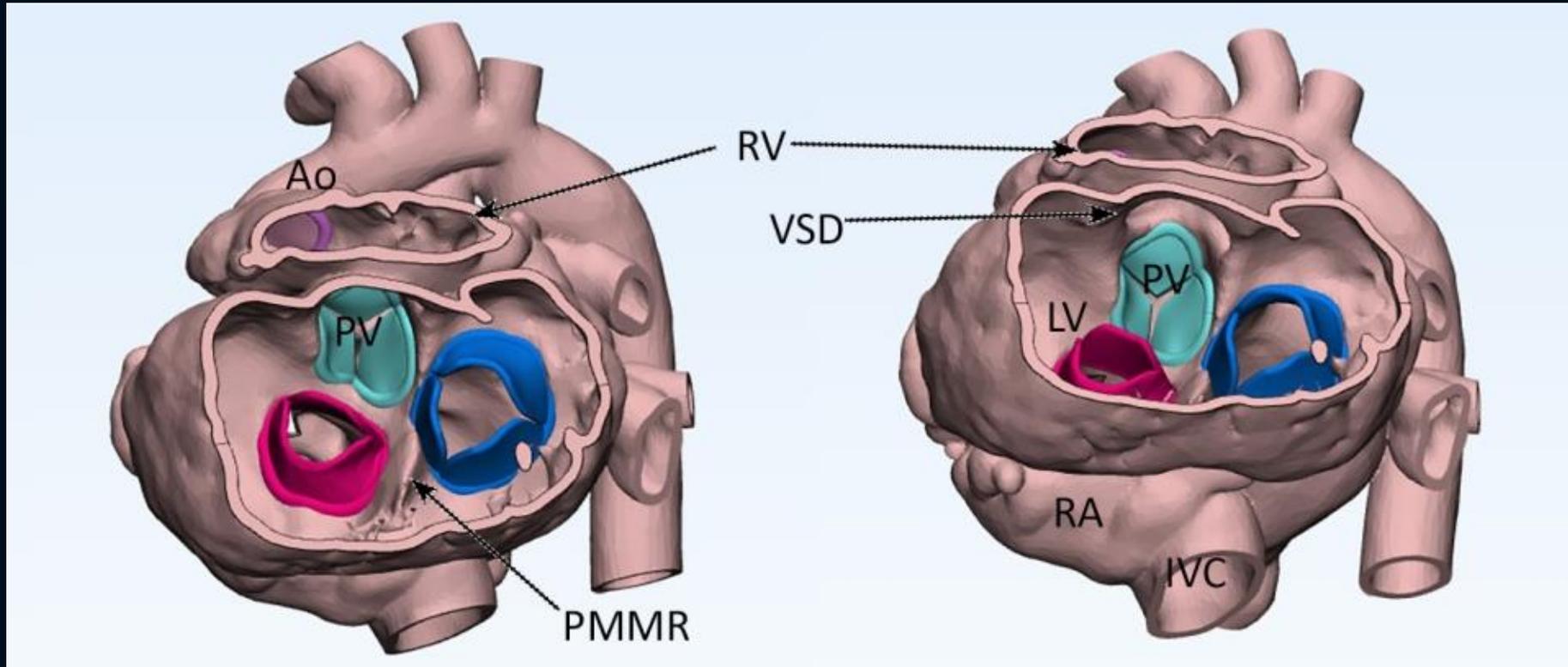
# ccTGA – Overriding / Straddling TV – DILV Spectrum



Double Inlet Left Ventricle



# DILV with transposition

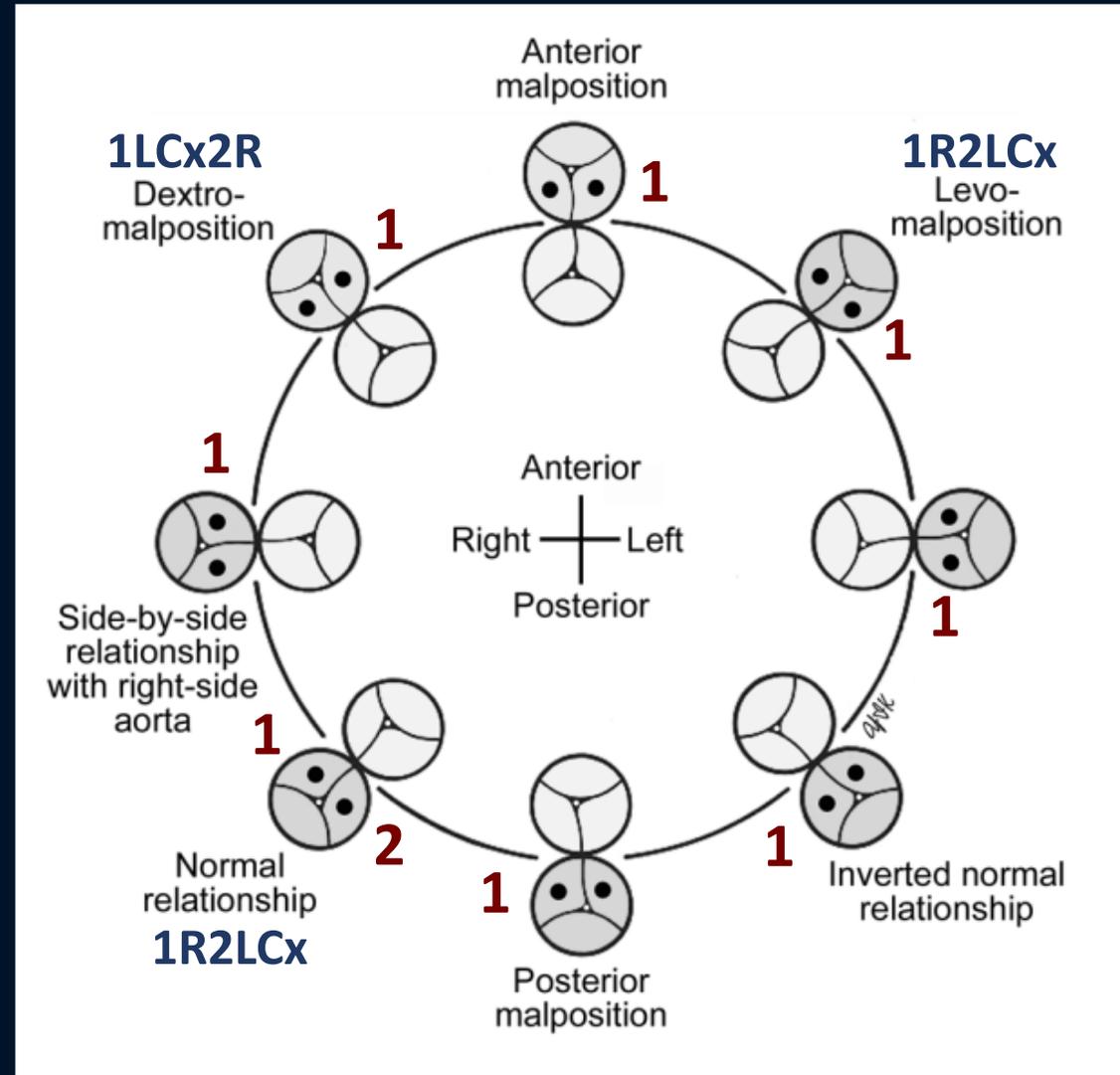


Basal View – Inlets

Basal View – Septum

# Leiden Classification for Coronary Arteries

Sinus 1 = To the right of a surgeon standing in non-coronary sinus  
 Sinus 2 = To the left



R = RCA  
 L = LAD  
 Cx = Circumflex

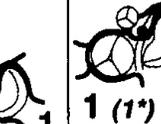
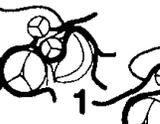
# Coronaries in Discordant AV Connections

## Coronary arteries

discordant VA connections  
(without pulmonary atresia)

Aorta arising from mRV  
& pulmonary atresia

double outlet mRV  
(without pulmonary atresia)

 4	 6	 3 (*)	 1 (*)
 1	 1	 1 (*)	 1
 1	 1	 1	 6
 2 (*)	 3	 3	 2
 1	 1	 1	 2
 1	 1	 1	 1

number of hearts, ( \* ) : hearts with mirror-imaged arrangement

➤ n=46

➤ Typical coronary pattern: 1R2LCx

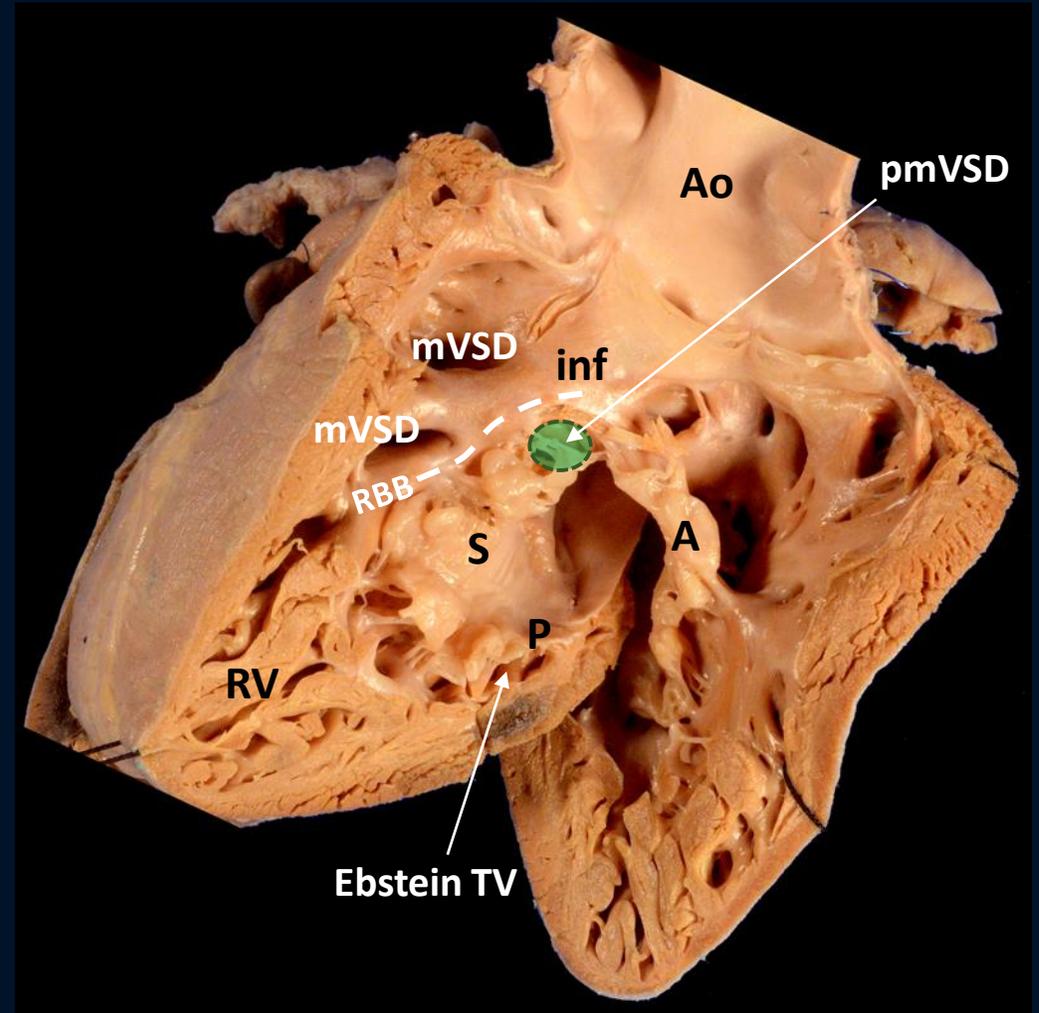
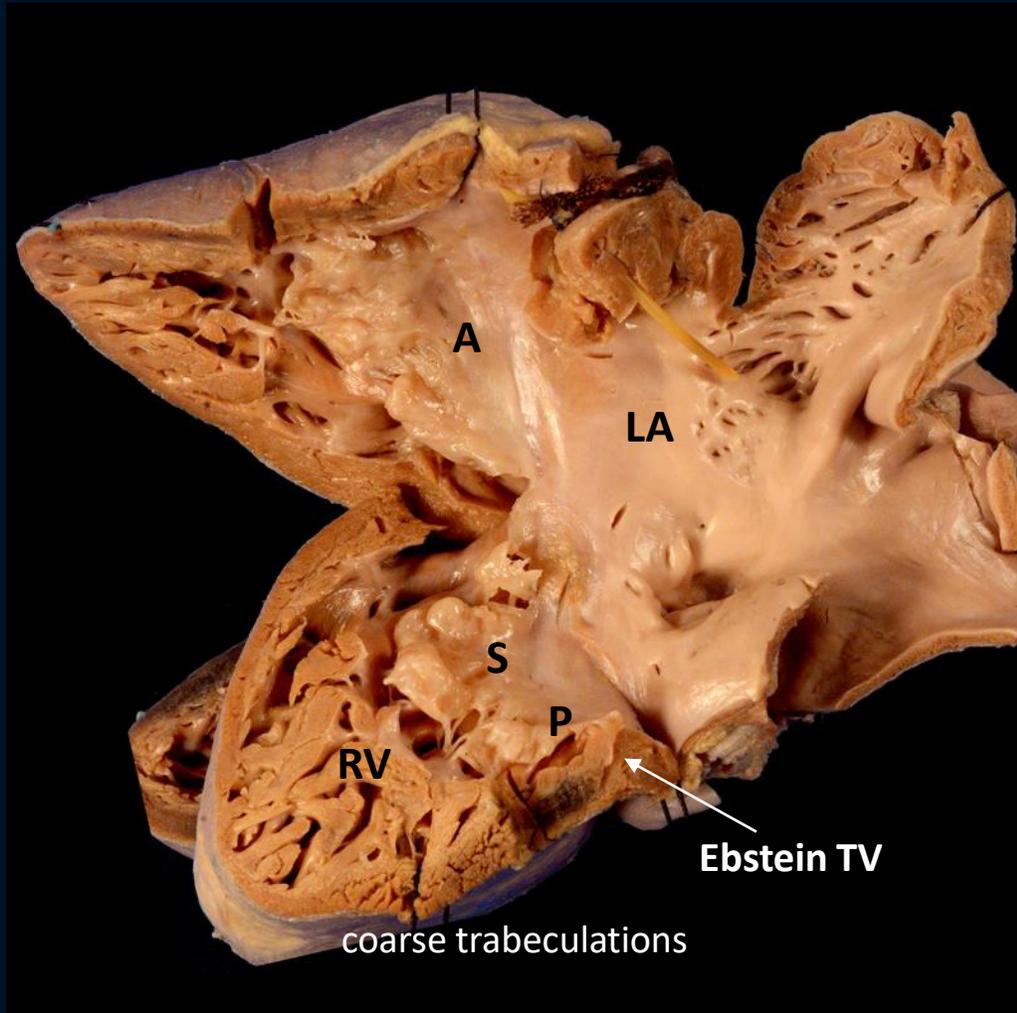
➤ ~50% with prominent conal branch

➤ 20-25% anomalous coronaries

Uemura H et al. Eur J Cardiothorac Surg. 1996; 10:194-200.

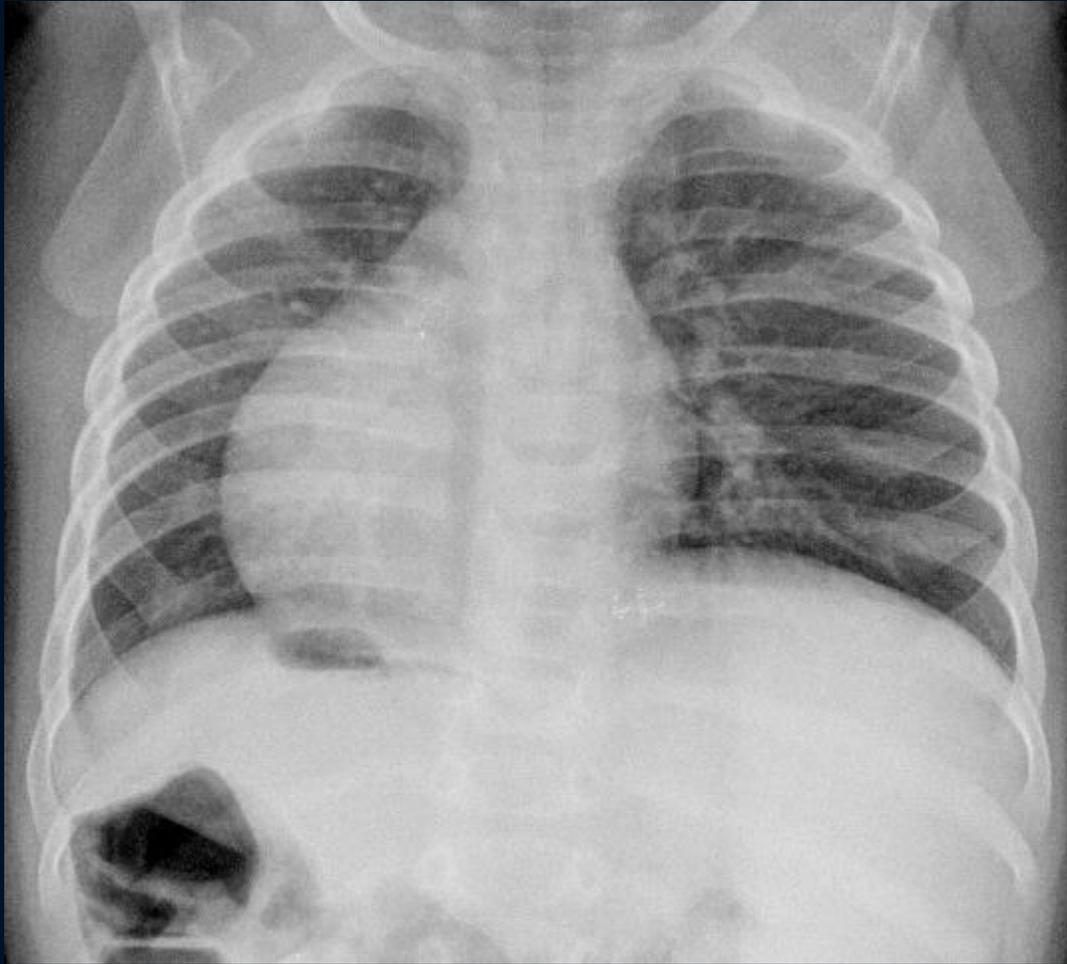


# ccTGA: Associated Lesions



# ccTGA: Atrial Situs

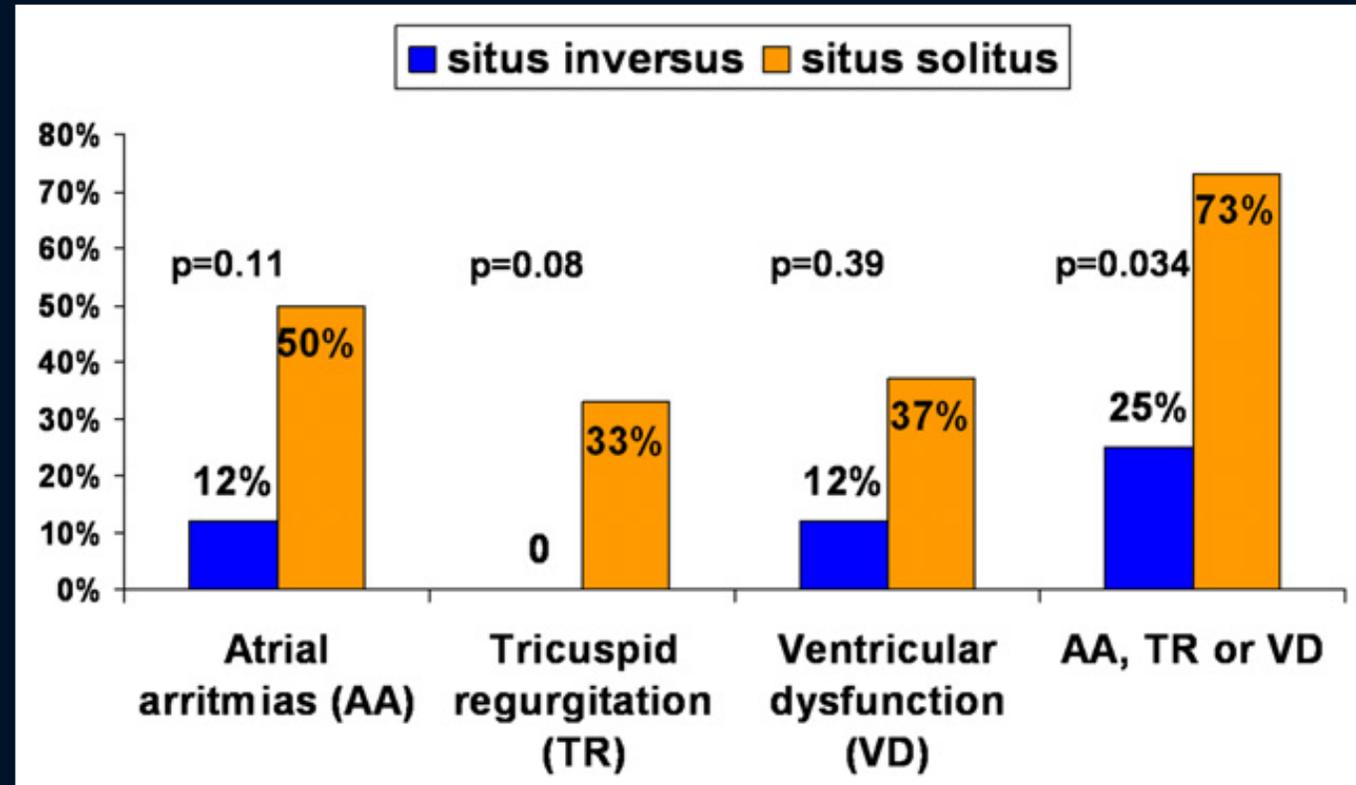
# How common are situs abnormalities in ccTGA?



- Atrial situs inversus in ~10%
- Dextrocardia / mesocardia with situs solitus also common

# What is the significance of inversus in ccTGA?

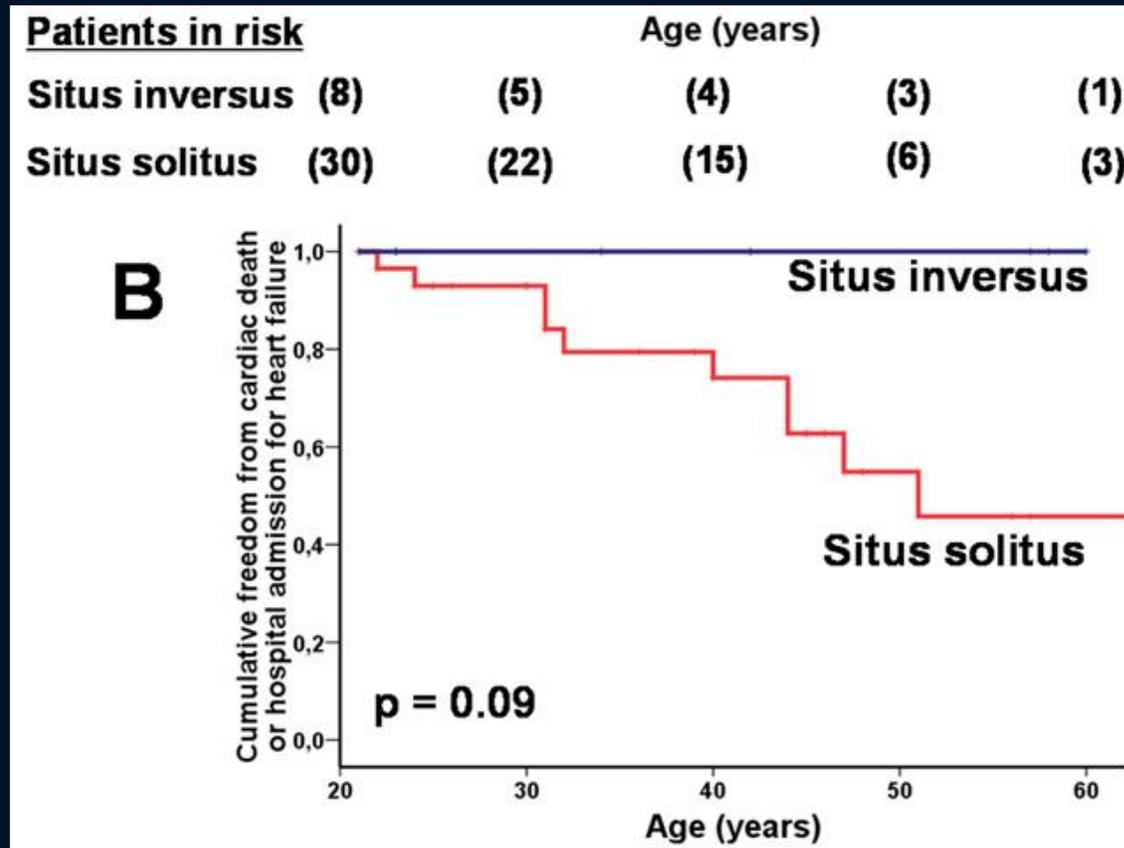
- n=38
- 8 situs inversus
- 1/8 complete AV block
  - More often normal conduction axis
- 0/8 Ebstein TV



Oliver JM et al. Am J Cardiol. 2012;110:1687-1691.

↓ atrial arrhythmia, TR, & systemic ventricular dysfunction in inversus

# What is the significance of inversus in ccTGA?

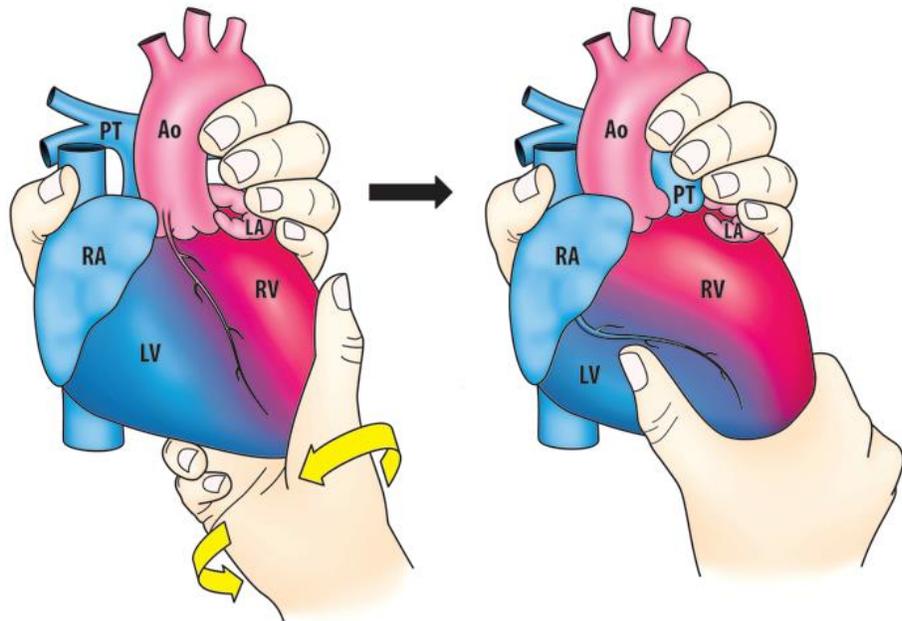


Oliver JM et al. Am J Cardiol. 2012;110:1687-1691.

**Situs inversus ccTGA is not just the mirror image of situs solitus ccTGA**

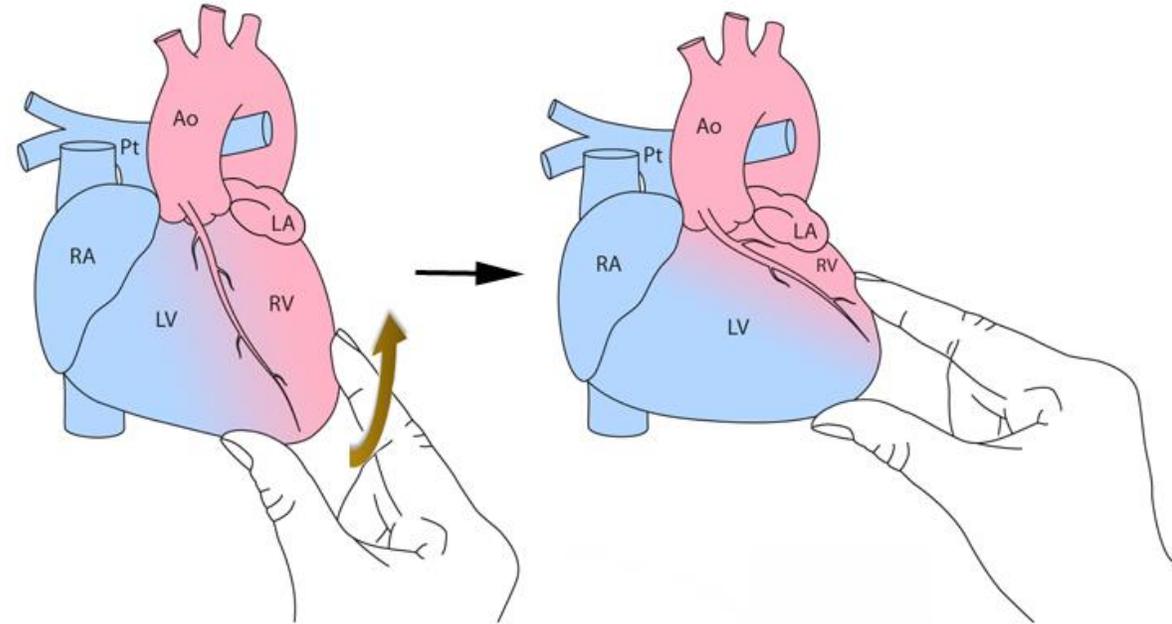
# ccTGA: Ventricular Relationship

# Abnormal ventricular relationships can be seen in ccTGA



**Twisted AV Connection / Criss-Cross Heart**

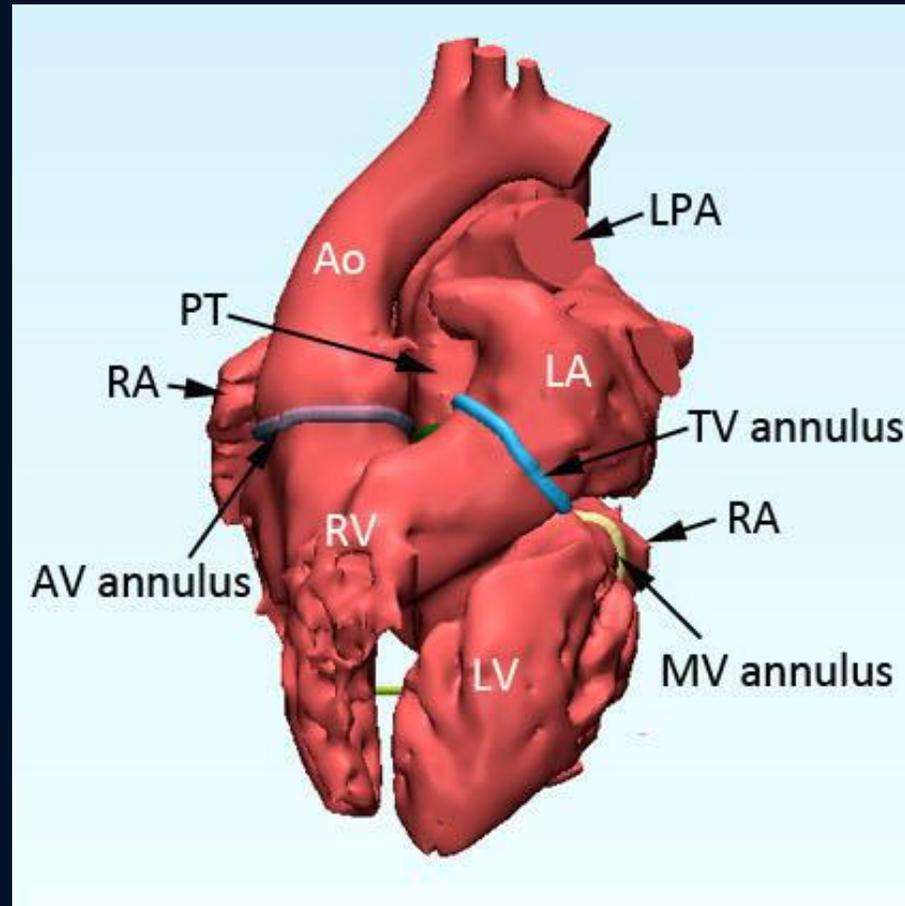
➤ rotation of ventricular mass



**Superior-Inferior Ventricles**

➤ tilting of ventricular mass

# ccTGA with twisted AV connection



# ccTGA: Surgical Strategy

# What are the main determinants of the natural history of ccTGA?

1. Systemic mRV function
2. Systemic tricuspid valve function
3. Development of arrhythmias



Modified by impact of associated anomalies

# ccTGA: Surgical Strategy

Surgical Strategy	Description	Issues
<b>Physiologic (“Classical”) Repair</b>	<b>Repair all associated anomalies</b> <ul style="list-style-type: none"> <li>➤ Close VSD</li> <li>➤ Relieve LVOTO or LV-PA conduit</li> <li>➤ Repair/replace tricuspid valve</li> </ul>	<ul style="list-style-type: none"> <li>➤ Systemic mRV failure</li> <li>➤ Progressive TR</li> </ul>
<b>Anatomic Repair</b>	<b>Double-Switch</b> <ul style="list-style-type: none"> <li>➤ Atrial Switch (Mustard/Senning)</li> <li>➤ Arterial Switch <ul style="list-style-type: none"> <li>➤ PS/PA: Rastelli, Nikaidoh, REV</li> </ul> </li> </ul>	<ul style="list-style-type: none"> <li>➤ Suitability for LV re-training <ul style="list-style-type: none"> <li>➤ Poor success &gt; 2 years-old</li> </ul> </li> <li>➤ Systemic mLV failure</li> <li>➤ Atrial baffle / RV-PA obstruction</li> <li>➤ Neo-aortic regurgitation</li> </ul>
<b>Palliation</b>	<ul style="list-style-type: none"> <li>➤ <b>PA Band:</b> theoretically ↓ TR</li> <li>➤ <b>Fontan:</b> ↓ reintervention rates, ↓ complete heart block</li> <li>➤ <b>1½:</b> anatomic repair + BCPC to unload mRV, ↓ baffle obstruction, theoretically ↓ strain on mLV</li> </ul>	<ul style="list-style-type: none"> <li>➤ Specific to chosen palliation</li> </ul>

# Other forms of Discordant AV Connections

# Other forms of Discordant AV Connections

## Discordant AV Connections with Pulmonary Atresia

- Relationships similar to ccTGA but with pulmonary atresia
  - With VSD: can baffle mLV to Ao
  - With IVS: hypoplastic mLV → single ventricle pathway

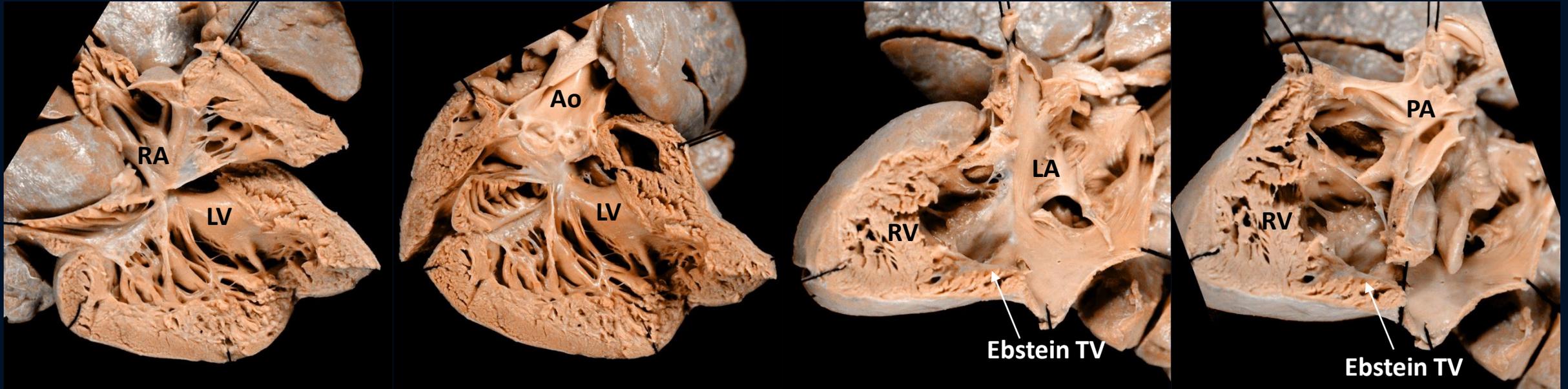
## Discordant AV Connections with Double Outlet Right Ventricle

- Usually with malposed great arteries and PT close to VSD; PS common
- Therefore, very similar to ccTGA with VSD

## Discordant AV Connections with Concordant VA Connections

- Variable infundibular morphology
- Physiology similar to transposition of the great arteries → atrial switch

# Discordant AV Connections with Concordant AV Connections



# Summary

- Most discordant AV connections will be ccTGA
- Morphology informs physiology & natural history of ccTGA:
  - Systemic RV failure
  - Systemic tricuspid valve regurgitation
  - Development of arrhythmia and AV block
- Modified by associated anomalies:
  - VSD: typically pmVSD, conduction axis runs anterosuperior
  - mLVOTO/PS: conduction axis at risk during repair
  - TV dysplasia / Ebstein malformation: TR is long-term risk factor
  - Coronary anomalies: may impact suitability for arterial switch
  - Inversus: more often normal conduction & less TV lesions = better prognosis
- Understanding morphology helps determine optimal surgical strategy



Thank you!

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### Special Acknowledgements

Shi-Joon Yoo, MD PhD  
Konstantin Krutikov, MD PhD  
David Chiasson, MD

