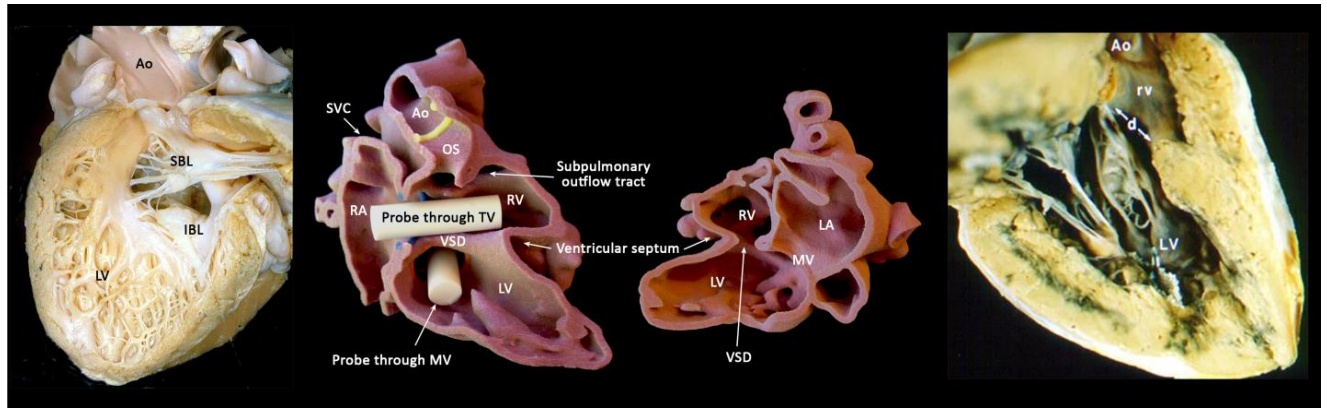
2nd Contemporary Morphology Course with Specimens and 3D Print Models
CONGENITAL HEART DISEASES IN YOUR HANDS
♥ Atrioventricular Septal Defects ♥



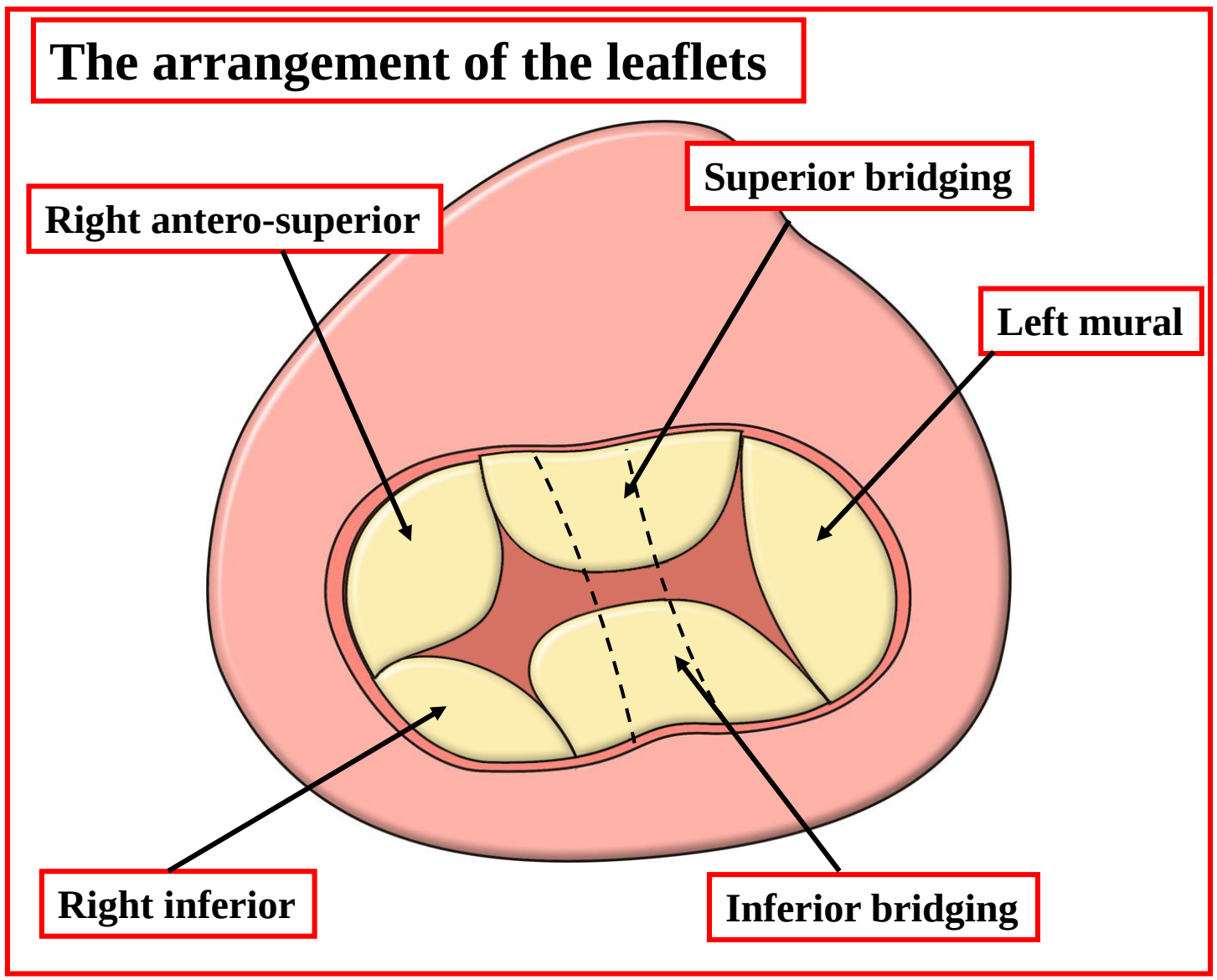
Mike Seed, MD

Head of Division of Cardiology

Atrioventricular septal defect - basics

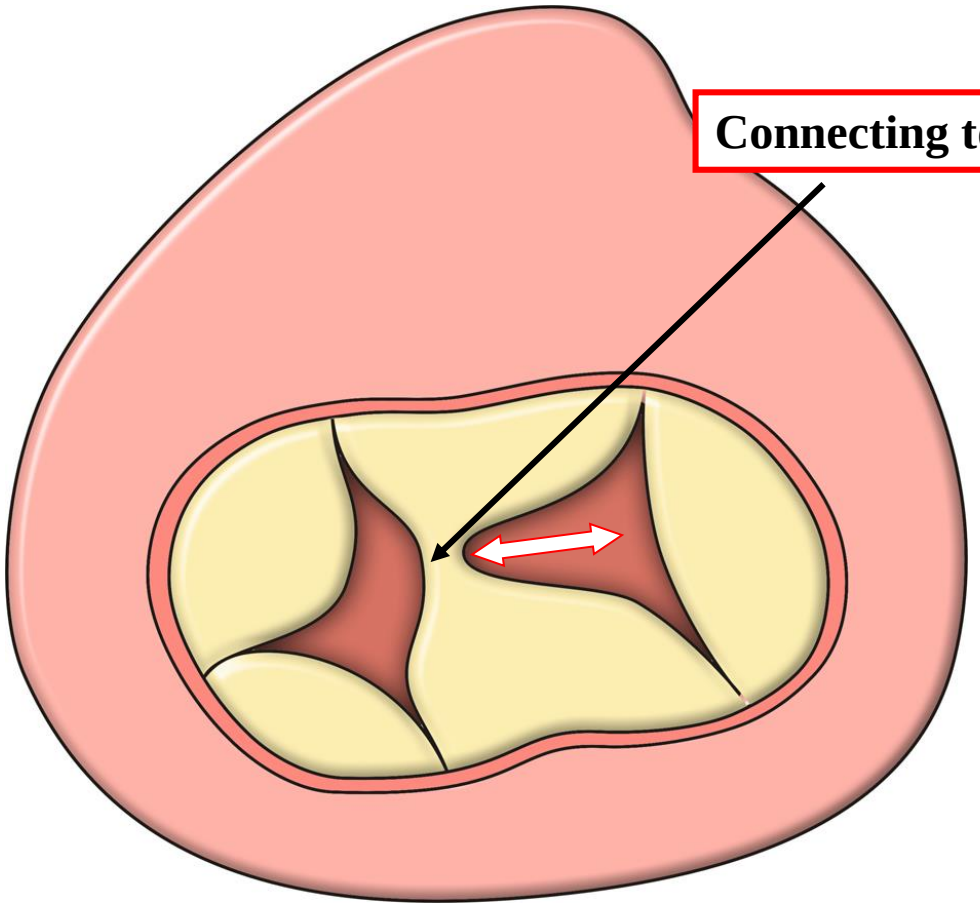
- A common versus separate atrioventricular connections
- The common valve has five leaflets, superior and inferior bridging leaflets a mural leaflet in the left ventricle and an antero-superior leaflet and an inferior leaflet in the right ventricle
- The orifice may also be single or partitioned
- Communication between the right and left heart may be at the atrial level (ostium primum defect), ventricular level or both
- Partial AVSD – communication just at atrial or ventricular level
- Complete AVSD – communication at both atrial and ventricular level
- Balanced or unbalanced – depending on commitment of the valve to the right and left ventricles
- Variable extent of obstruction and valvar regurgitation

The arrangement of the leaflets



Courtesy Robert Anderson

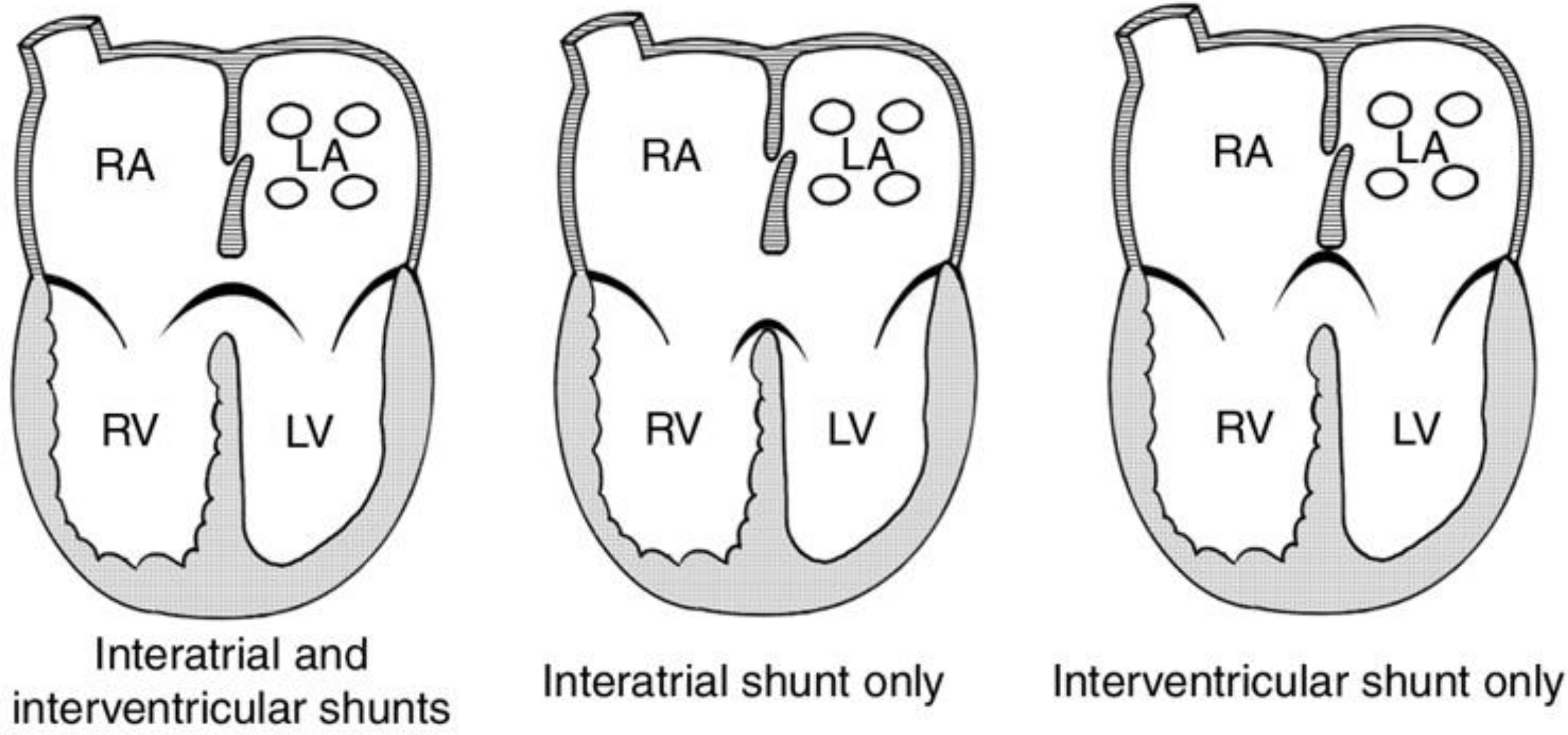
Partial atrioventricular septal defect

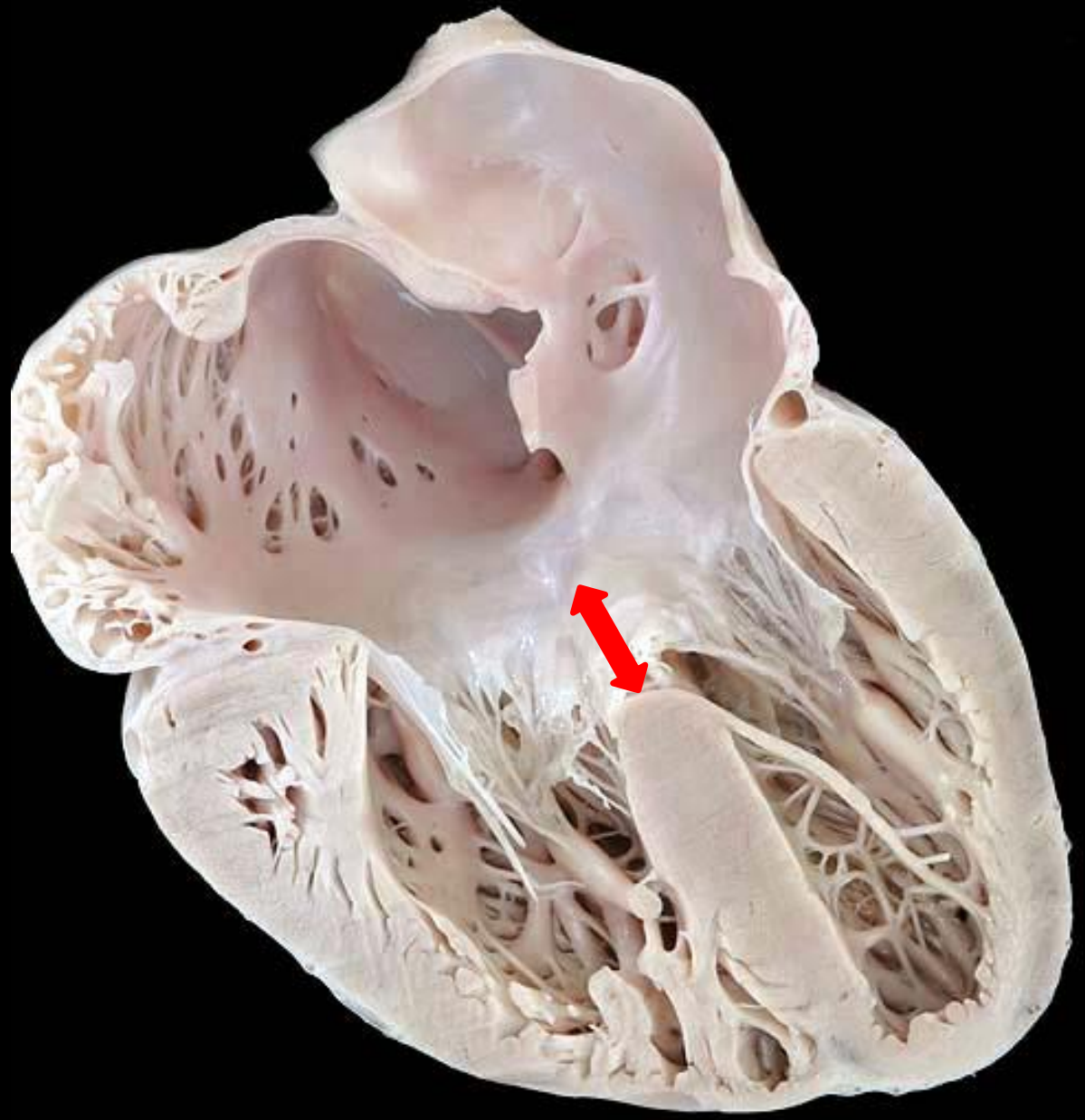


Separate valvar orifices within common junction

Courtesy Robert Anderson

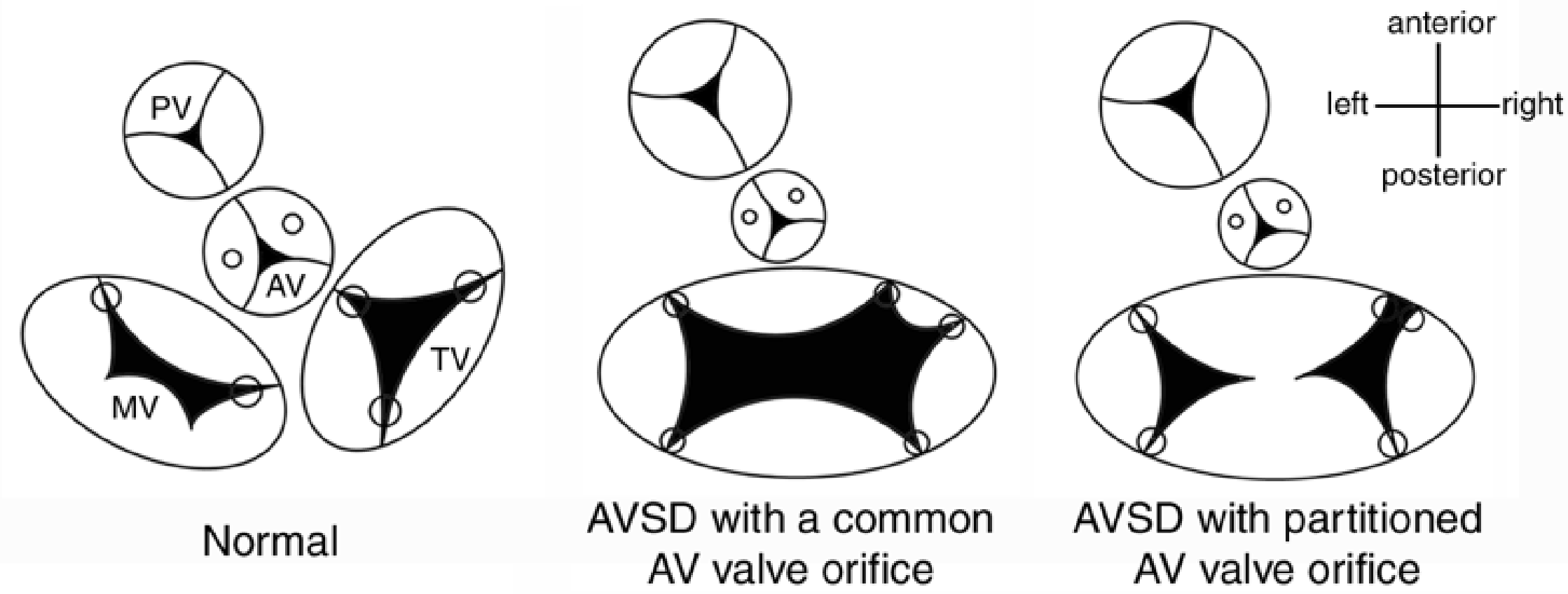
Complete vs partial atrioventricular septal defects





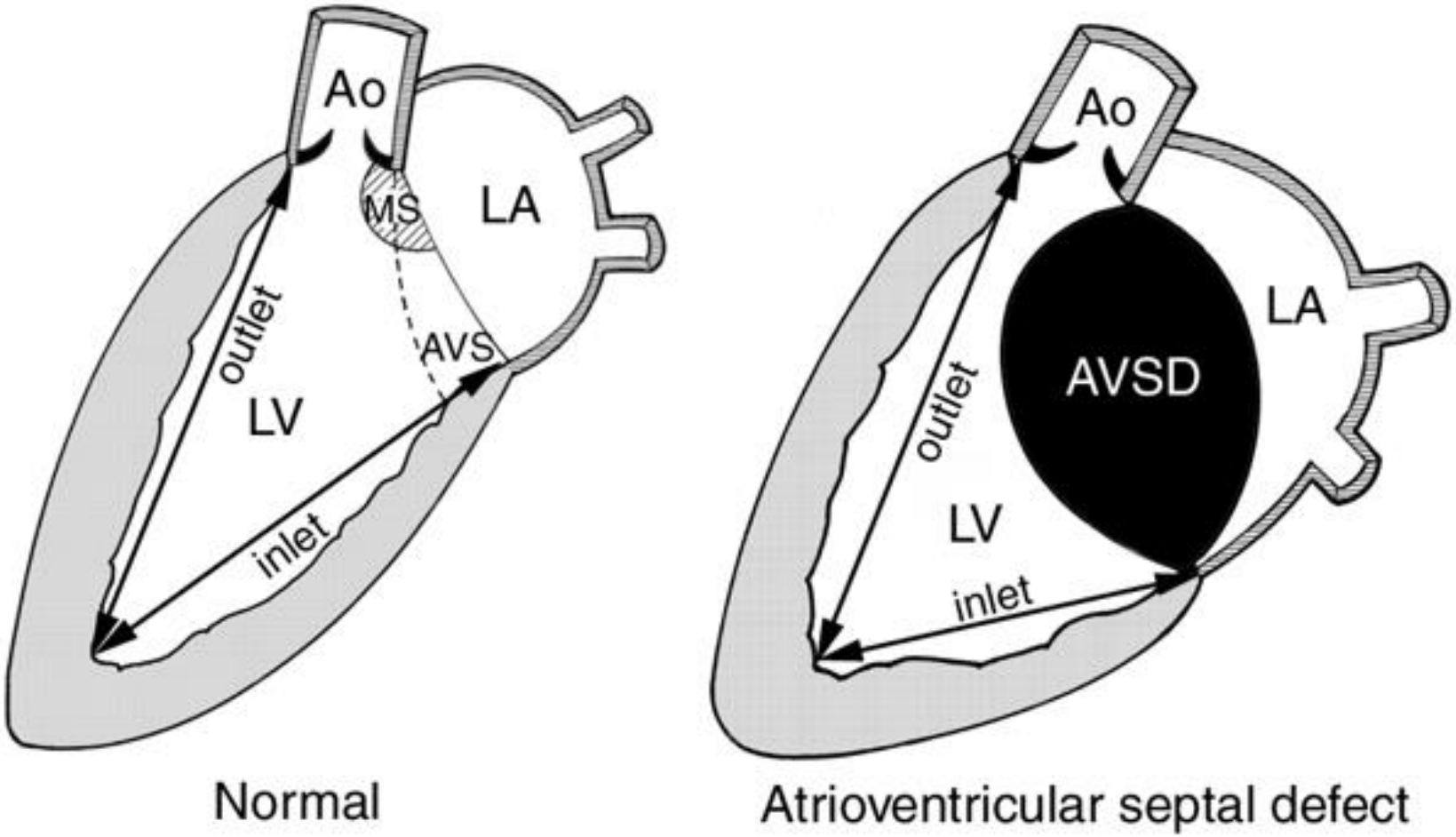
Courtesy Diane Spicer

”Unwedging” of the aortic root

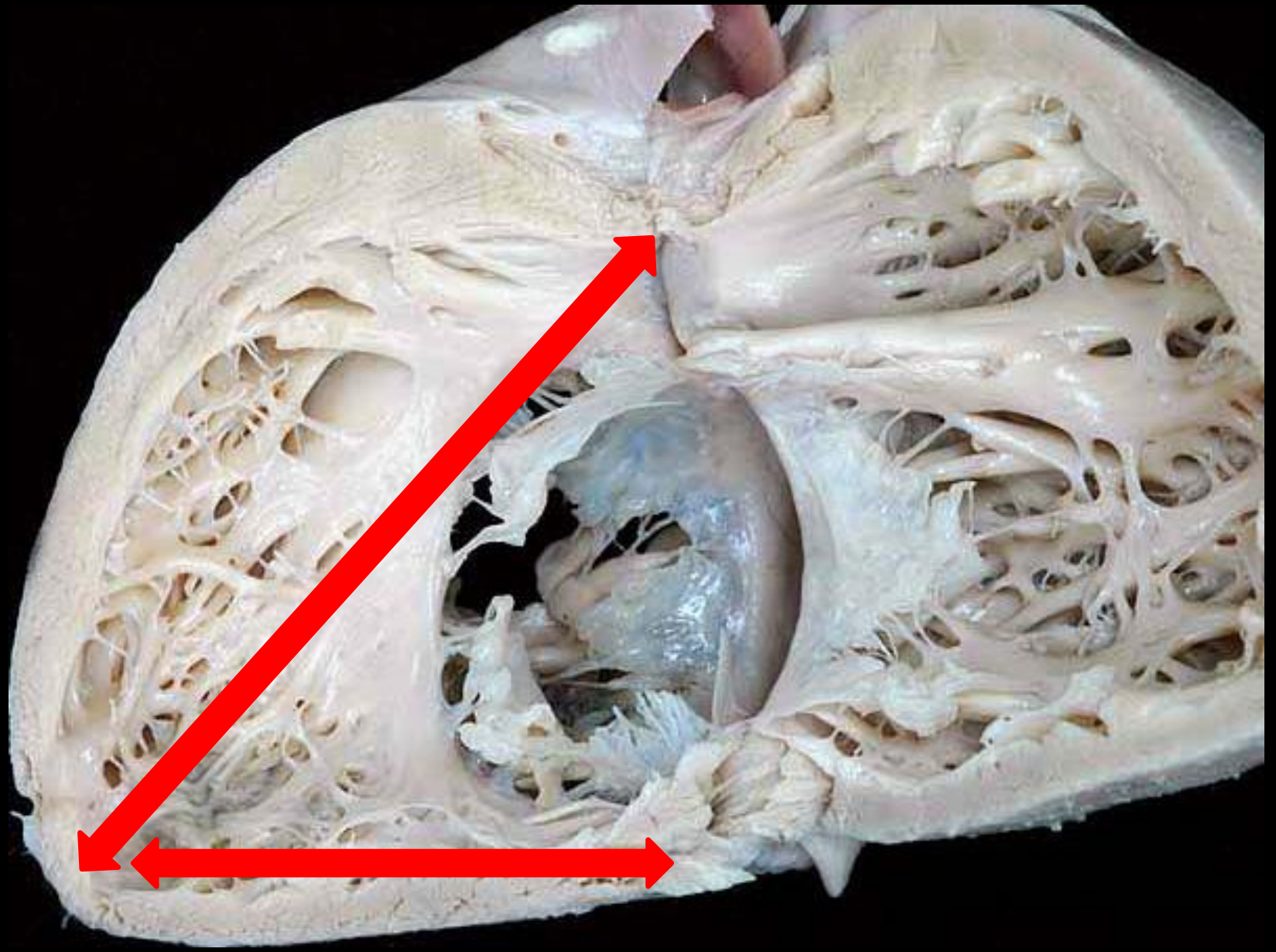


Yoo et al. Chest Radiographic Interpretation. Thieme, 2010

Left ventricular inlet/outlet disproportion



Yoo et al. Chest Radiographic Interpretation. Thieme, 2010



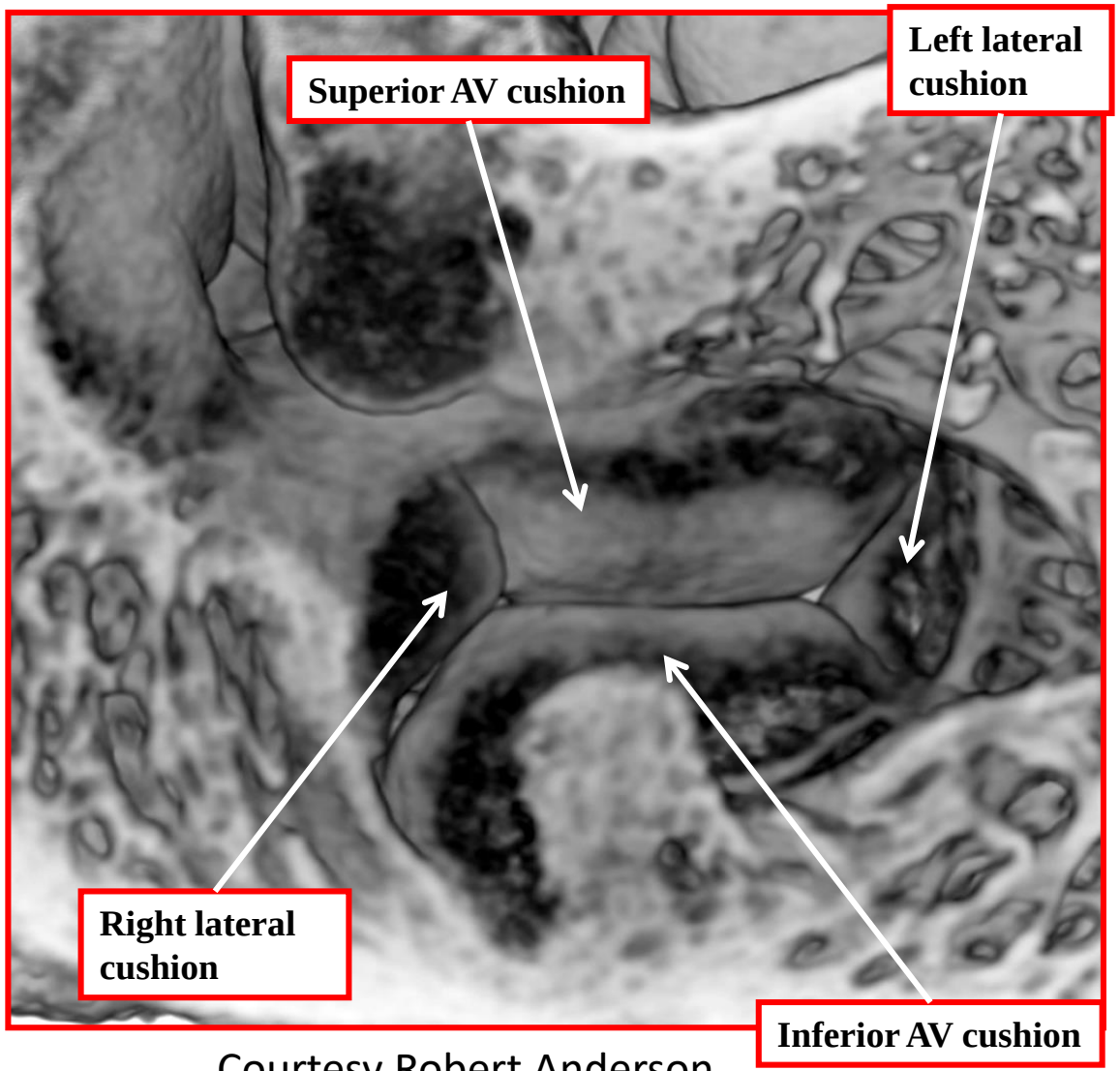
Courtesy Diane Spicer

Gooseneck deformity of the LVOT

https://thoracickey.com/atrioventricular-septal-defects-2/

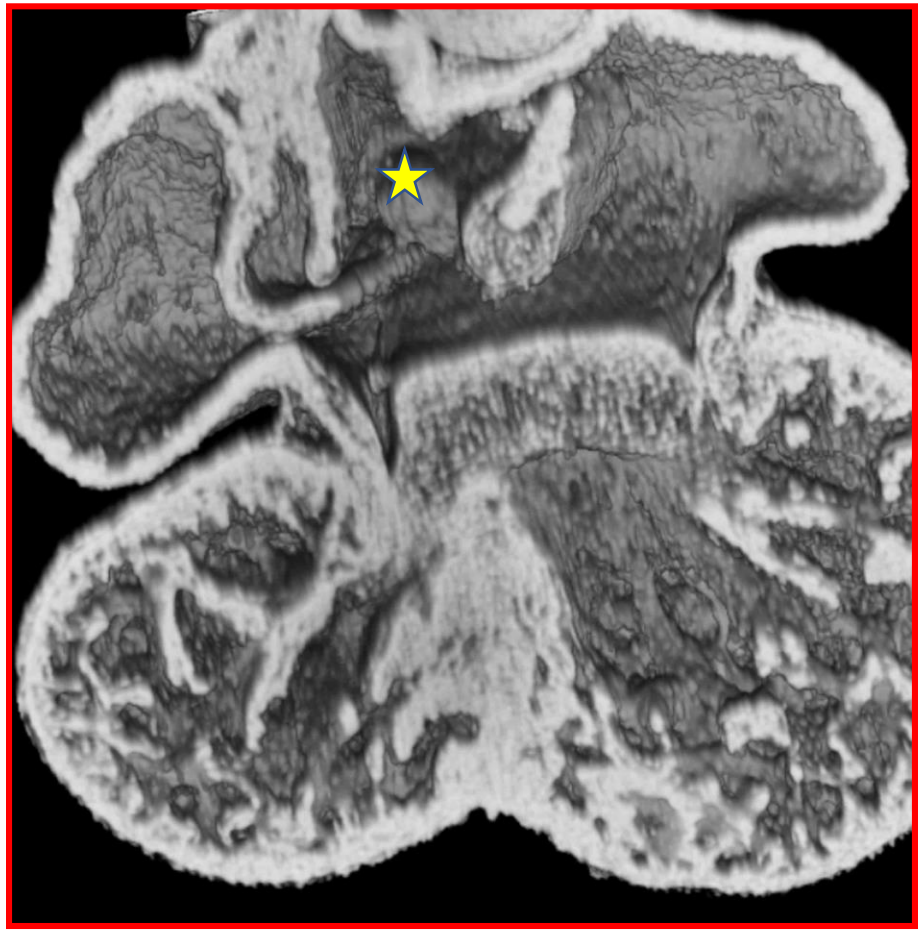
Atrioventricular septal defect – mouse embryology

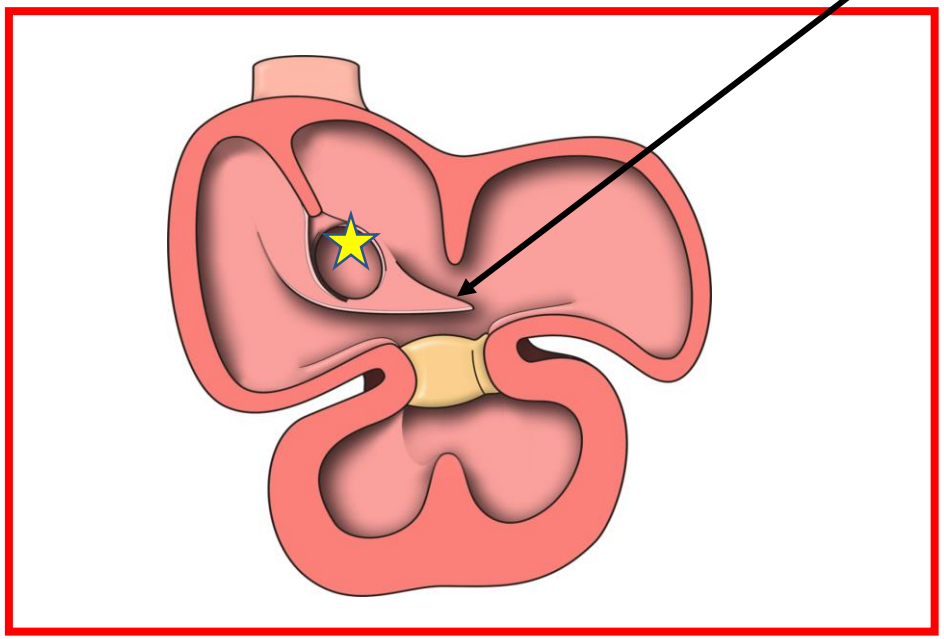
- At the end of E11.5, the arrangement of the cushions is reminiscent of the definitive pattern seen in AVSD
- With formation of separate right and left AV junctions, the cushions produce a bifoliate valve in the left ventricle
- These processes influence how the aorta is transferred to the left ventricle



Courtesy Robert Anderson

Separation of atrioventricular junctions

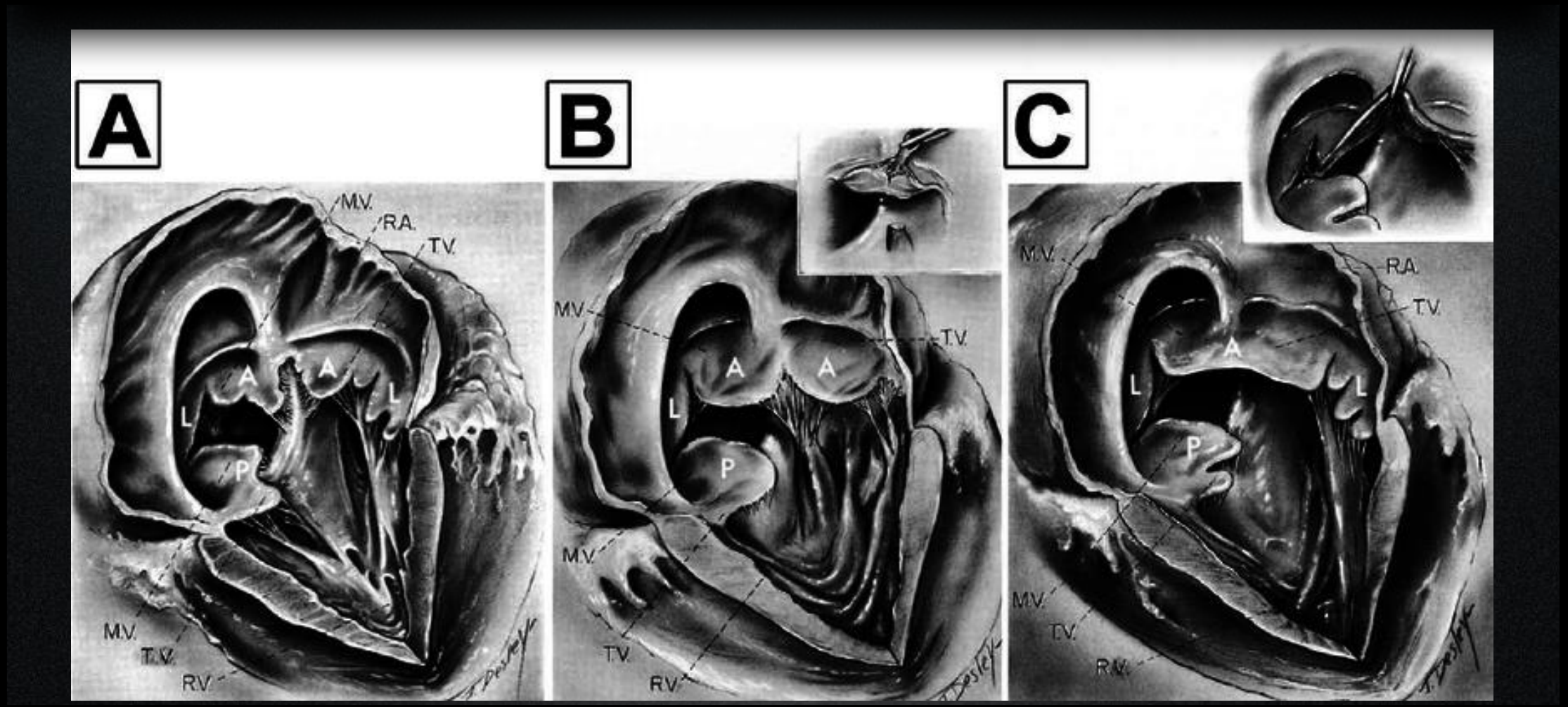




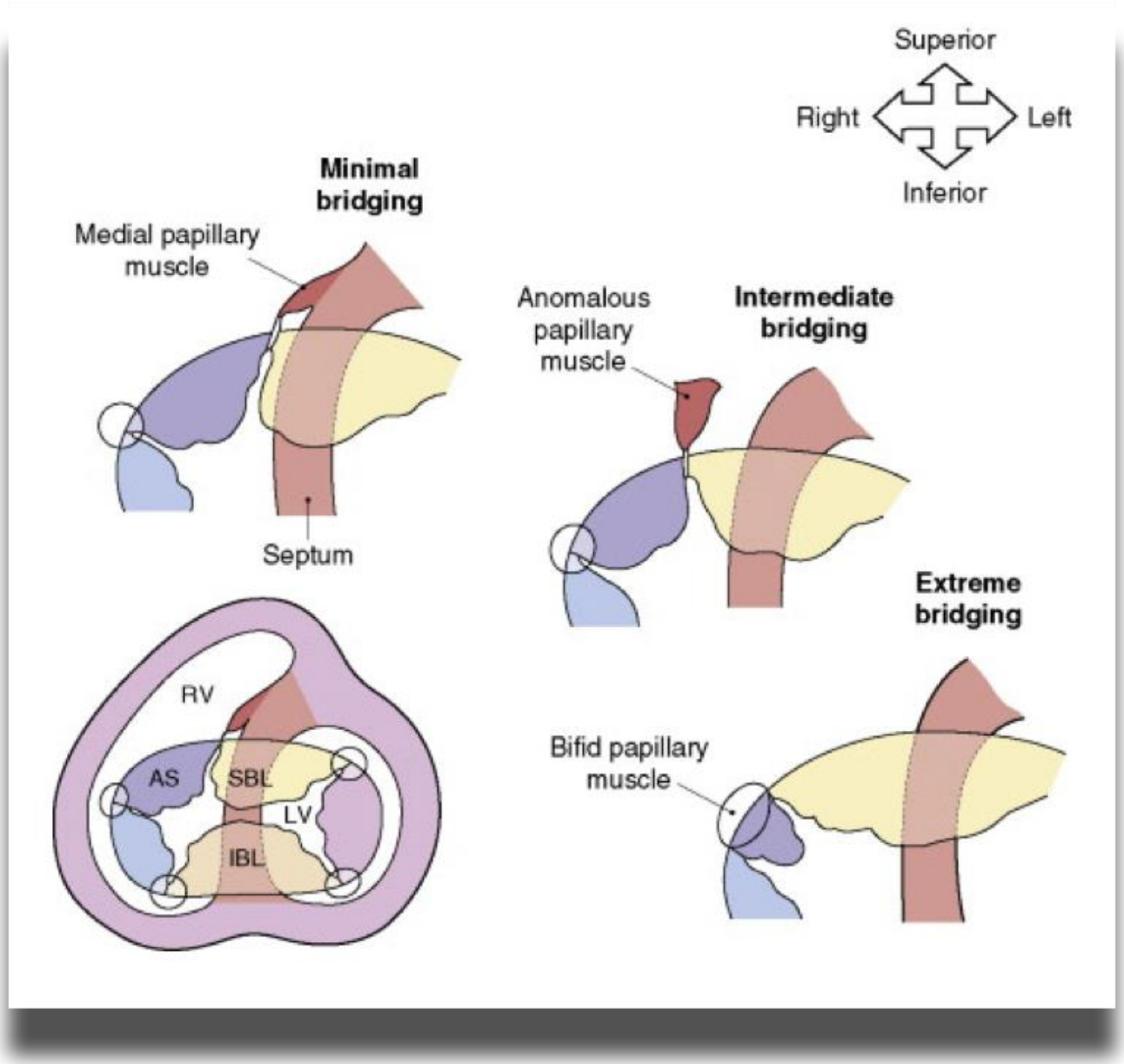
- The key feature is growth through the right pulmonary ridge of the vestibular spine, also known as the dorsal mesenchymal protrusion

Courtesy Robert Anderson

Superior bridging leaflet – Rastelli classification



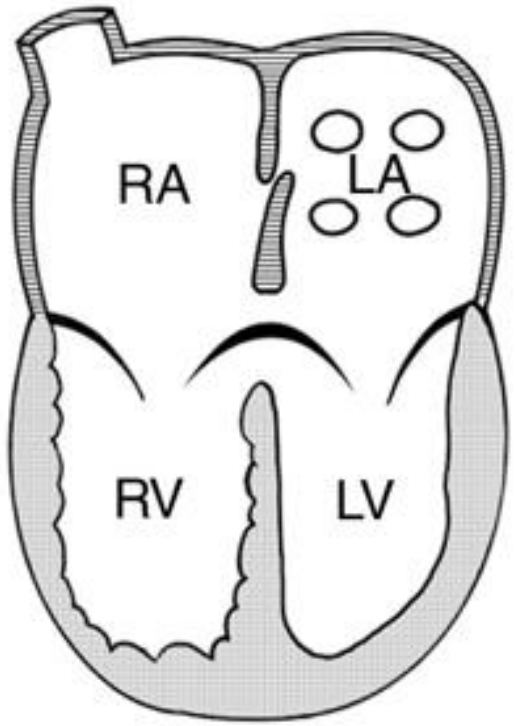
Rastelli et al. *Mayo Clinic Proc* 1967



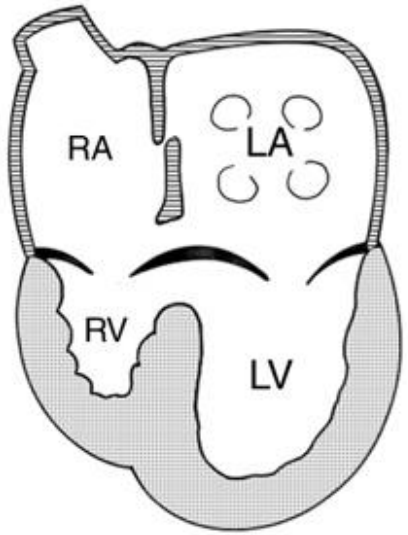
Rastelli classification

Shinebourne and Ho *Science Direct* 2010

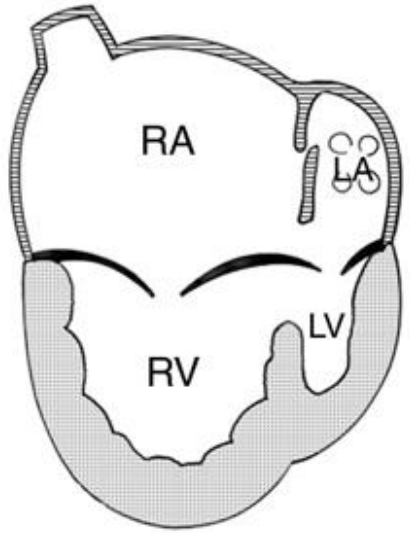
Balanced vs unbalanced
Aligned vs misaligned septums



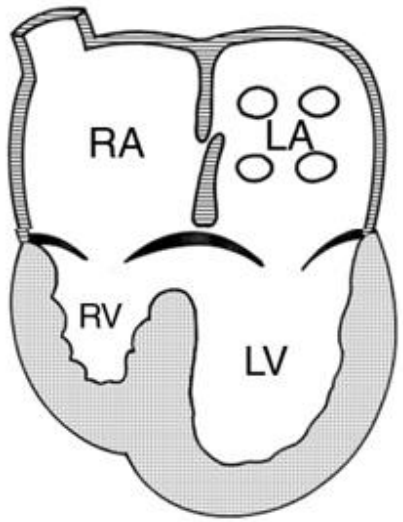
Balanced type



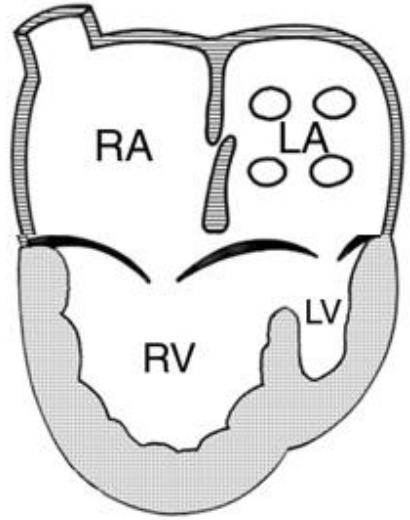
Unbalanced / left-side dominant type



Unbalanced / right-side dominant type

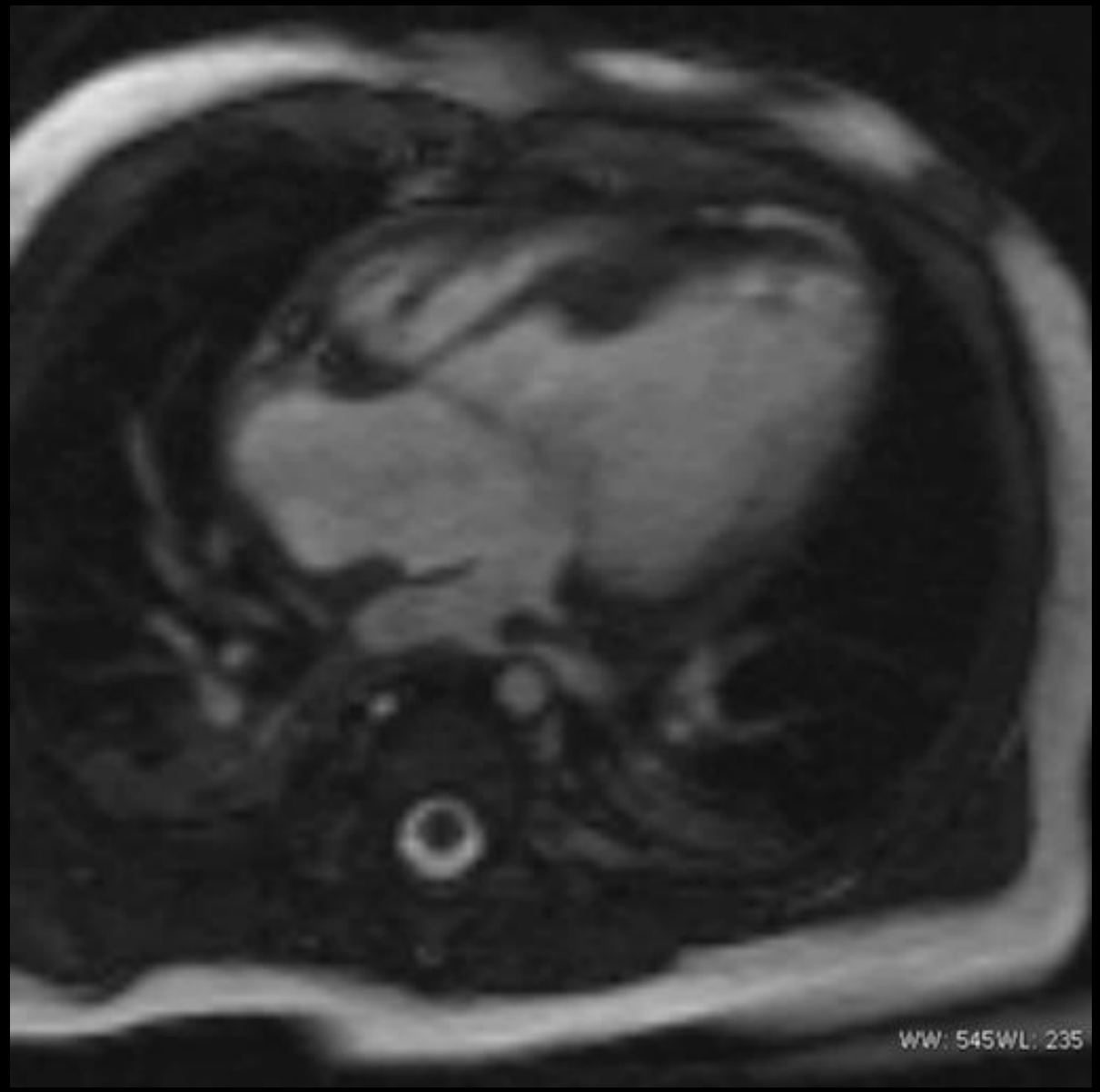


Unbalanced / left-side dominant type



Unbalanced / right-side dominant type

Yoo et al. Chest Radiographic Interpretation. Thieme, 2010


WW: 545WL: 235


WW: 497WL: 208

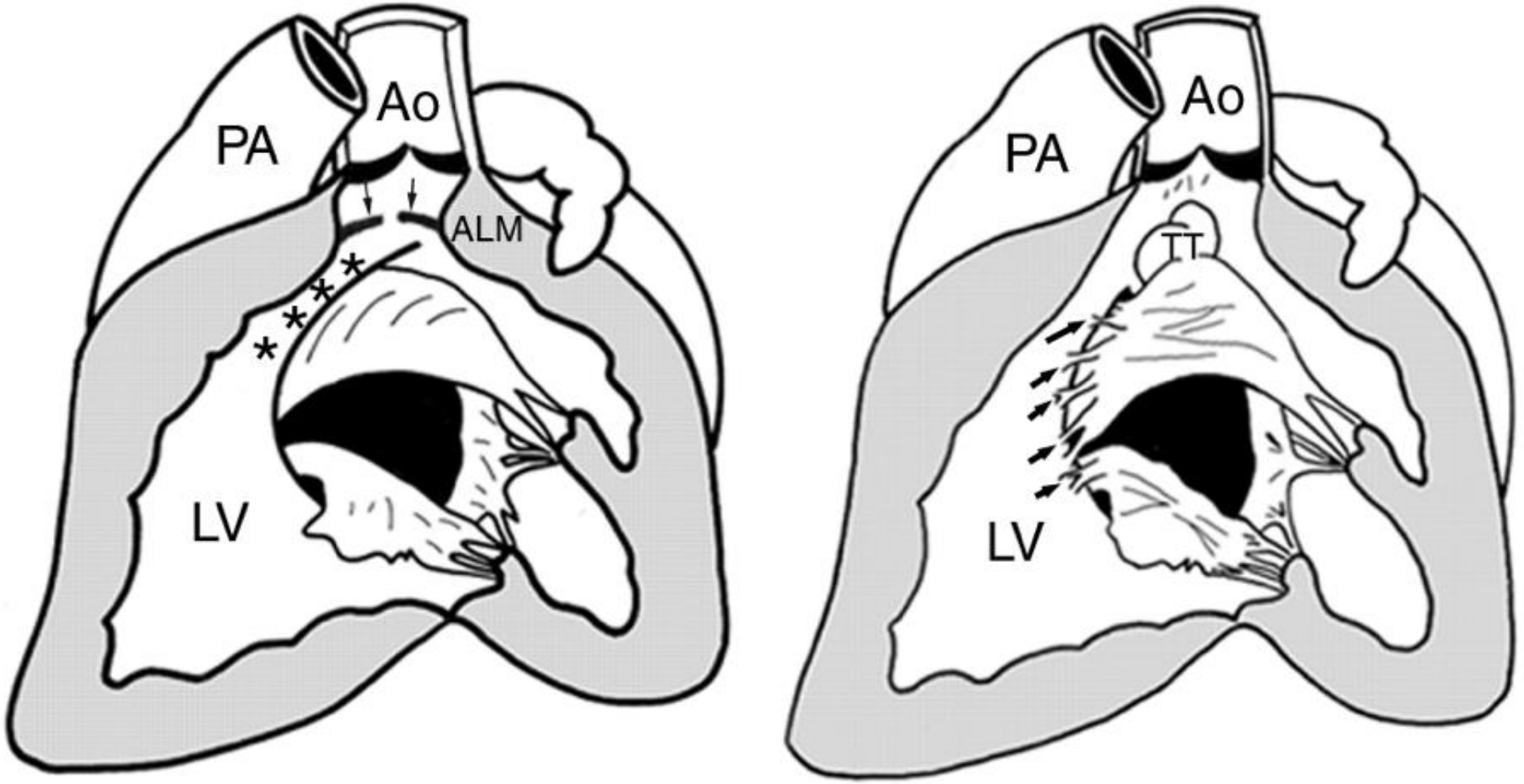
AVSD – associated abnormalities

- Syndromes
  - Trisomy 21 in 30-50%
  - Right isomerism in >10%
  - Left isomerism in >20%
- Left sided obstructive lesions usually non-Downs
- Tetralogy of Fallot in 5%, more common in T21
- Atrioventricular block > 10%, particularly in left isomerism
- Extracardiac abnormalities present in 15% of non-Downs

AVSD – mechanisms of LVOTO

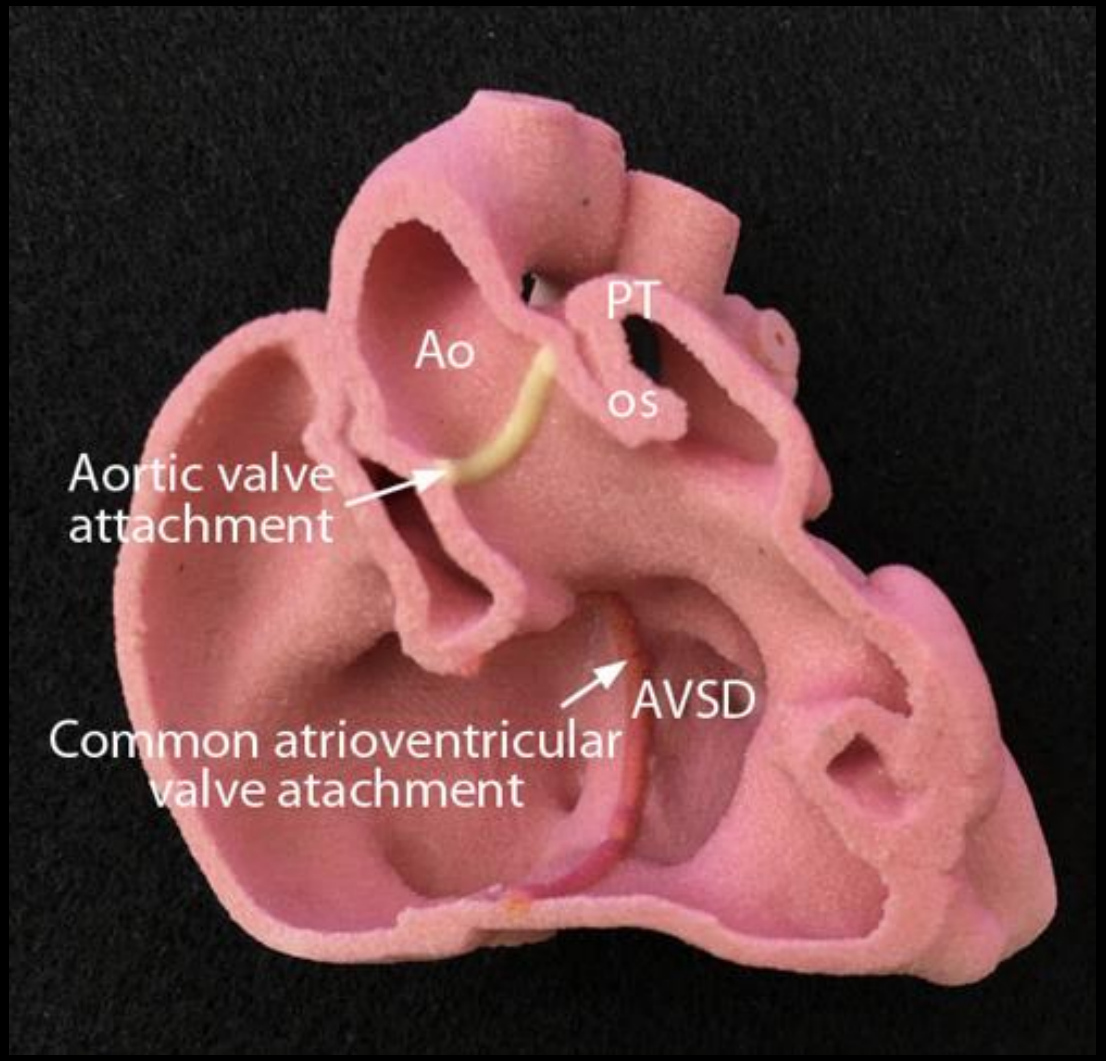
- Excessive scooped-out ventricular septum with narrowing and elongation of LVOT
- Mal-attachment of superior bridging leaflet to the ventricular septal crest or septum
- Prominent antero-lateral muscle bundle
- Diplaced insertion of papillary muscles
- Tissue tags from AV valve or membranous septum
- Fibromuscular tunnel
- Subaortic membrane

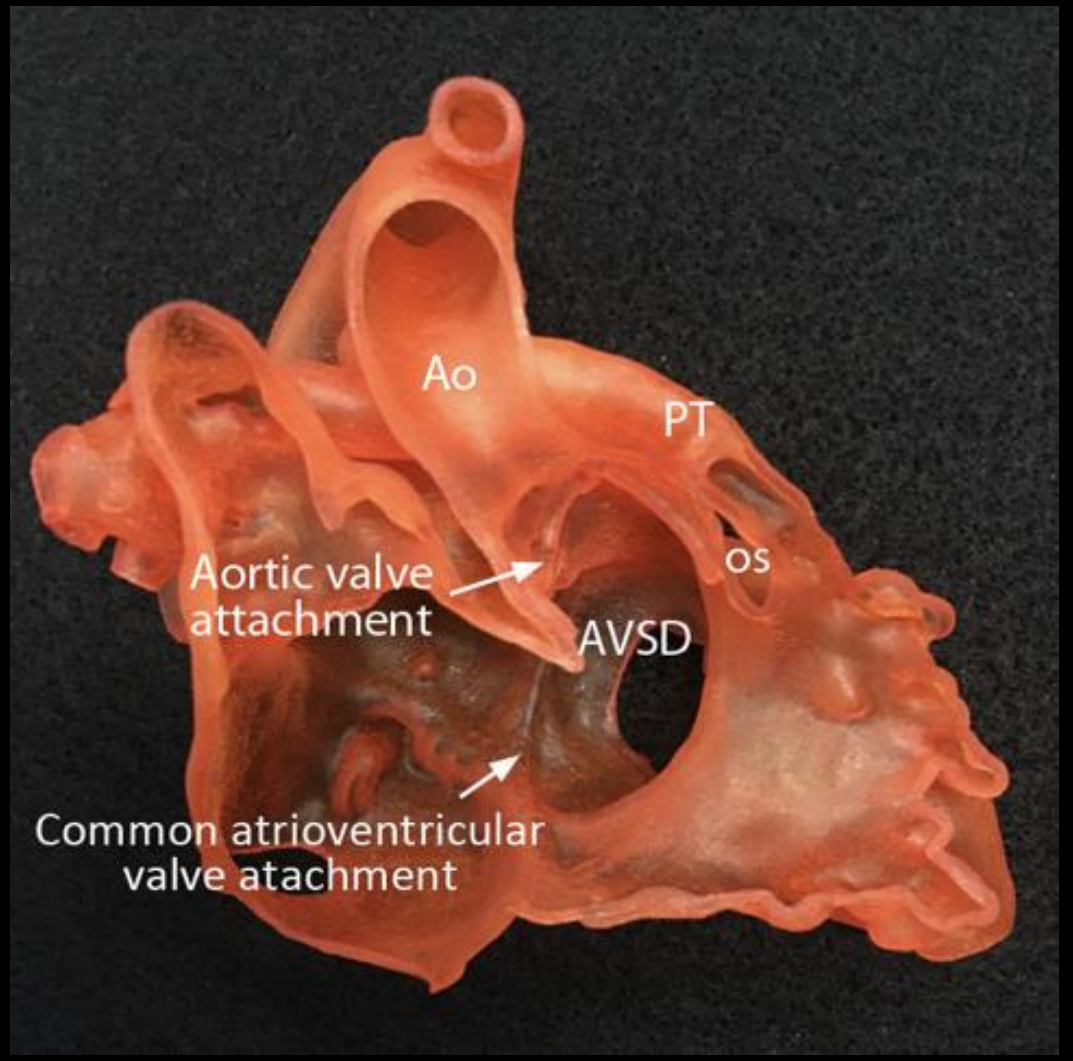
Left ventricular outflow tract obstruction in AVSDs



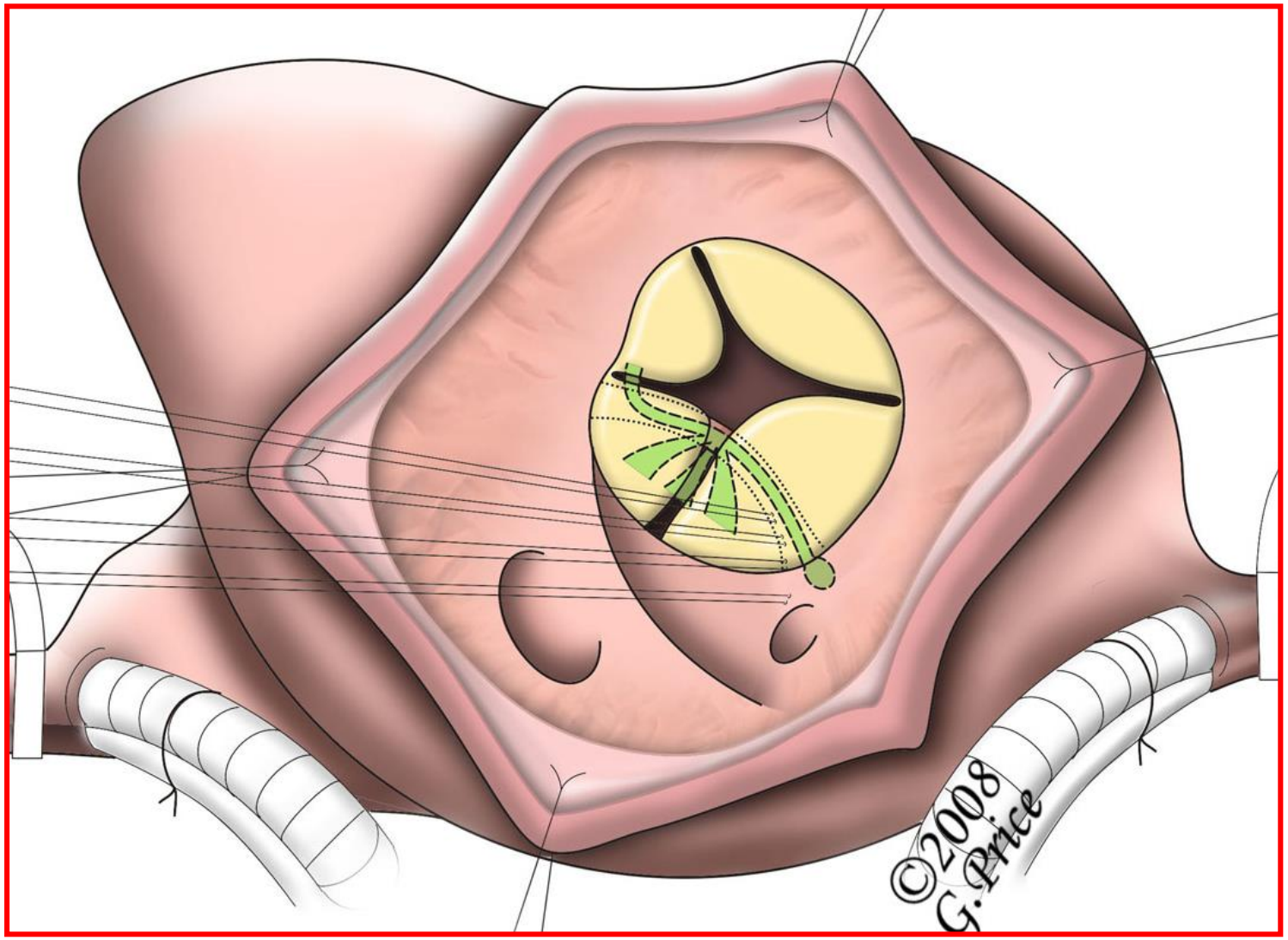
Freedom and Yoo et al. The Natural and Modified History of Congenital Heart Disease. Futura, 2003

AVSD with tetralogy of Fallot and DORV





Conduction tissue in AVSD



Courtesy Robert Anderson

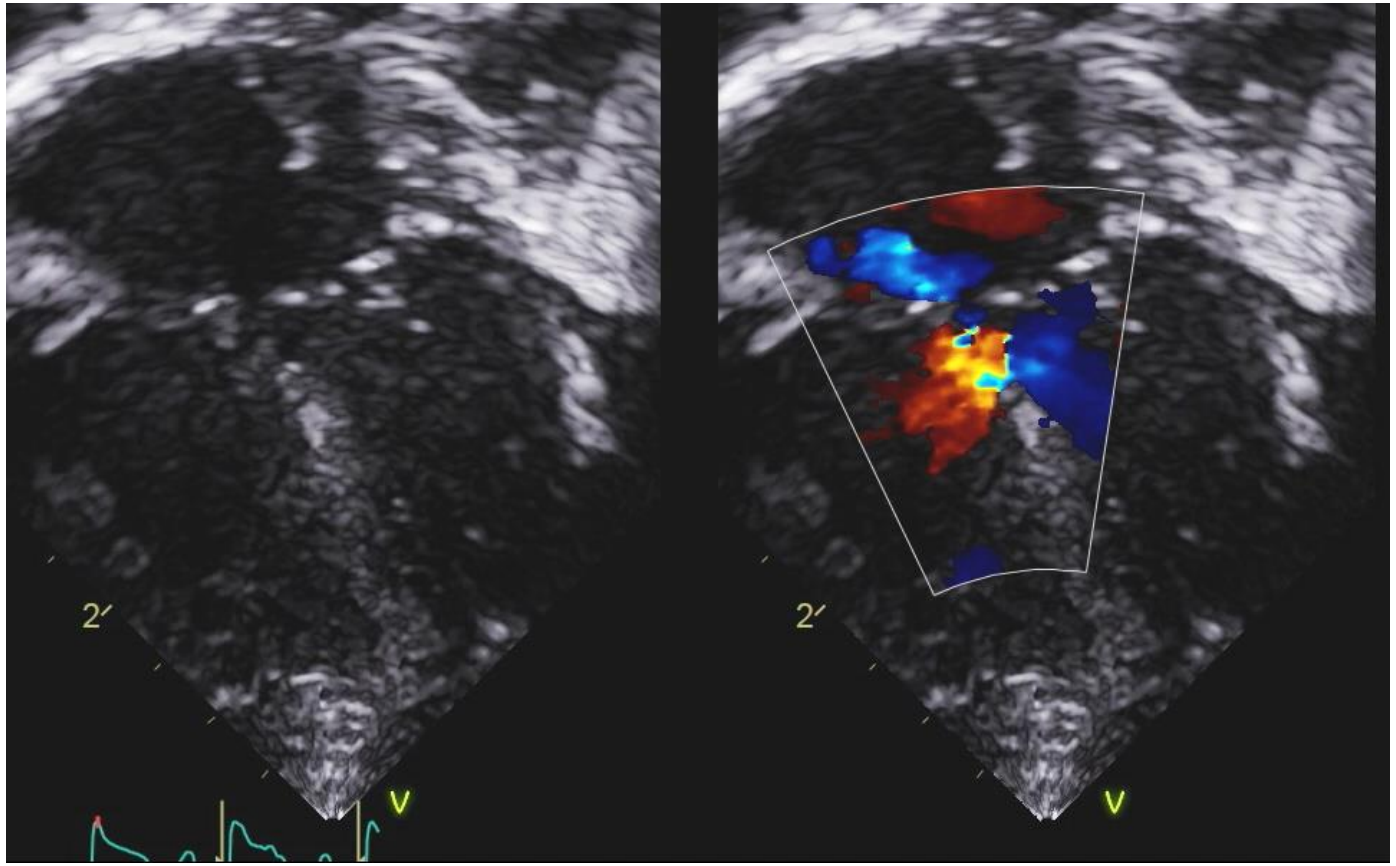
Physiologic considerations

- Heart block or severe AV valve regurgitation may result in fetal hydrops or demise
- Ostium primum: left-to-right shunt at atrial level driven by more compliant right ventricle, leads to right atrial and ventricular enlargement over time
- AV valve regurgitation frequently results in left ventricle to right atrial shunt
- Complete AVSD or partial AVSD with ventricular level shunt: results in high pulmonary blood flow, diminished systemic blood flow, pulmonary edema i.e. heart failure + risk of pulmonary vascular disease with extended exposure
- T21 patients have less heart failure due to higher pulmonary vascular resistance
- Unbalanced AVSD: single ventricle physiology with parallel circulation, risk of pulmonary over-circulation, particularly in the setting of significant AV valve regurgitation
- AVSD with TOF or AVSD with right isomerism with pulmonary stenosis/atresia may result in inadequate PBF

Fetal diagnosis and counselling

- Diagnosis of AVSD
- Genetic testing
- Associated abnormalities
- Heart block in left isomerism
- Pulmonary venous obstruction in right isomerism
- AV valve regurgitation may result in hydrops, fetal demise

Postnatal assessment



- Segmental sequential analysis
- Extent of ventricular shunting, and gradient
- Commitment of AV valve (AV valve ratio)
- Mural leaflet – spacing of LAVV papillary muscles
- AV valve regurgitation
- Ventricular hypoplasia
- Outflow tract obstruction
- Coarctation
- Pulmonary arteries and veins

Management

- Ostium primum: usually manage as for ASD with elective repair at around 4 years of age to address shunt and prevent progressive AV valve regurgitation – earlier if progressive AV valve regurgitation
- Complete AVSD or partial AVSD with unrestrictive ventricular level shunt: medical management of heart failure and repair at 4-6 months to address left to right shunt and avoid pulmonary vascular disease – earlier if progressive AV valve regurgitation
- When there is severe ventricular hypoplasia then single ventricle palliation according to the anatomy – i.e. Norwood or Hybrid in the setting of left ventricular hypoplasia and hypoplastic aortic arch versus pulmonary artery band when both outflows unobstructed or BT shunt when there is RVOT obstruction
- BCPS and Fontan may be precluded in T21 patients due to elevated PVR

Surgery

- Aim of surgery is to septal the heart and create two functional atrioventricular valves
- AV valve repair (closure of zone of apposition) also performed during closure of ostium primum defect
- Two patch technique the usual approach for complete AVSD
- Single patch (Australian repair) for complete AVSD with a small interventricular communication
- Severity of pre-operative AV valve regurgitation correlates with post-op AV valve regurgitation
- Leaflet plasty/extension for dysplastic and dysfunctional valves
- Re-operation or valve replacement may be needed for severe AV valve regurgitation
- Borderline LV hypoplasia in AVSD may be particularly amenable to LV recruitment with pulmonary artery banding and atrial septation

AVSD surgery

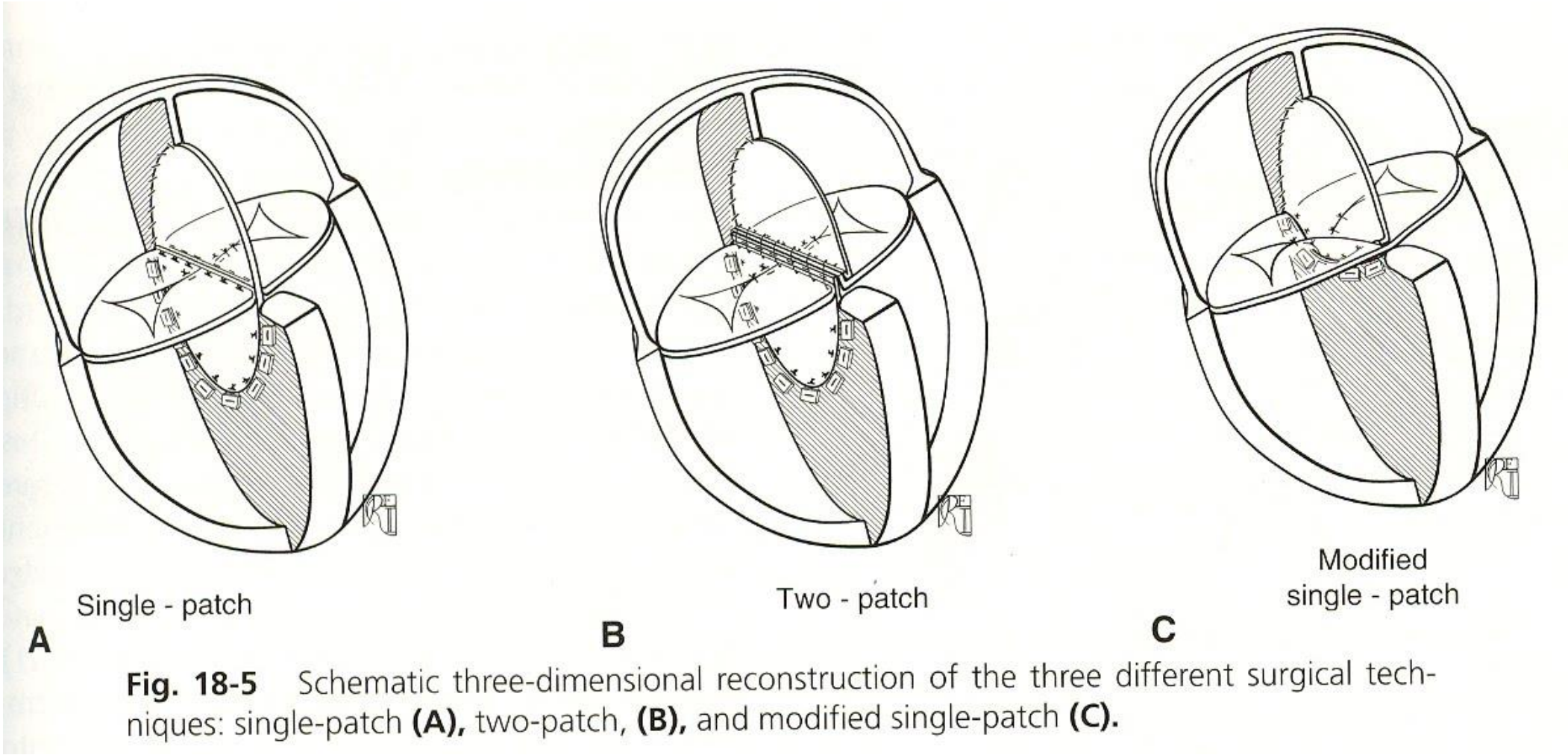


Fig. 18-5 Schematic three-dimensional reconstruction of the three different surgical techniques: single-patch **(A),** two-patch, **(B),** and modified single-patch **(C).**

Nunn GR. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2007:28-31

AVSD surgical outcomes

- Perioperative mortality: <5%
- Survival following surgery:
  - 85% at 10 years
  - 70% at 30 years
- Reoperation: 10-15%
  - Left AV valve regurgitation in ~ 10%
  - LVOT obstruction in ~ 5%
  - Residual shunt/coarctation

- Anatomical risk factors
  - Malaligned septums
  - Right of left ventricular hypoplasia
  - LVOTO
  - Double orifice left or right AV valve
  - Parachute left AV valve
  - High origin of papillary muscles and short chords
  - Dysplastic left AV valve leaflets
  - Associated major cardiac defect
  - Heterotaxy, TOF, DORV etc