

PAIVS

The morphologic spectrum of **Pulmonary Atresia with Intact Ventricular Septum**

Yen Ho FRCPath, FESC, FHEA
 Cardiac Morphology
 Royal Brompton & Harefield Hospitals
 Imperial College London
 UK

BCM

LinkedIn: **Brompton Cardiac Morphology**
 Website: www.rbht.nhs.uk/morphology

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Pulmonary Atresia with Intact Ventricular Septum

Chamber connections

- Almost always found with concordant AV & VA connections
- Very rarely with other connections
 - complete transposition
 - corrected transposition

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Pulmonary Atresia with Intact Ventricular Septum

Anatomic features

- Nature of atresia
- Hypoplasia of ventricular cavity
- Morphology of tricuspid valve
- Fistulous communications to coronary arteries

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Pulmonary Atresia with Intact Ventricular Septum

Nature of atresia

- Imperforate valve
- Muscular atresia

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Pulmonary Atresia with Intact Ventricular Septum

Imperforate valve dome

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Pulmonary Atresia with Intact Ventricular Septum

Muscular atresia

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Imperforate valve



Muscular atresia



Cavity of RV
Range of sizes
Not always hypoplastic

- o Right ventricular cavity size
- o Spectrum of cavity hypoplasia from tiny to dilated
- o Interpret in terms of overgrowth of cavity by wall hypertrophy
 - o apical component
 - o apical and outlet components



A spectrum of RV size

Tiny Regular Dilated



Tricuspid valve

- o Valve is often dysplastic
 - o thickened leaflets and cords
 - o rolled edges
- o May show Ebstein malformation
- o Absent leaflets (unguarded orifice)

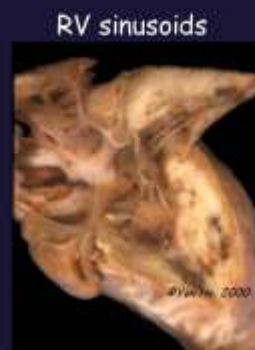


- o Dilated right atrium /ventricle
- o "Wall-to-wall" heart
- o Found with either
 - o Ebstein's malformation
 - o Partial or complete absence of leaflets of tricuspid valve

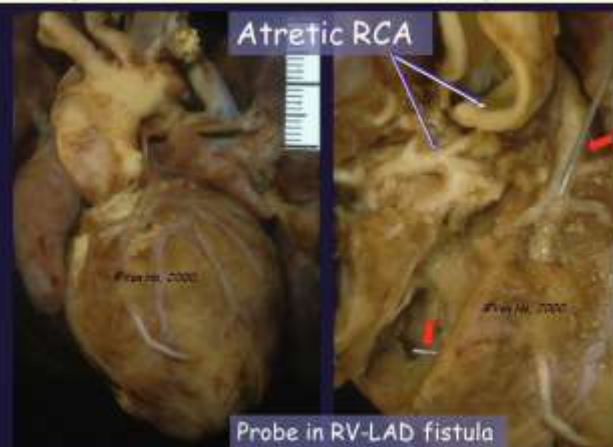
Associated malformations

- Atrial septal defect (usually within oval fossa)
- Coronary arterial malformations
 - anomalous origin
 - sinusoids: fistulous communications (most frequent with small RV cavity)
 - atretic segments
 - intimal obliteration of lumen

Coronary artery: anomalous origin from pulm trunk



Coronary artery: sinusoids



Groupings within overall spectrum

- ♥ with dilated right heart - very poor prognosis
- ♥ with muscular atresia - sinusoids, small ventricle, etc
- ♥ with imperforate pulmonary valve - best anatomy

Summary

Morphologic spectrum

- Chamber connections
- Right ventricular morphology
 - cavity size
 - tricuspid valve
 - ventriculo-arterial junction
- Pulmonary arteries and blood supply
- Associated malformations

