Pulmonary Atresia With Intact Ventricular Septum -

Anatomy, Physiology, and Diagnostic Imaging

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Disclosure Information

Pulmonary Atresia with Intact Ventricular Septum (PAIVS)–

Anatomy, Physiology, and Diagnostic Imaging

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As a faculty member for this program, I disclose the following relationships with industry:

None pertaining to this presentation

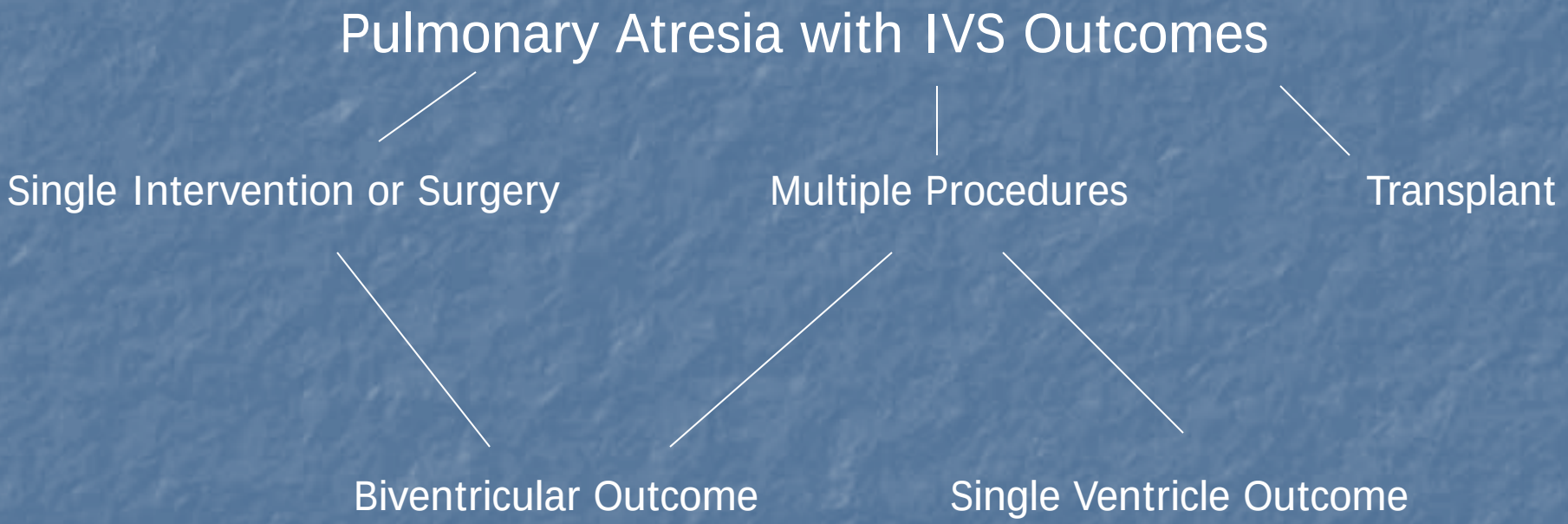
Goals of Presentation

- Review anatomy of PAIVS
- Give examples of diagnostic imaging – echo and angiography - to illustrate anatomy
- Review aspects of physiology important to clinical care and anesthesia management

PAIVS Overview

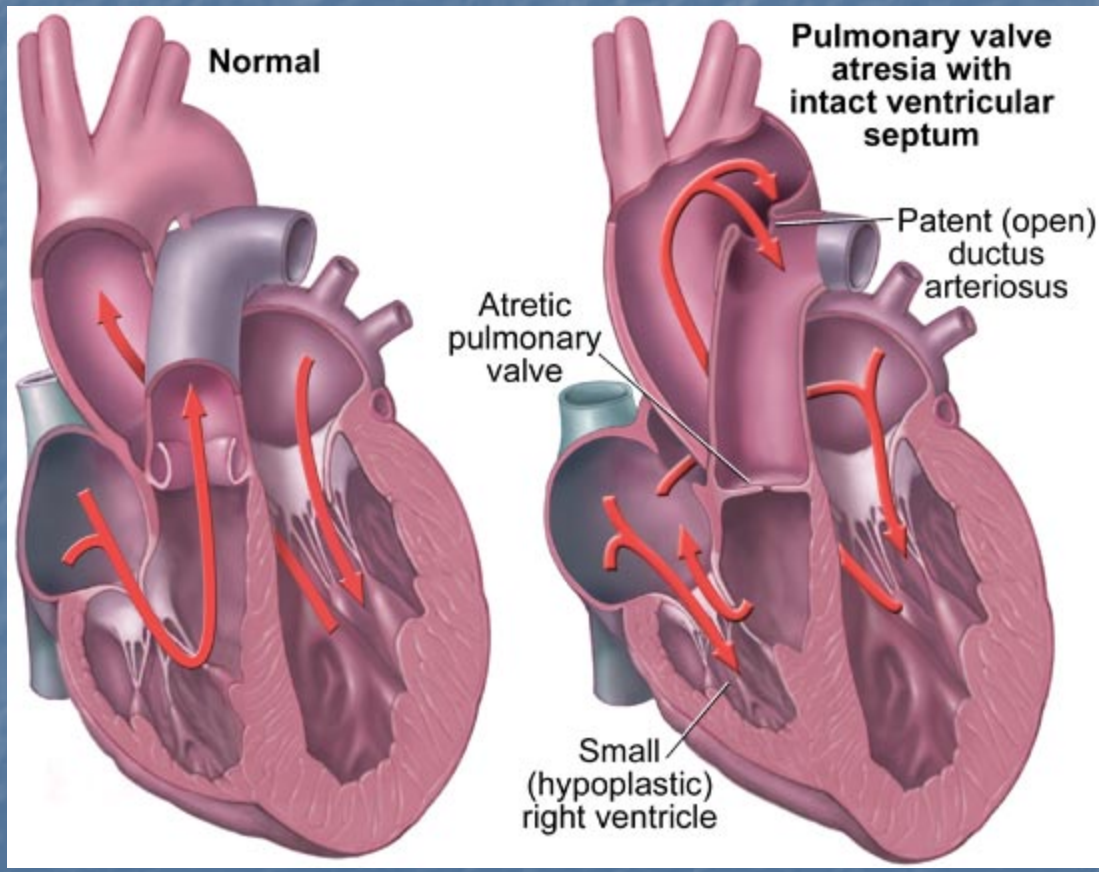
- About 3% of CHD
  - 4-8 / 100,000 live births
  - One of the more common cyanotic lesions
- Probably occurs later in morphogenesis than PA VSD
  - Ventricular septum formed
  - PA's often near normal size
- Cause is unknown

Why is Anatomy and Physiology Especially Important in PAIVS ?



Major Anatomic Considerations

- Atretic valve
- RV
- Tricuspid valve
- ASD
- PDA
- Pulmonary arteries
- Coronary arteries



Atretic Pulmonary Valve

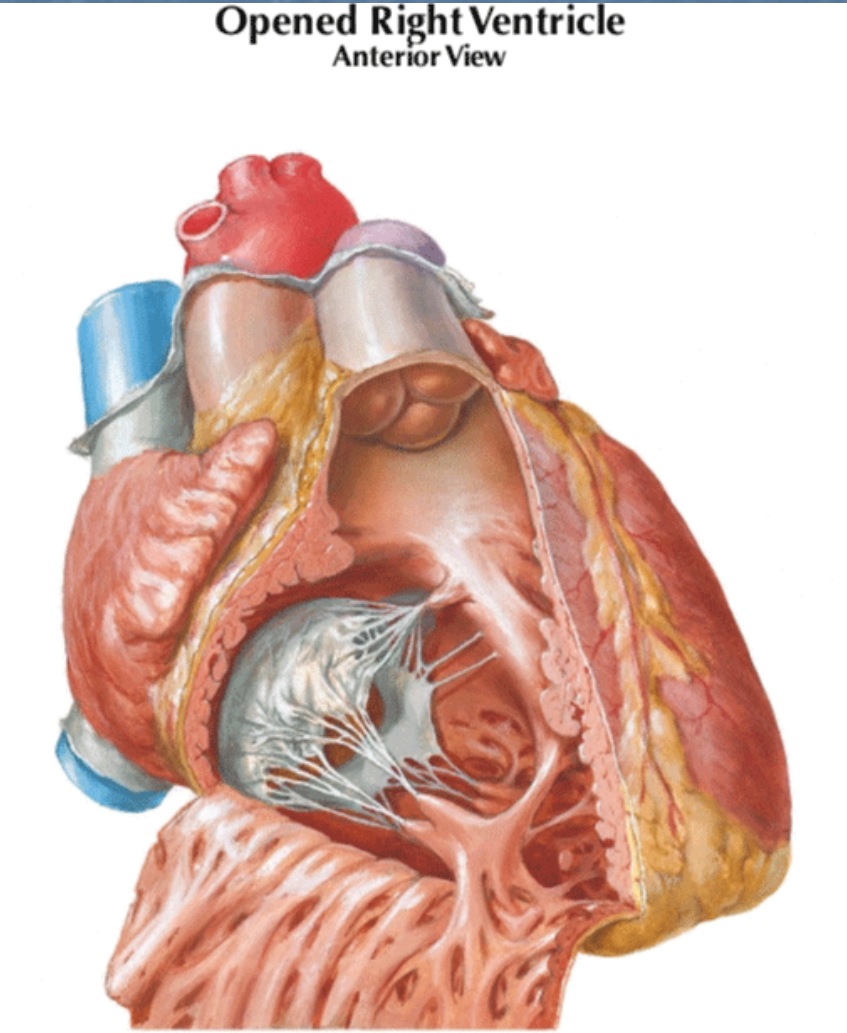
- May be well formed with fused leaflets
- Degree of development tends to correlate with development of RVOT



Nykanen D; Moss and Adams

RV Size and Features

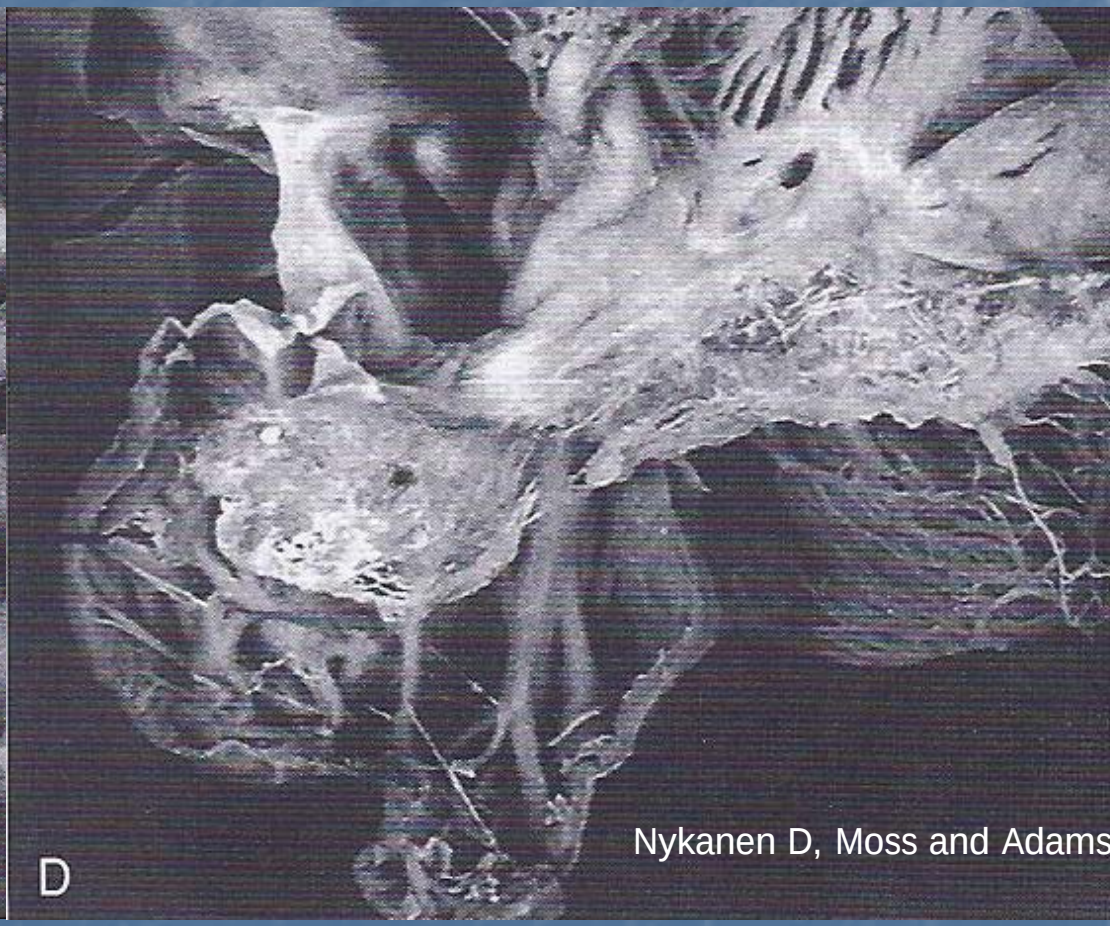
- Many feel RV is tripartite
  - All parts tend to be hypoplastic, but variably
- Tricuspid valve correlates best with overall RV size
- Outlet correlates best with PA anatomy and biventricular repair



Tricuspid Valve is Usually Abnormal

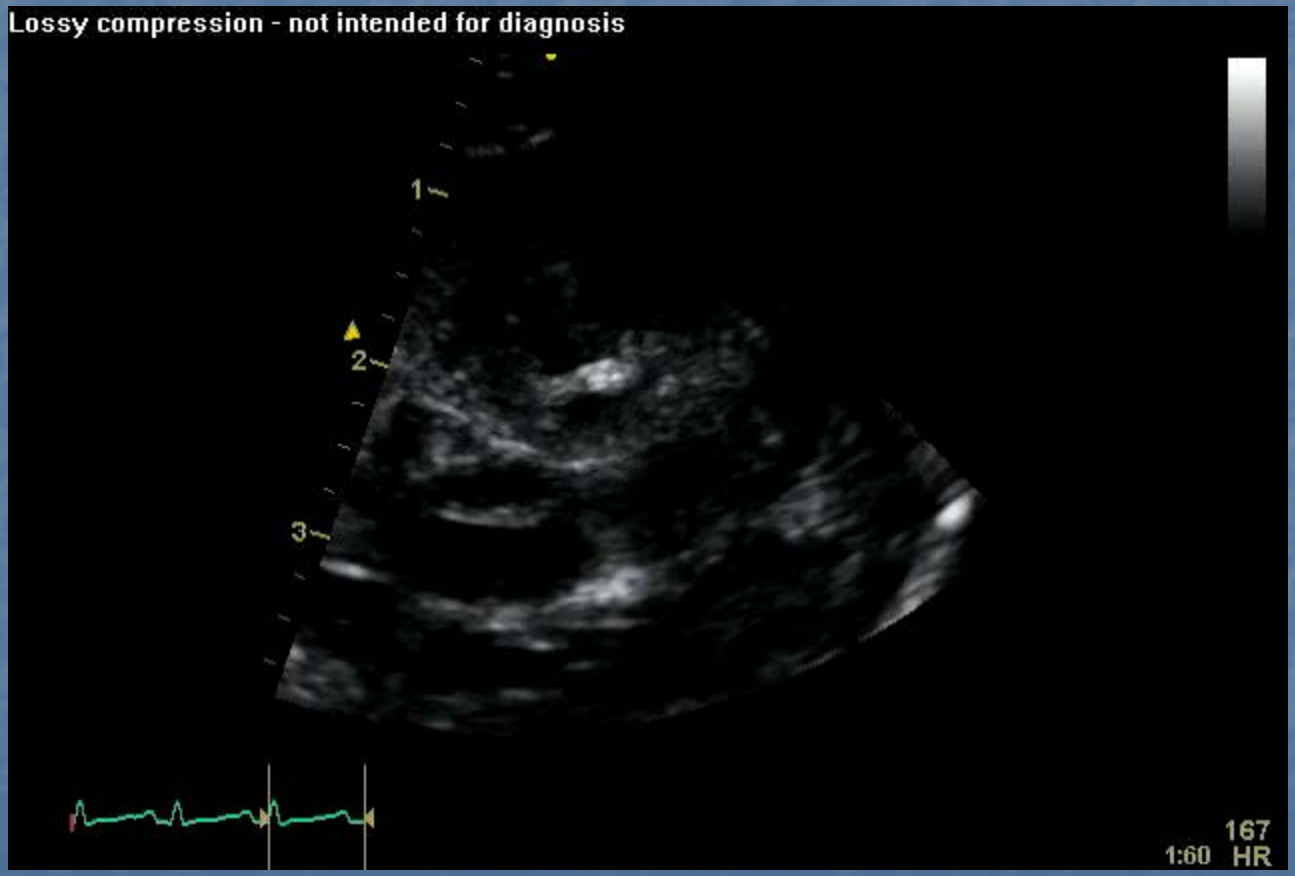


Stenotic and Hypoplastic Tricuspid Valve

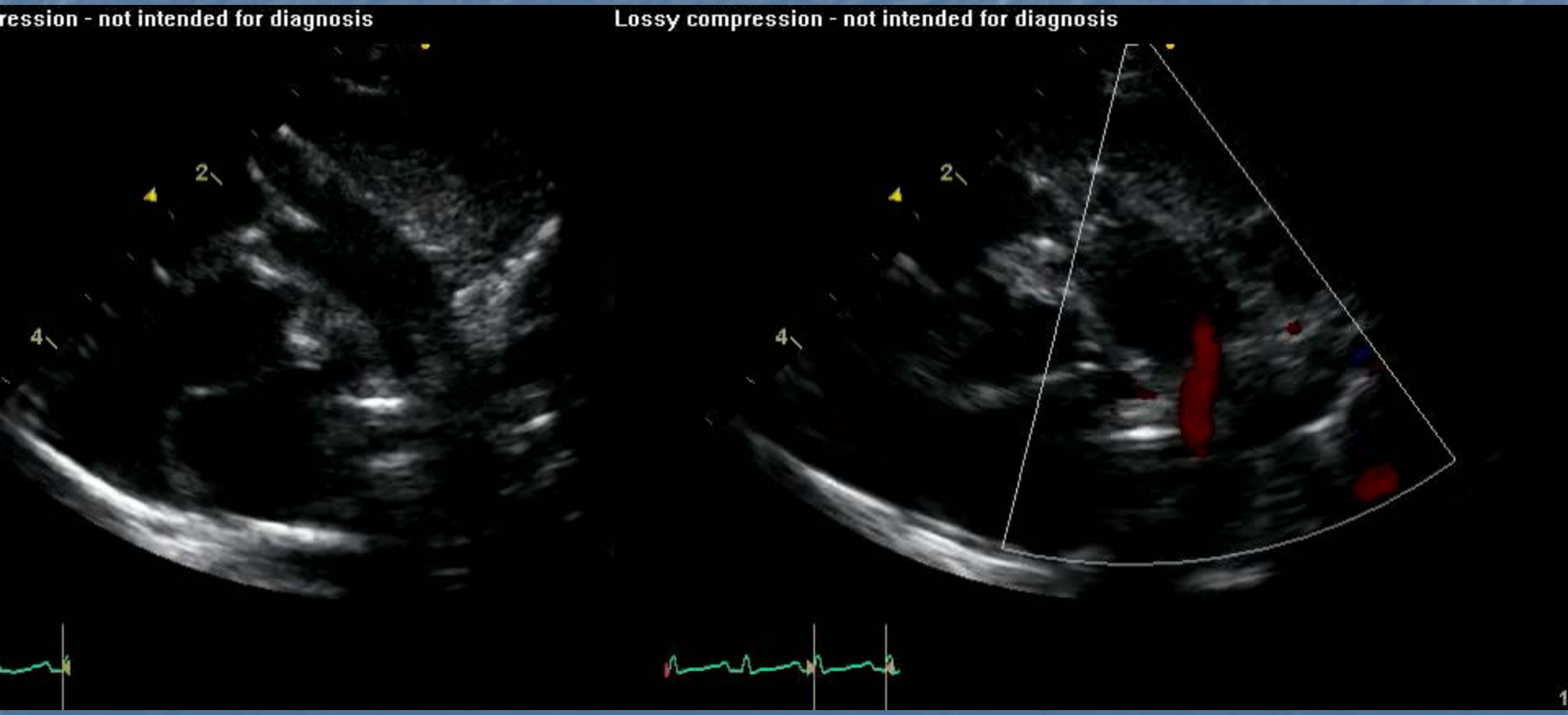


Severely Dysplastic TV with Severe TR and Enlarged RV

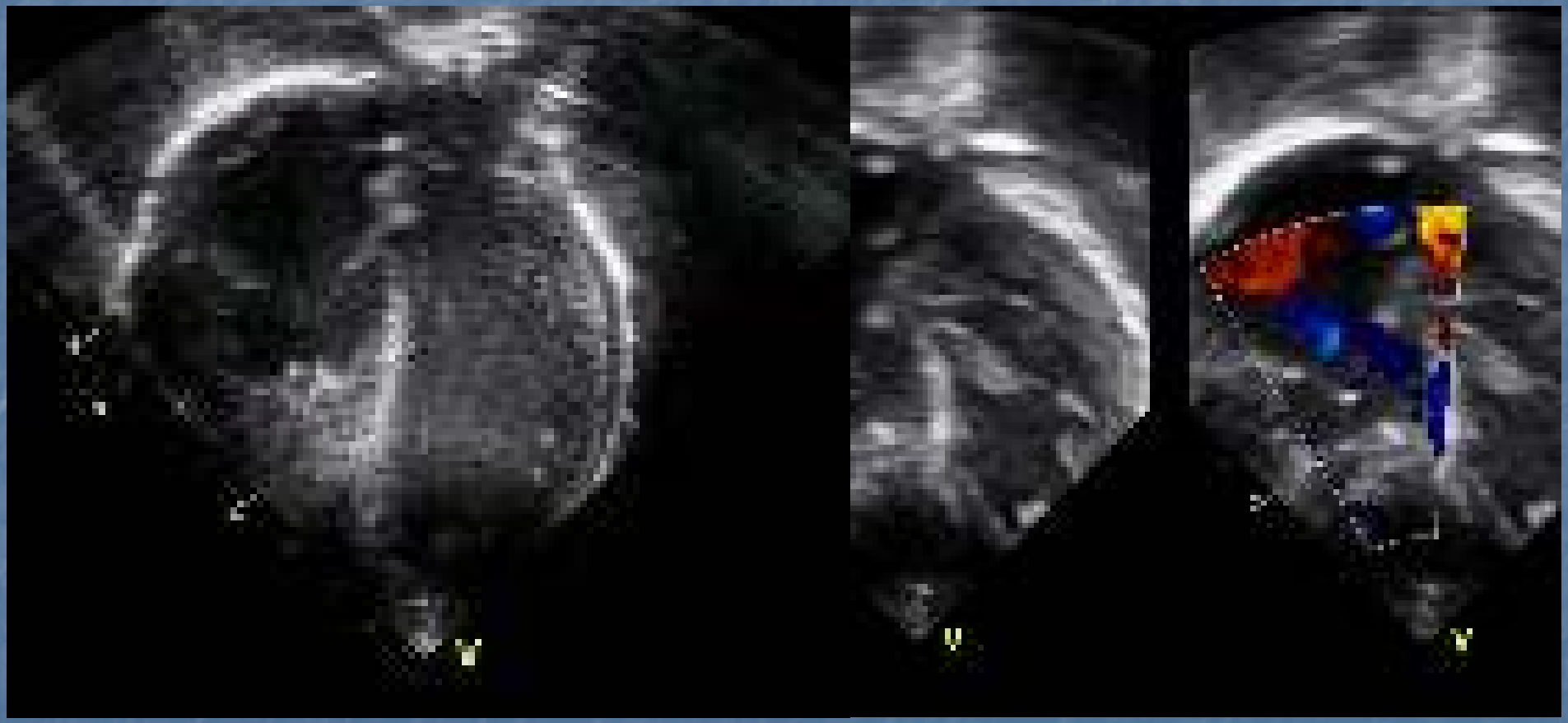
Echo Short Axis Pul Vlv Pre



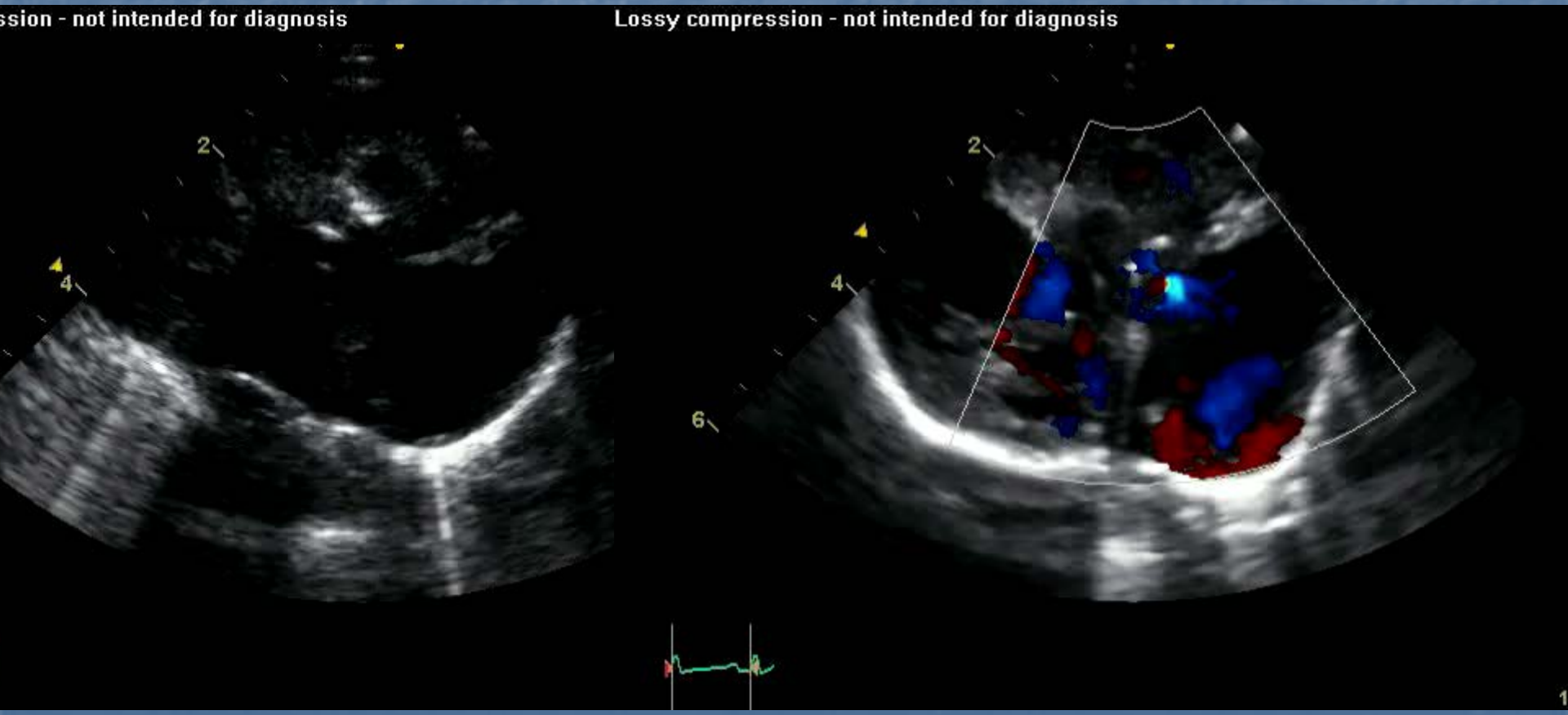
RVOT and Pul Vlv Echo Pre



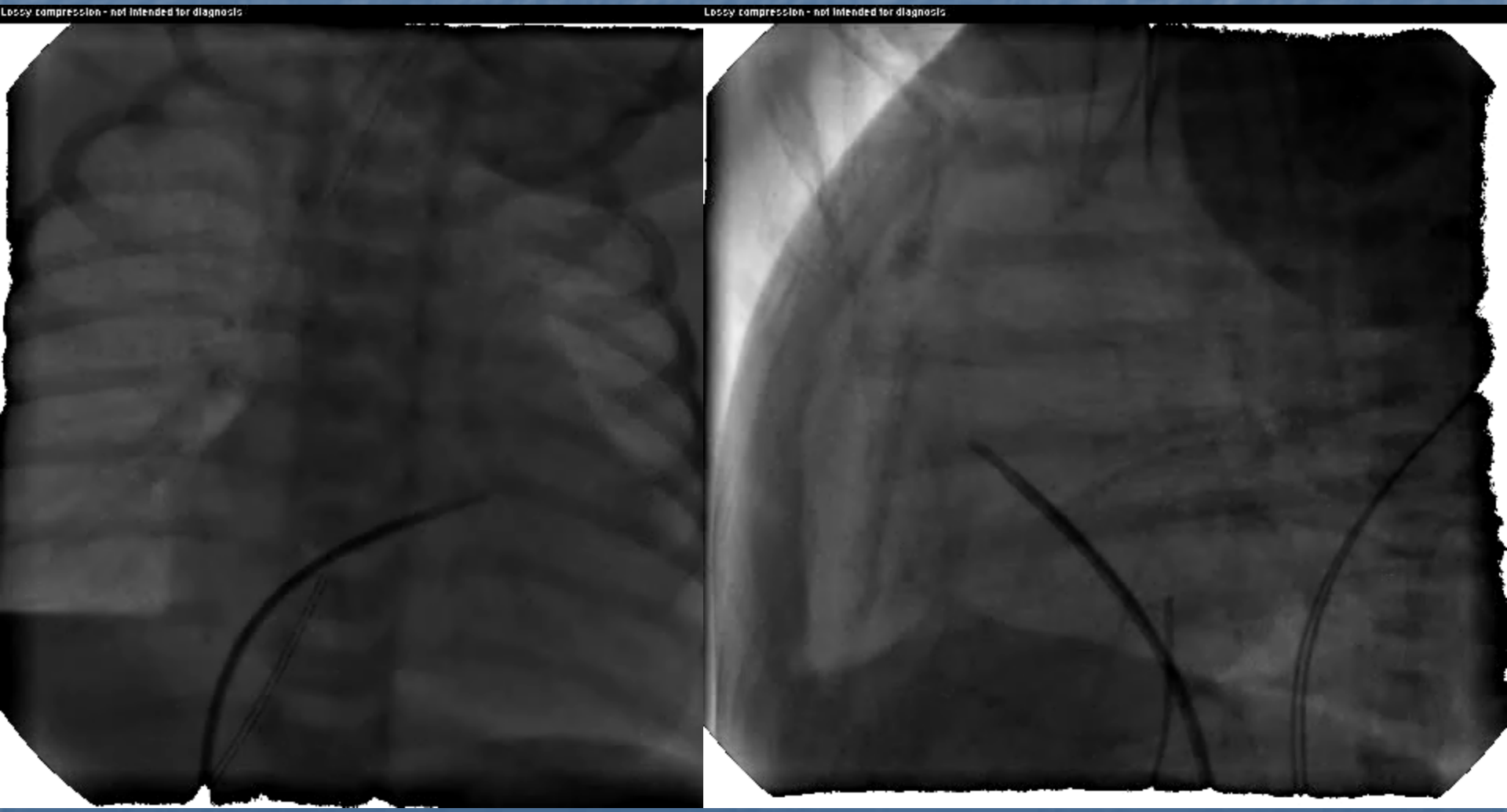
Echo of TV and RV



Echo RV Long Axis

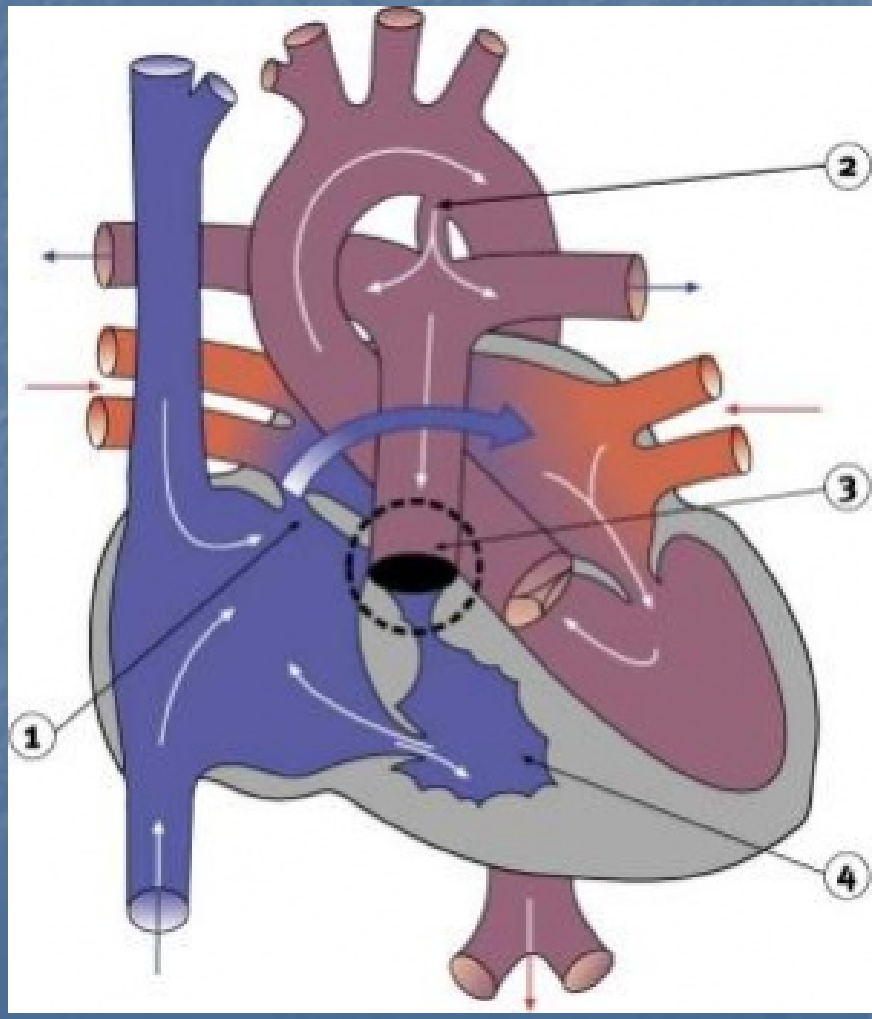


RV Pre- AP and Lat



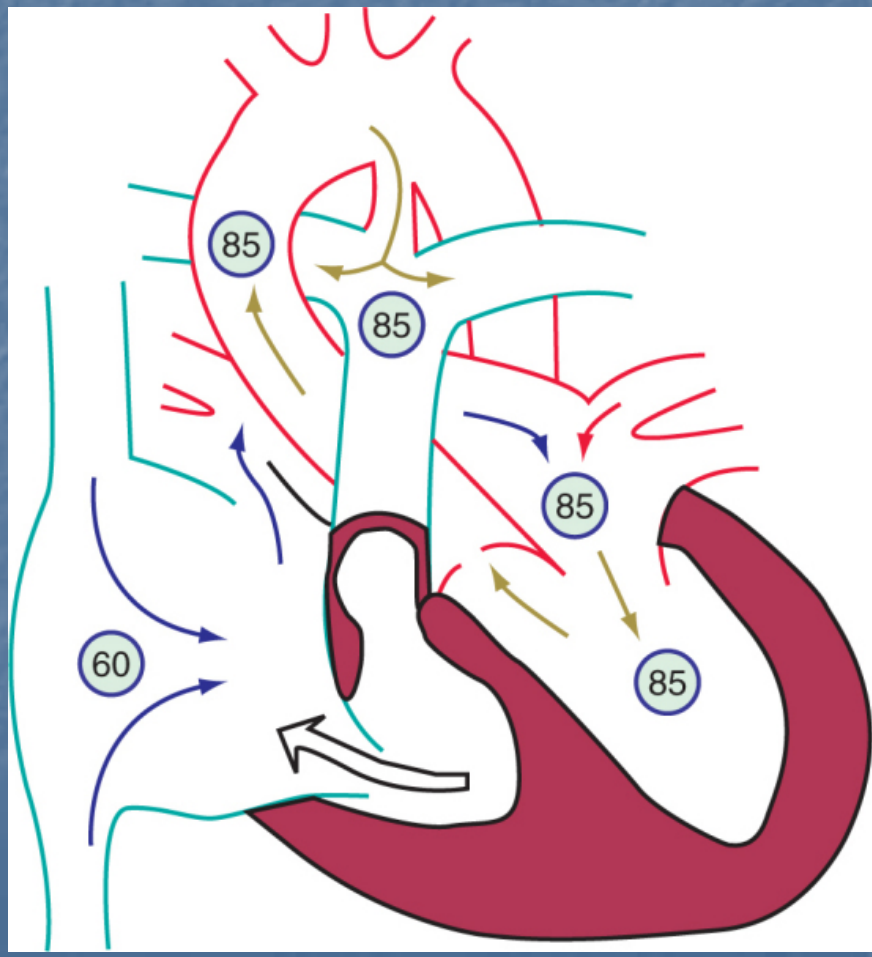
Interatrial Communication in PAIVS

- Usually PFO or small ASD
- Essential for survival if no flow through RV
- For Single V pathway, want it wide open
- For Bi V pathway, want it small / closed



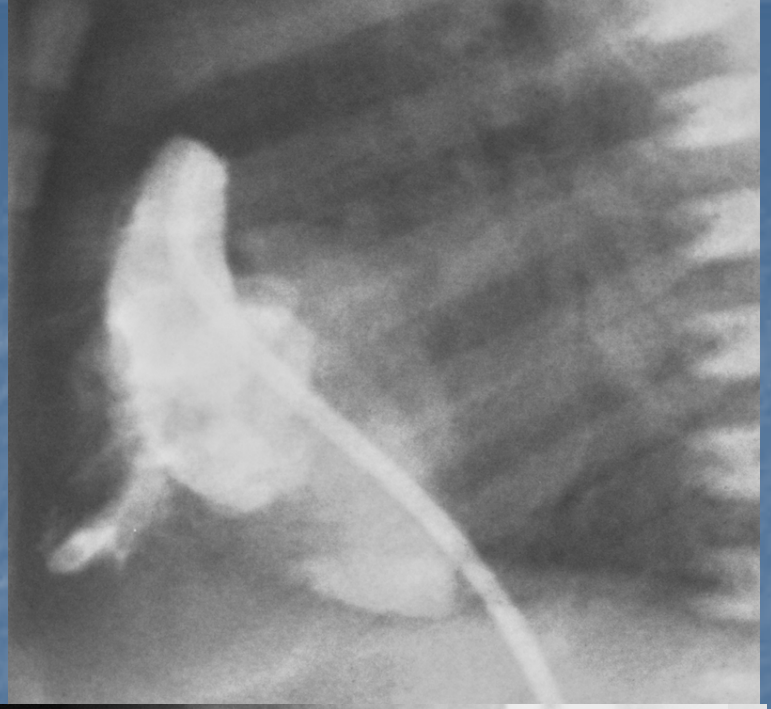
PDA in PAIVS

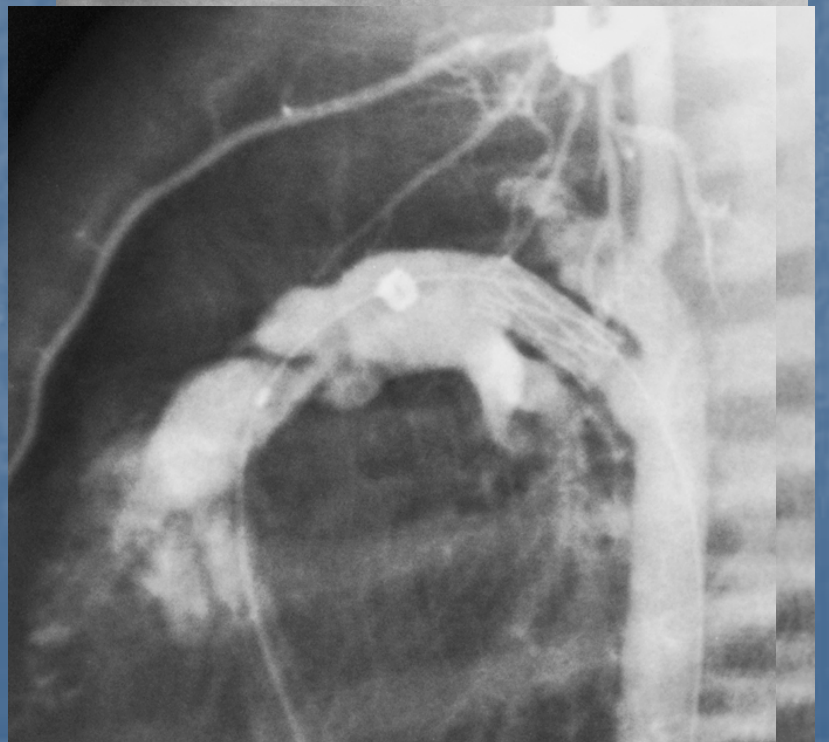
- Essential pathway for pulmonary blood flow early >> PGE
- For Single V pathway want PDA or shunt till Glenn
- For Bi V pathway, may want open for while, but eventually closed



PDA Stents in PAIVS

- PDA often helpful temporarily even if Bi - V pathway
- Anatomy usually amenable to stent if good RV
- Stented PDA can be closed later by cath procedure if necessary





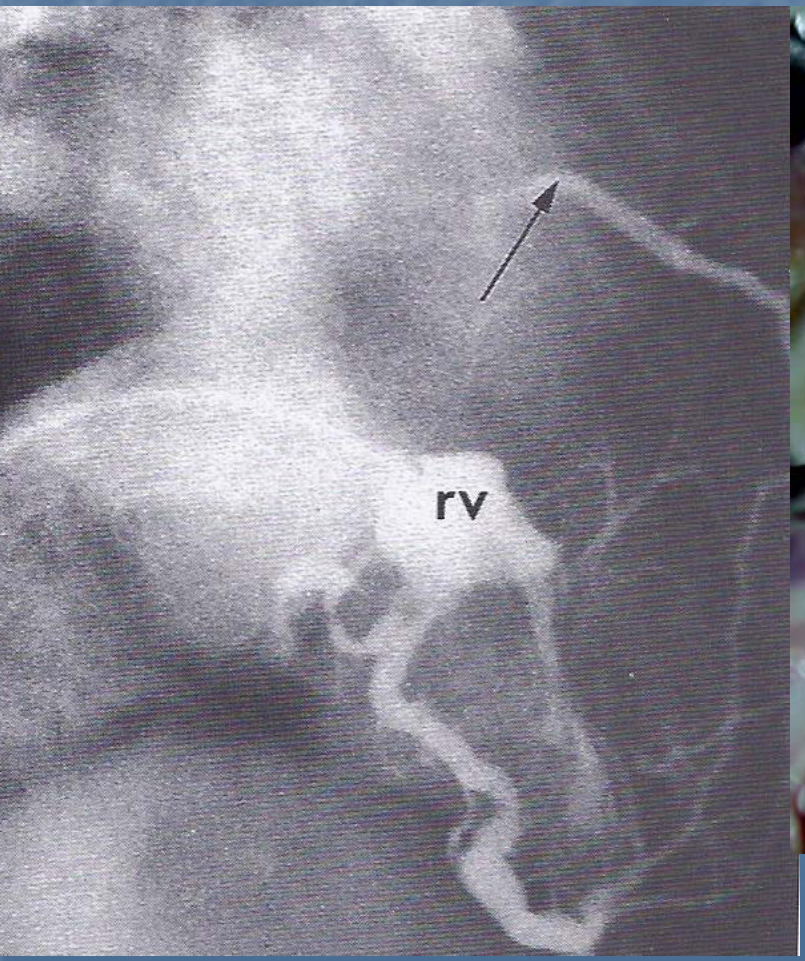
Pulmonary Arteries in PAIVS

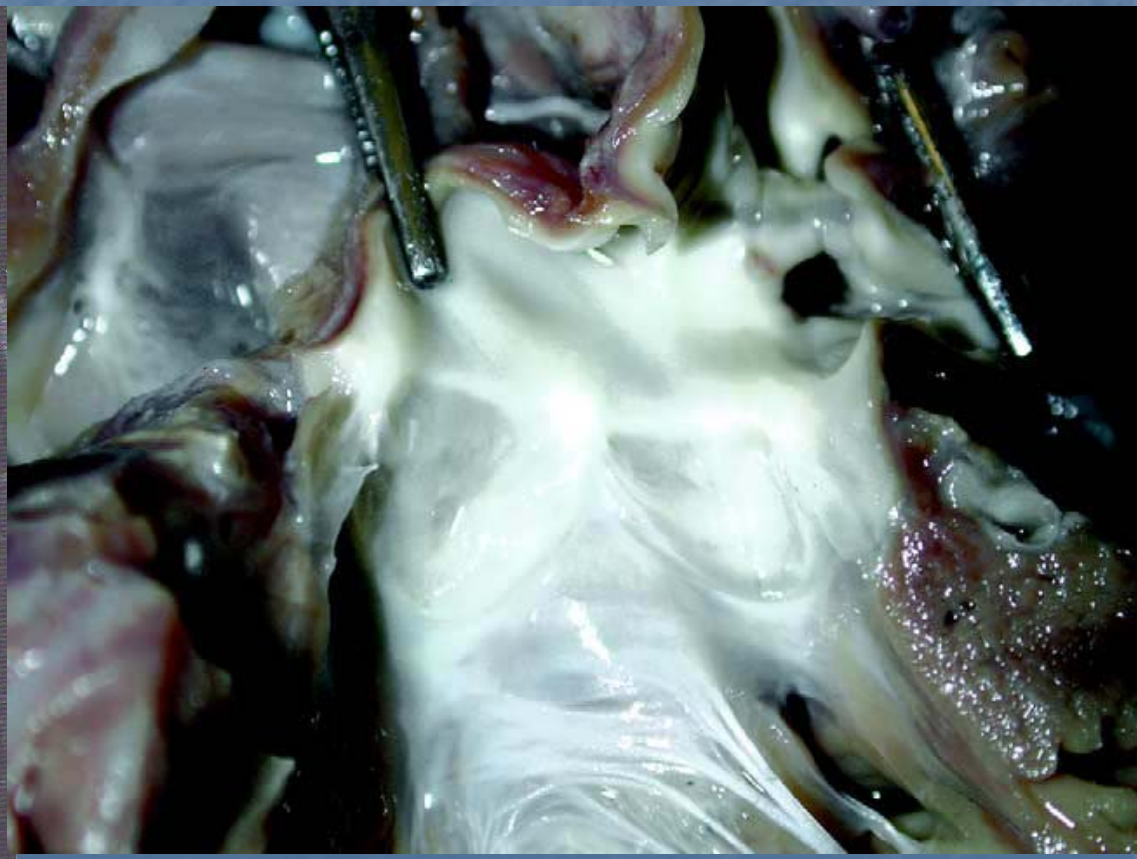
- Different from Pas in PA VSD
  - Usually near normal size
  - MAPCAs rare
  - Only rarely a major determinant of outcome
- Coarctation of LPA at ductal insertion is sometimes seen

Coronary Arteries in PAIVS

- RV blood needs to go somewhere
- If no TR, then coronary artery connections open up
  - Highest pressure RVs tend to be small to very small with little or no outflow tract
  - Really tiny RV may have little inflow and not large coronary connections
- Severe TR associated with large RV, low RV pressure and no coronary connections

RV Dependent Coronary Circulation – Ostia May Be Atretic



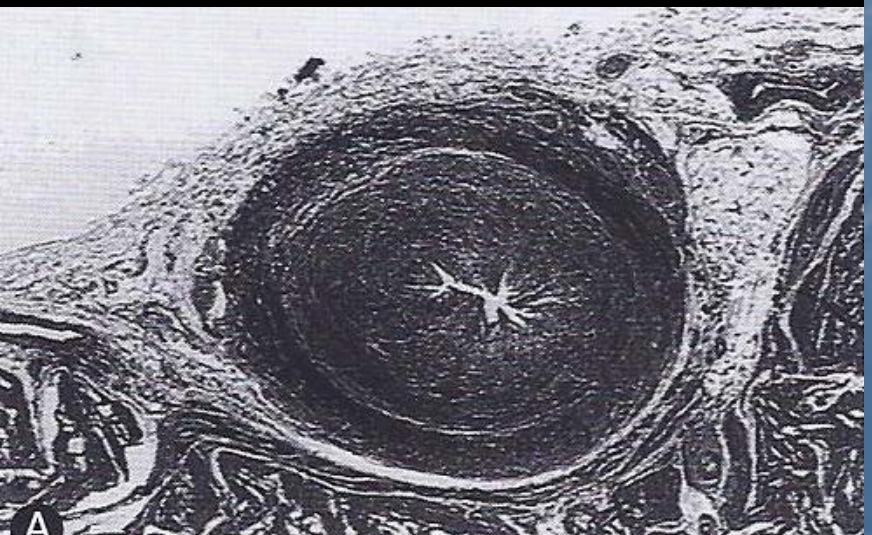


Nykanen D, Moss and Adams

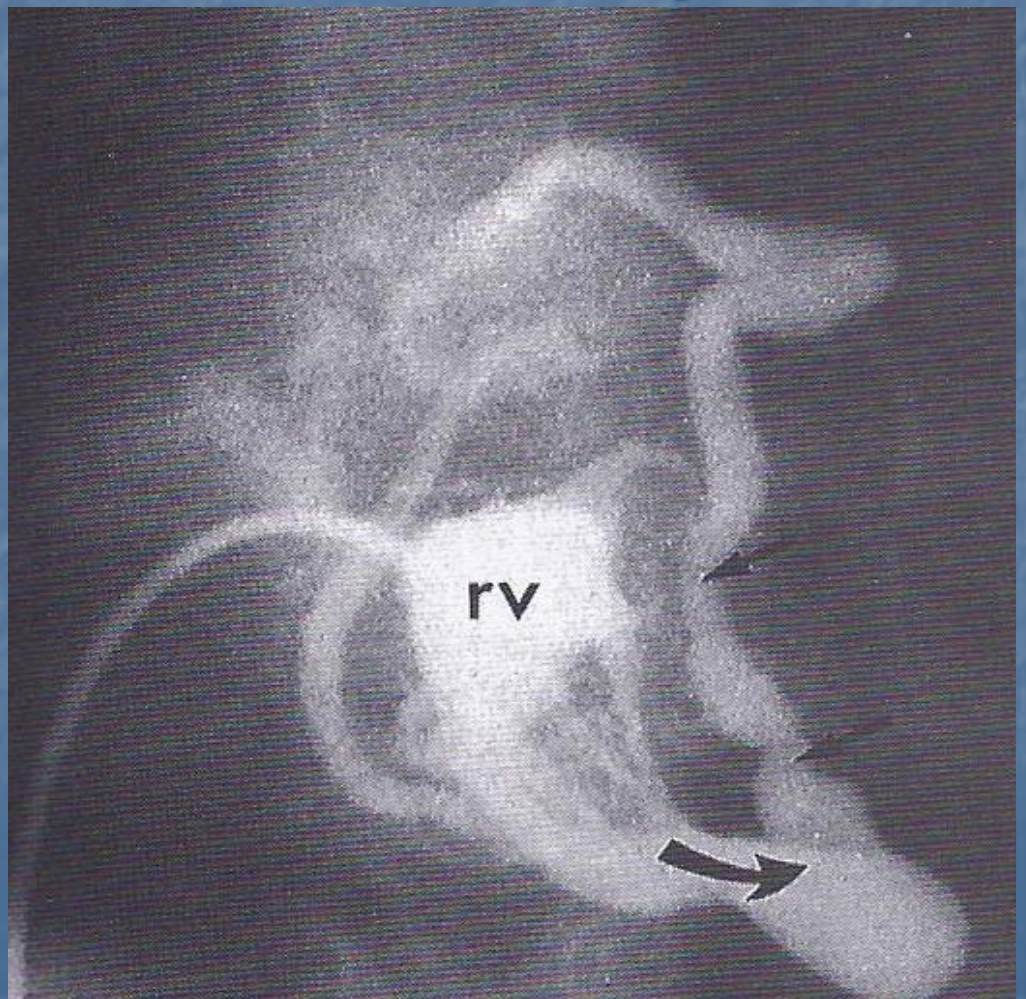
Lajos et al;An Thor Surg 2004

Pulmonary Atresia IVS





Coronary Dependence May Develop



Coronaries in PAIVS

- Sudden marked RV decompression can cause “steal” phenomenon (like coronary artery fistula)
- RV-coronary connections tend to regress if RV pressure moderately reduced

RV Growth Post Valvotomy

- Restrictive RV physiology common early after opening RVOT
- Diastolic filling improves over time and R-L atrial level shunting improves
- ASD flow often R-L early, but becomes L-R in weeks to months if good relief of RVOT obstruction

Summary PAIVS

- Wide spectrum of associated issues
  - RV size, tricuspid valve, coronaries
- Echocardiography and angiography are primary imaging modalities
- Physiology highly variable
- Primary anesthesia issues
  - Pulmonary flow
  - Coronary flow
  - Blood pressure