

# ATRIOVENTRICULAR CANAL DEFECTS

*Pathologist's Perspective*

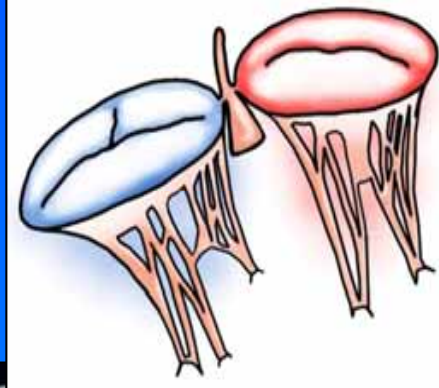
Debra L. Kearney M.D.  
Texas Children's Hospital  
Baylor College of Medicine

# OBJECTIVES

- Brief overview of endocardial cushions/embryology
- Nomenclature:
  - Synonyms
  - Complete vs Partial vs Transitional AV canal
- Major Anatomic features
- Rastelli classification
- Associated features:
  - Balanced vs Unbalanced
  - LV outflow tract obstruction
- Associated anomalies
- Conduction system with AV canal defects

No disclosures

# AV CANAL SEGMENT

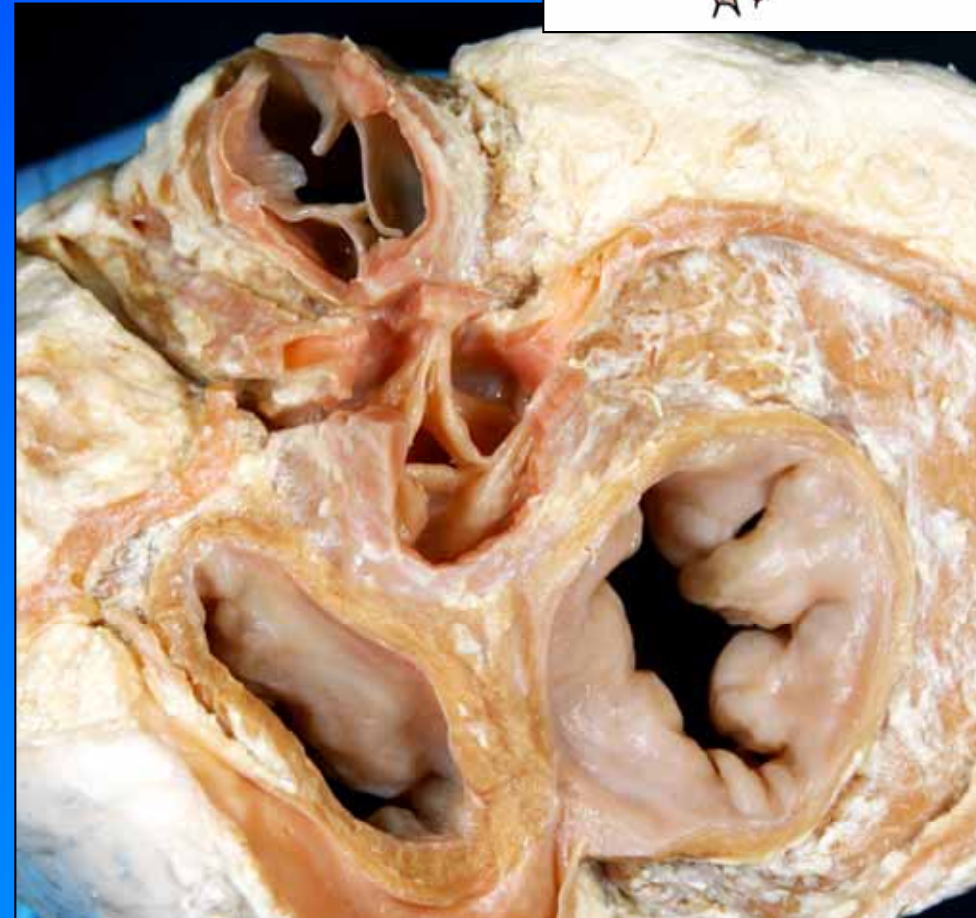


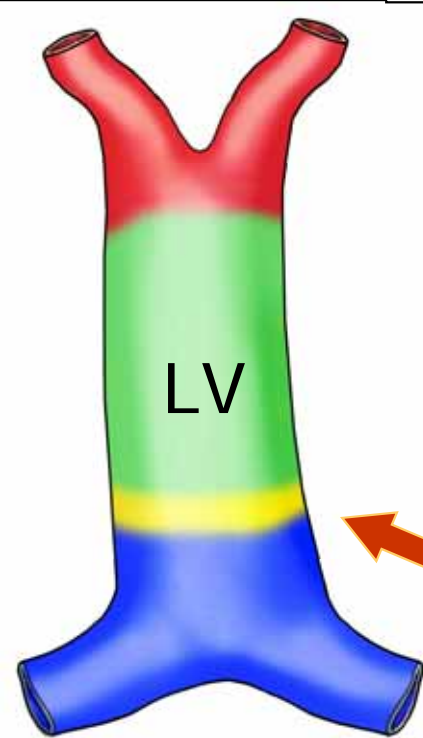
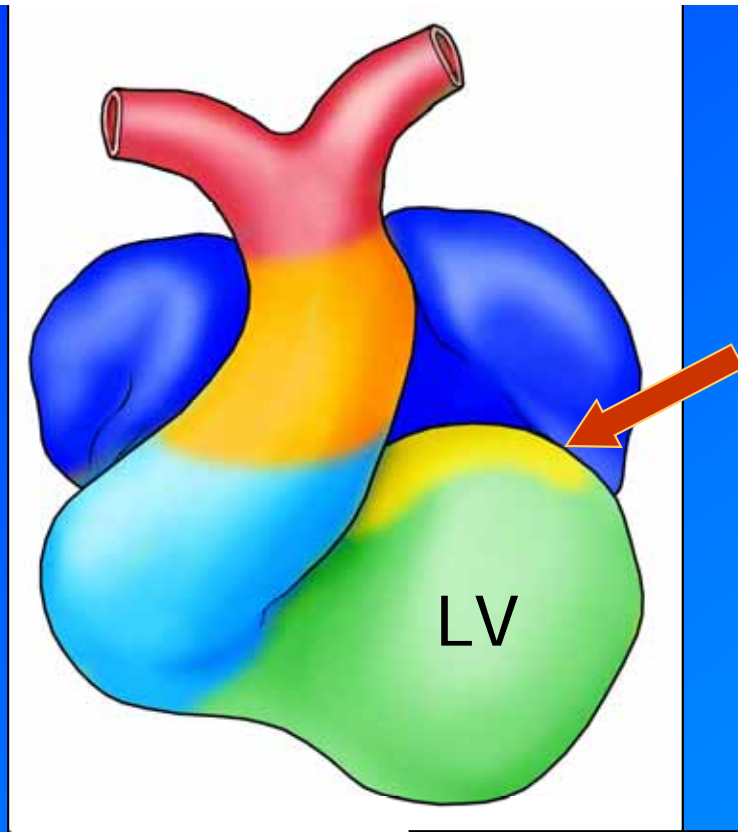
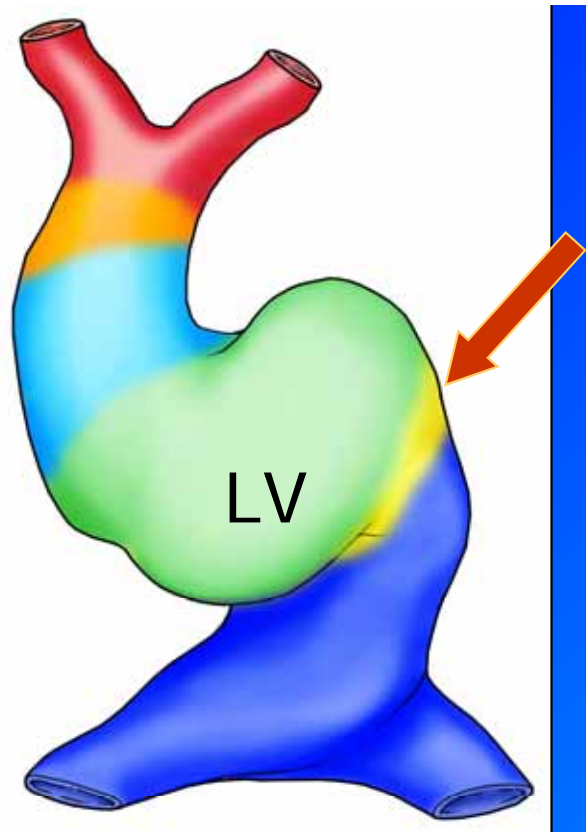
Segment connecting atria with ventricles

- AV valves
- Lowermost atrial septum
- Crest of inlet ventricular septum

AV canal development:

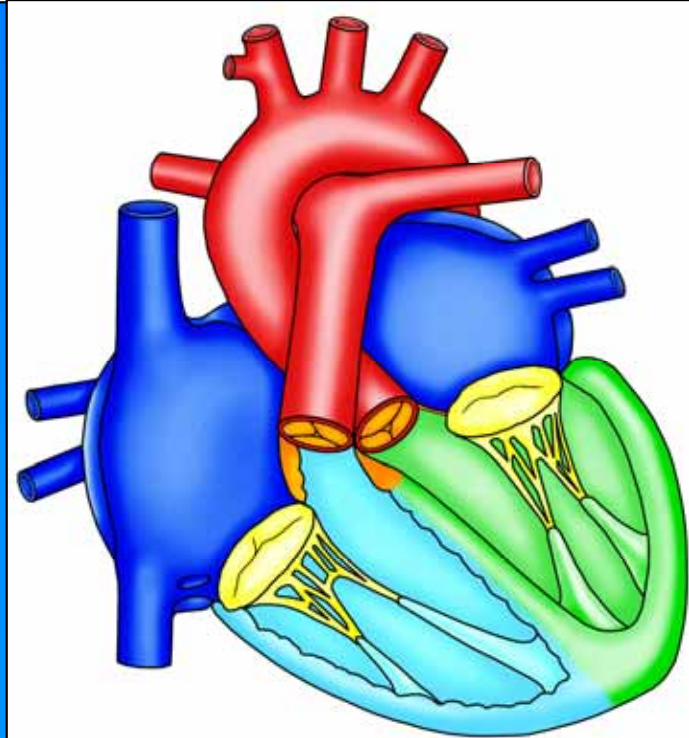
- Develop appropriate morphology
- Septate into right and left components
- Align/connect to appropriate chambers



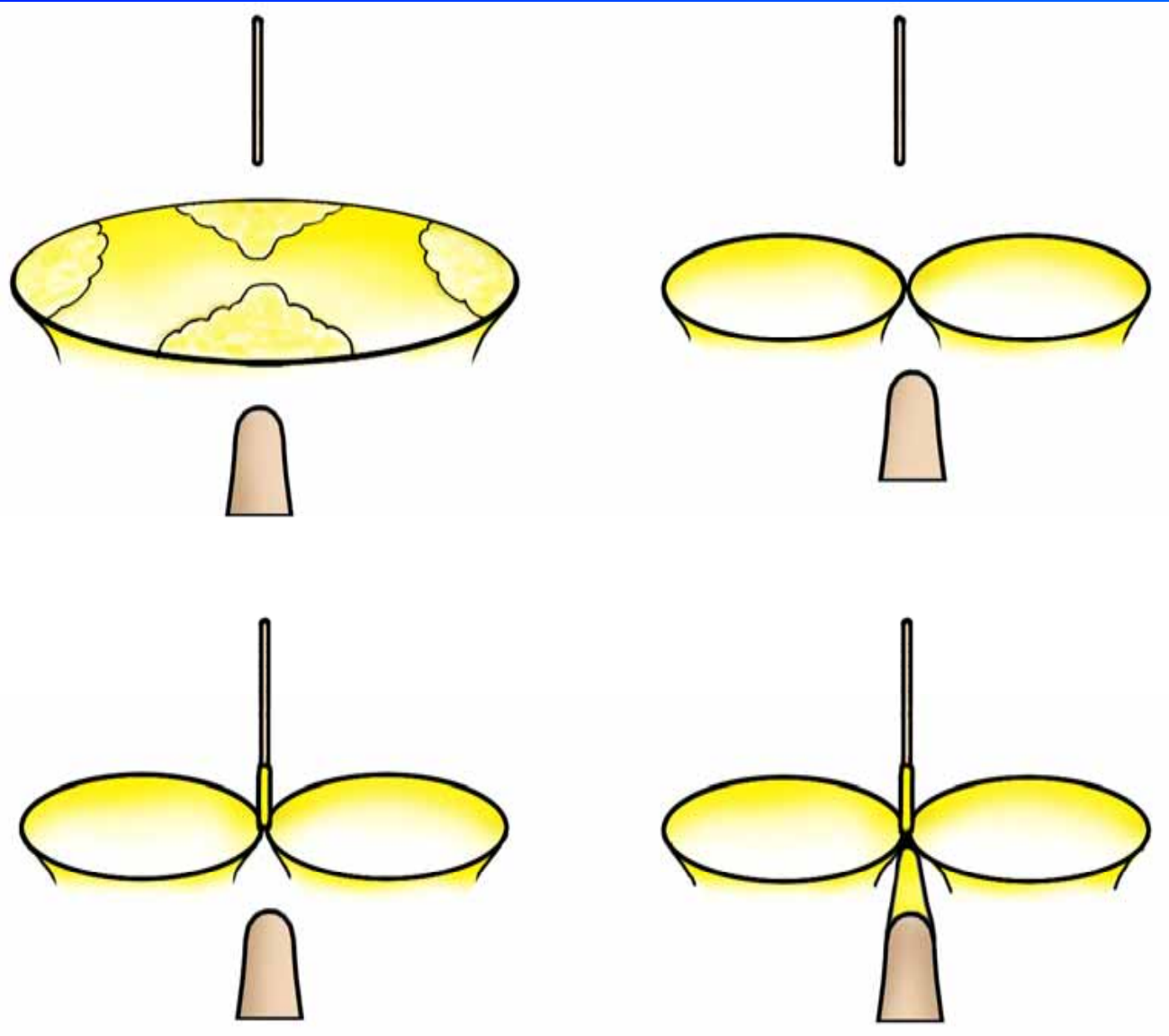
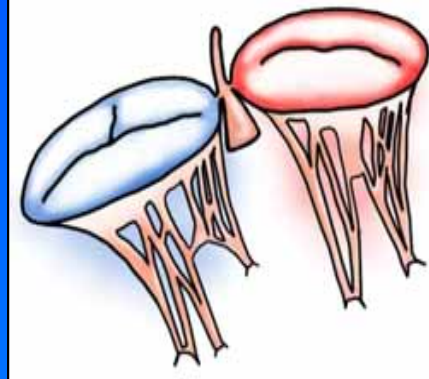


## AV canal

- Part of initial primary heart tube
- Initially aligned with LV



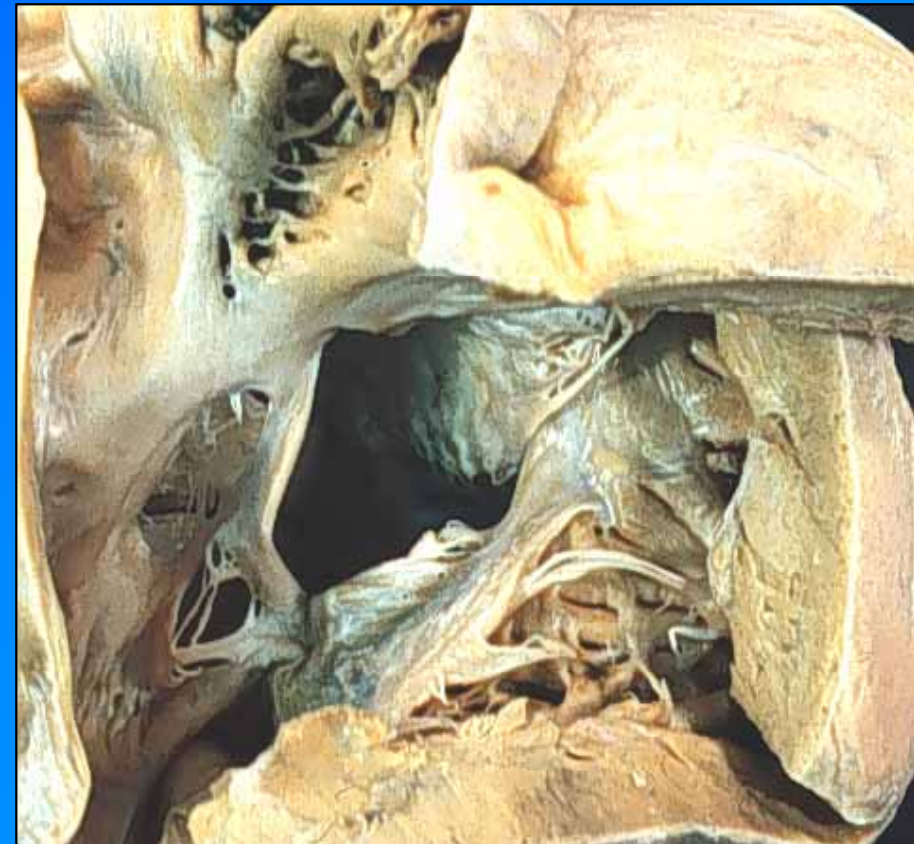
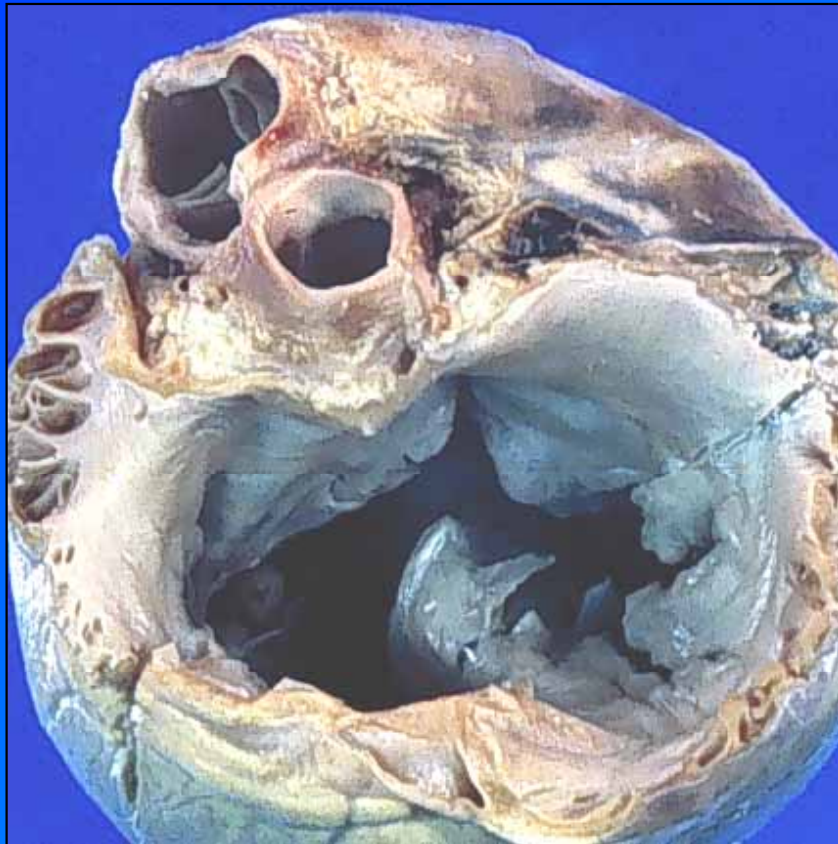
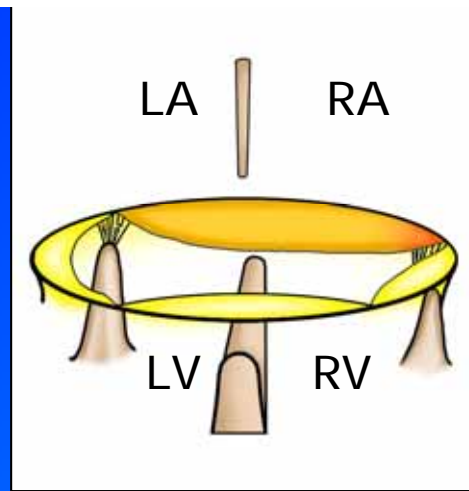
# AV CANAL SEPTATION



- Endocardial cushions partition AV canal into TV and MV orifices and help form valve leaflets.
- AV canal must connect to atrial septum
- AV canal must connect to ventricular septum

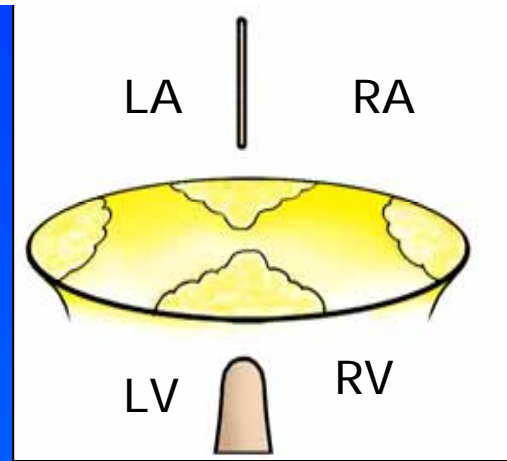
# Complete AV CANAL DEFECT

- No AV canal septum
- Common AV orifice
- Atrial & ventricular septa not connected



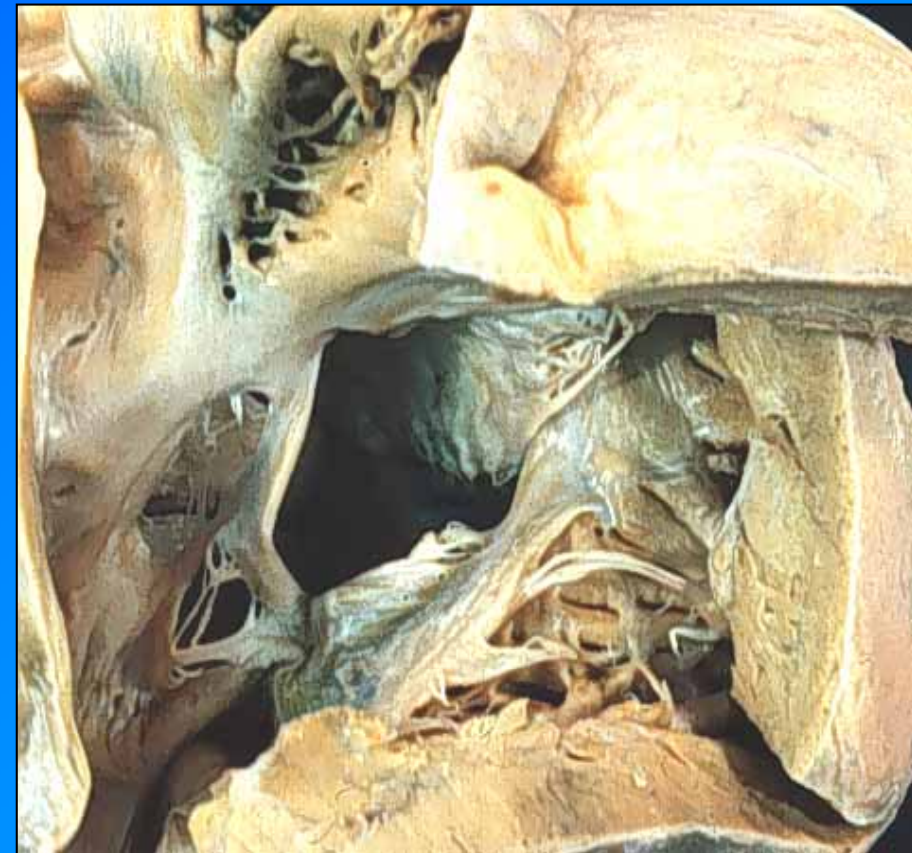
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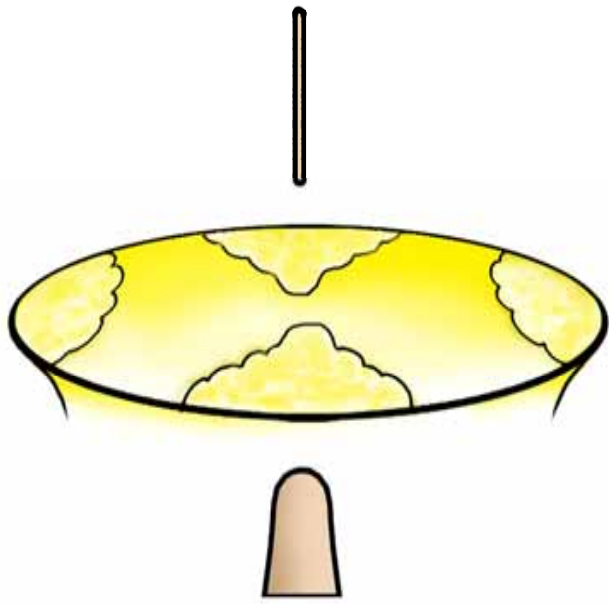


## Also known as:

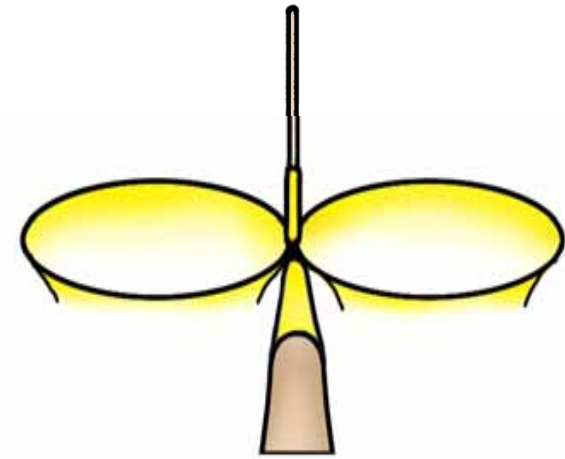
- Complete endocardial cushion defect (embryologic)
- Atrioventricular septal defect (morphologic)



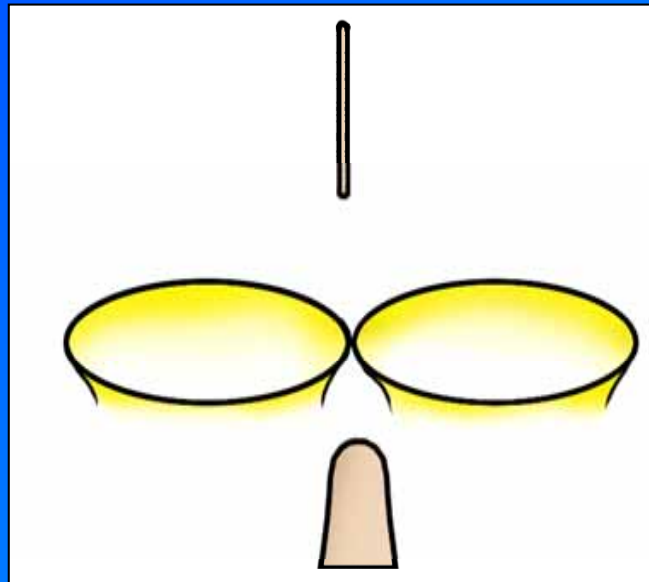
# Types of AV canal defects



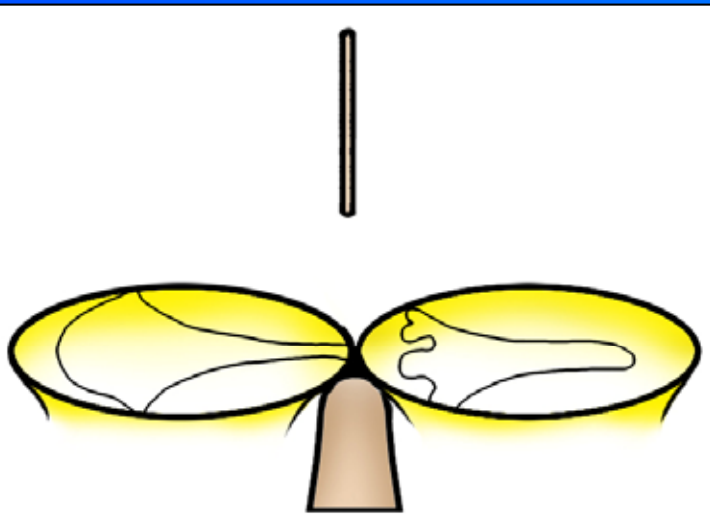
Transitional AV canal  
"ASD/small VSD"



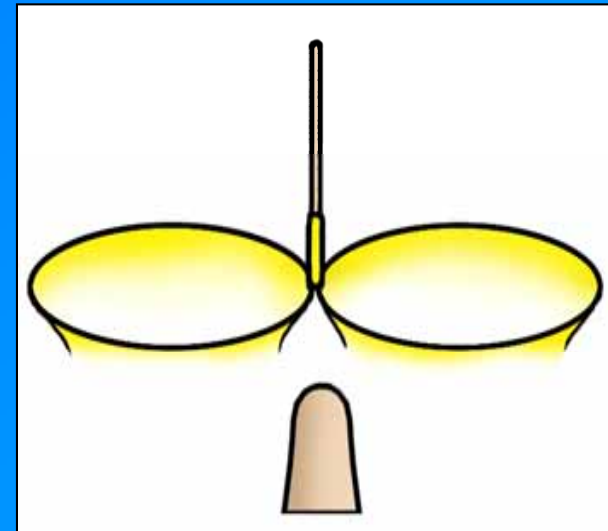
Partial AV canal  
"ASD"



Partial AV canal  
"VSD"



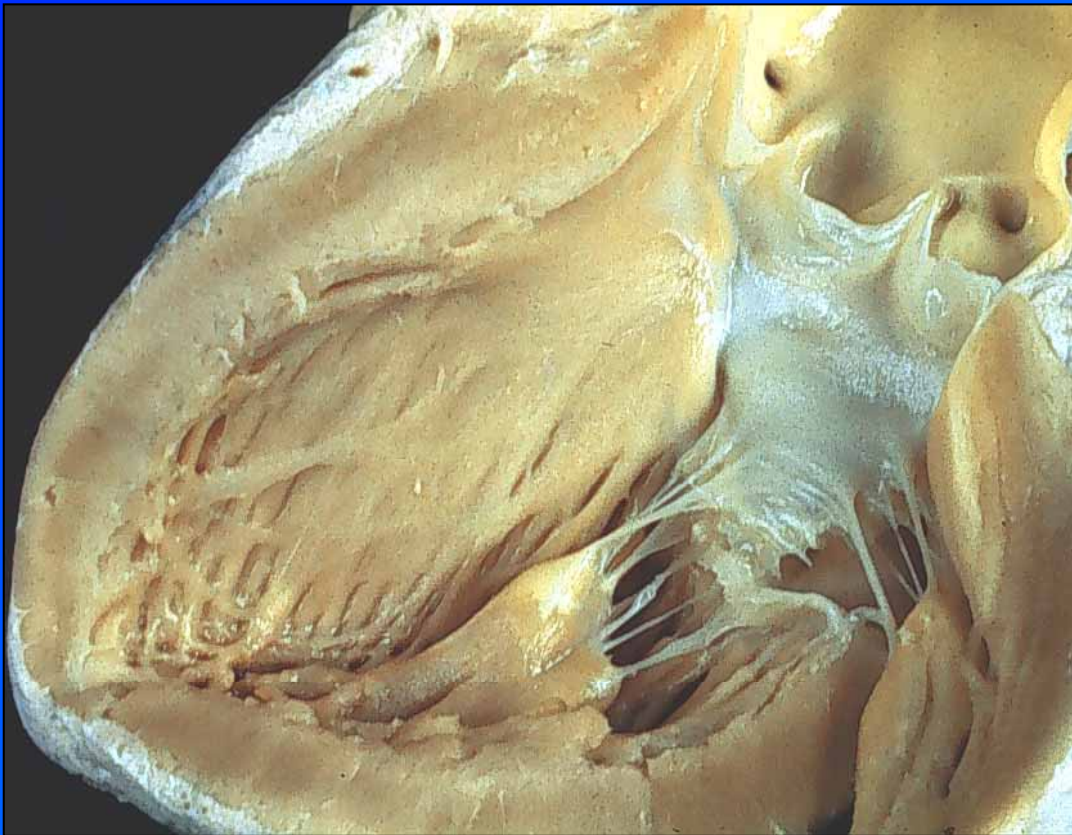
• Isolated cleft MV  
"rare"



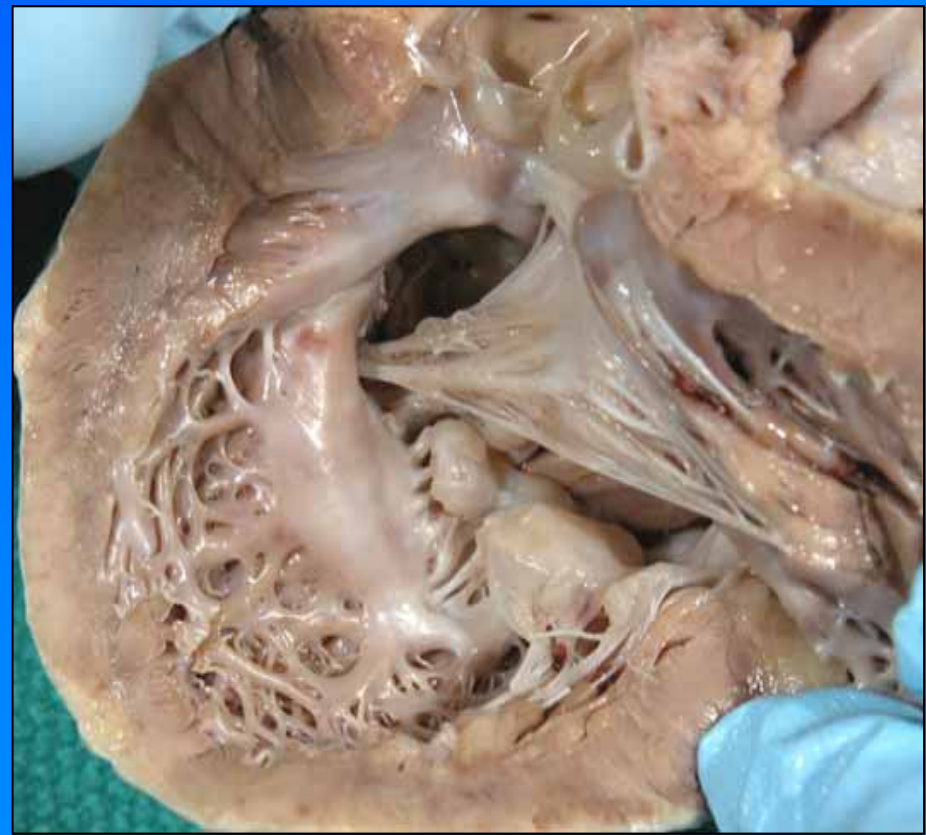
# AV CANAL DEFECTS-Anatomy

## Goose-neck deformity

Normal LVOT



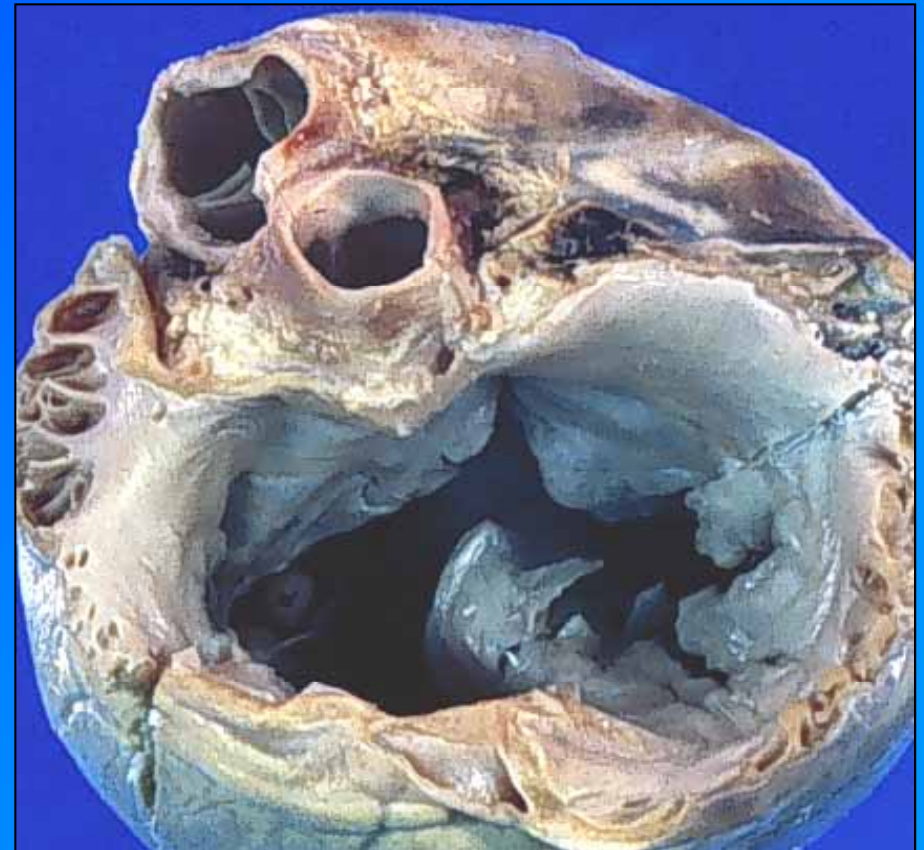
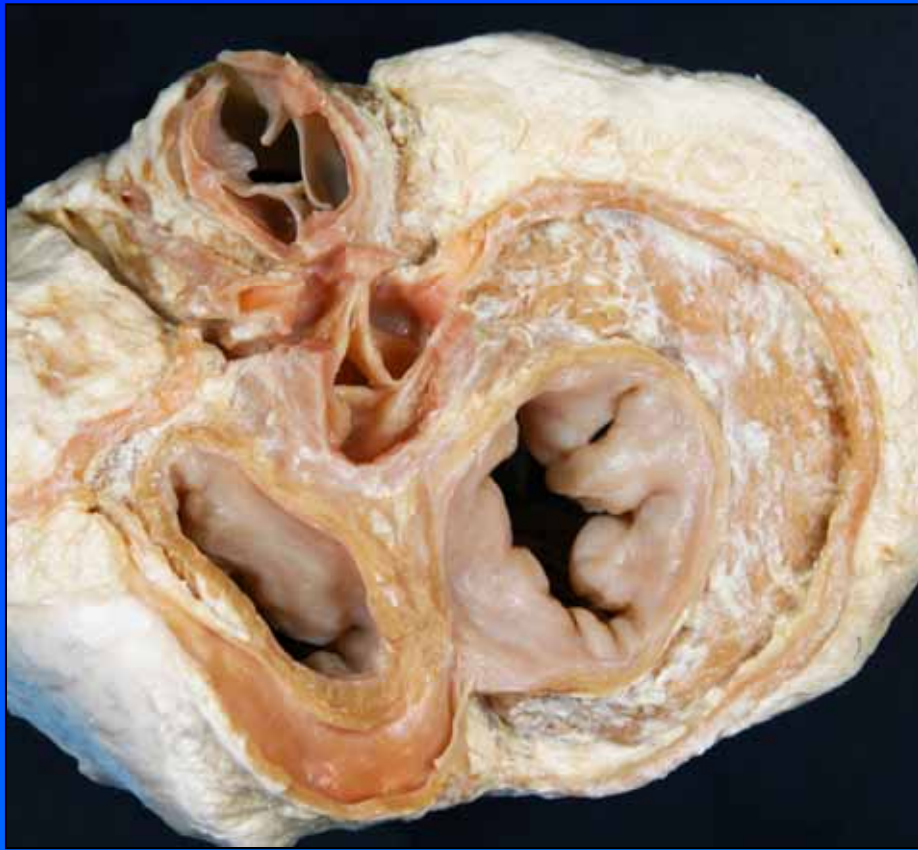
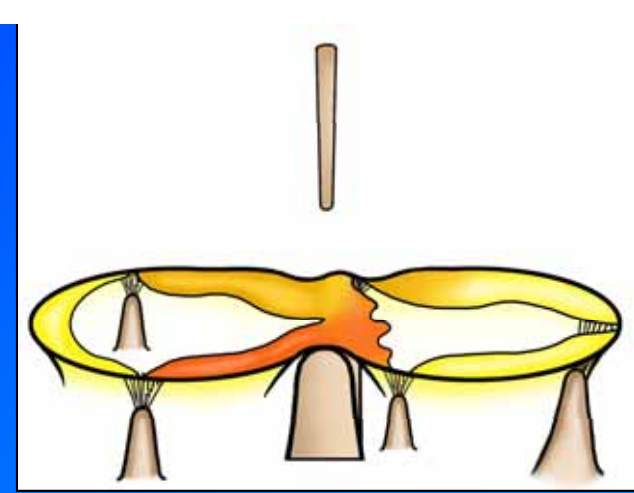
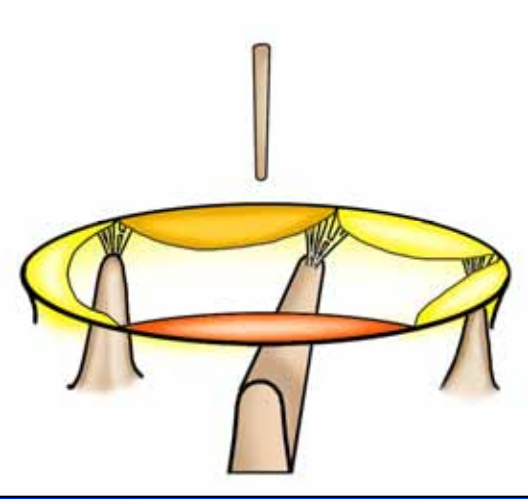
AV canal LVOT



- Elongated LV outflow tract.
- Deficient "scooped out" LV inflow tract.

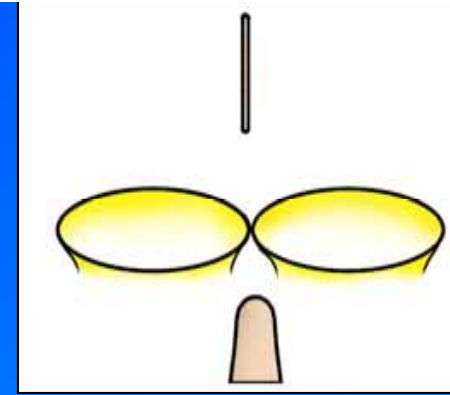
# ANATOMY

## Common AV junction



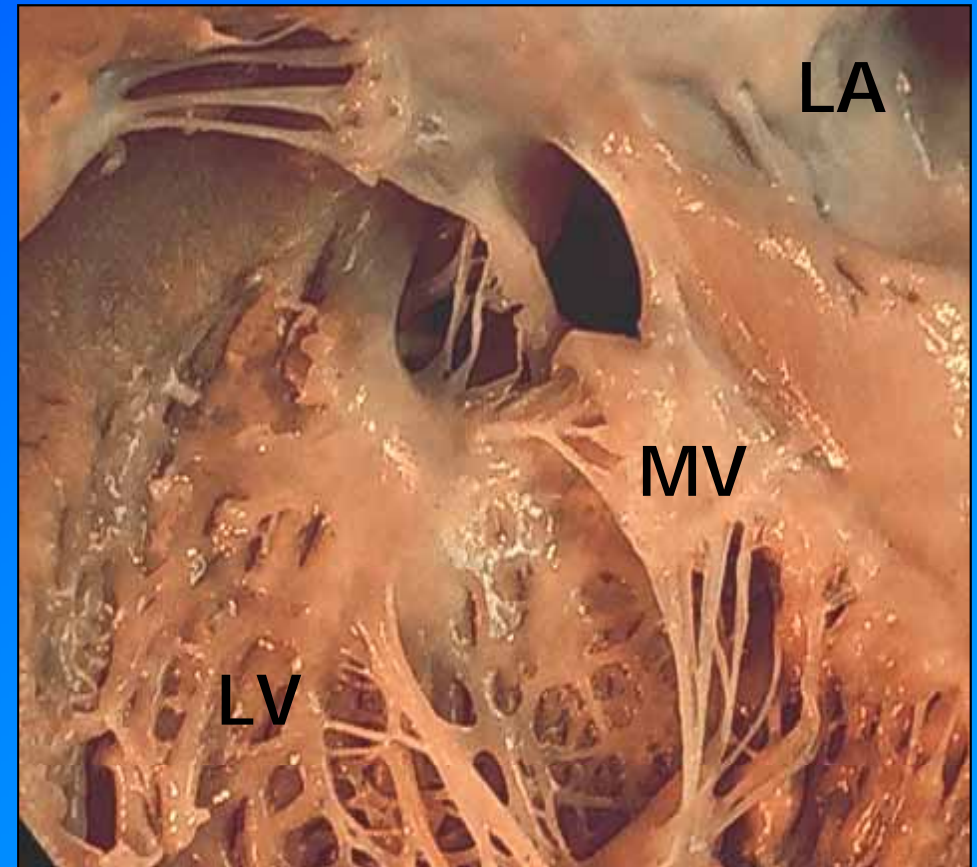
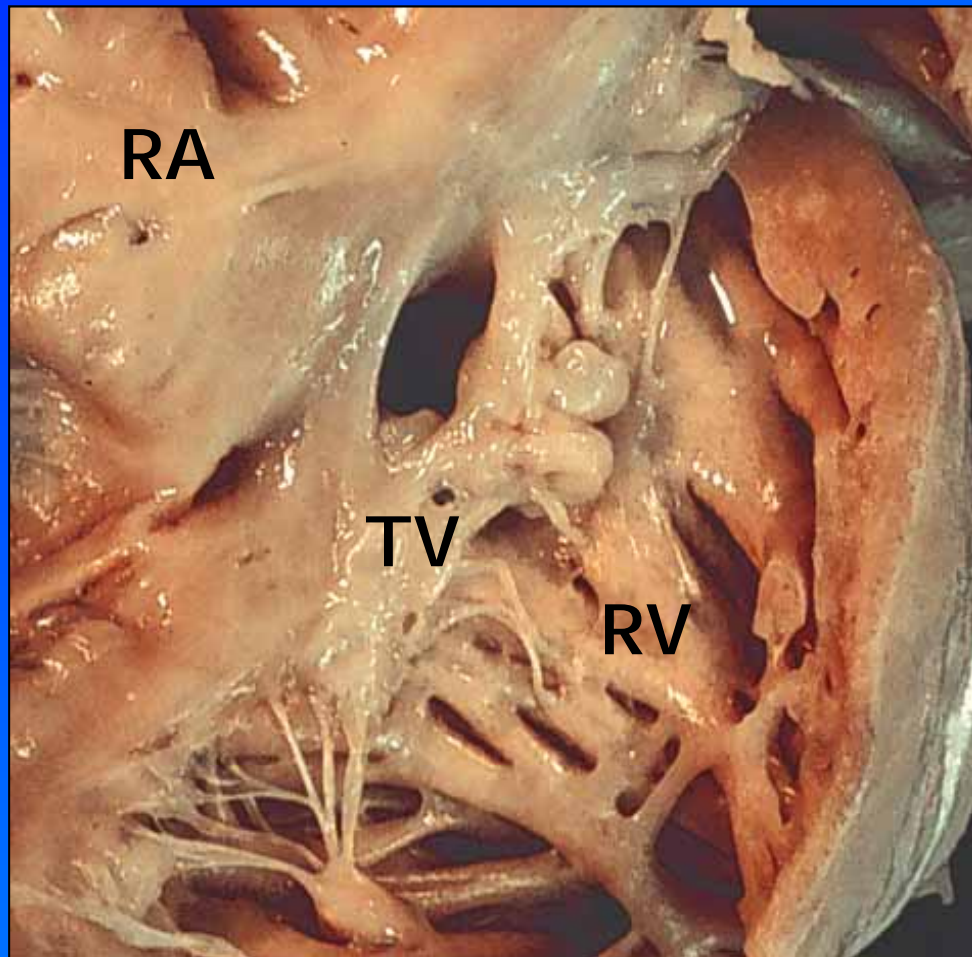
- Aorta not wedged between MV-TV valve rings
- Contributes to elongated LV outflow tract

# Transitional AV Canal Defect



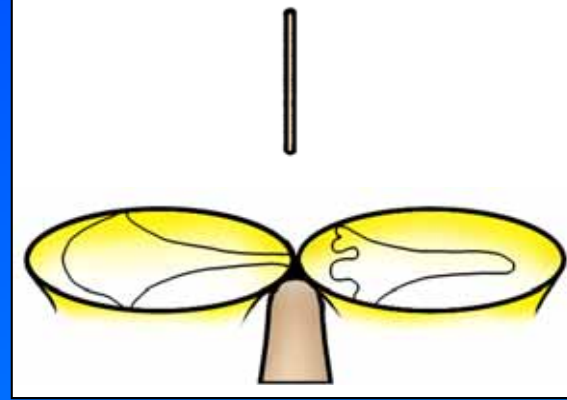
"Rare"

- Primum ASD
- Malformed TV and cleft MV.
- Small VSD

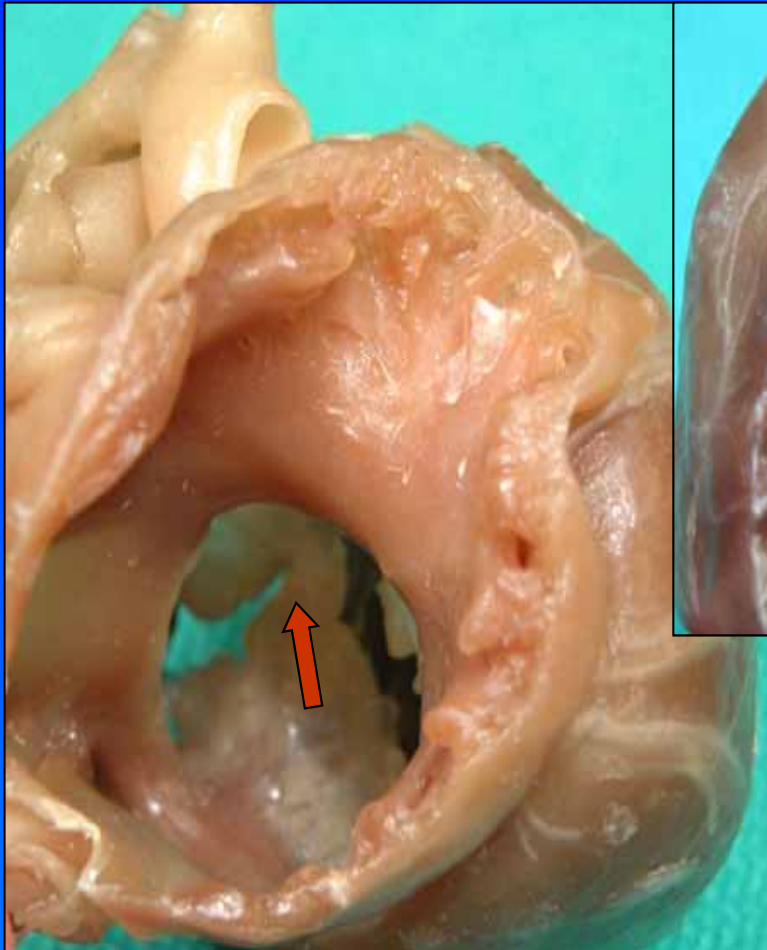
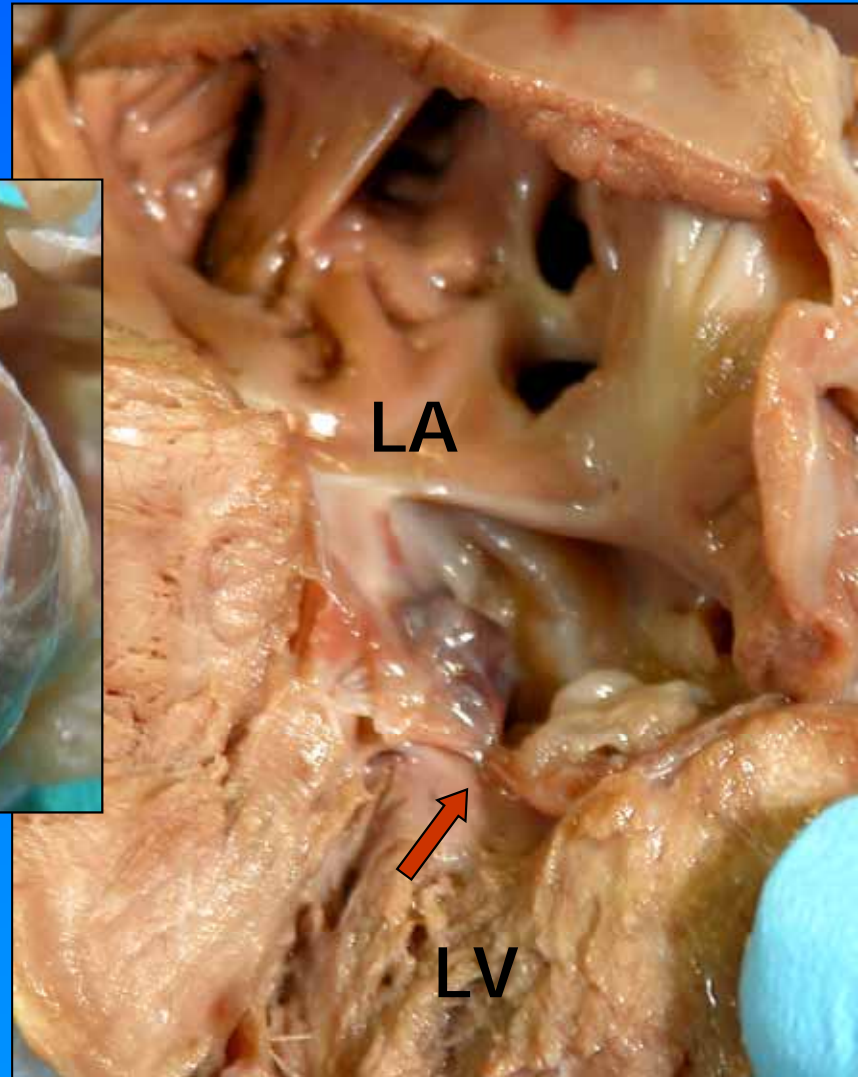


Cleft mitral valve

# Partial AV Canal Defect

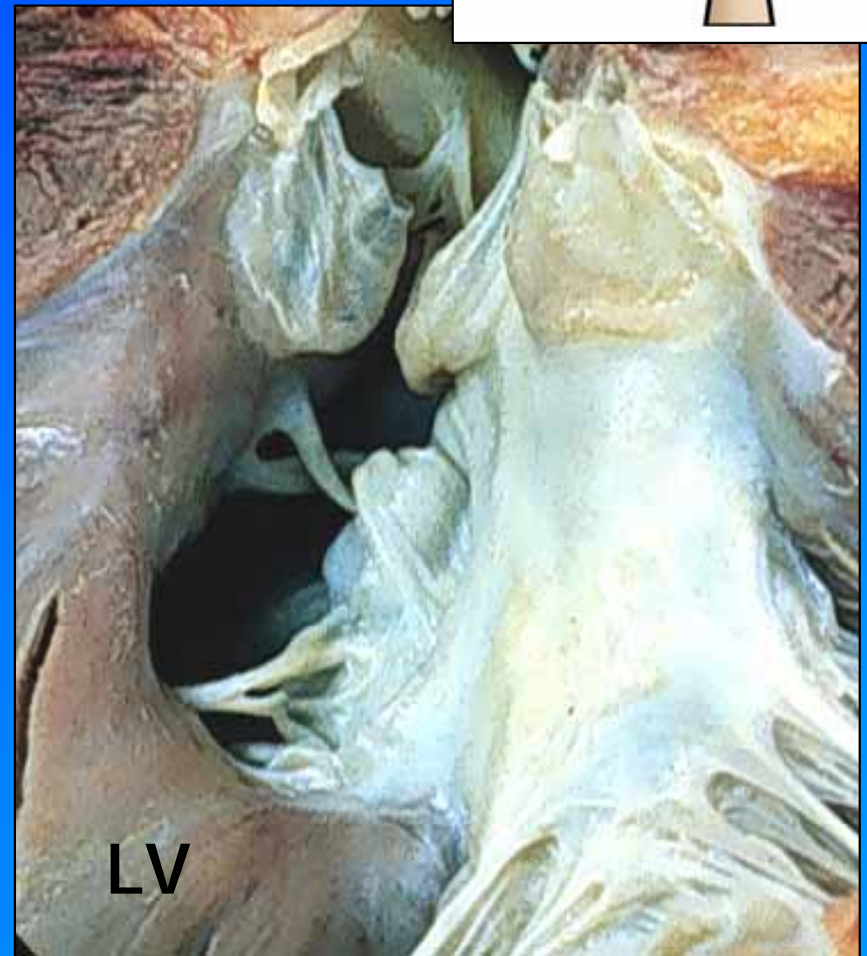
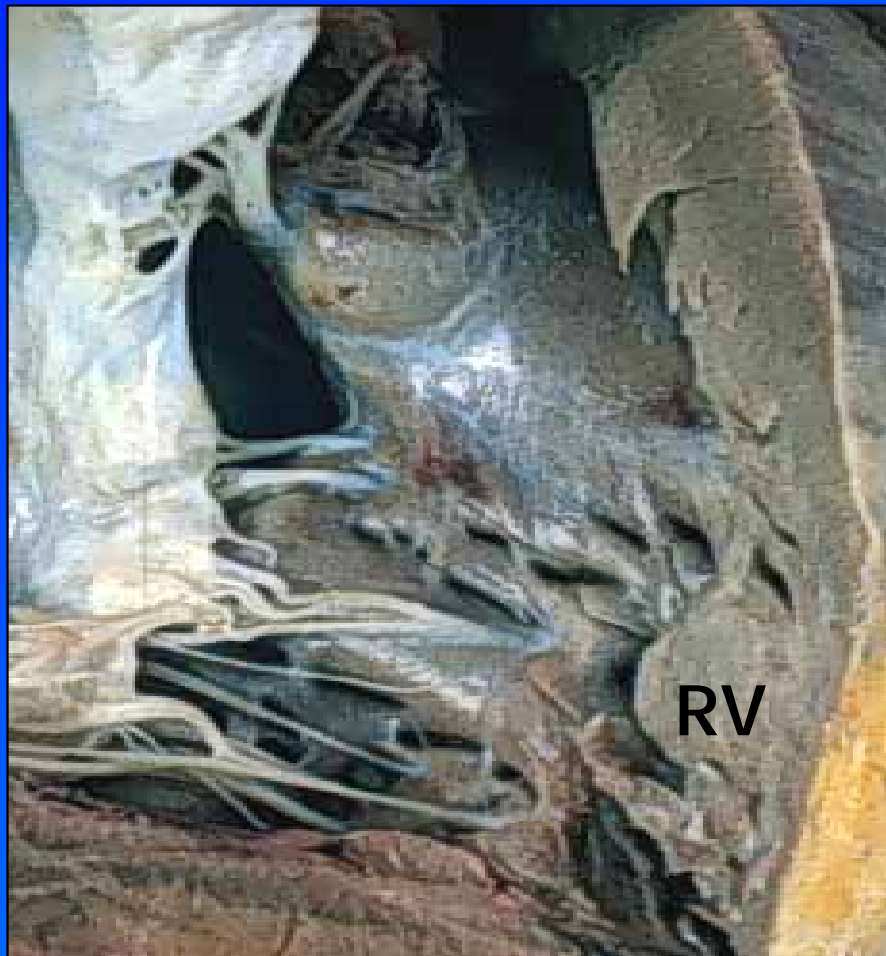
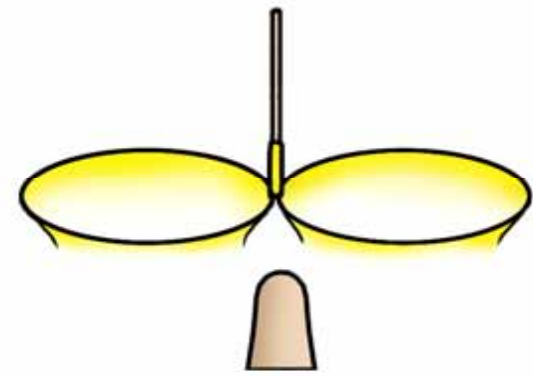


- Primum ASD
- Cleft MV
- Abnormal TV
- Deficient ventricular septum



# Partial AV Canal Defect

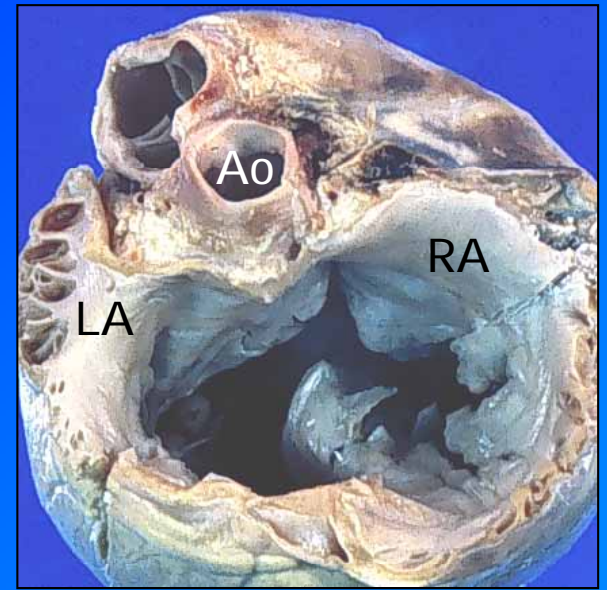
## VSD of AV canal type



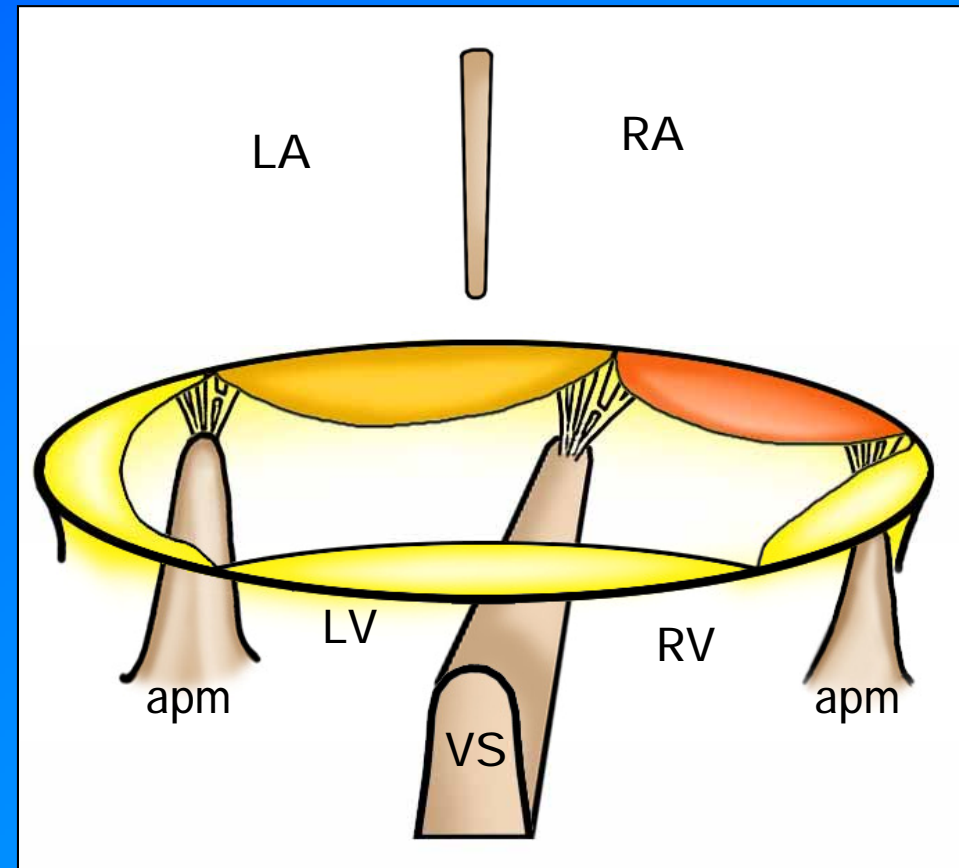
- VSD extends beneath TV septal leaflet
- Deficient muscle supporting MV & TV annuli

# Complete AV Canal Defect

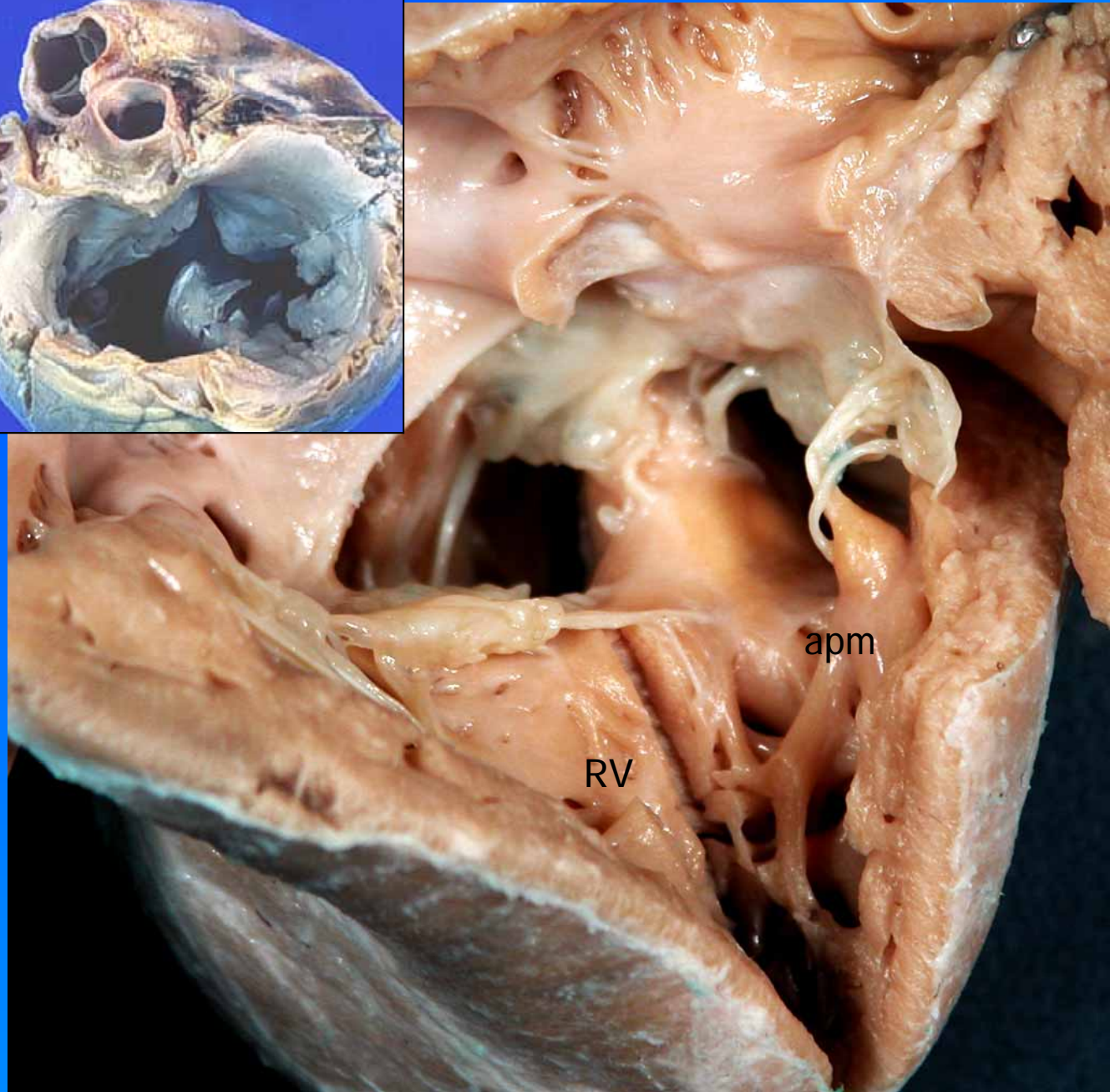
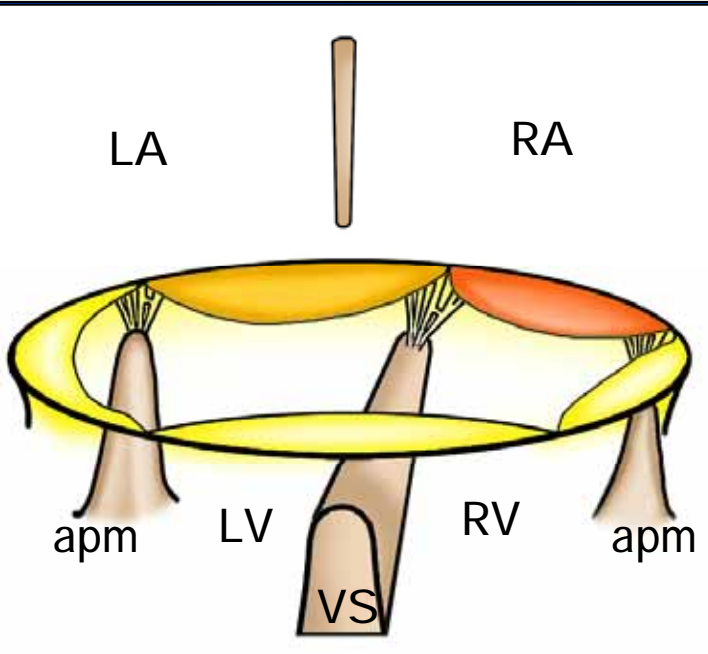
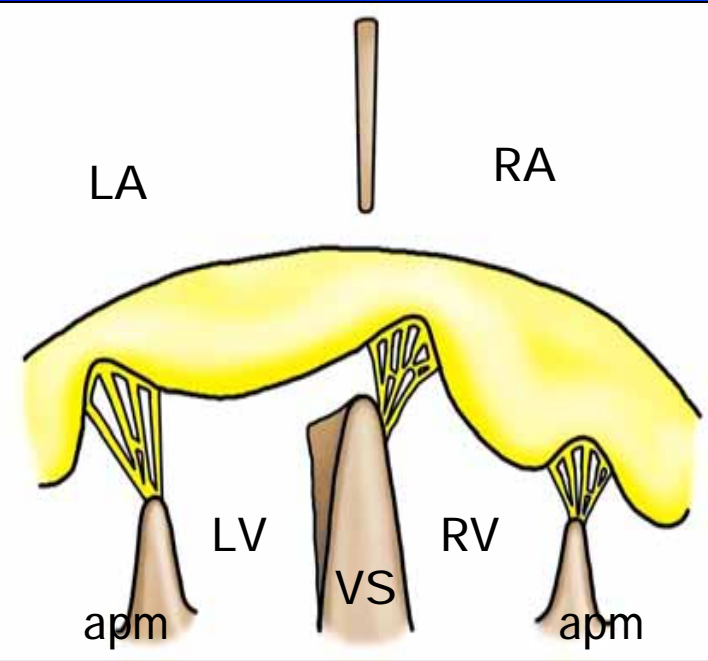
## Rastelli "A"



- *Anterior Bridging Leaflet: minimally crosses VS*
- *Chordae tendineae attach to crest of VS*
- *Normal RV anterior leaflet*

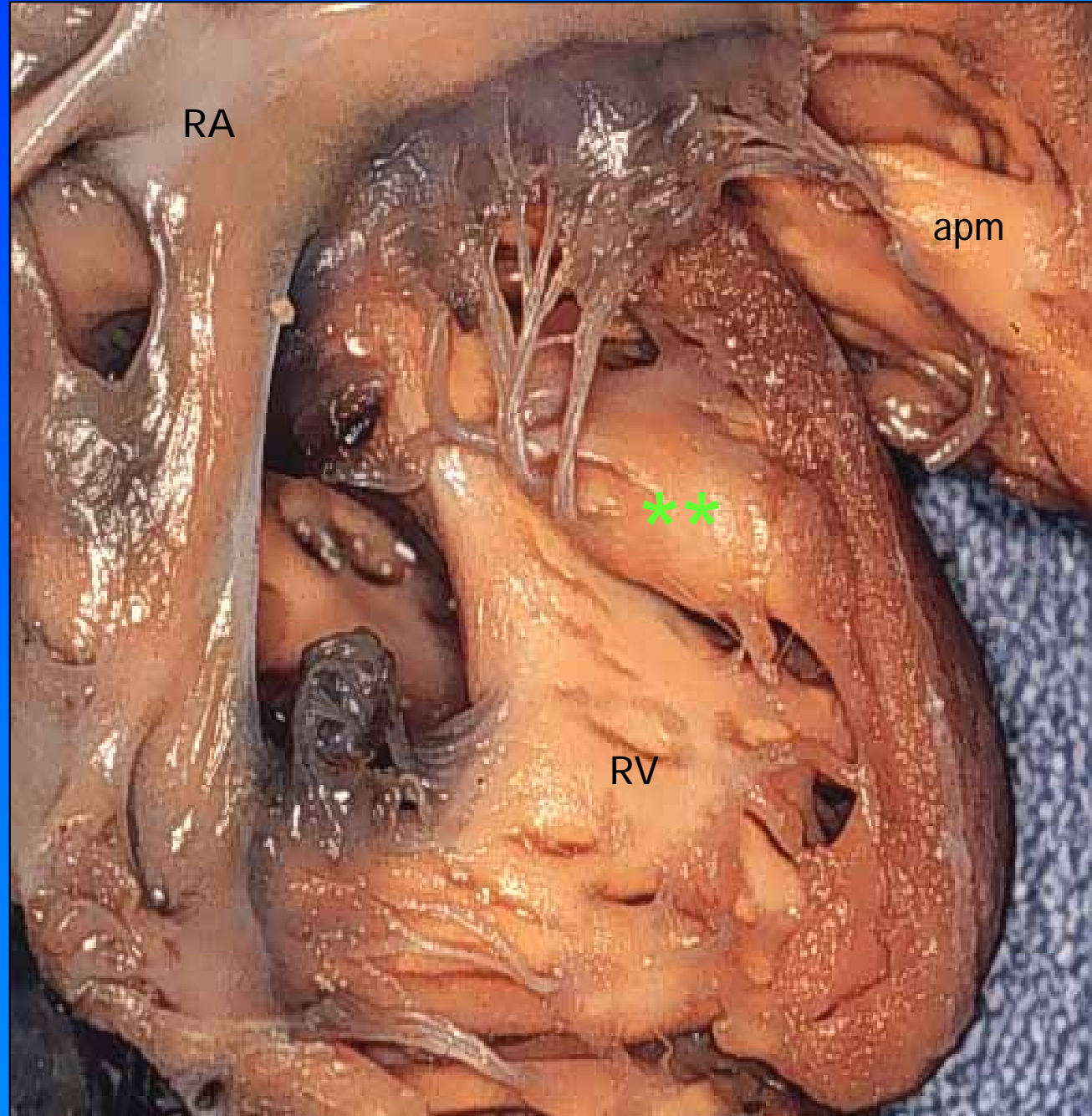
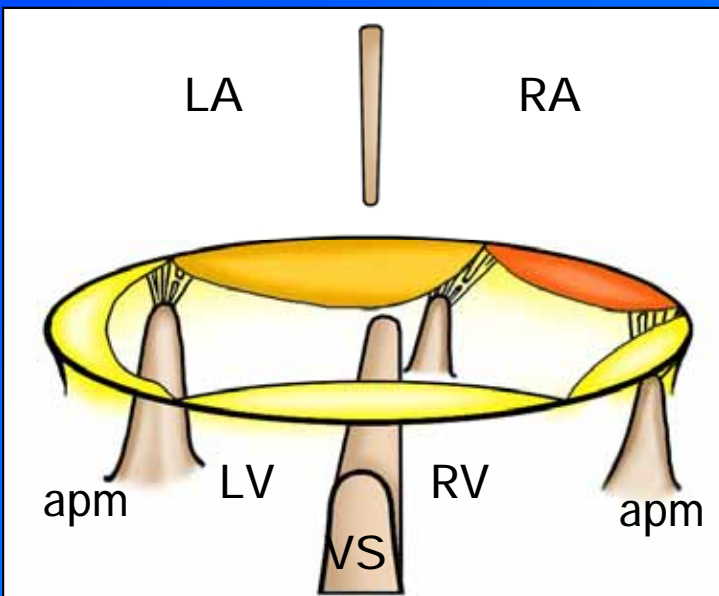
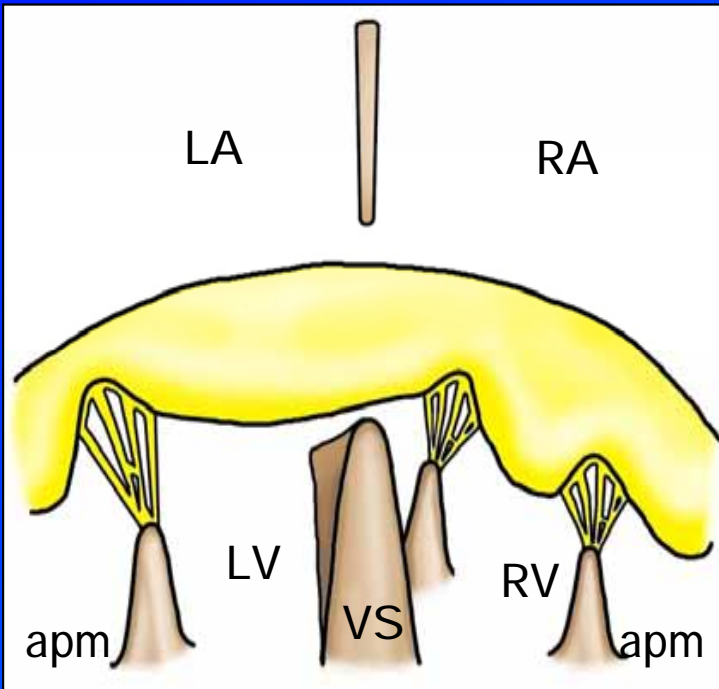


# Rastelli "A"



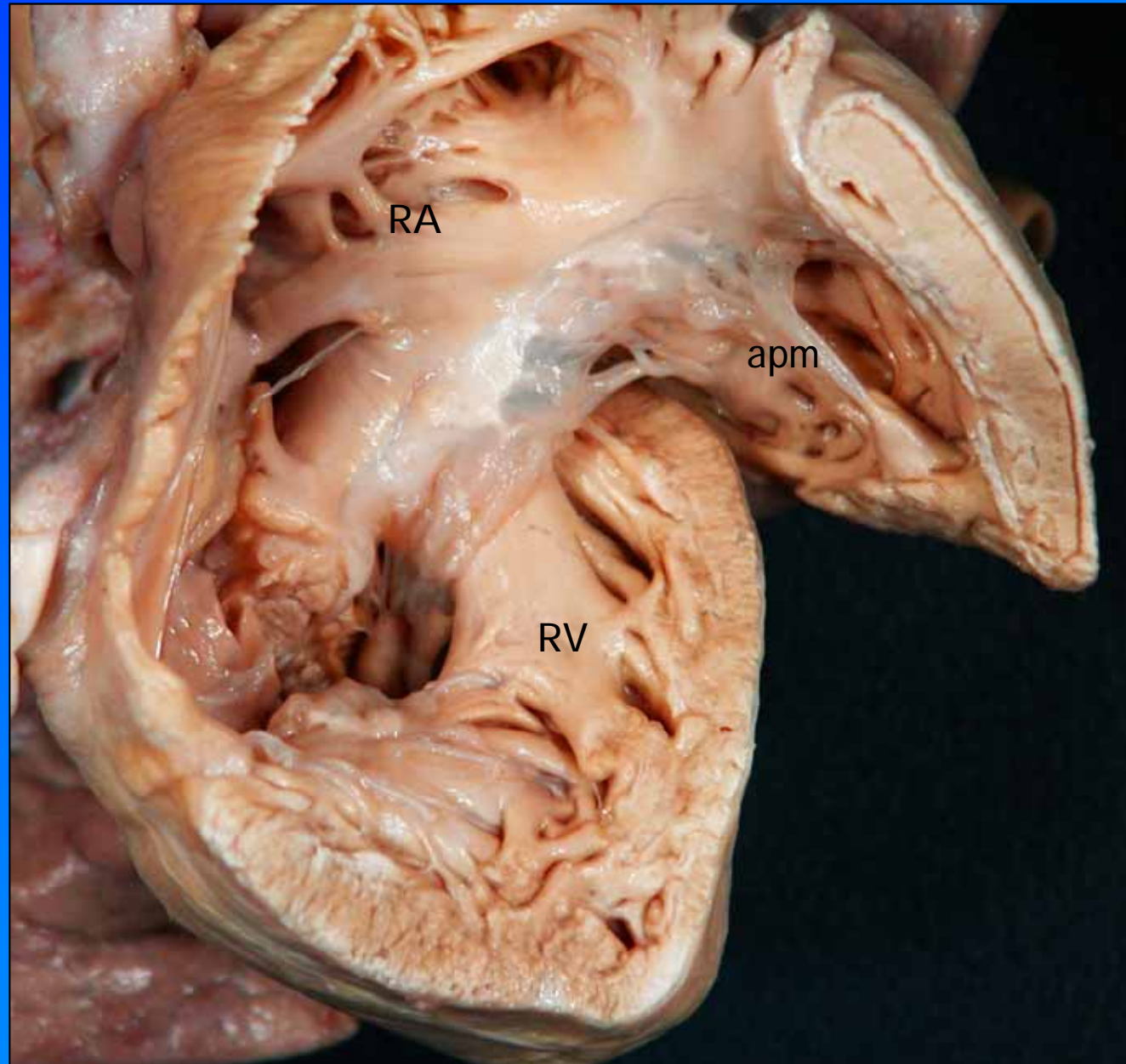
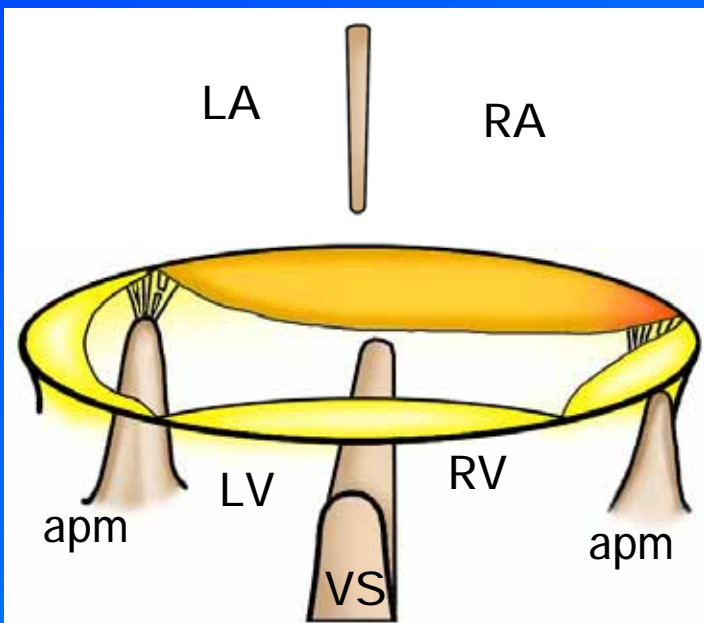
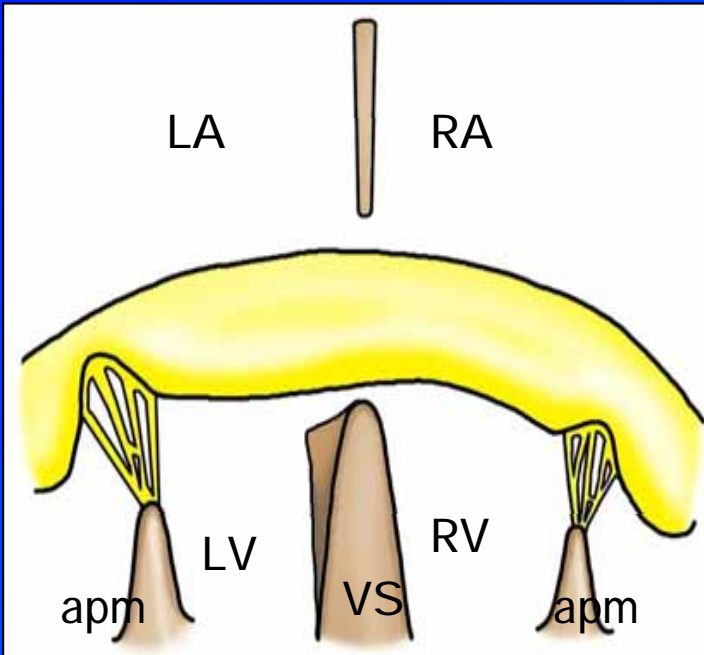
# Rastelli "B"

*Anterior bridging leaflet connects to anomalous RV pap muscle (\*)*

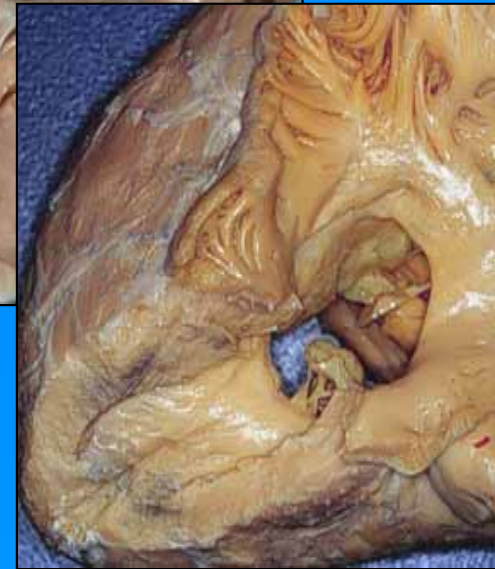
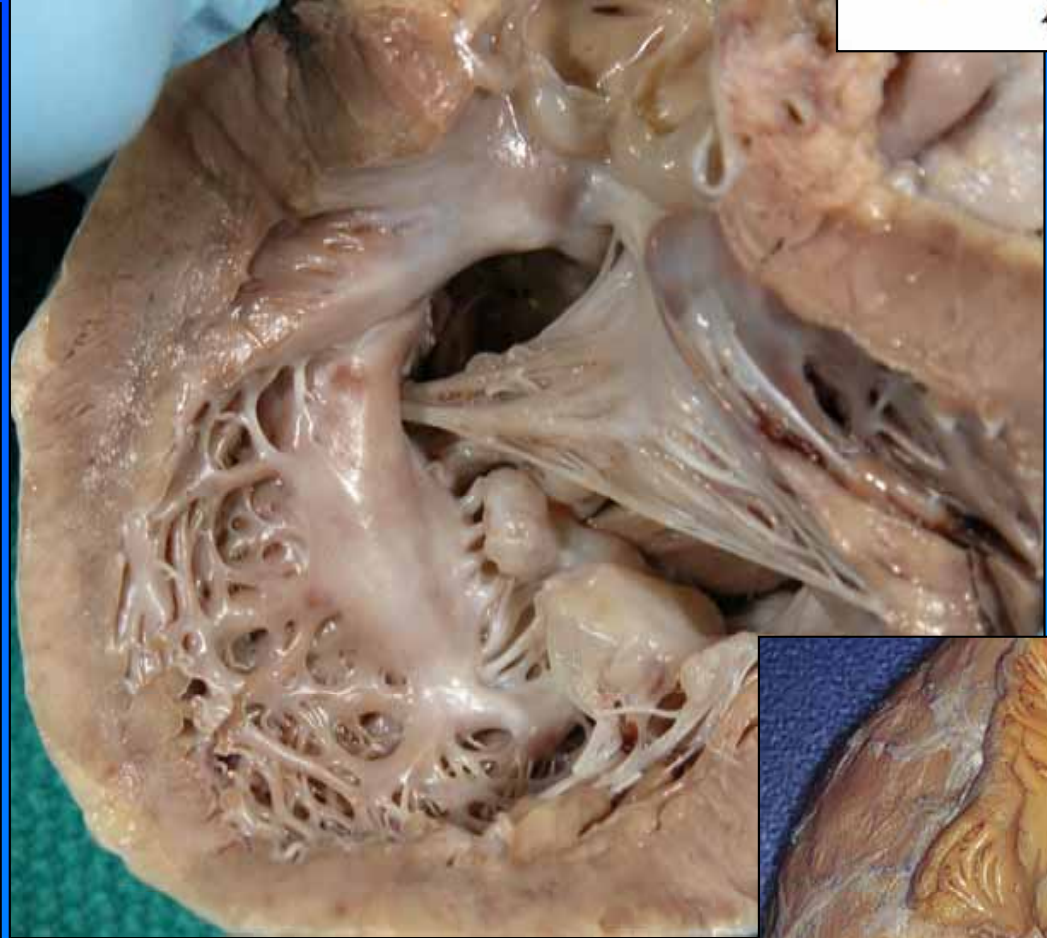
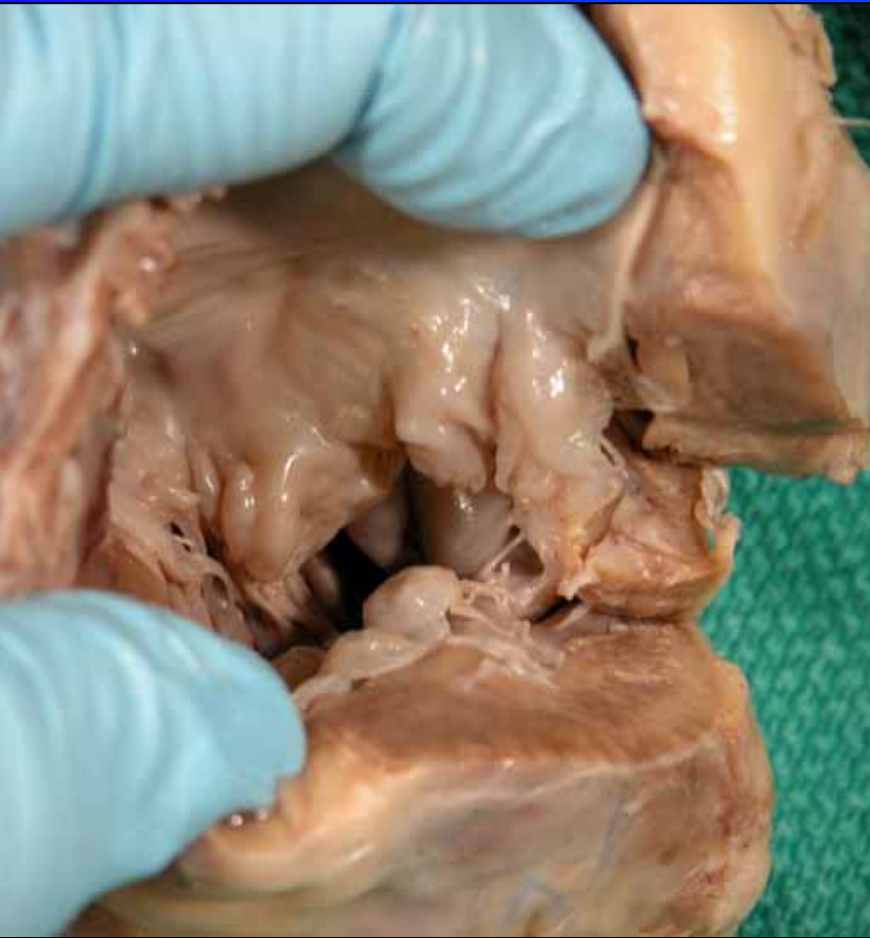
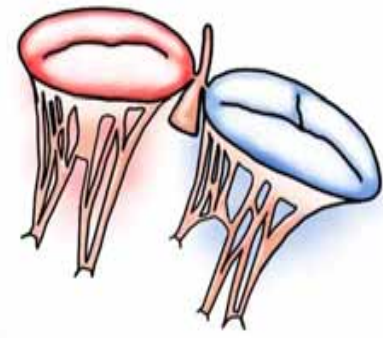


# Rastelli "C"

- *Large anterior bridging leaflet*
- *Virtually absent RV anterior leaflet*



# Unbalanced AV CANAL



- LV dominant AV canal
- Hypoplastic RV inlet
- Dysplastic AV valve leaflets

- RV Dominant

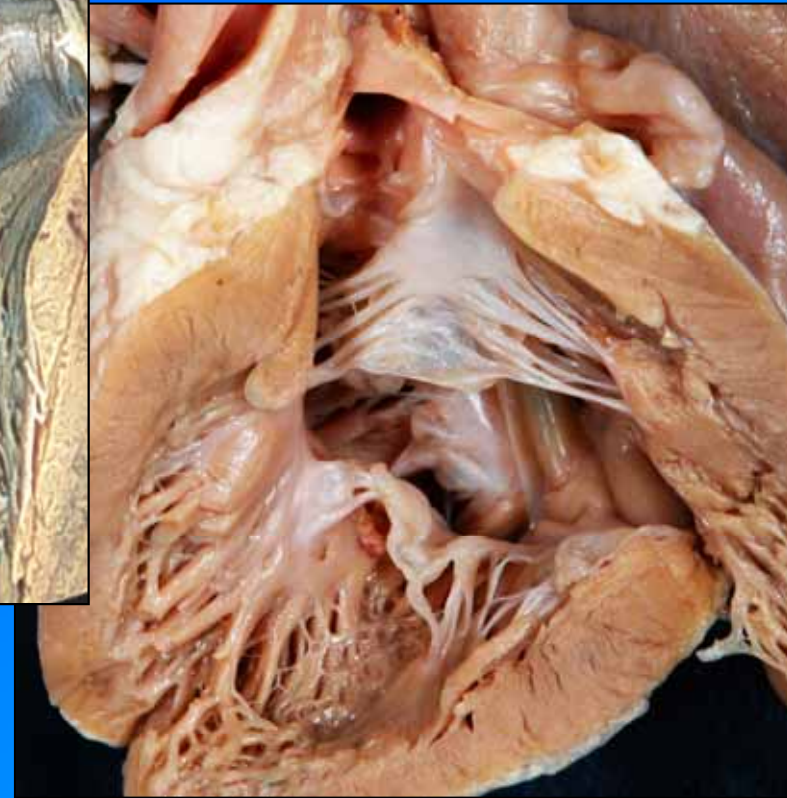
# LVOT obstruction

Primum ASD



Rastelli "C"

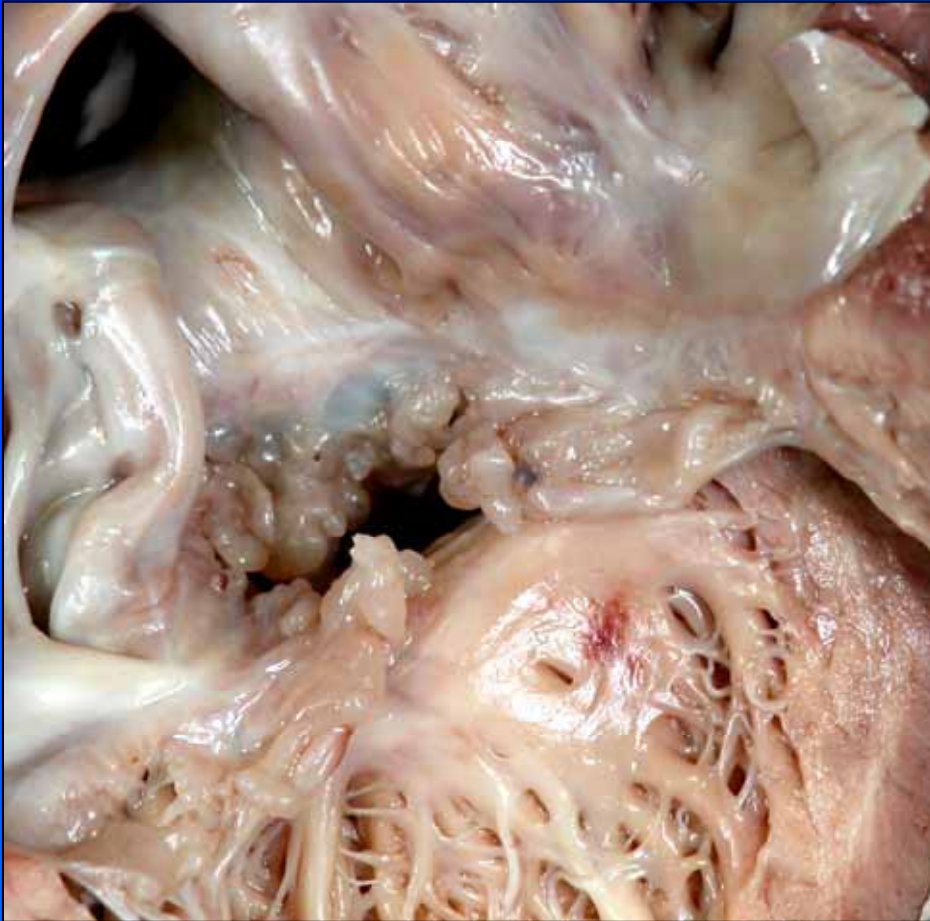
Rastelli "A"



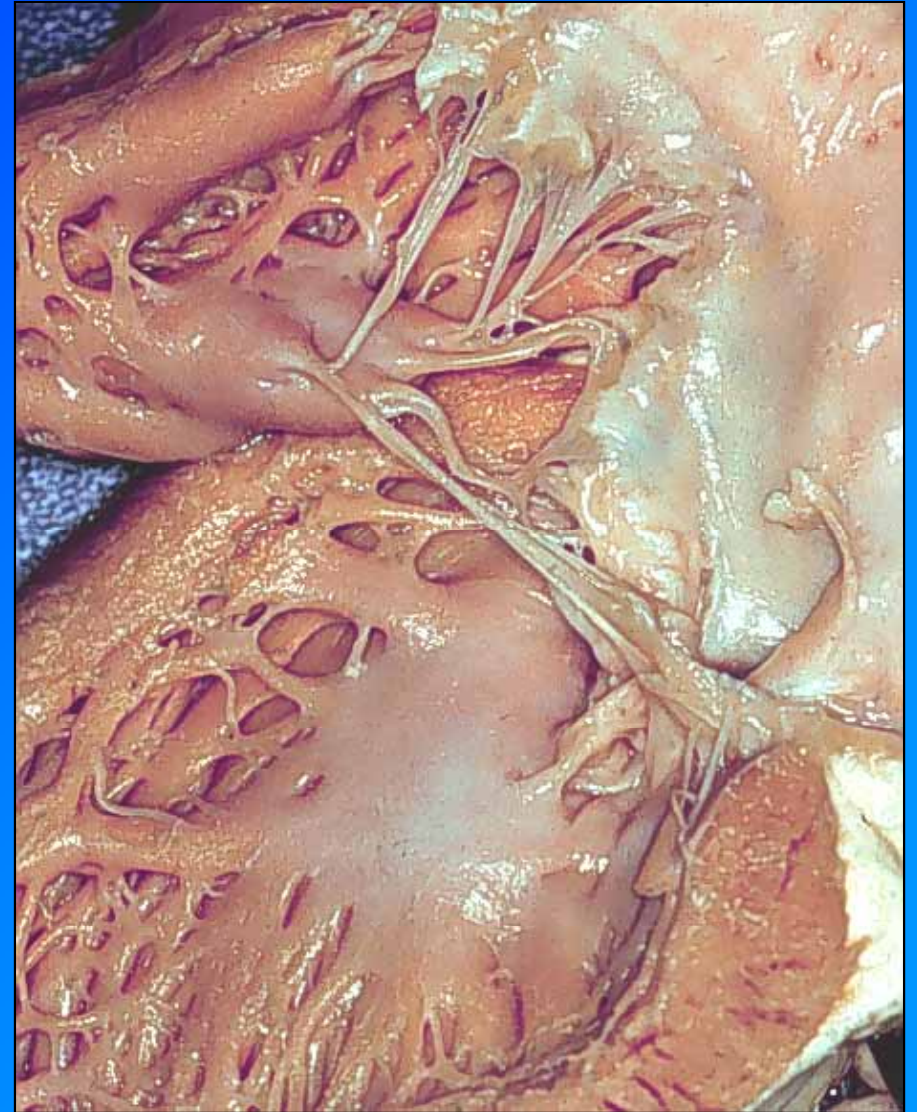
- Narrowed LVOT with tethered valve leaflet/chordae
- Partial AV canal (Primum ASD) highest risk
- Rastelli "C" (unattached bridging leaflet) low risk

# AV CANAL: Associated Issues

## Inflow tracts

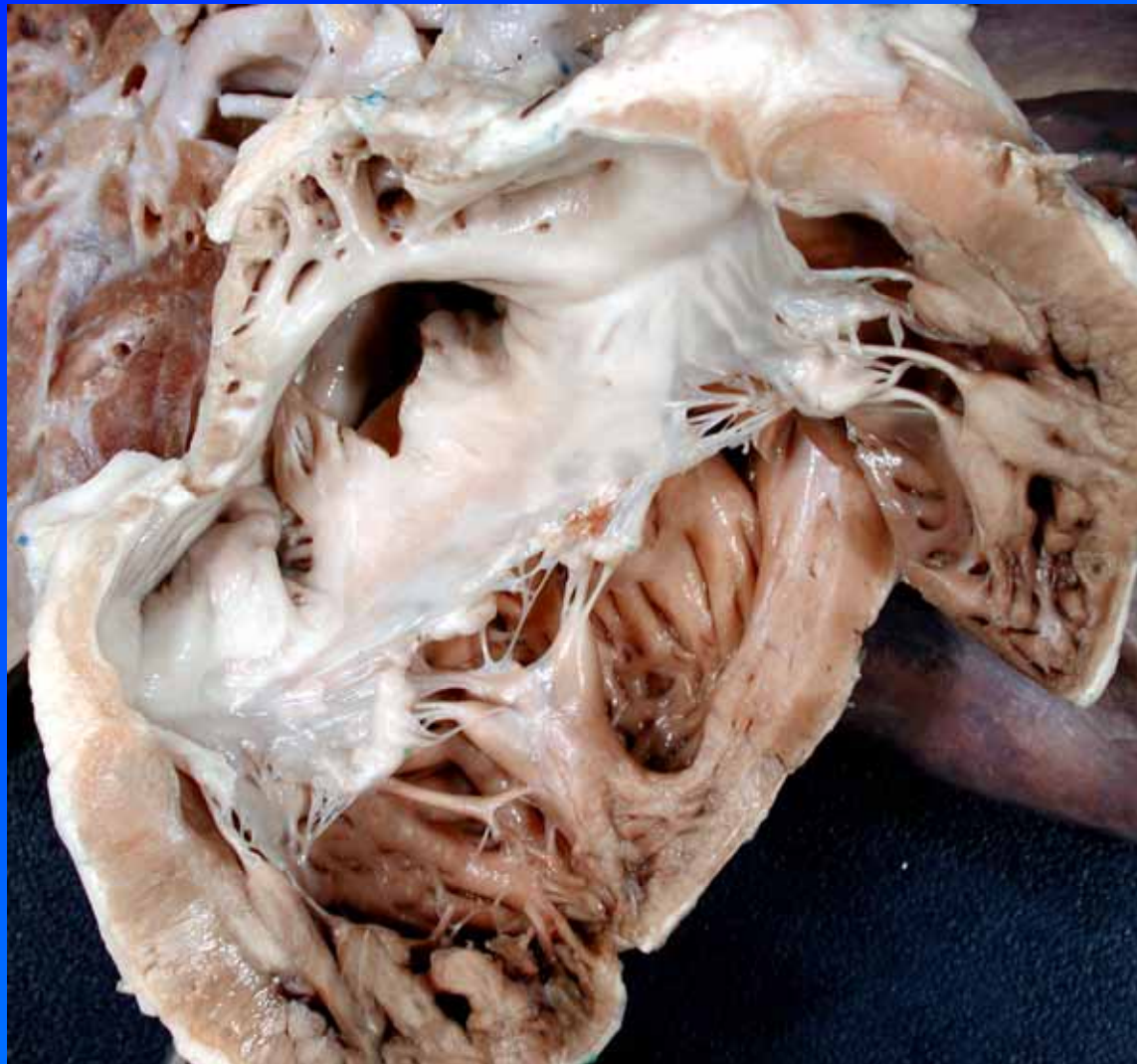
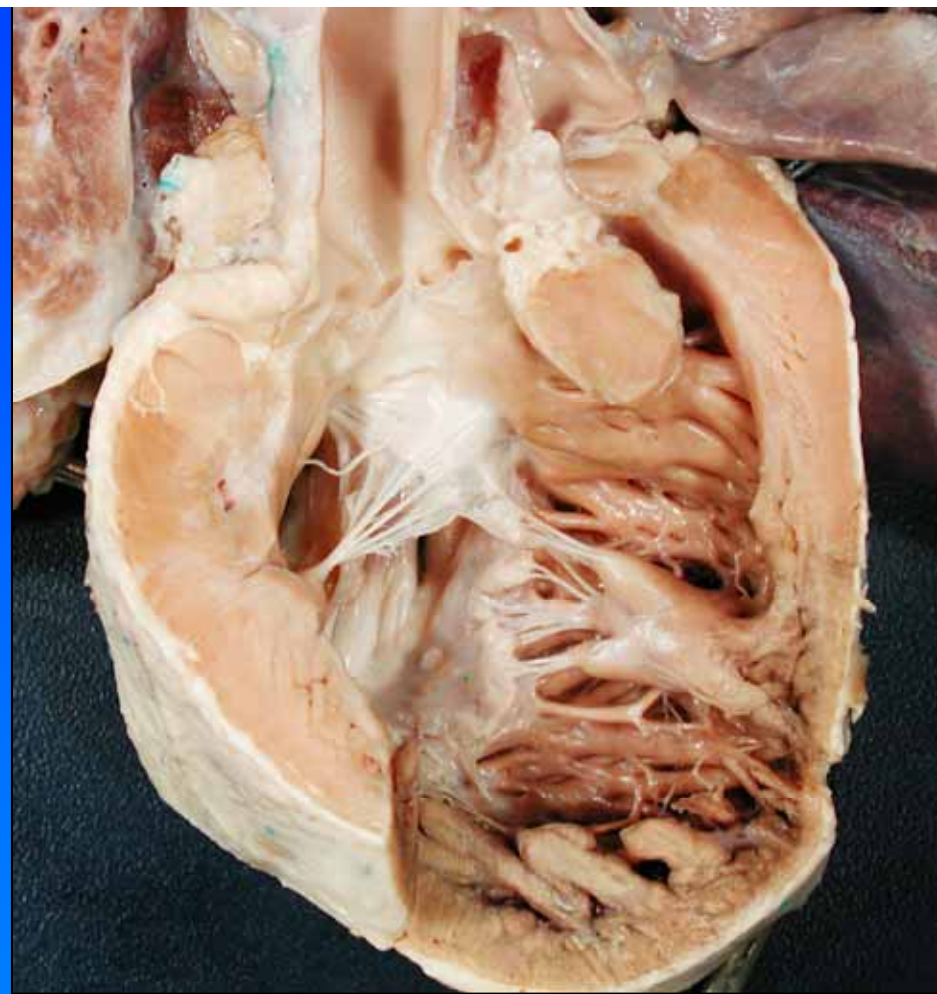


Dysplastic leaflets

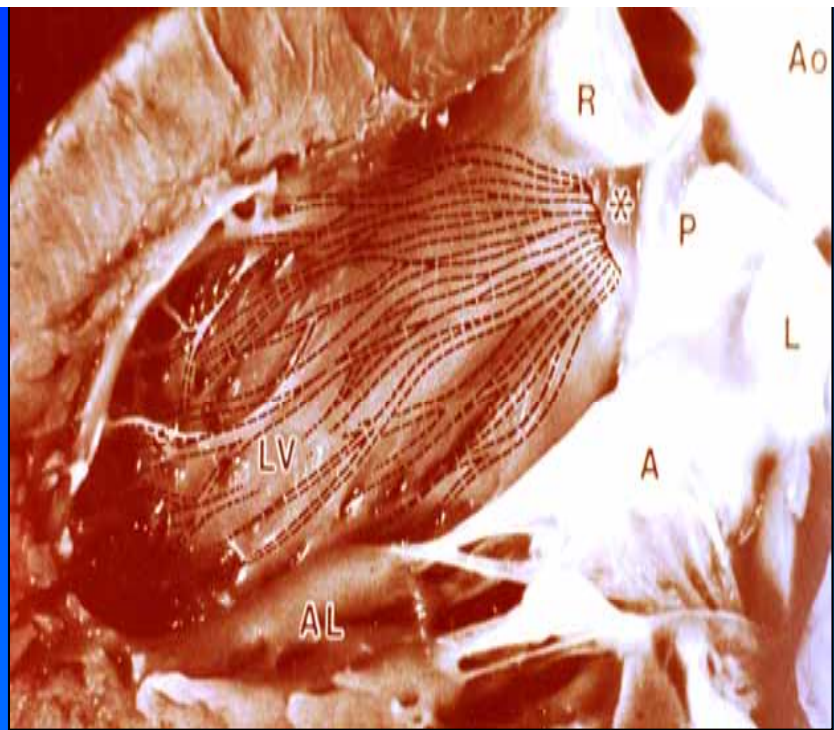
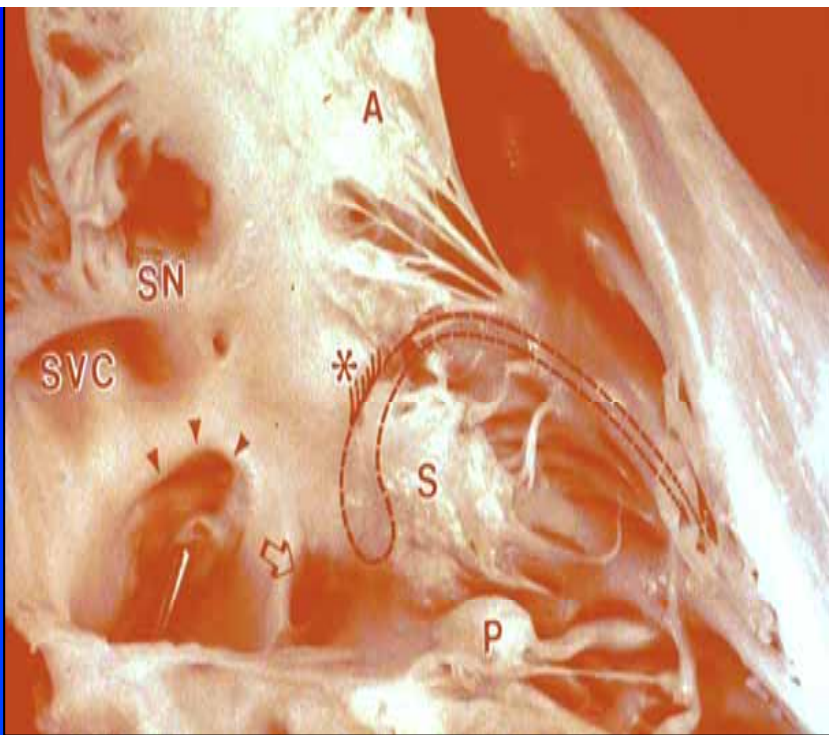


Single LV papillary muscle

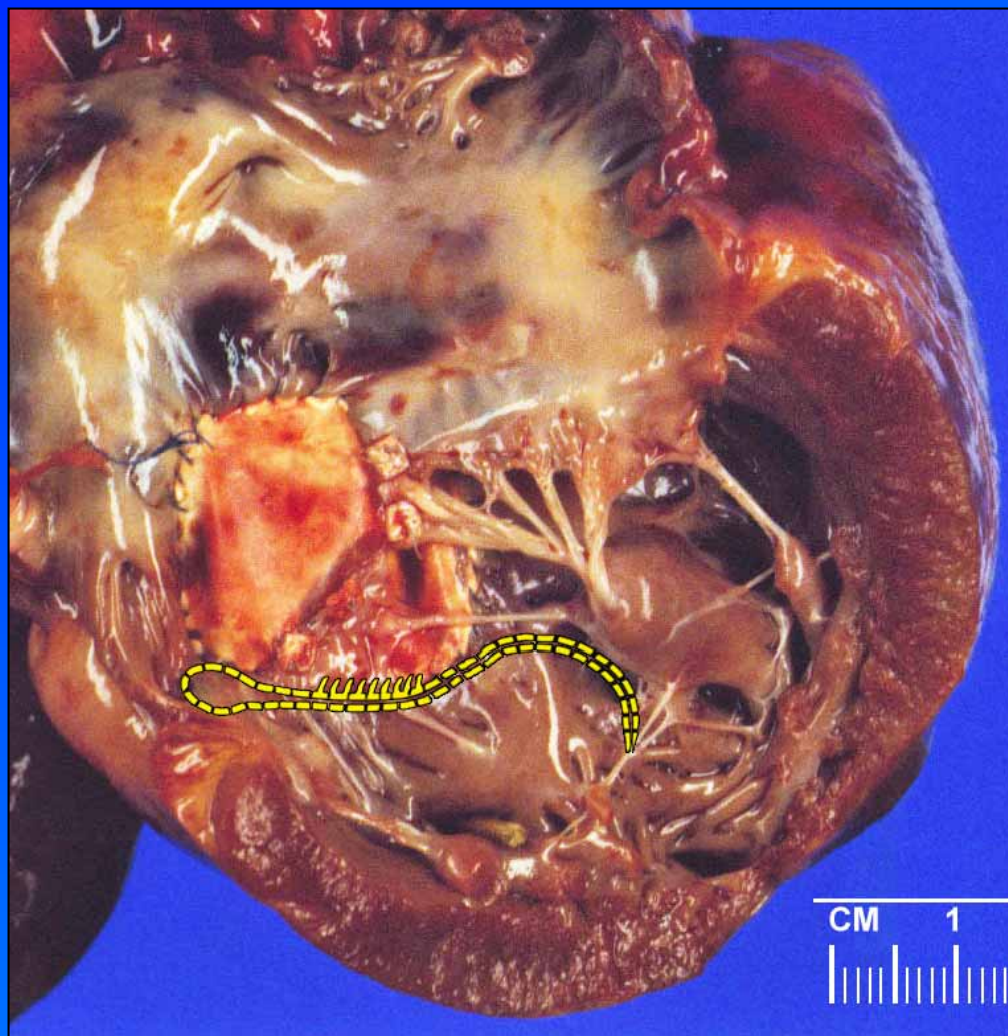
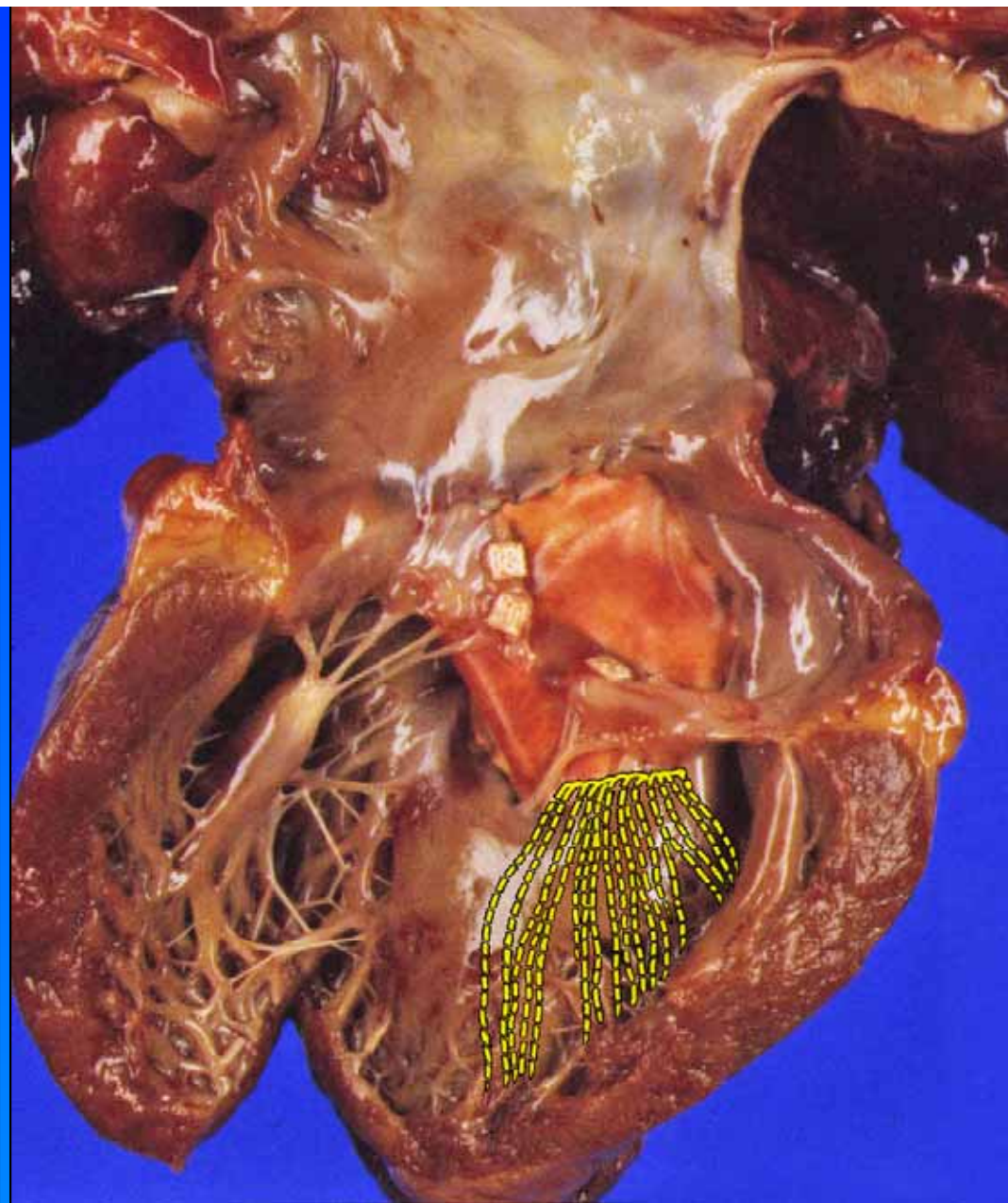
Common inlet RV with  
common AV valve and DORV  
"Heterotaxy"



- Other assoc. anomalies
- Tetralogy of fallot



# Conduction system Repaired AV canal



AV canal defects result from abnormalities in morphologic development, septation, & alignment of the embryonic AV canal

