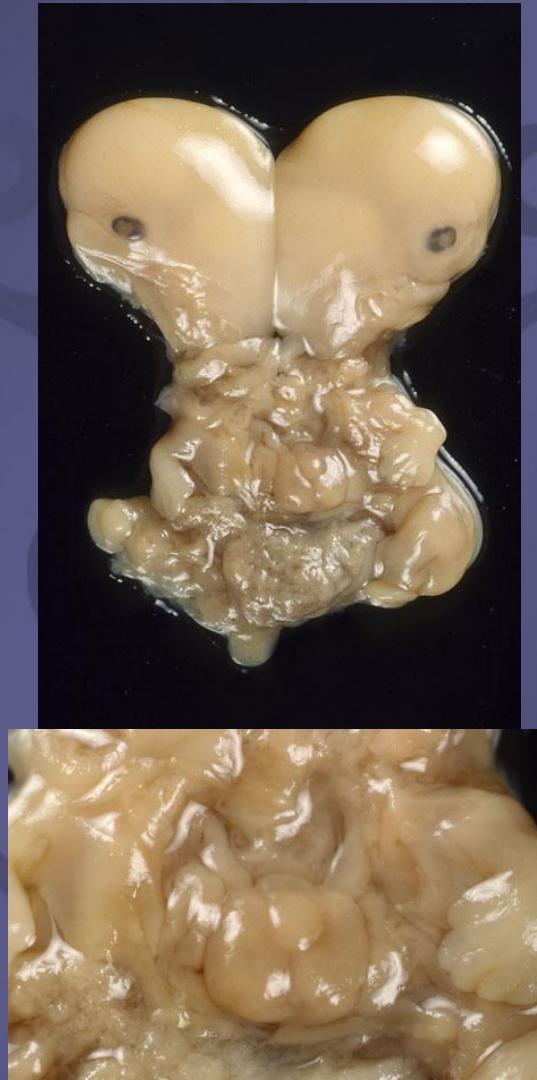
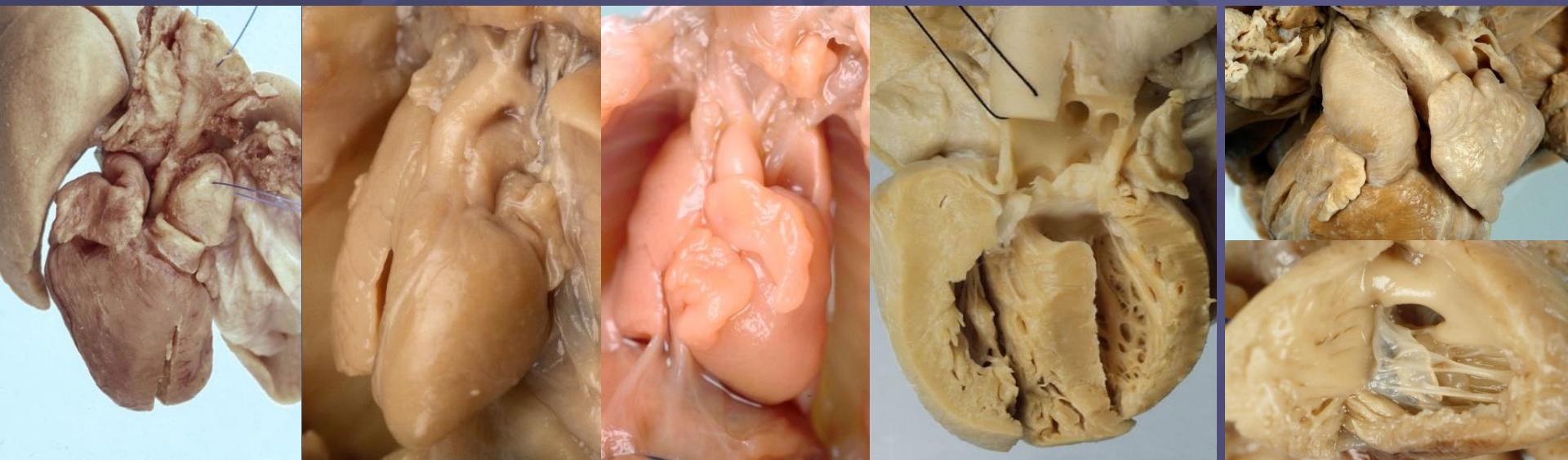


# Morphology of Congenital Heart Disease



# Morphology of Congenital Heart Disease



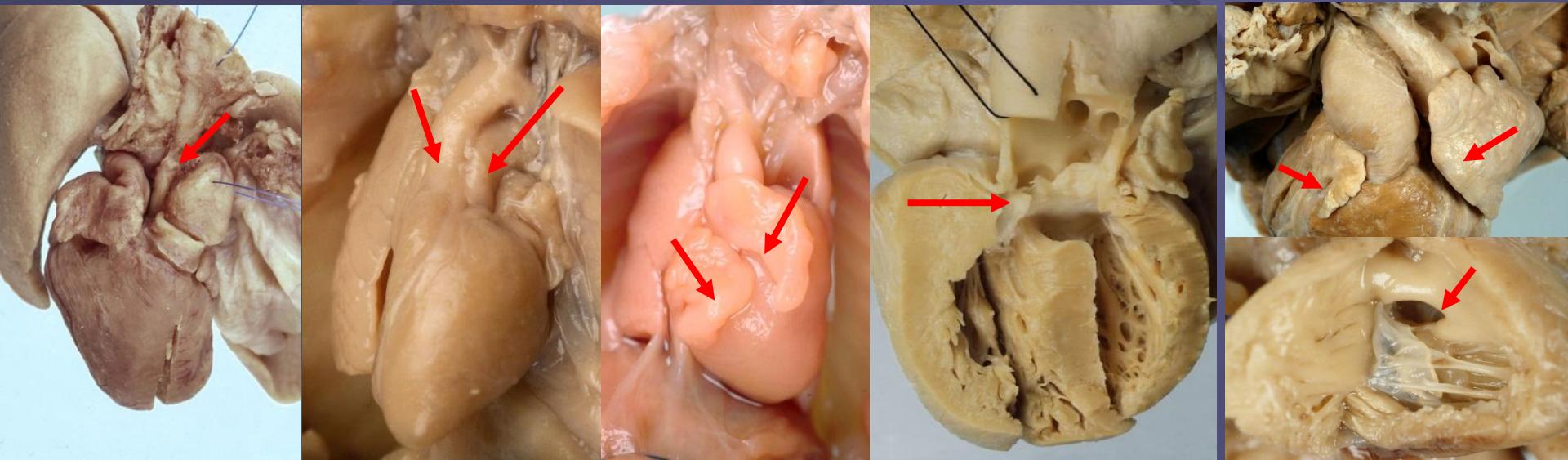
## NOMENCLATURE AND CLASSIFICATION OF CONGENITAL HEART DISEASE

By

**MICHAEL J. TYNAN, ANTON E. BECKER, FERGUS J. MACARTNEY,  
MANUEL QUERO JIMENEZ, ELLIOT A. SHINEBOURNE, AND  
ROBERT H. ANDERSON**

*Reprinted from British Heart Journal*  
*Volume XLI, No. 5, pages 544-553, May 1979*

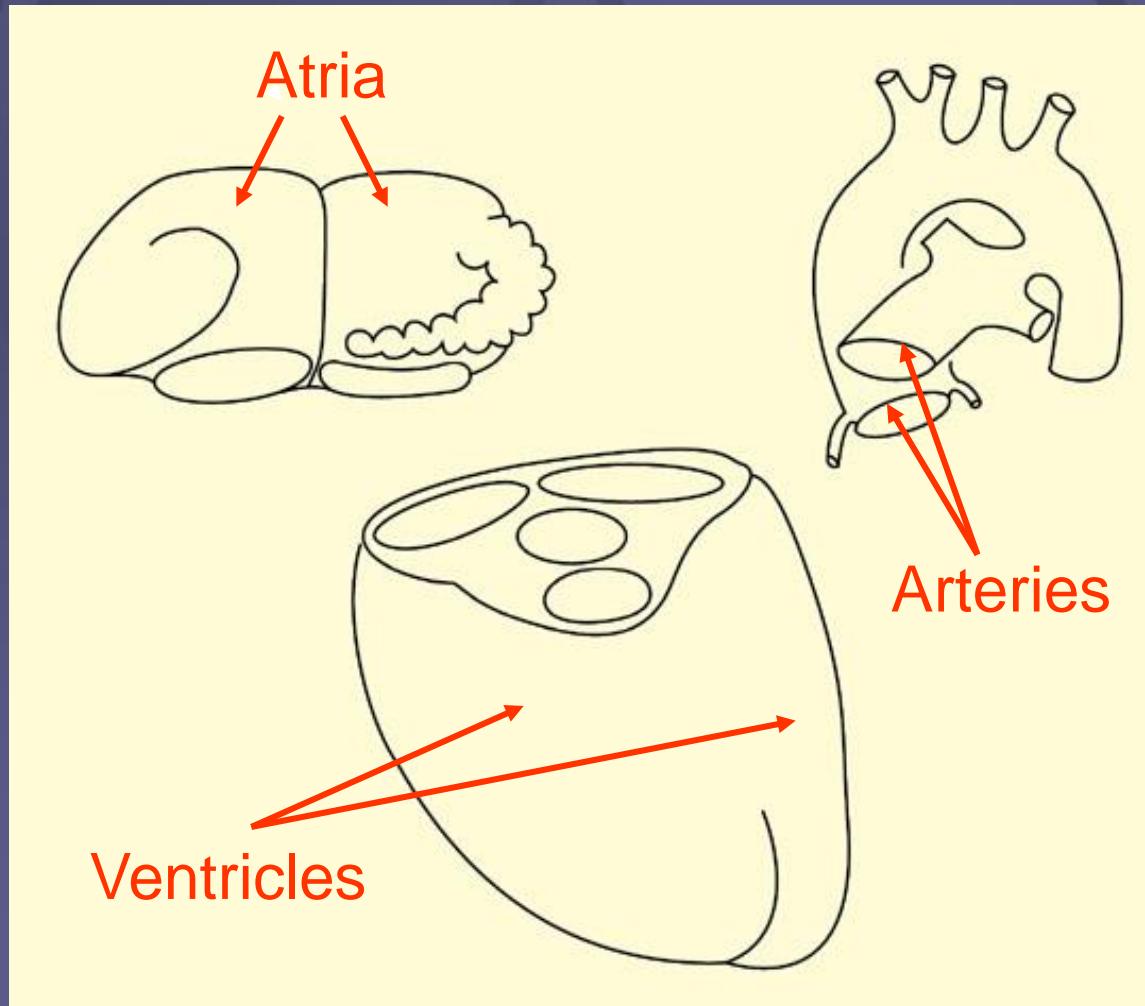
# Morphology of Congenital Heart Disease



## Sequential Segmental Analysis

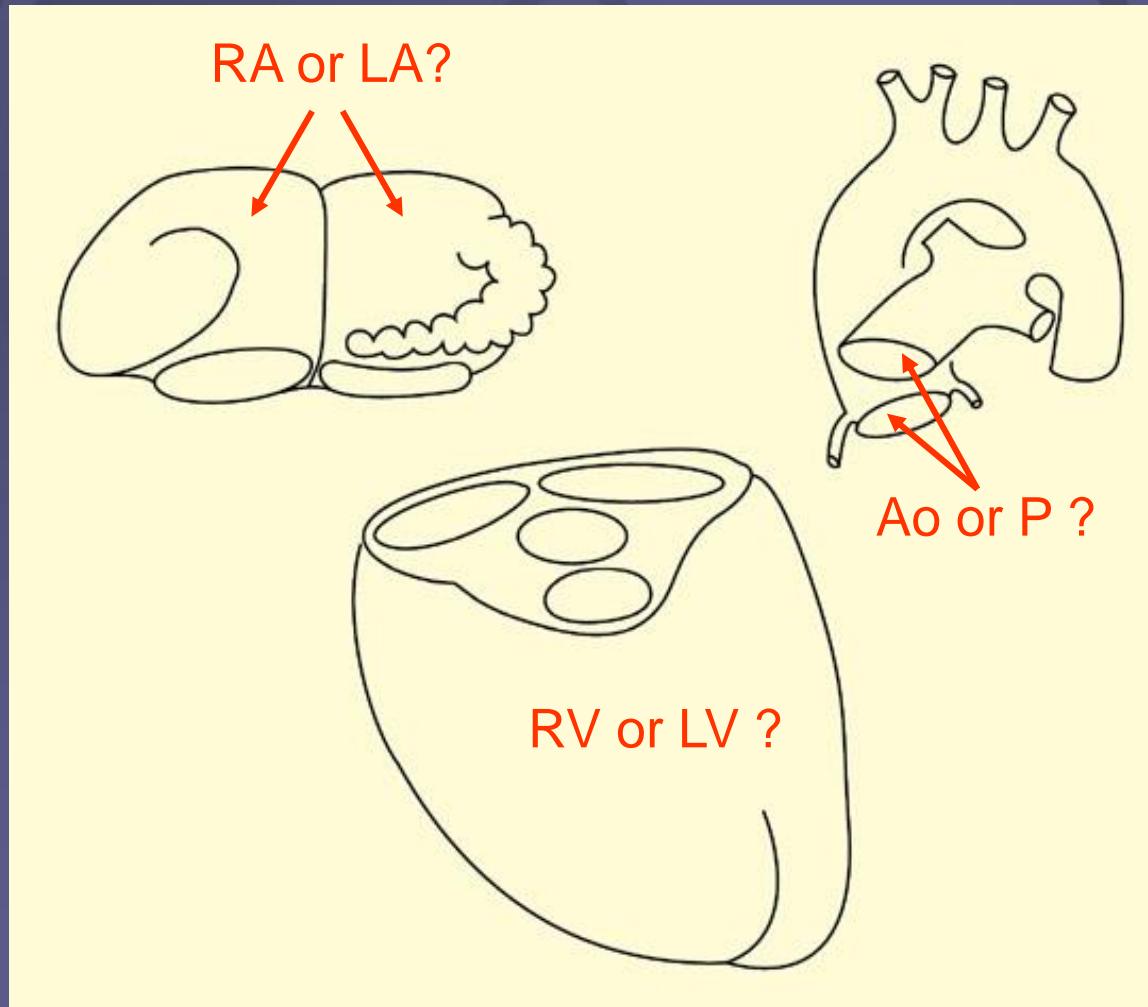
# Sequential segmental analysis

## Basis:segments



# Sequential segmental analysis

## Identification of the segments

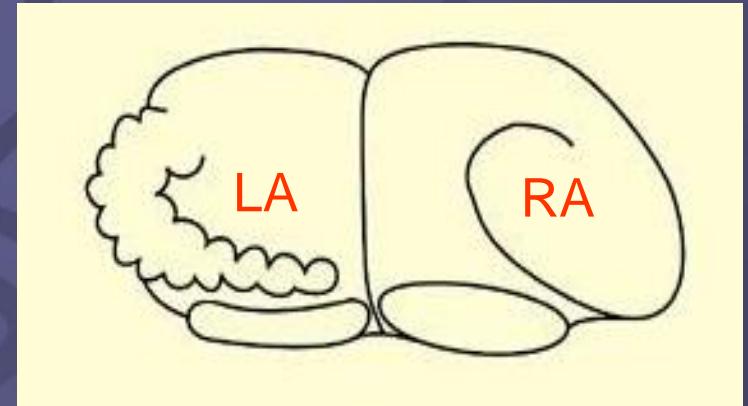


# Sequential segmental analysis

## Determination atrial situs

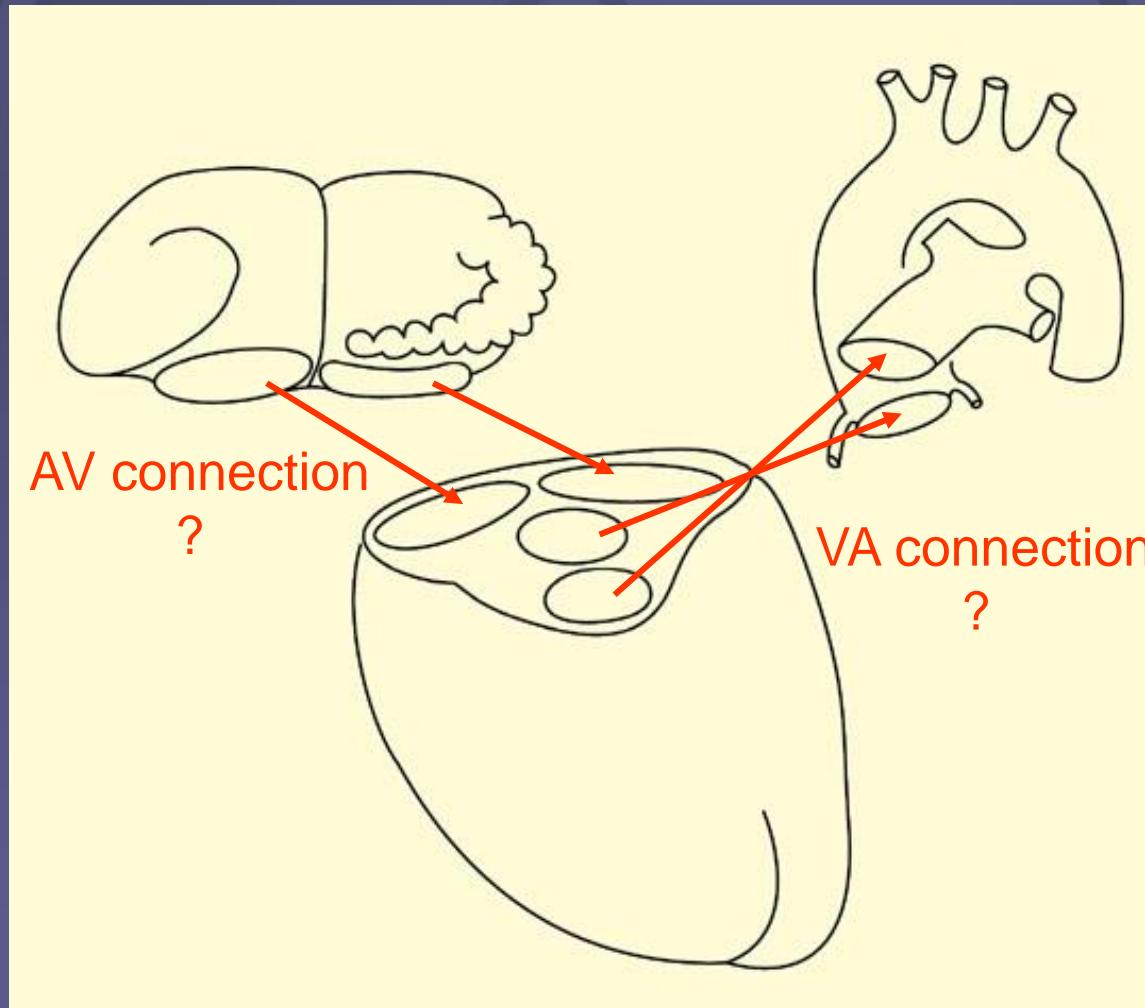


?



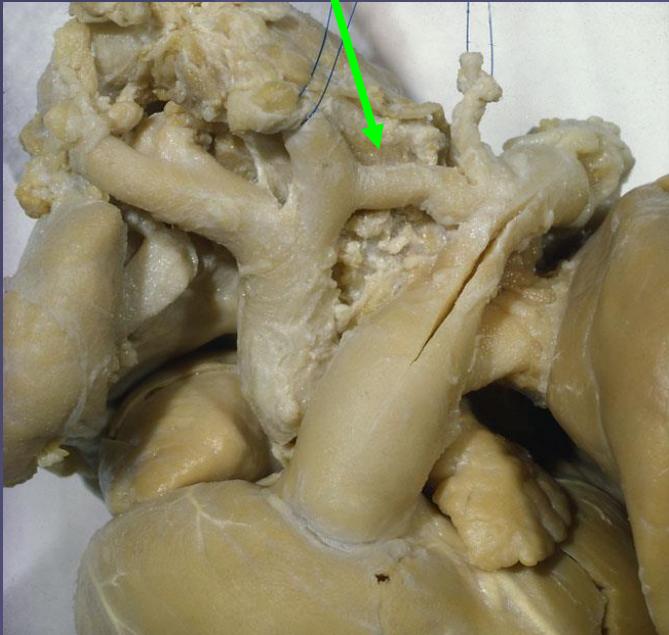
# Sequential segmental analysis

## Analysis AV and VA connections

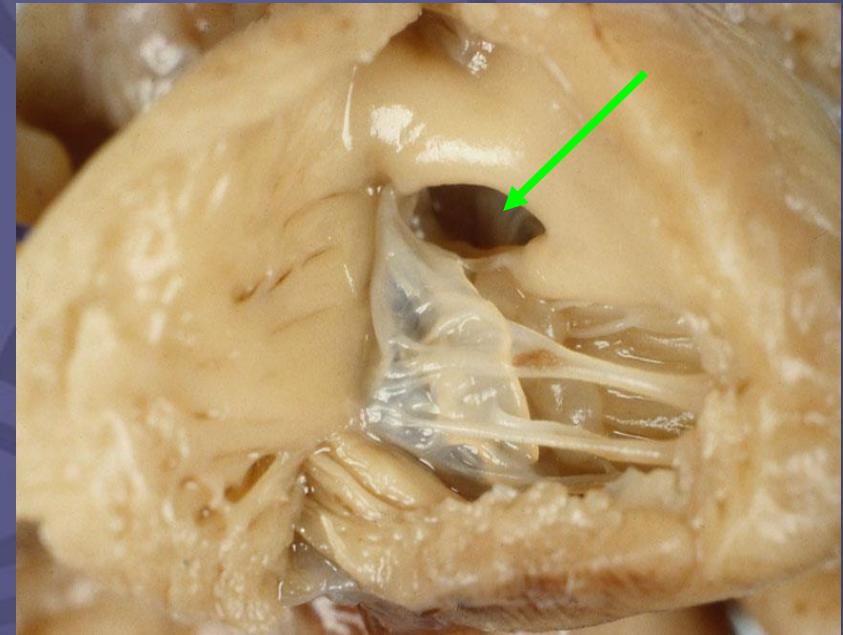


# Sequential segmental analysis

## Analysis associated lesions



Hypoplasia aortic arch



VSD

# Identification of the atria

always 2 atria

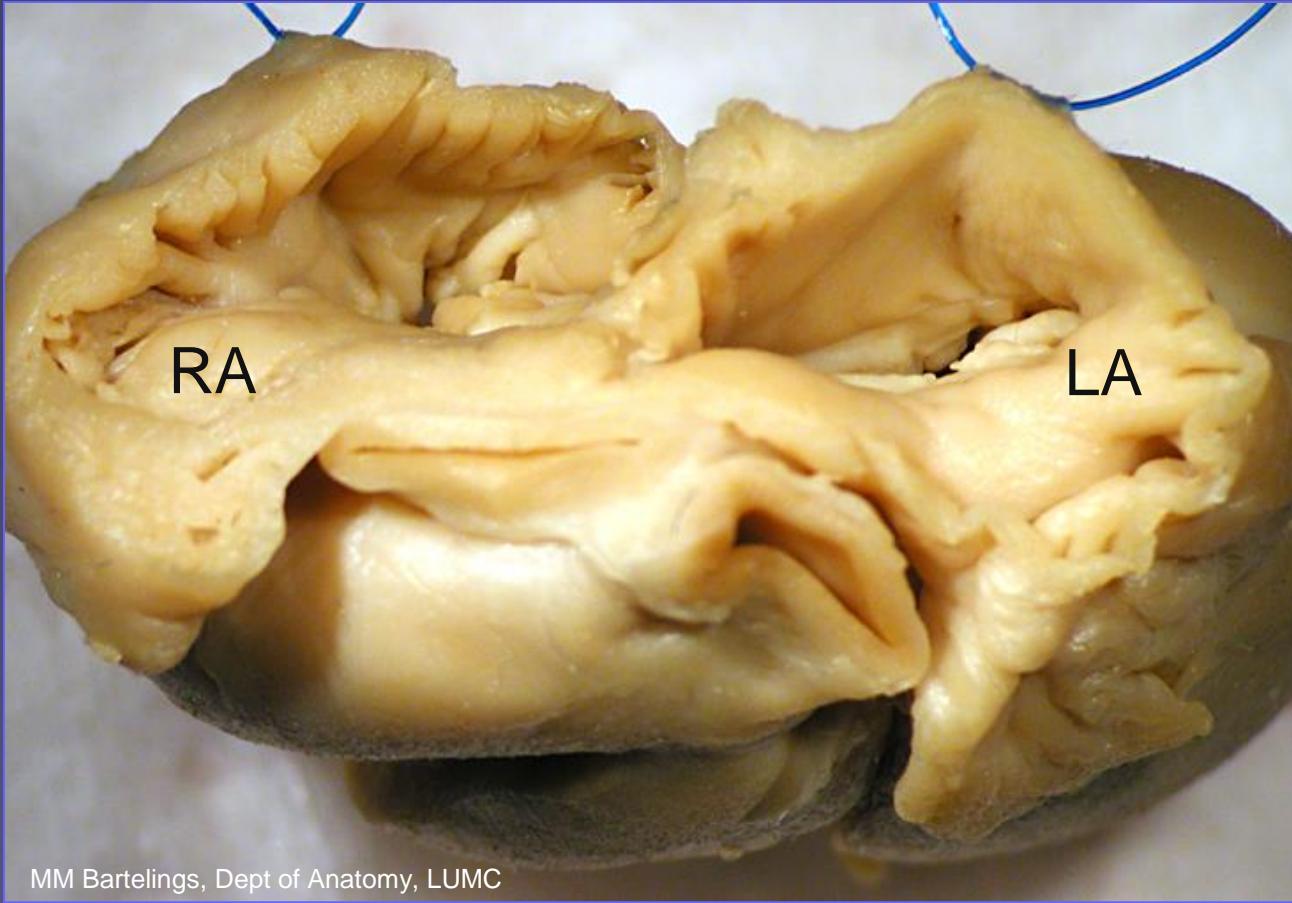
morphologically right (green) or left (orange)



appendages

right: blunt

left: tubular

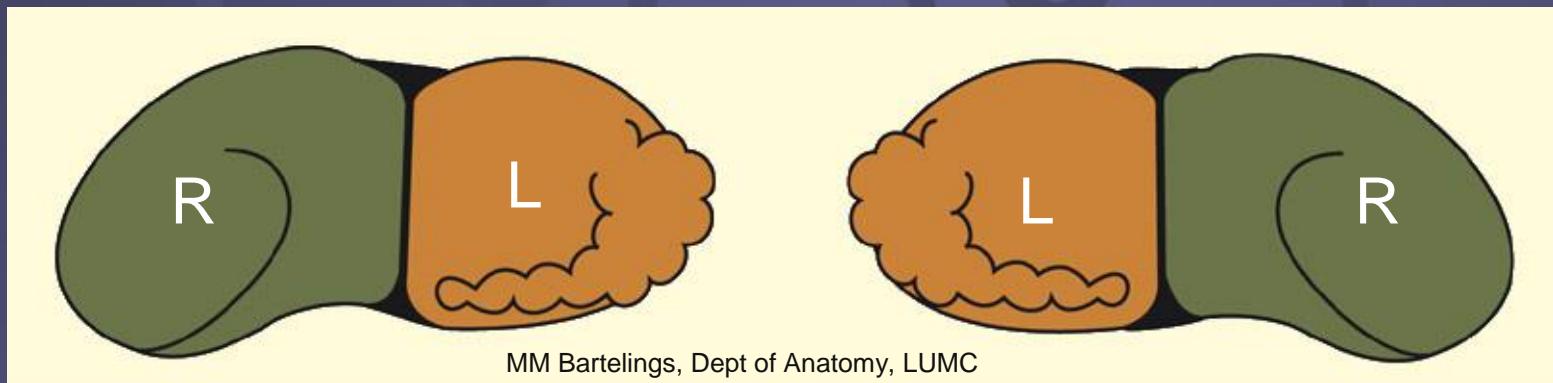


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Right atrium:  
mm. pectinati over the  
entire posterior wall

Left atrium:  
smooth posterior wall

# Atrial arrangement (lateralized)



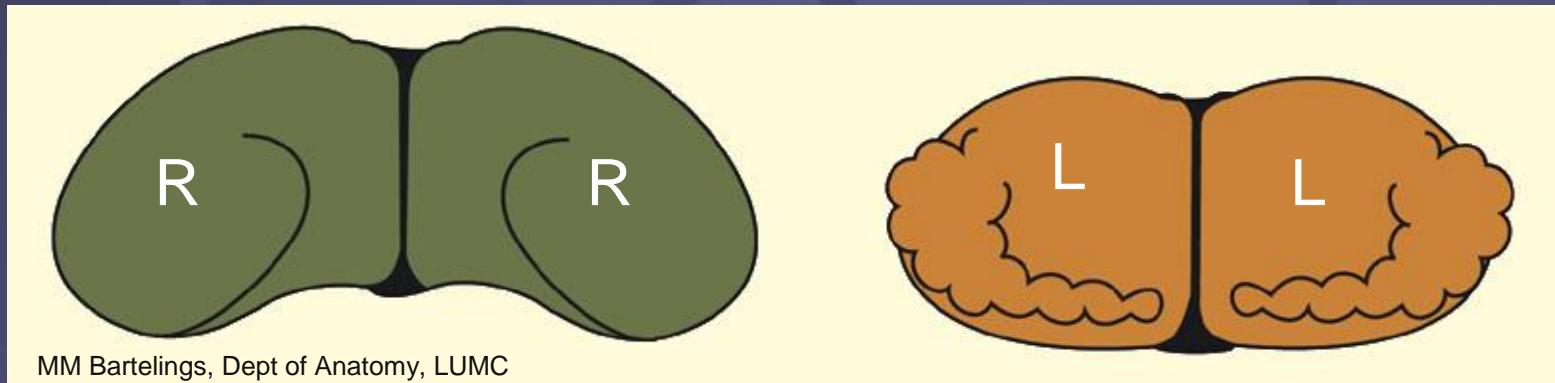
situs solitus

normal

situs inversus

mirror image

# Atrial arrangement (isomeric)



right-isomerism

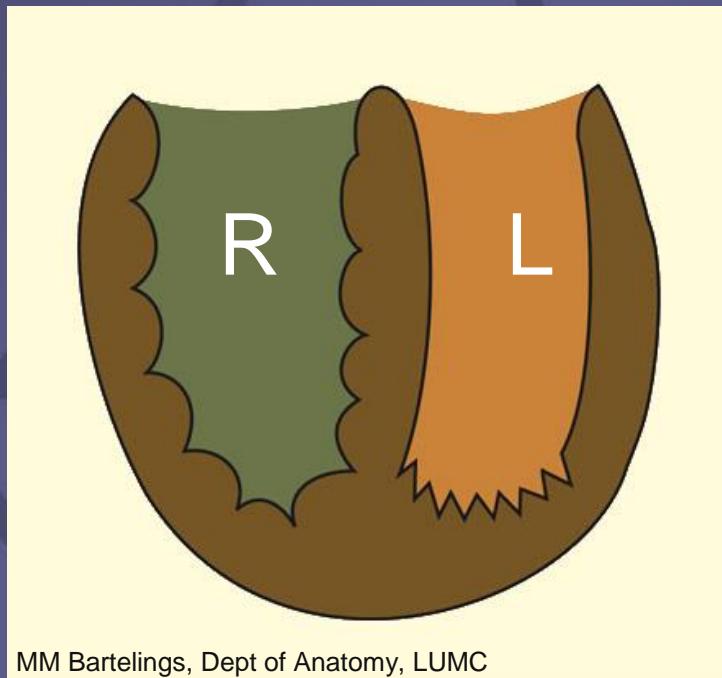
left-isomerism

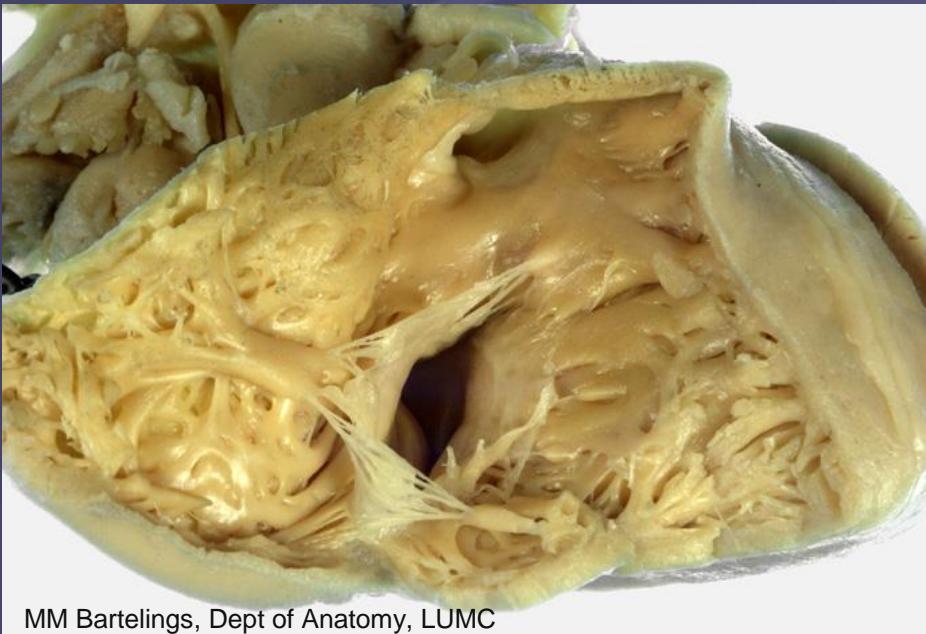
situs ambiguus

# Identification of the ventricles

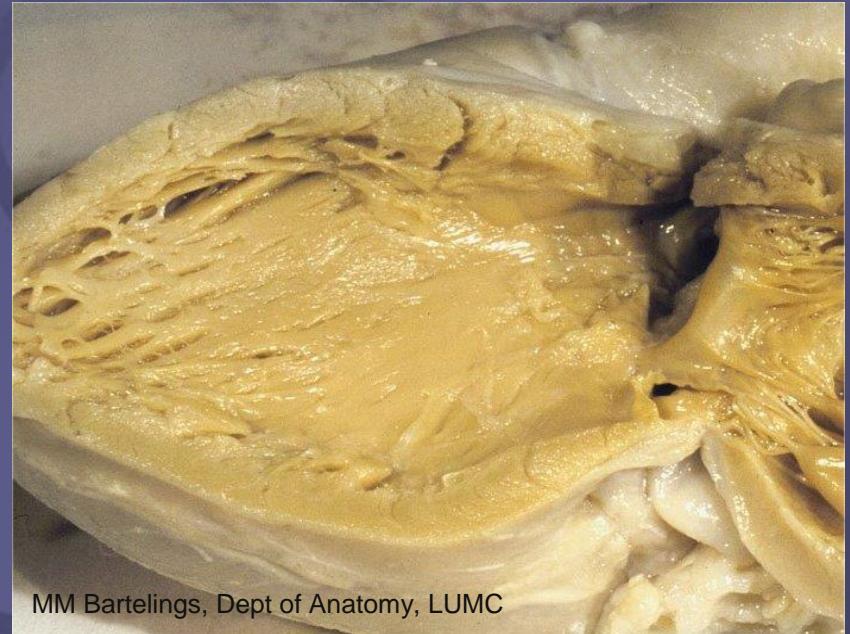
usually 2 ventricles

morphologically RV and LV





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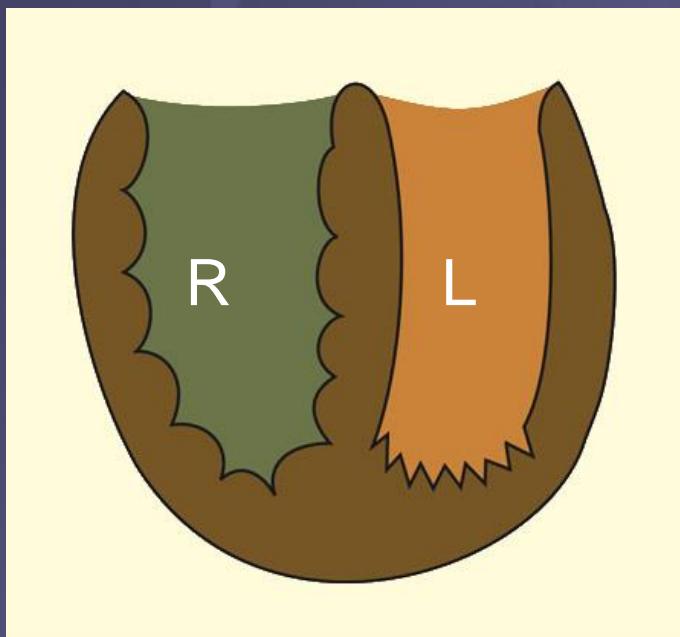


MM Bartelings, Dept of Anatomy, LUMC

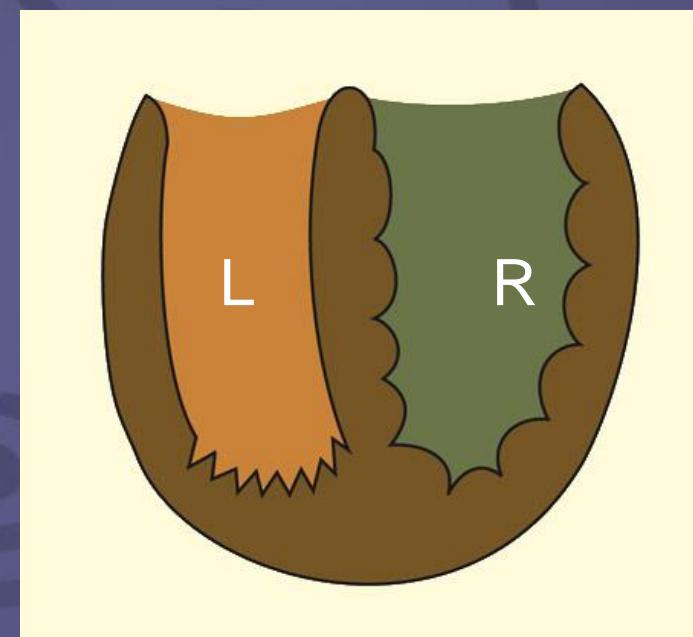
RV:  
coarse apical trabeculations  
TSM and moderator band  
muscular infundibulum

LV:  
fine trabeculations  
smooth septal surface

# Identification of the ventricles

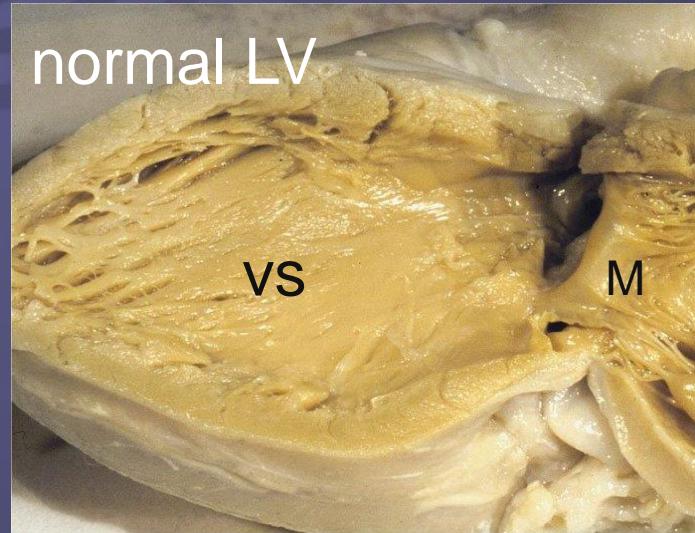
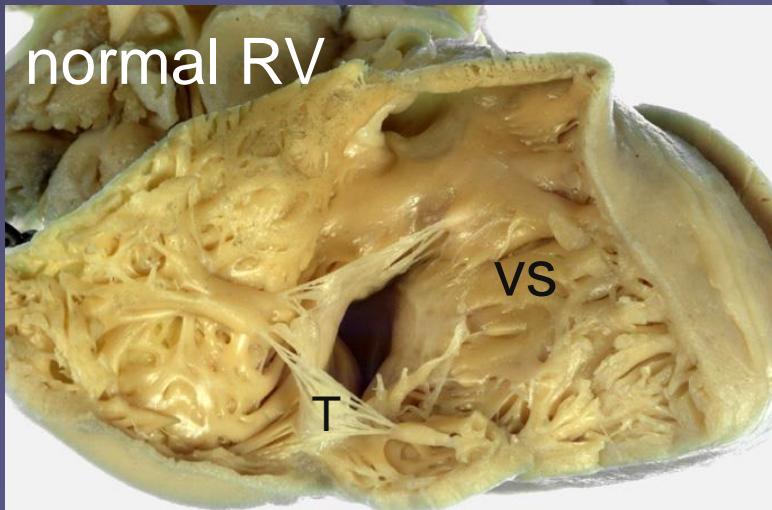
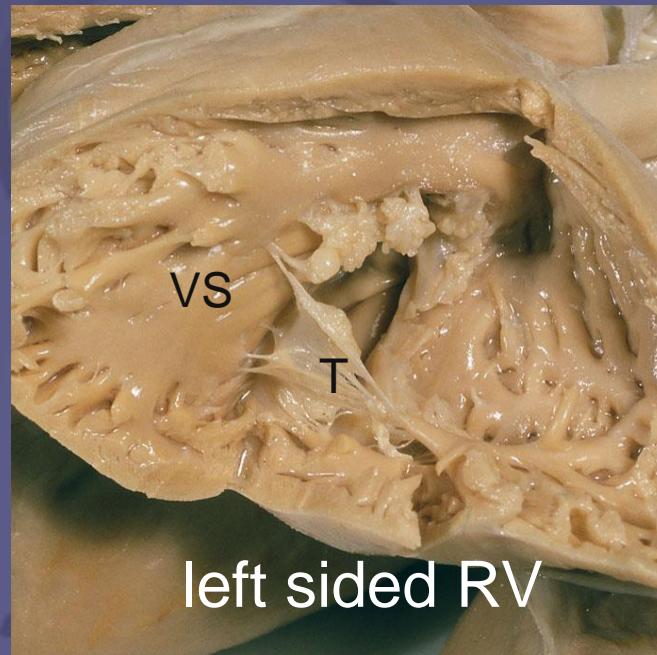
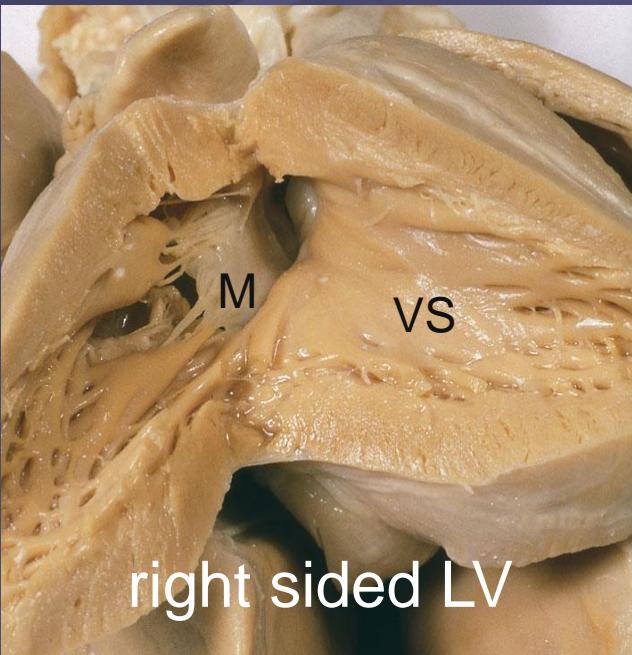


normal



ventricular inversion  
mirror image

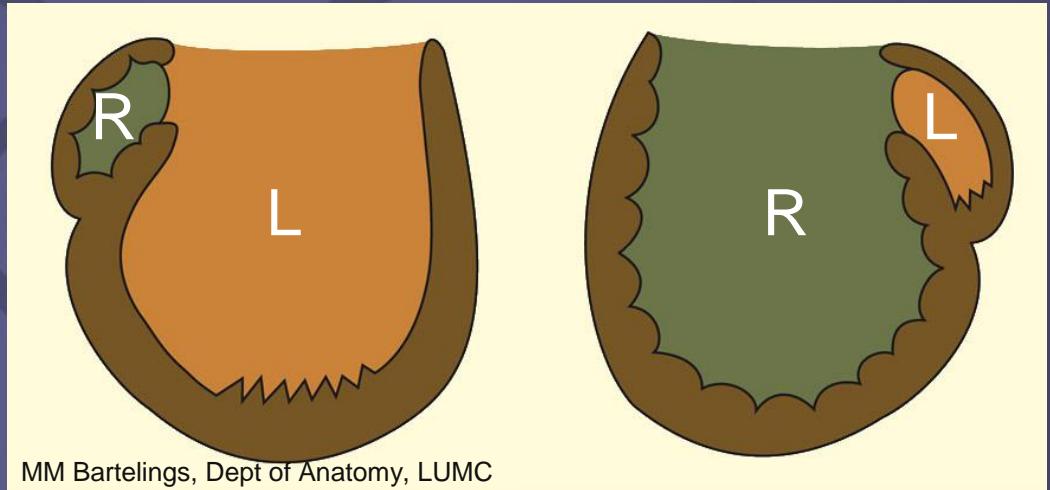
# ventricular inversion



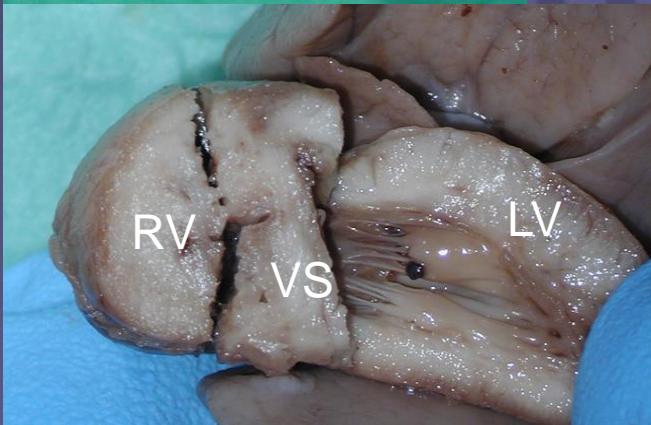
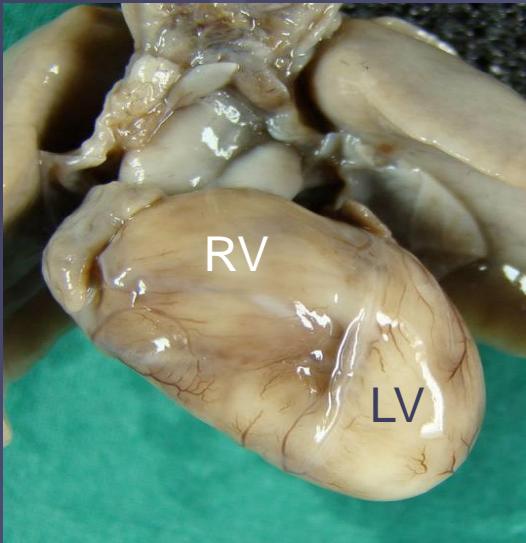
# Identification of the ventricles

balanced

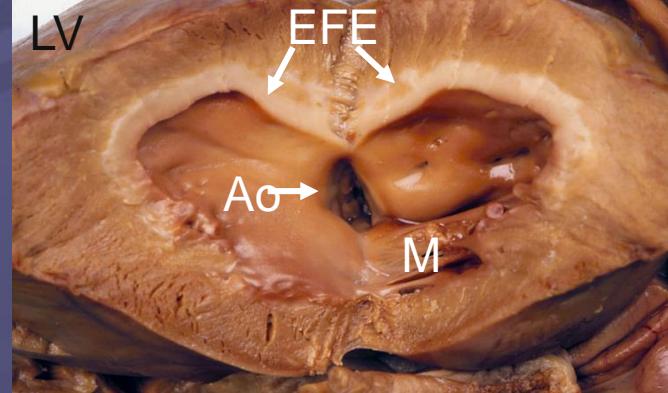
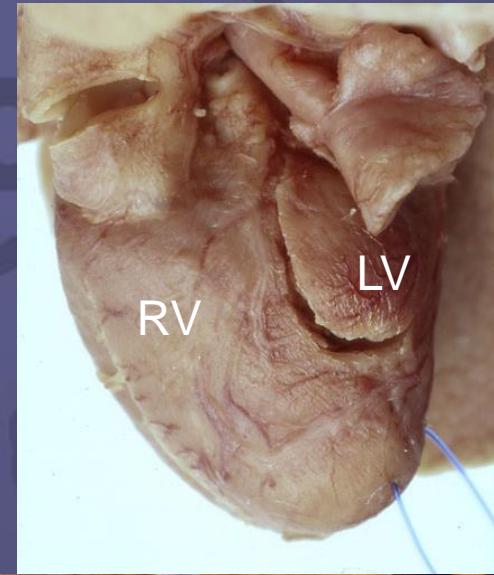
RV hypoplasia LV



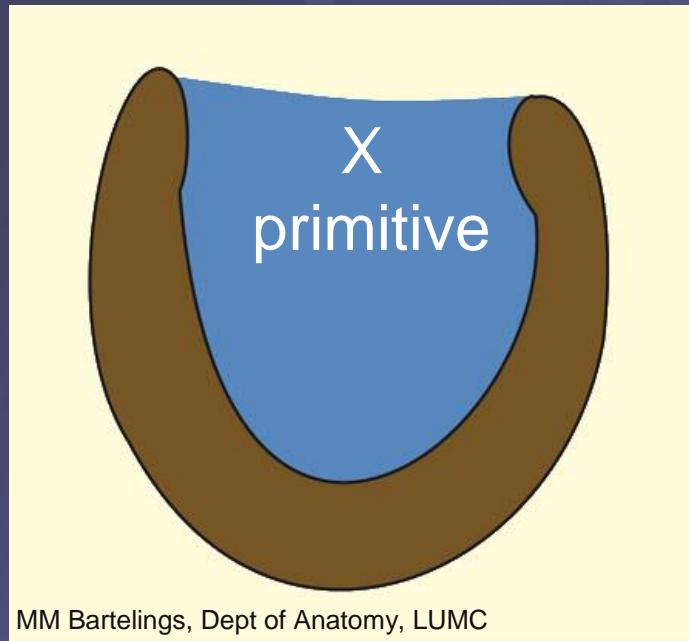
hypoplastic RV



hypoplastic LV



# Identification of the ventricle(s)



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single ventricle  
(rare)

# Atrial situs

- solitus
- inversus
- ambiguous  
isomerism R/L

# Ventricular pattern

- normal +/- hypoplasia RV/LV
- inversion +/- hypoplasia RV/LV
- single ventricle

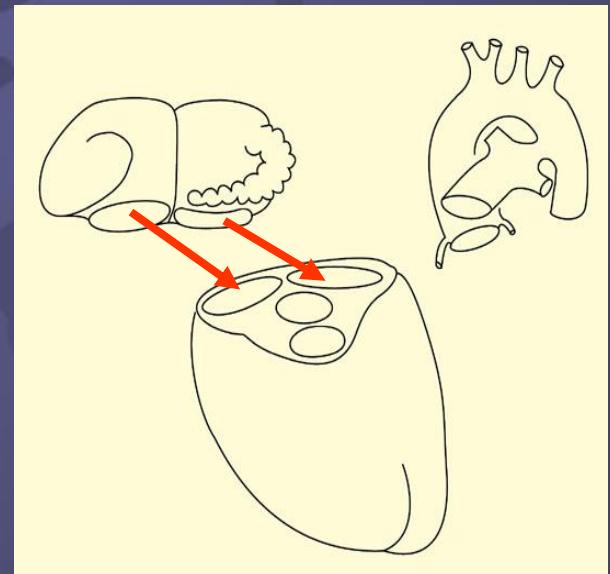
## Atrioventricular (AV) junction

# The atrioventricular (AV) junction

- **type** of AV connection
  - which atrium connected to which ventricle
  - biventricular or univentricular
- **mode** of AV connection
  - details of the valve / the orifice

# 5 types of AV connection

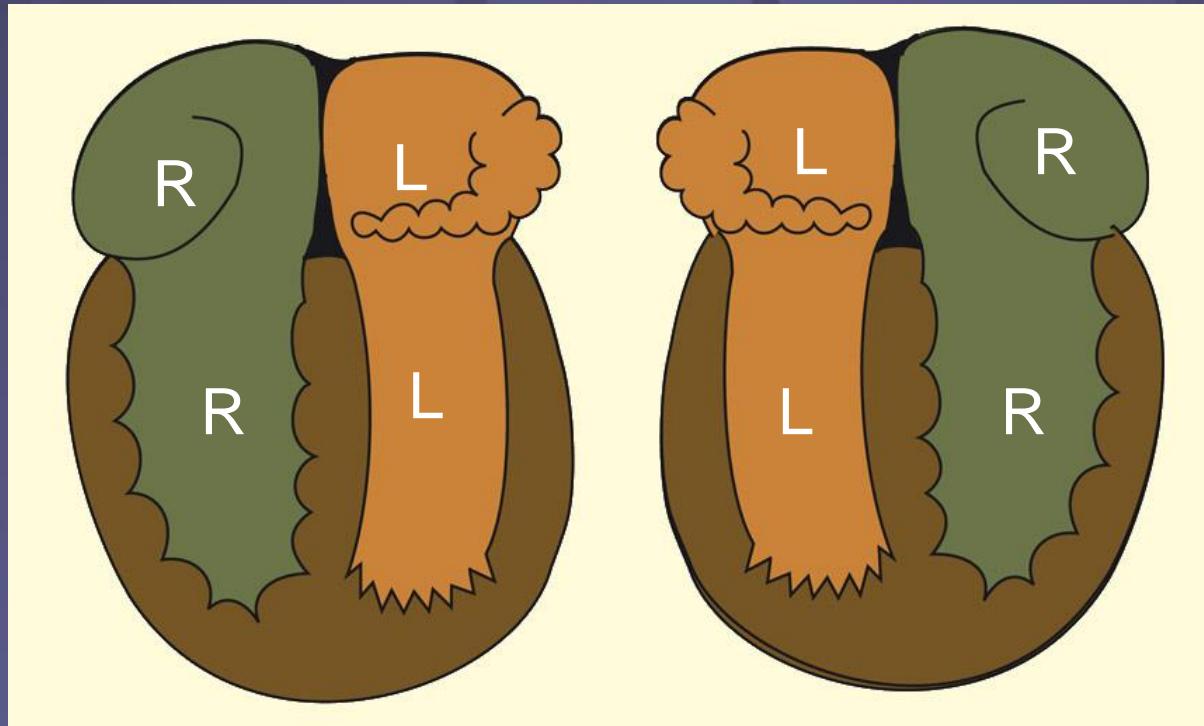
- concordant
  - discordant
  - ambiguous/mixed
  - double inlet
  - absent
- } either / or



# Type of AV connection (biventricular)

## Concordant

situs  
solitus

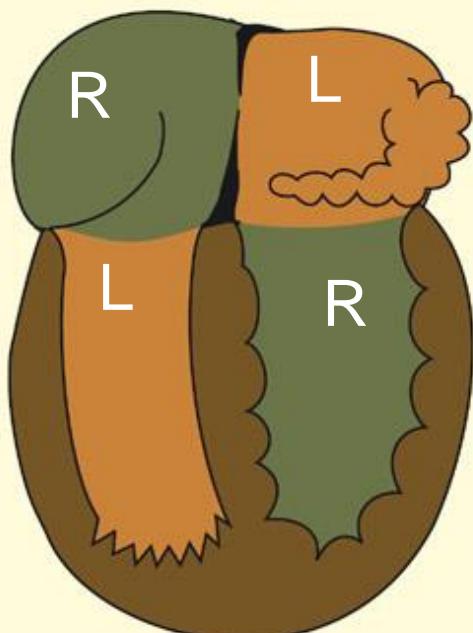


situs  
inversus

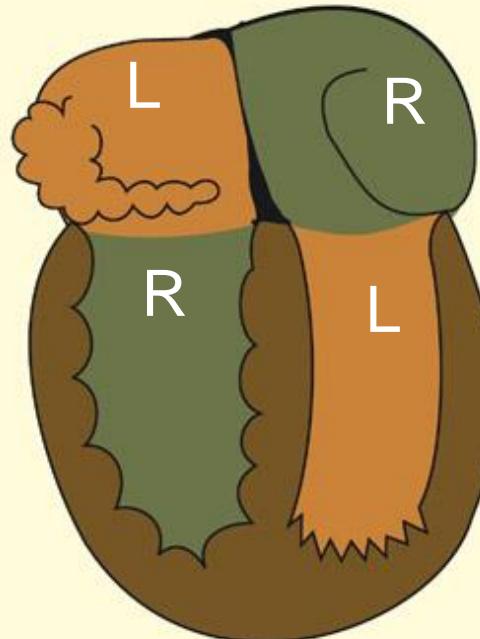
# Type of AV connection (biventricular)

## Discordant

situs  
solitus



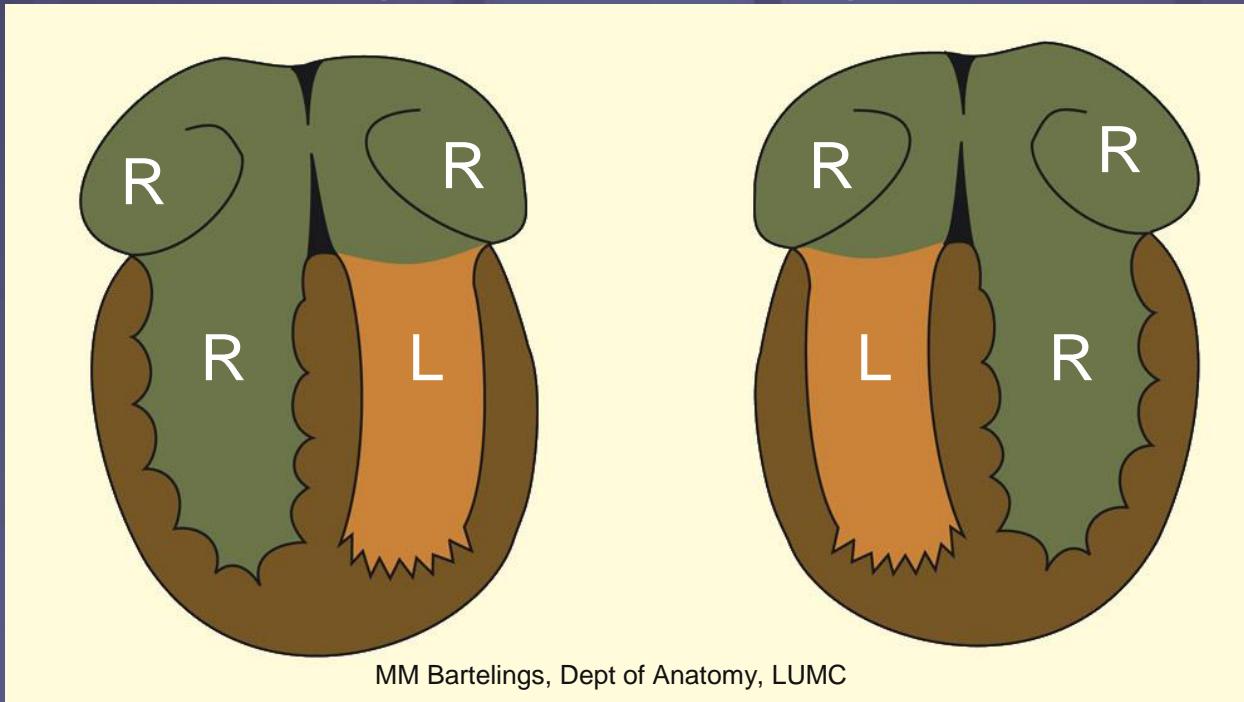
situs  
inversus



# Type of AV connection (biventricular)

## Ambiguous/mixed

situs ambiguus atria with right isomerism

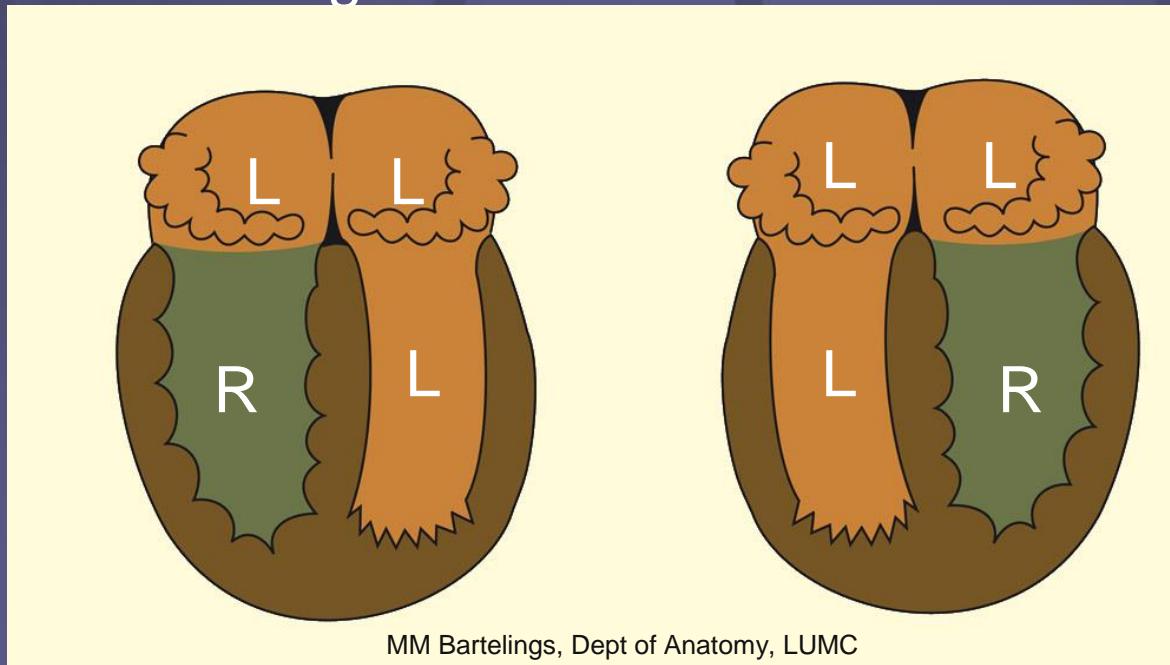


describe ventricles!

# Type of AV connection (biventricular)

## Ambiguous/mixed

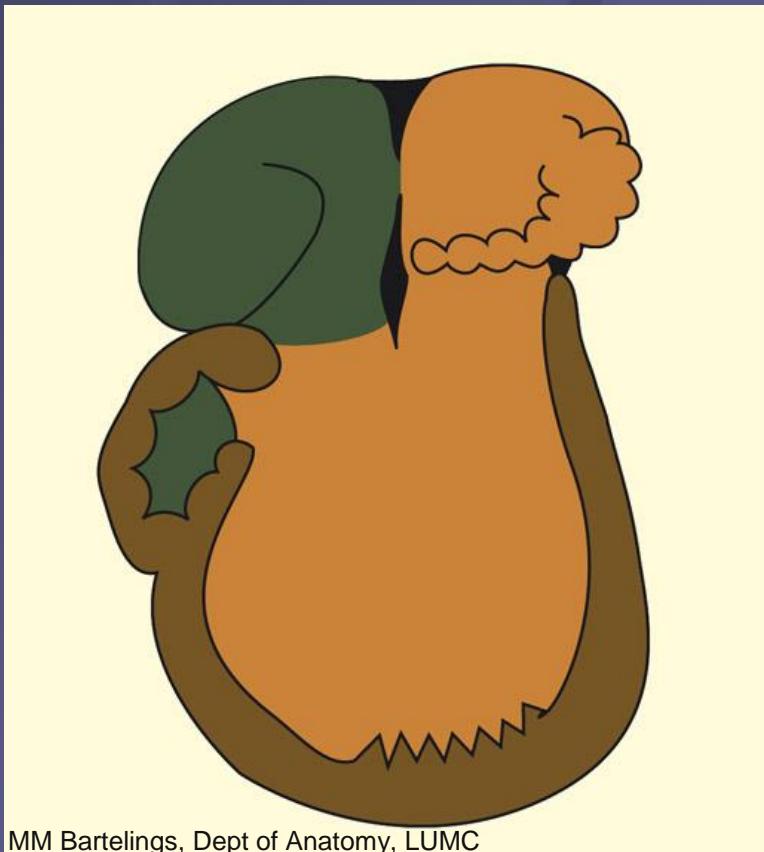
situs ambiguus atria with left isomerism



describe ventricles!

# Type of AV connection (univentricular)

## Double inlet

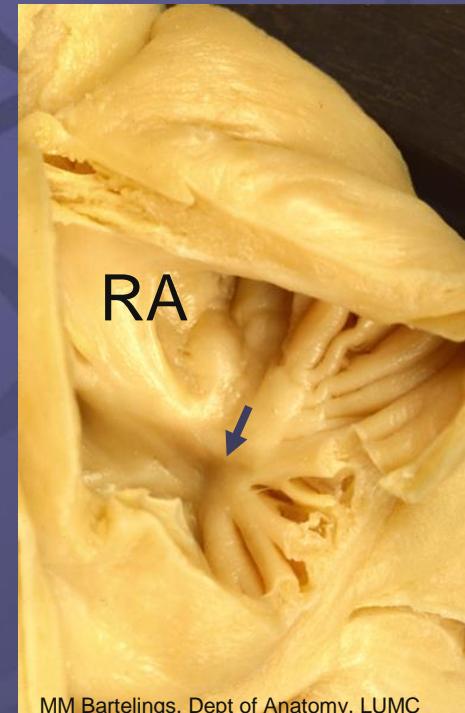
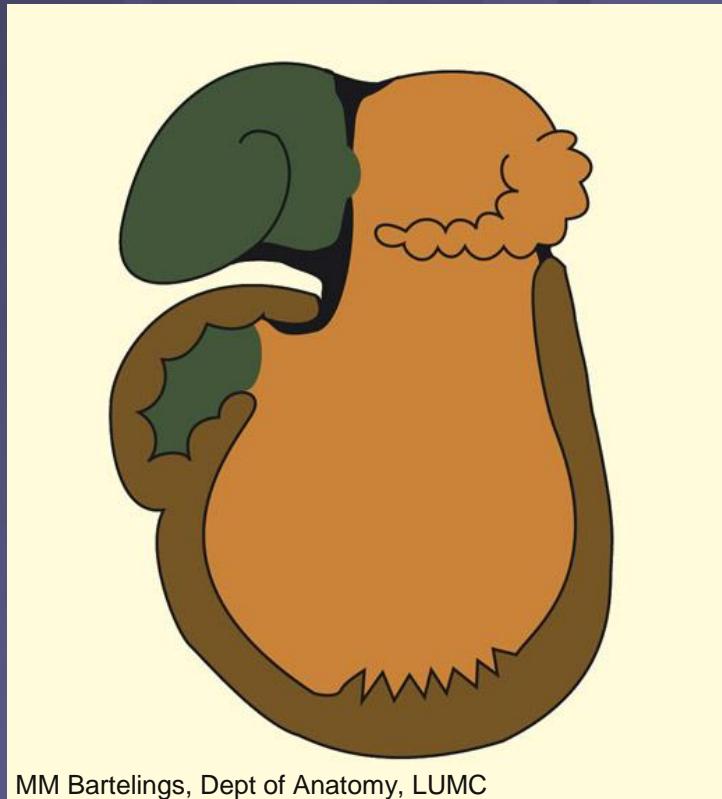


(LV or single ventricle)

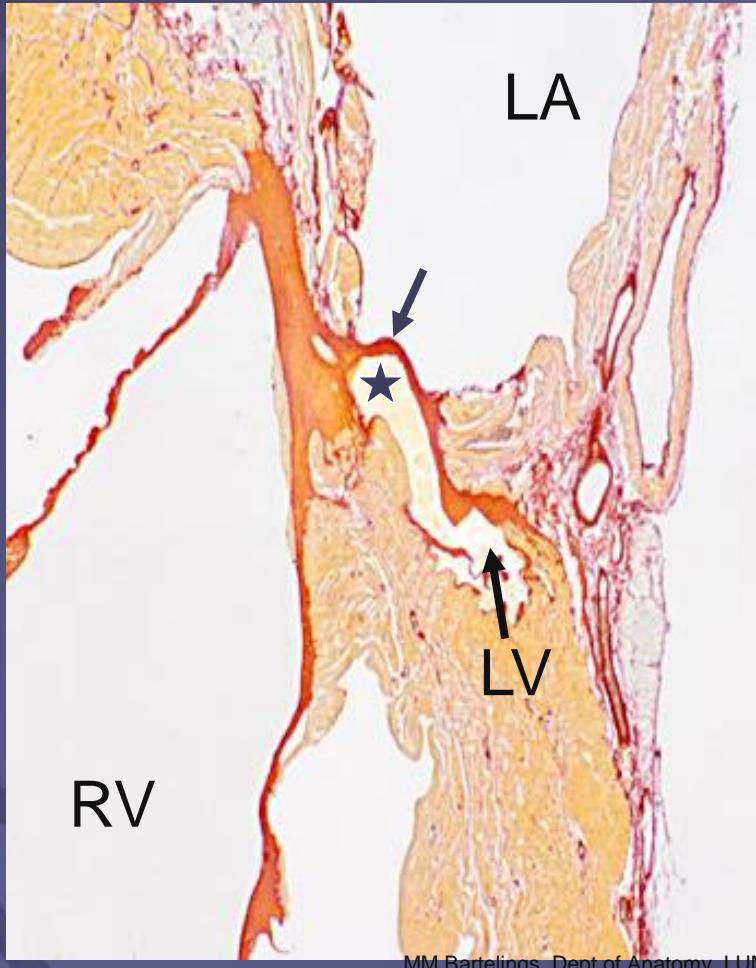
# Type of AV connection (univentricular)

## Absent connection

(R)



only with muscular tricuspid atresia

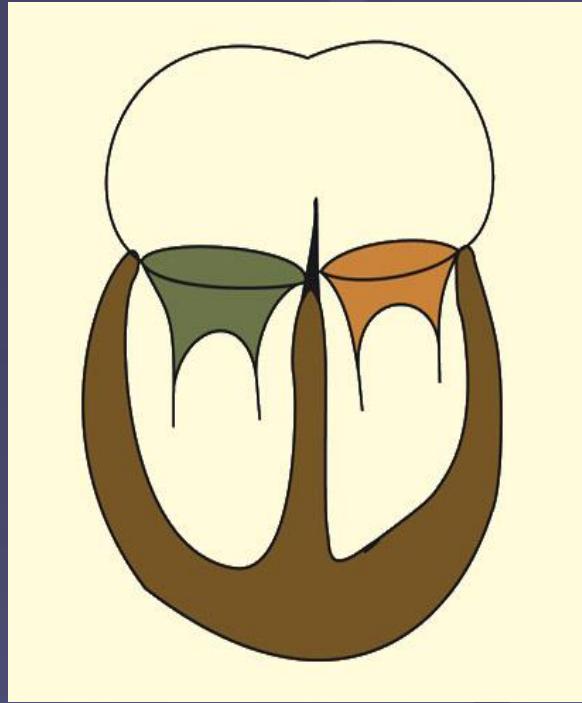


concordant AV

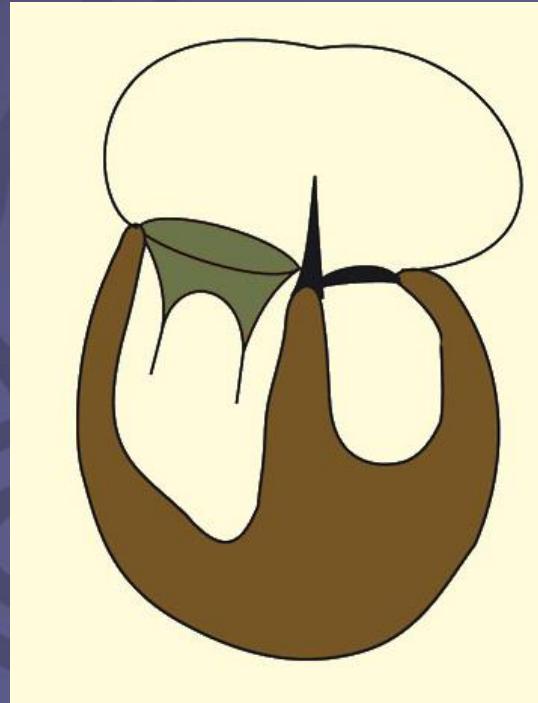
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mitral atresia – never absent AV connection

# Mode of AV connection



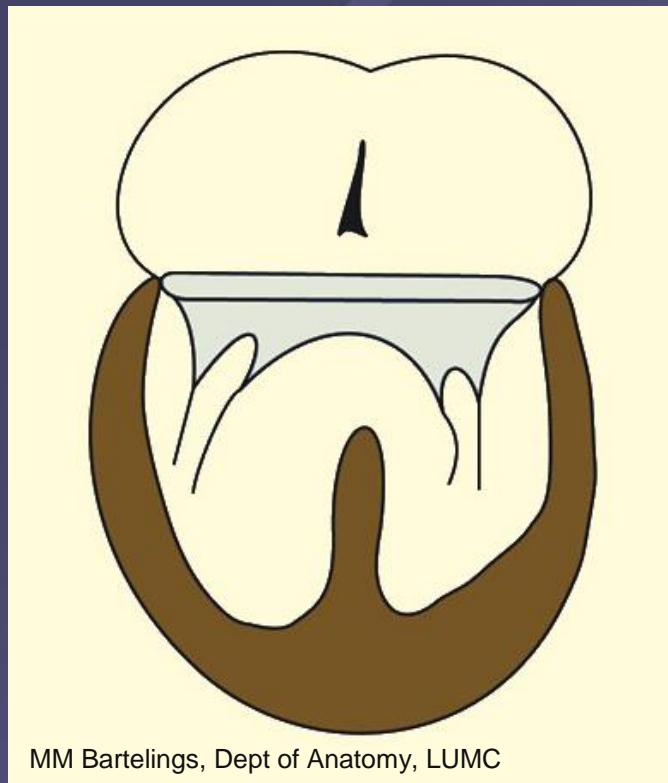
perforate valves



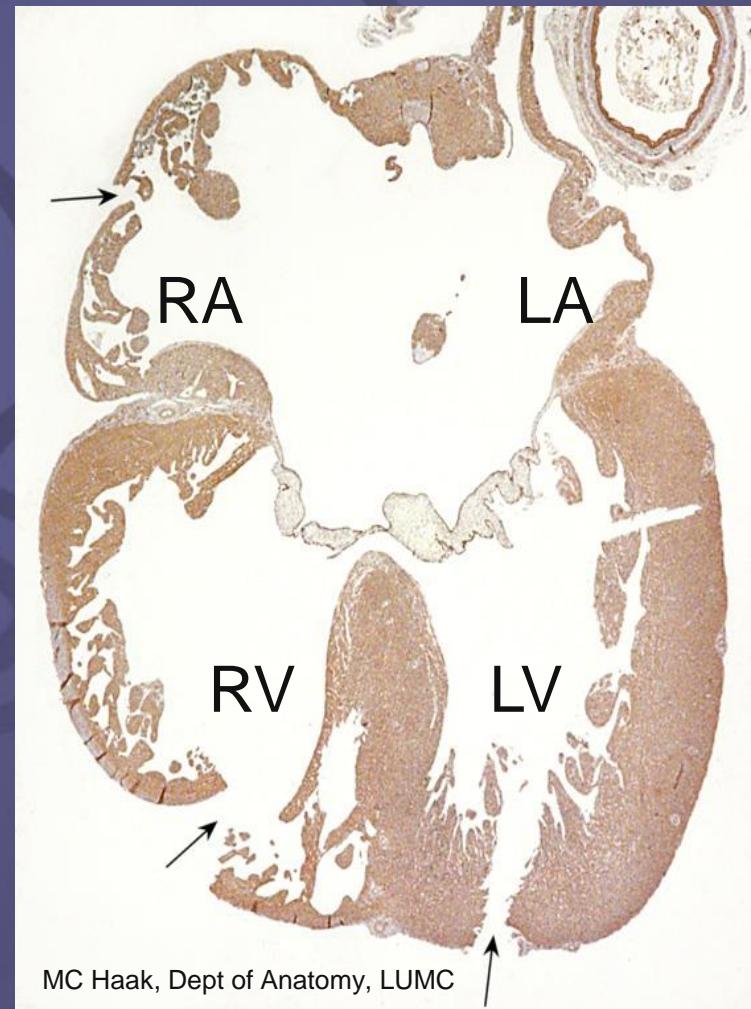
imperforate valve  
valvular (membranous) atresia



# Mode of AV connection

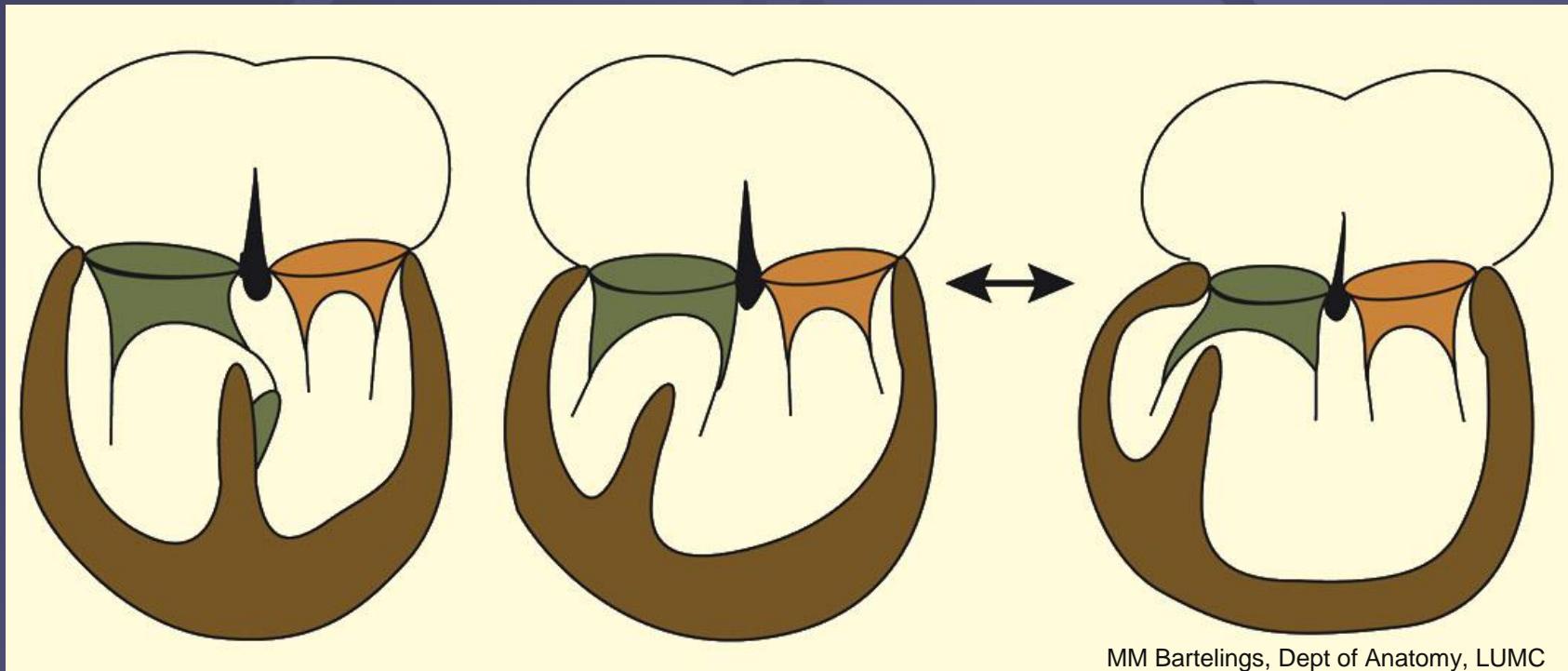


common valve



trisomy 21

# Mode of AV connection



straddling

overriding  
 $< 50\%$

biventricular

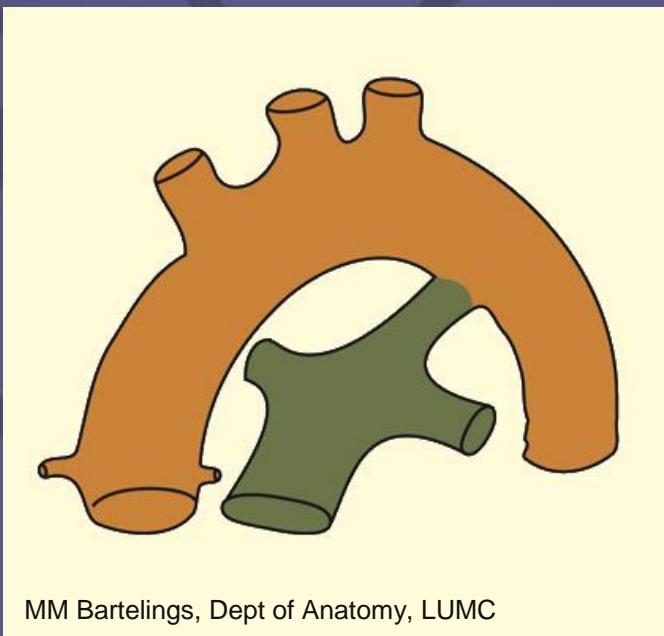
overriding  
 $> 50\%$

univentricular

# Identification great arteries

Ascending aorta continuous with proximal aortic arch + branches and usually coronary arteries

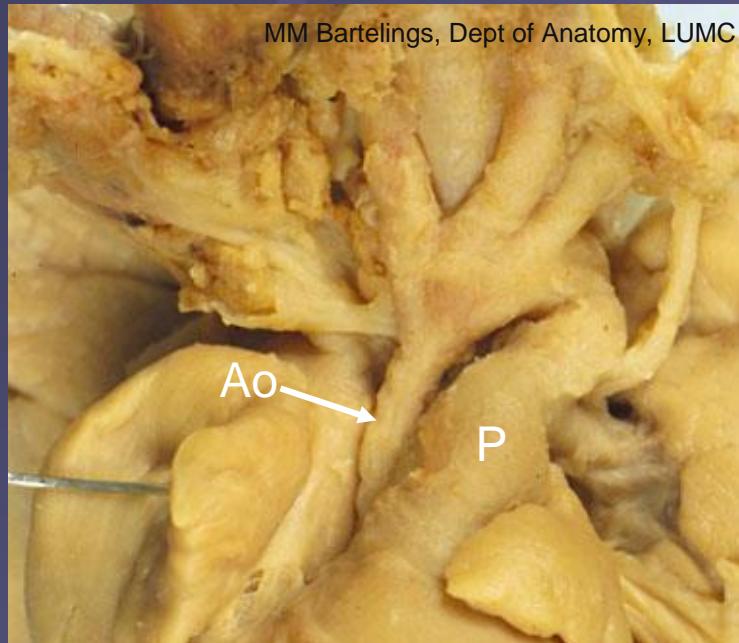
Pulmonary trunk continuous with at least 1 pulmonary artery and usually the ductus arteriosus



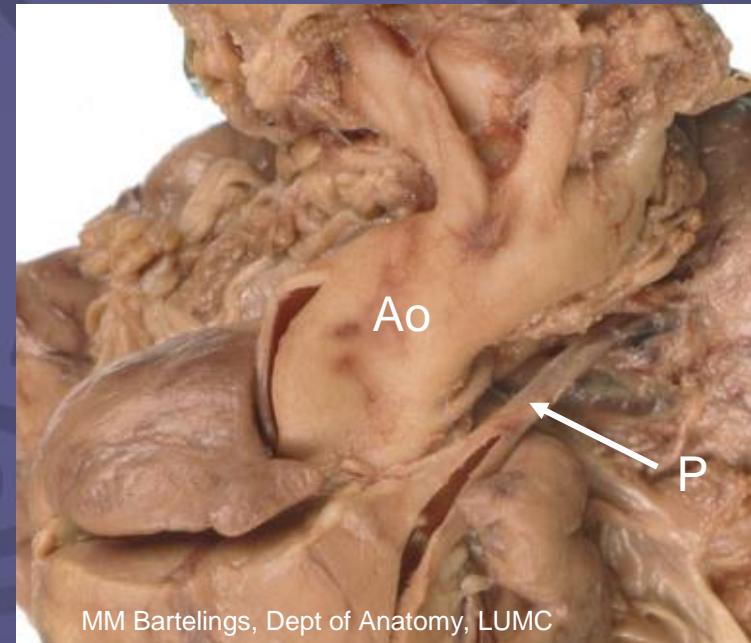
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# Identification great arteries

## hypoplastic great artery



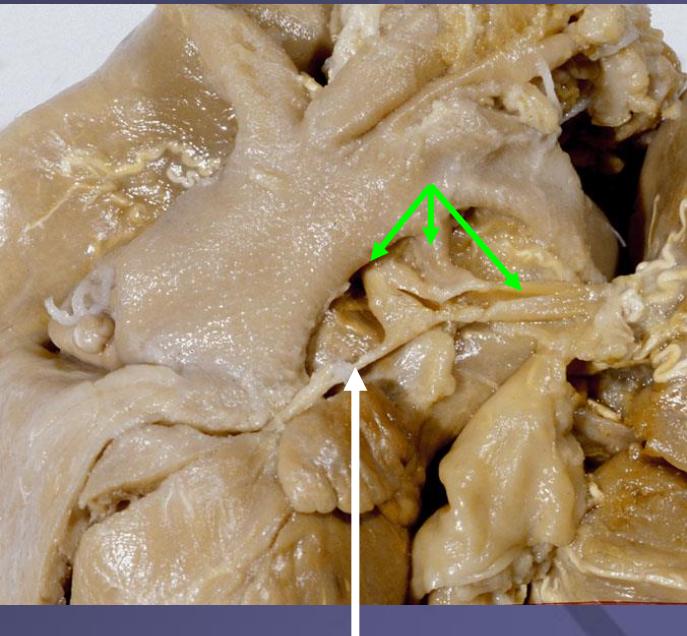
hypoplastic ascending aorta



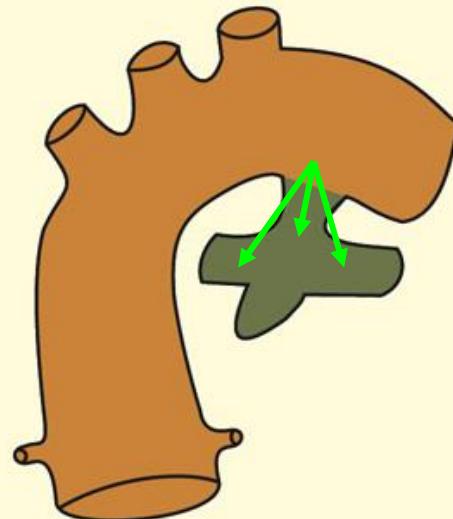
hypoplastic pulmonar trunk

# Identification great arteries

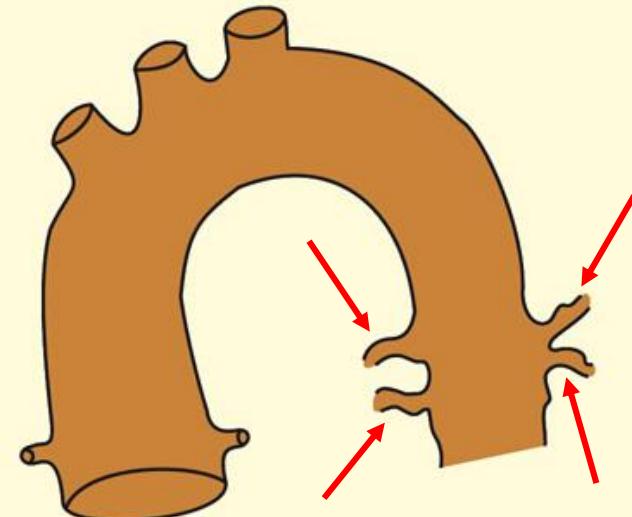
## single great artery



atresia pulm. trunk



absent pulm. trunk

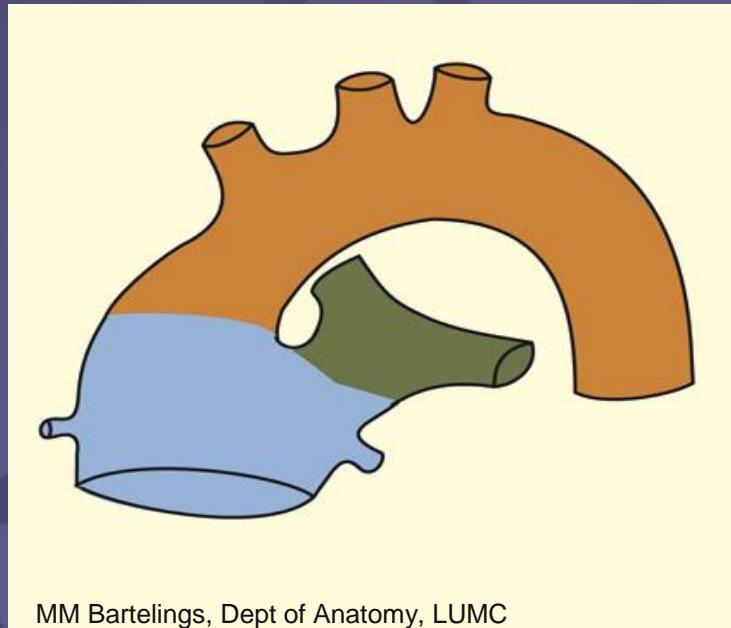


absent pulm. trunk + aa.

SPCA's

# Identification great arteries

single great artery



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common arterial trunk

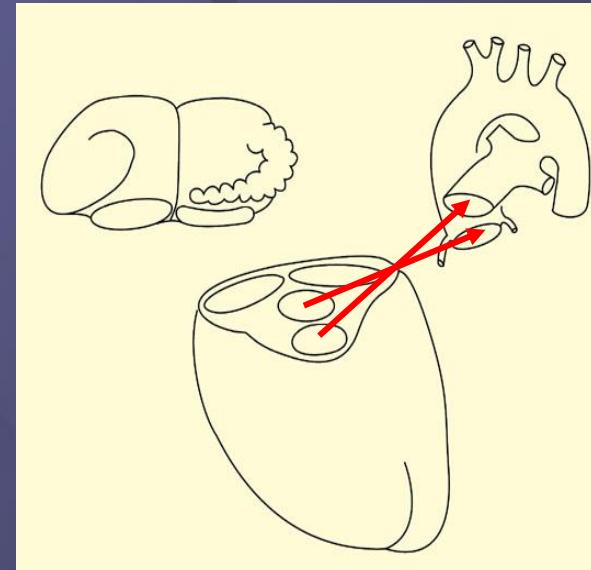
# The ventriculo-arterial (VA) junction

- **type** of VA connection  
which ventricle connected to which artery
- **mode** of VA connection  
details of the valve / the orifice

# 4 types of VA connection

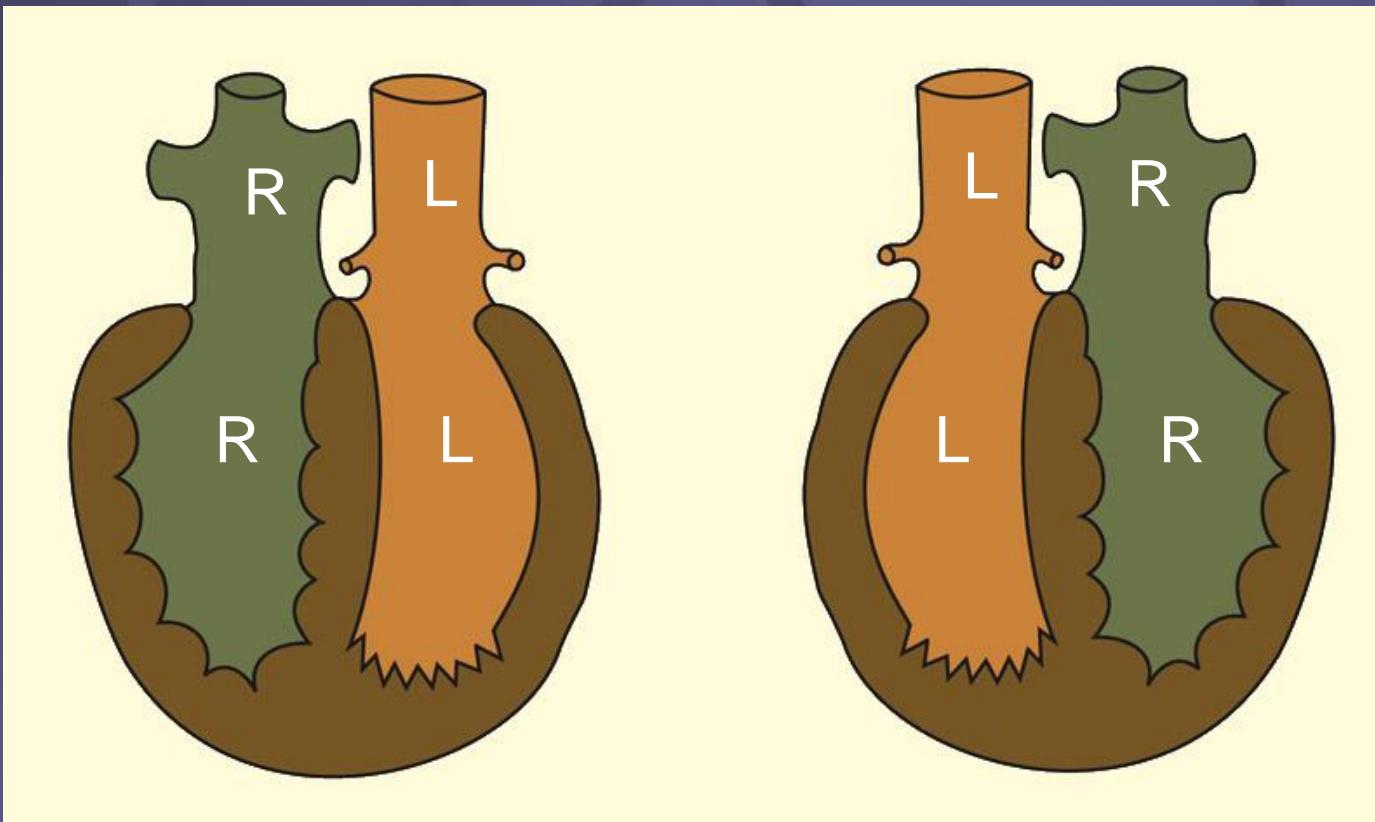
- concordant
- discordant
- double outlet
- single outlet

} either / or



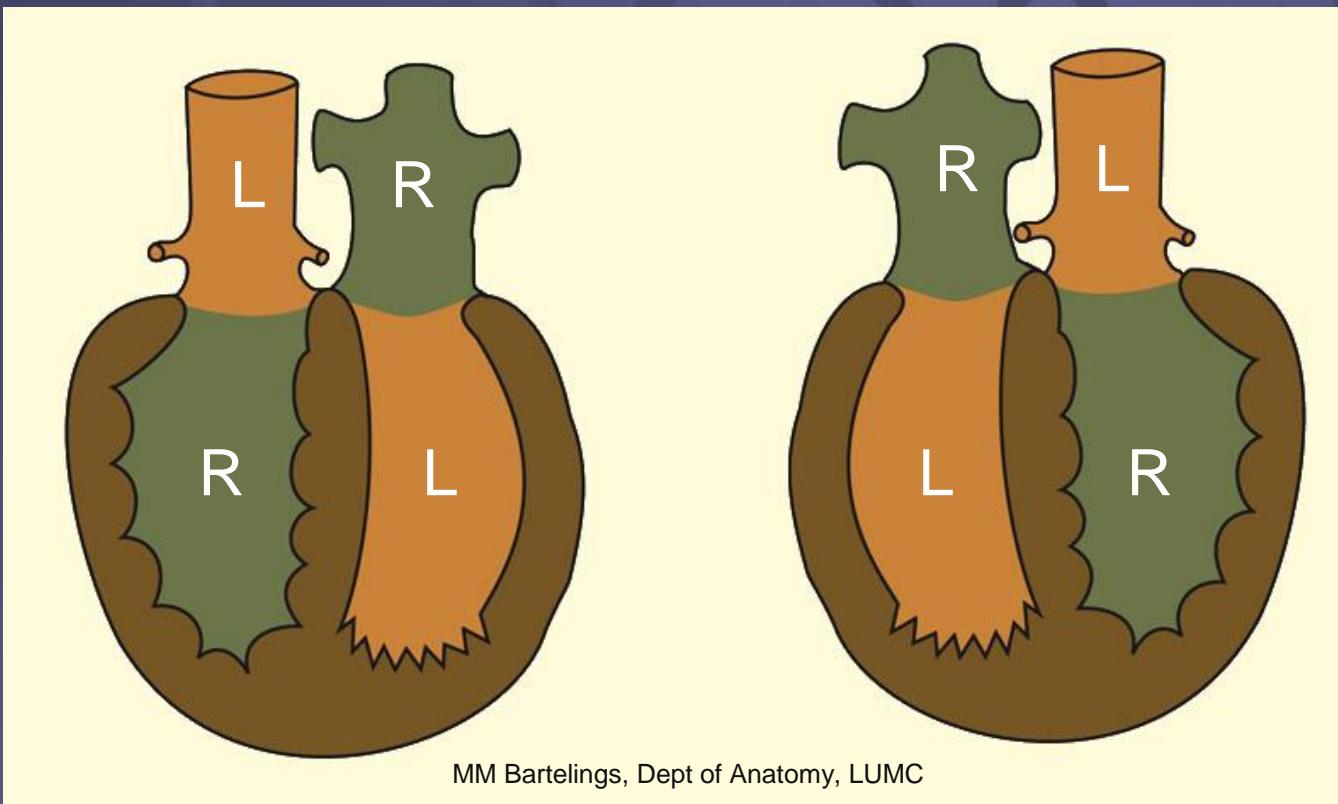
# Type of VA connection

## Concordant



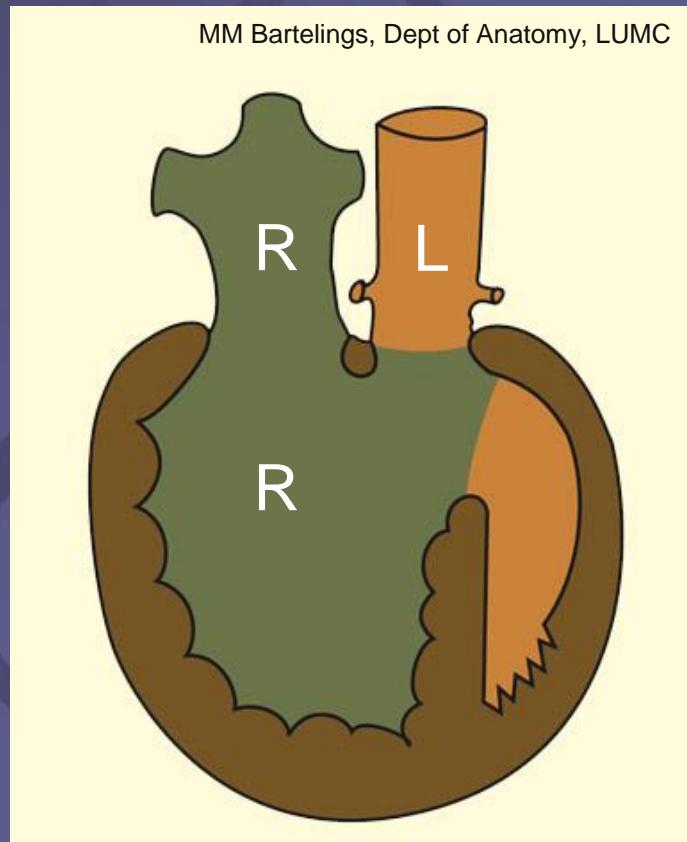
# Type of VA connection

## Discordant



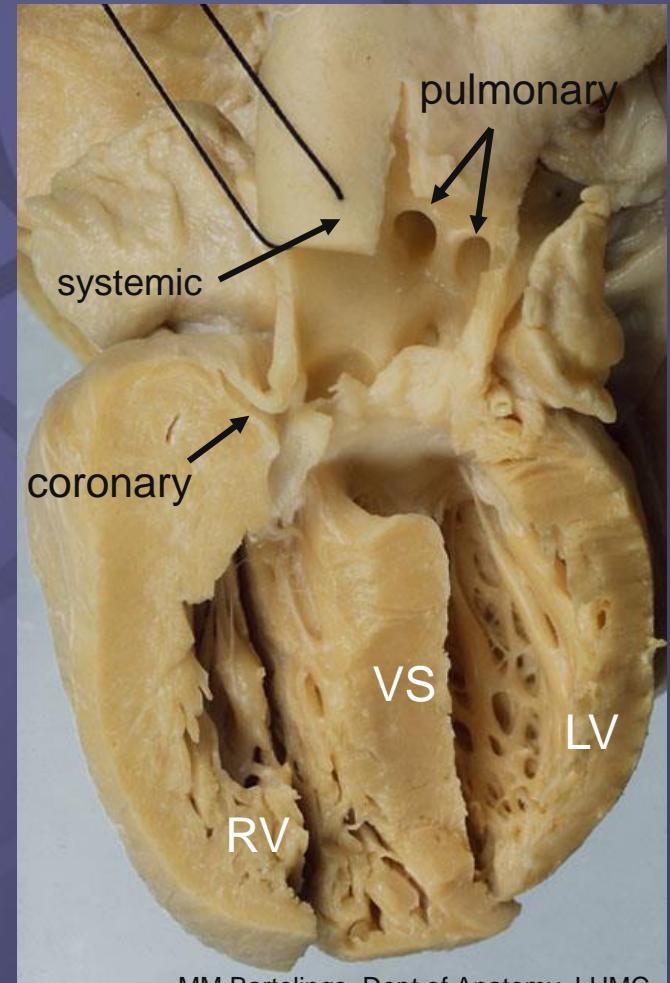
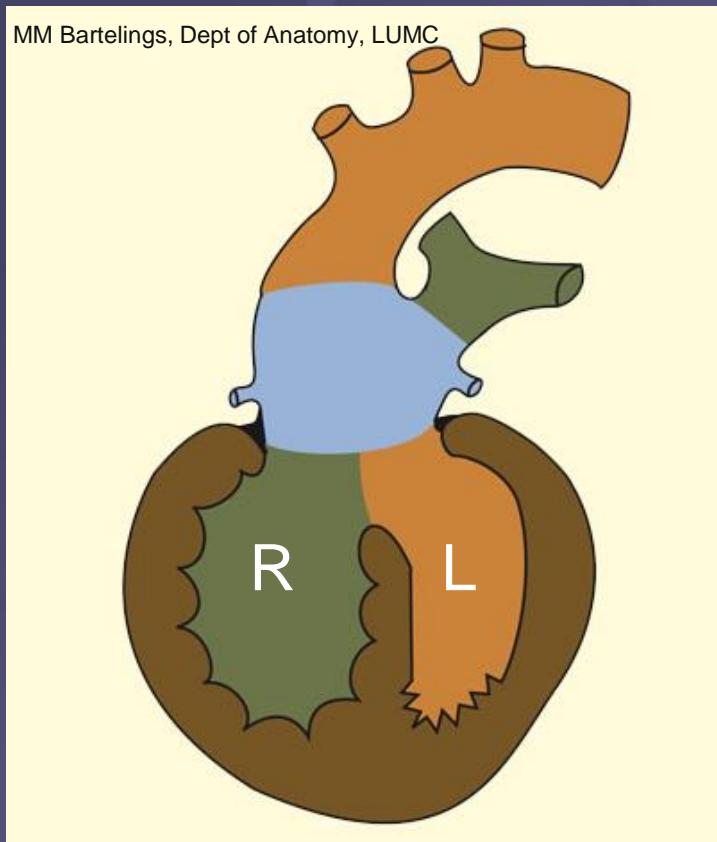
# Type of VA connection

## Double outlet



# Type of VA connection

## Single outlet



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# Type of VA connection

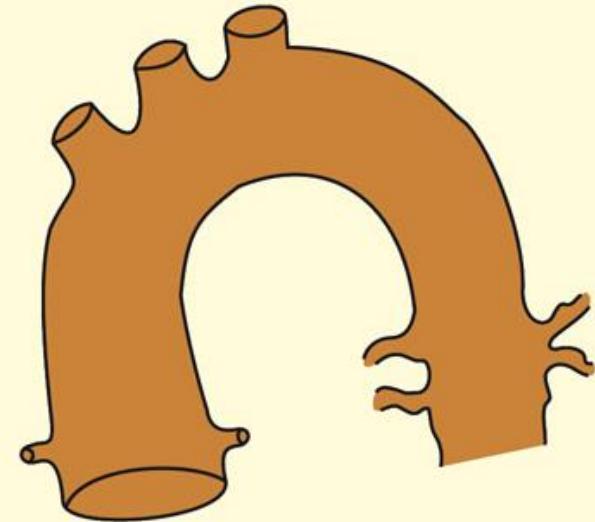
## single outlet



atresia pulmonary trunk



absent pulm. trunk



absent pulm.trunk + aa.

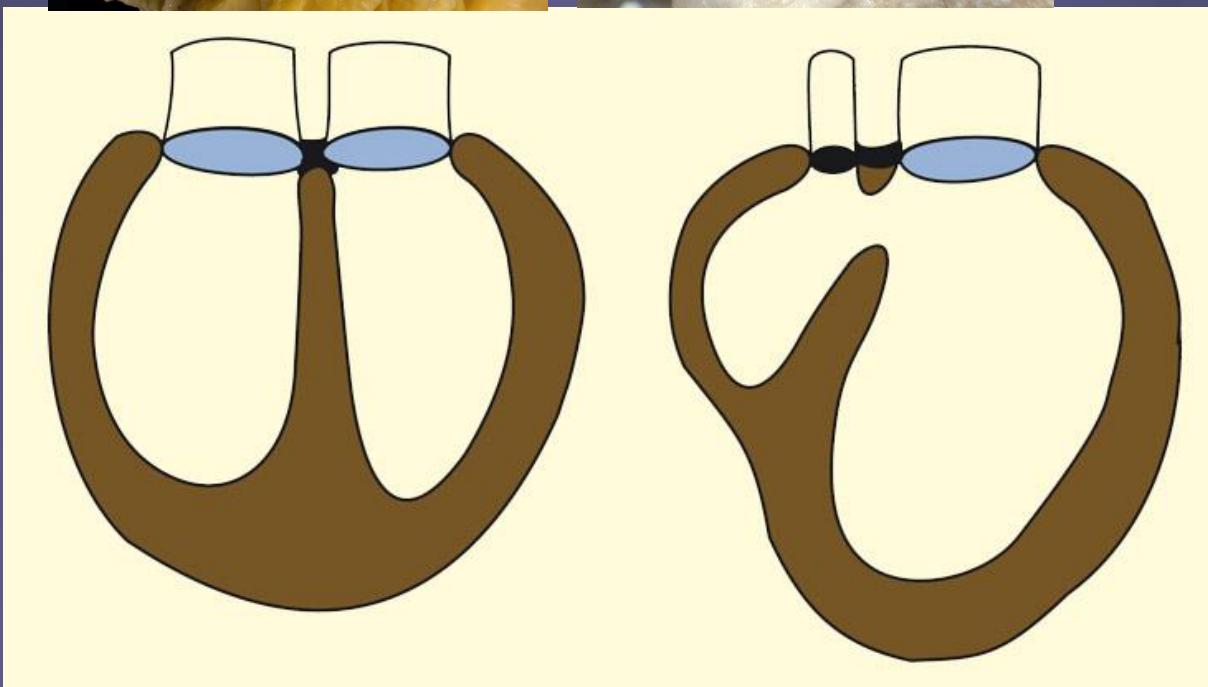
# Mode of VA connection

2 perforate valves

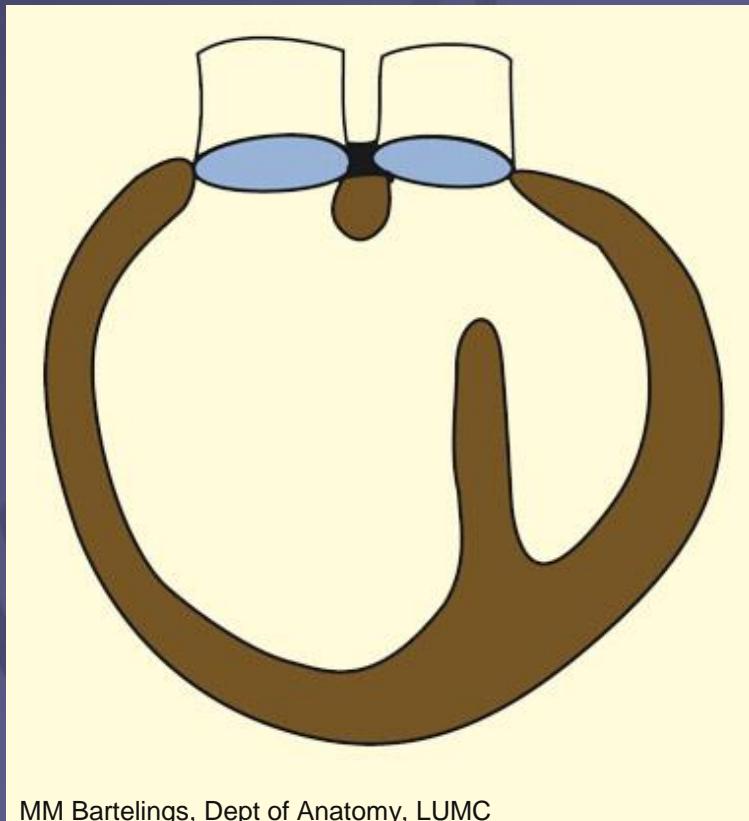


imperforate valve

(valvular atresia)



# Mode of VA connection

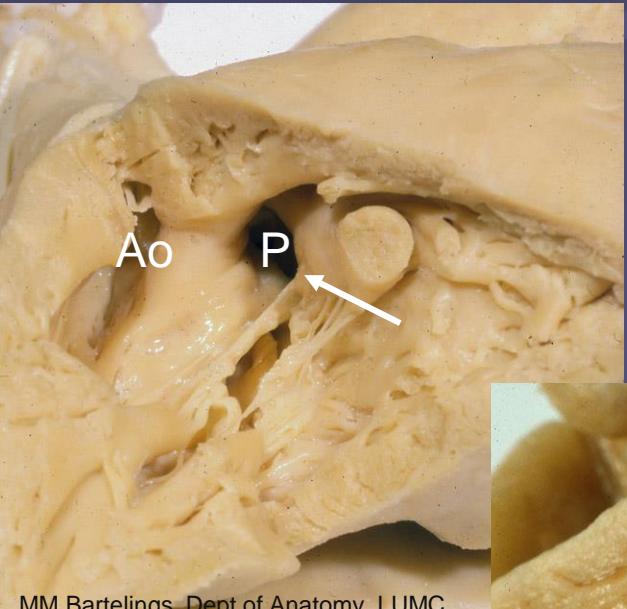


MM Bartelings, Dept of Anatomy, LUMC

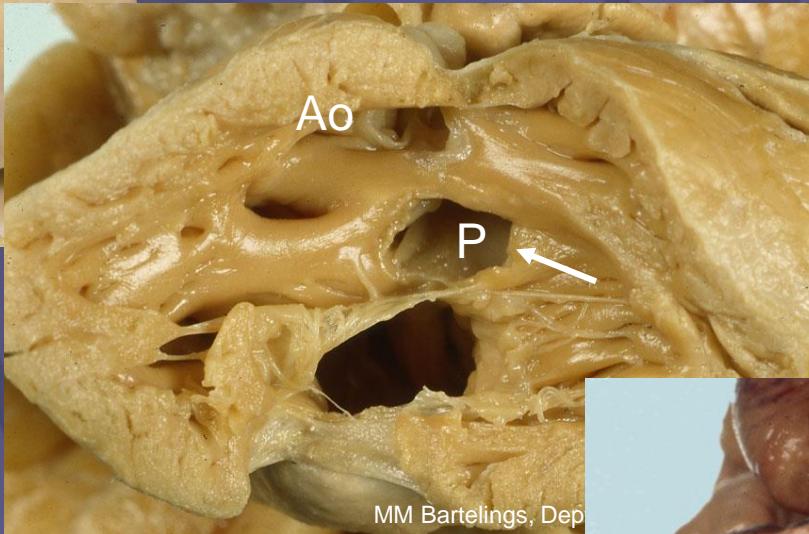
overriding orifice

50% rule

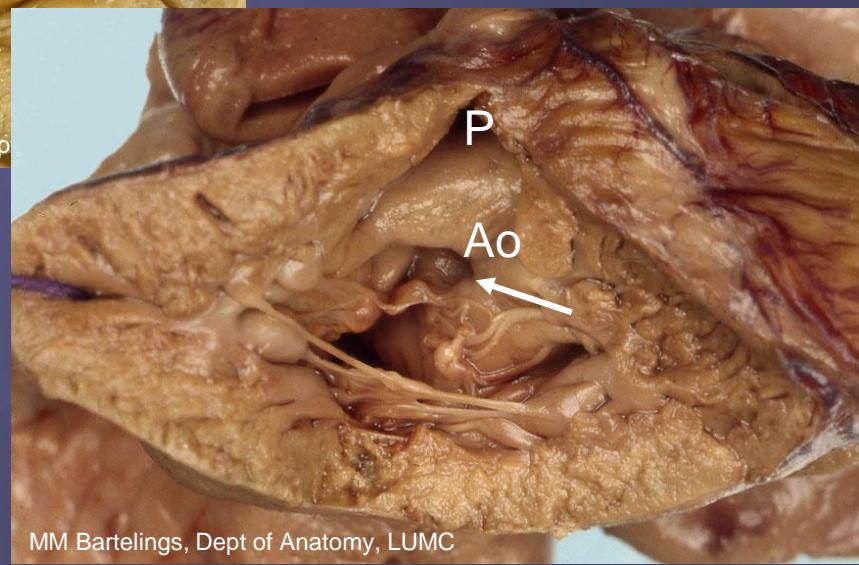
# Mode of VA connection: overriding



30% overriding P: discordant VA



65% overriding P: DORV



80% overriding Ao: DORV

# Sequential segmental analysis

I

situs of atria

type and mode of AV and VA connection

II

analysis all associated lesions

# Associated lesions

- venous connections
- atrial septum
- ventricular septum
- pulmonary arteries, ductus arteriosus
- coronary arteries, aortic arch
- myocardium, valves
- etc.

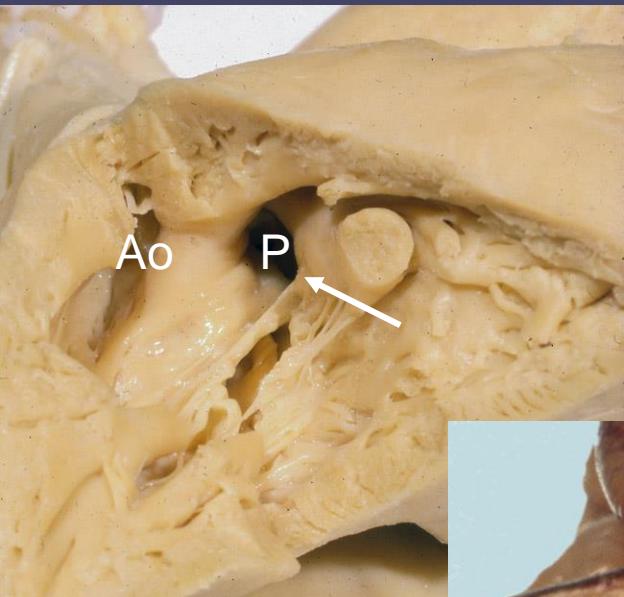
# Congenital Heart Disease

In diagnosing CHD, try to study the heart  
systematically,  
segmentally,  
sequentially !

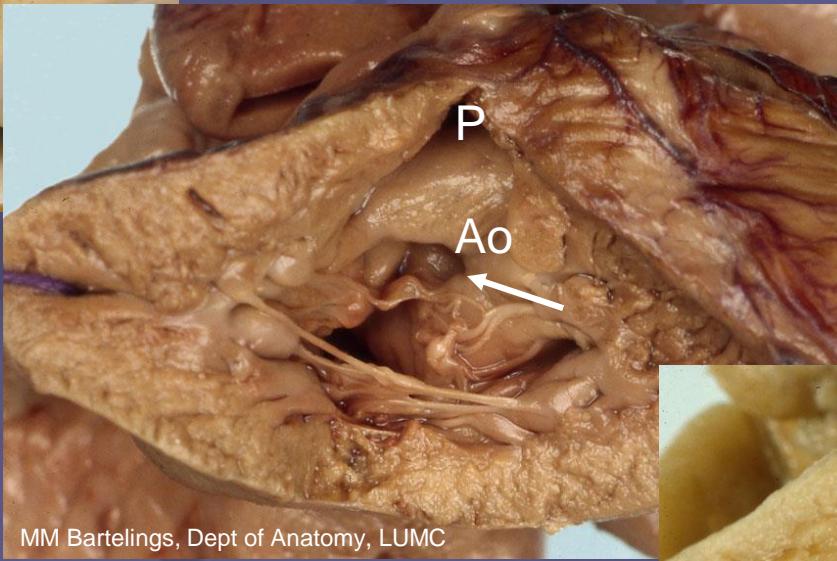


L U  
M C

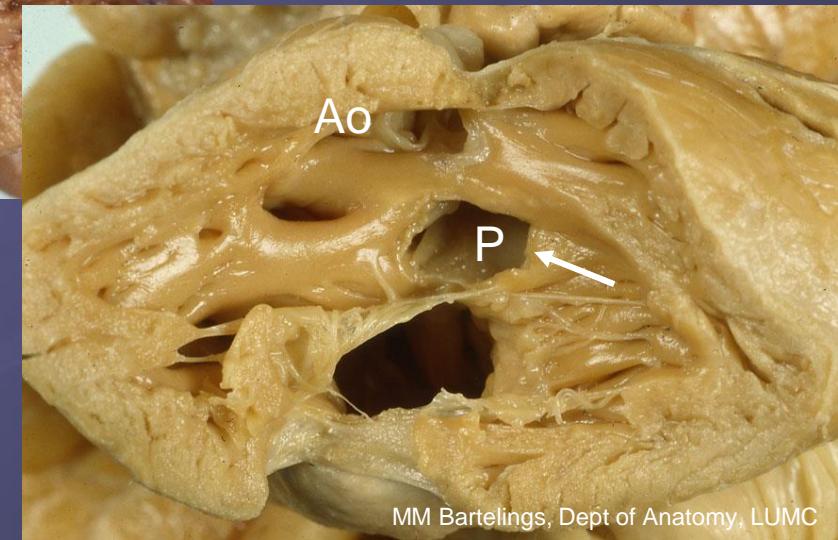
# Mode of VA connection: overriding



30% overriding P: **discordant VA**



80% overriding Ao: **DORV**



65% overriding P: **DORV**