대한흉부심장혈관외과학회 제10차 전공의 연수교육
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Atrioventricular Septal Defect

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The First Cardiac Operation Using Cardiopulmonary Bypass

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The first open heart operation ever performed was by Clarence Dennis on April 5, 1951, at the University of Minnesota. For several years he had worked developing a heart-lung machine. The operation was on a young girl considered to have an ostium secundum atrial septal defect, but at the procedure a large ostium primum was identified instead. It could not be repaired, and the child died. The anatomic details of this heart have not been previously presented. For historical purposes, the clinical history, intraoperative course, and pathologic details will be discussed and shown.

(Ann Thorac Surg 2017;103:e339–40)

Atrioventricular Septal Defect

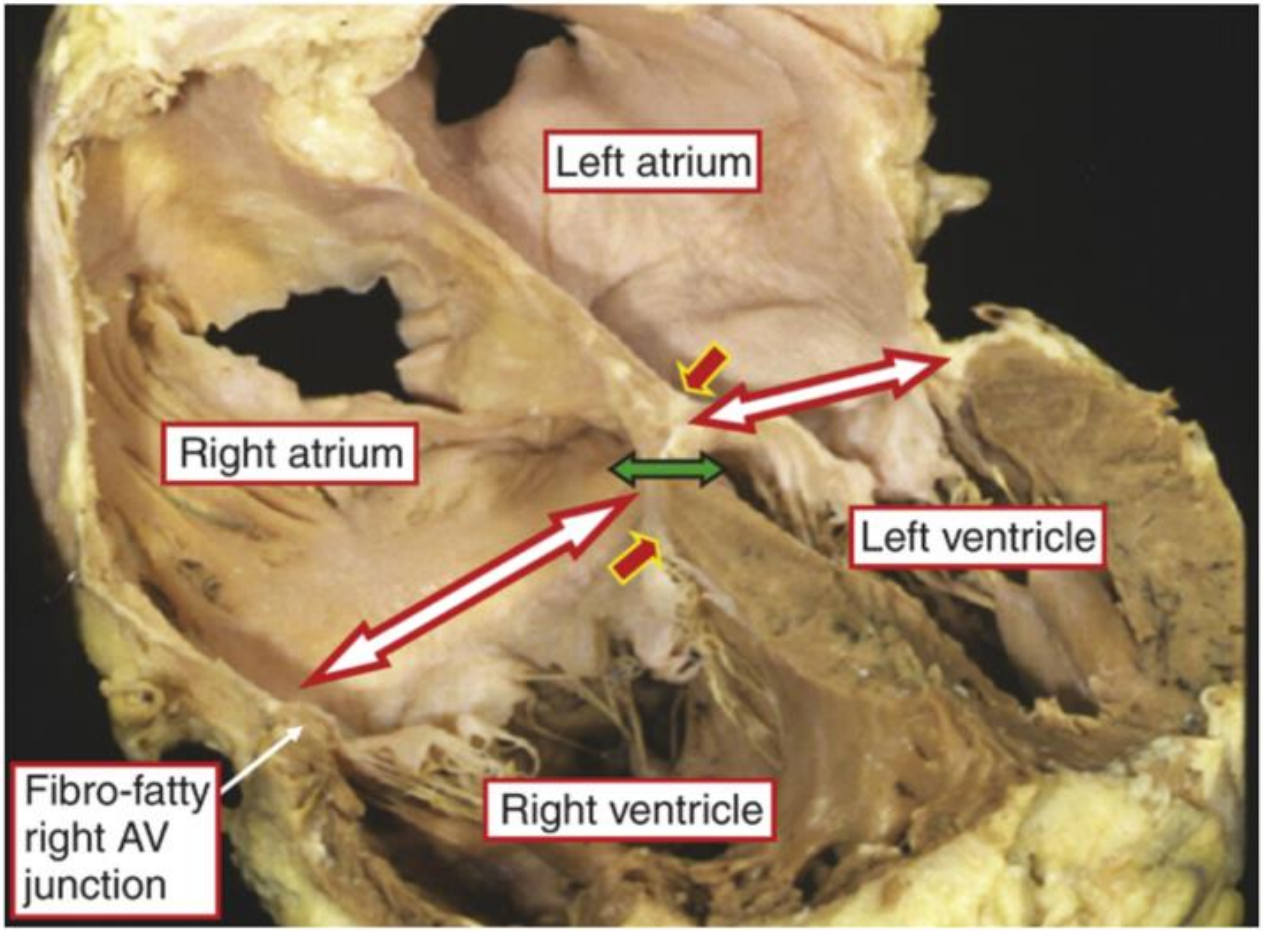
- A group of lesions unified by the anatomical hallmark of **a common atrioventricular junction** co-existing with **deficient atrioventricular septation**

- Synonyms
  * Atrioventricular canal defect
  * Endocardial cushion defect

- Approximately 4% of all congenital heart diseases

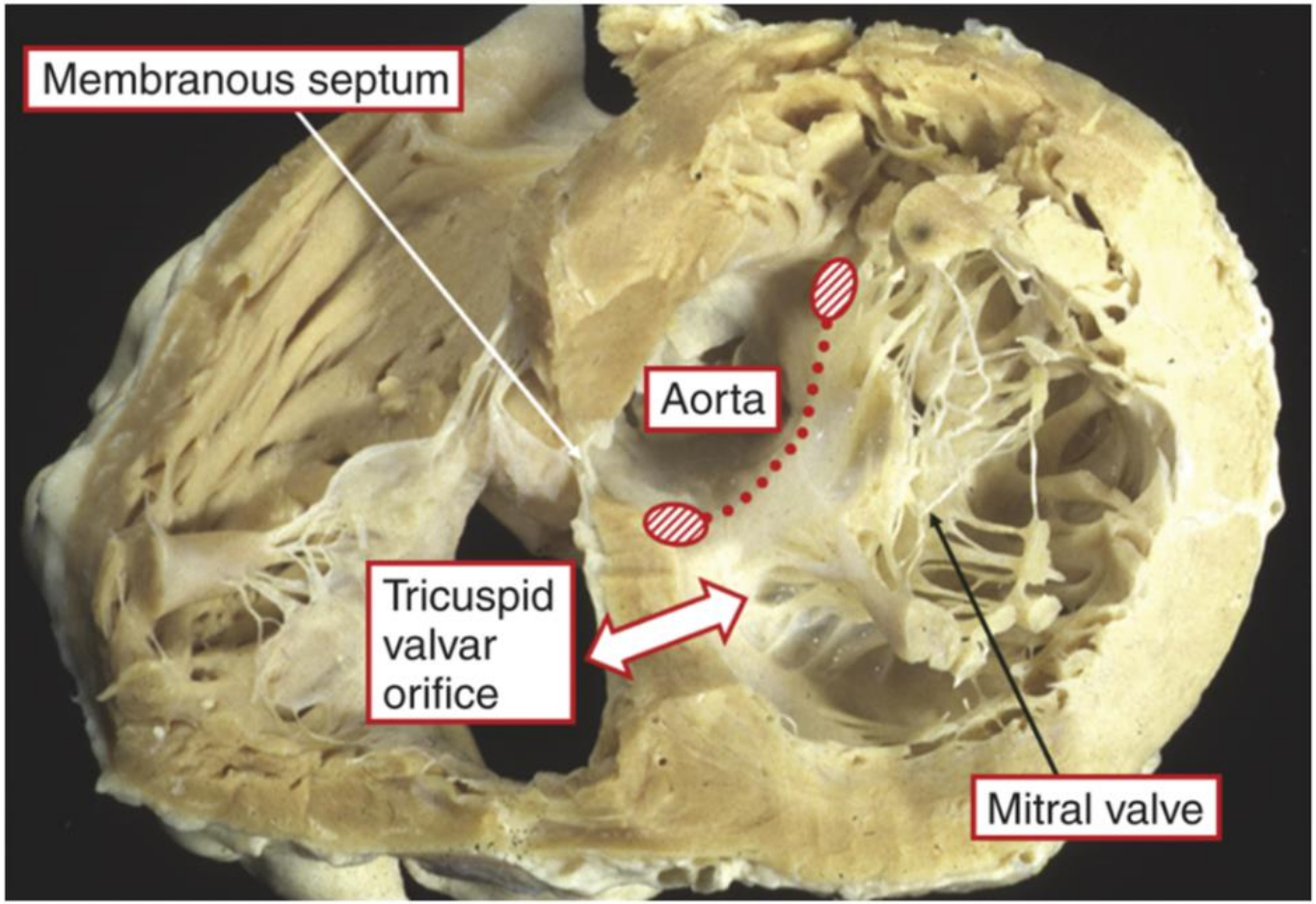
Atrioventricular Septal Defect

Normal Atrioventricular Junctions



Anderson RH, et al. Paediatric Cardiology. 3rd ed.

Normal Atrioventricular Junctions



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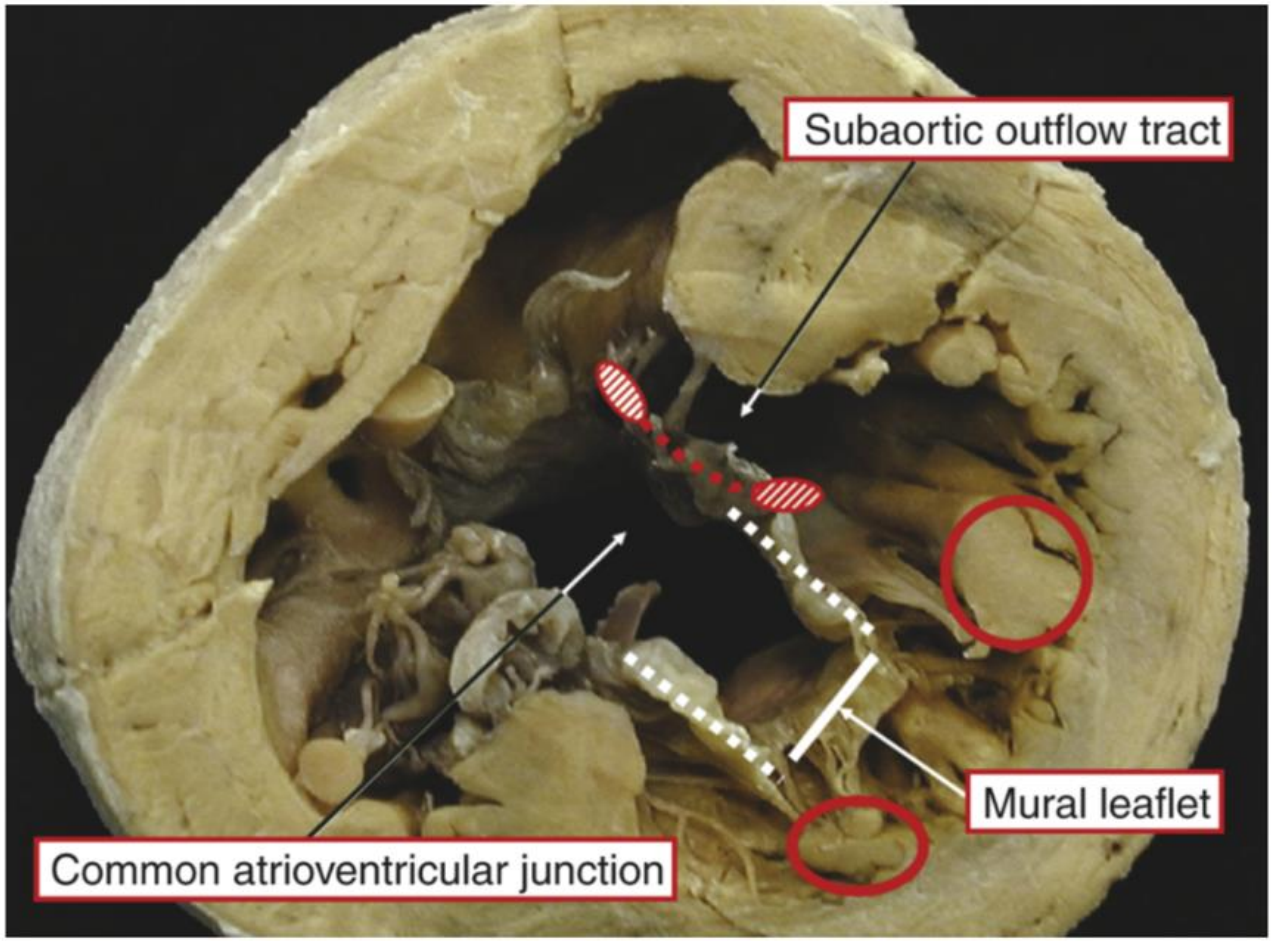
Common Atrioventricular Junction



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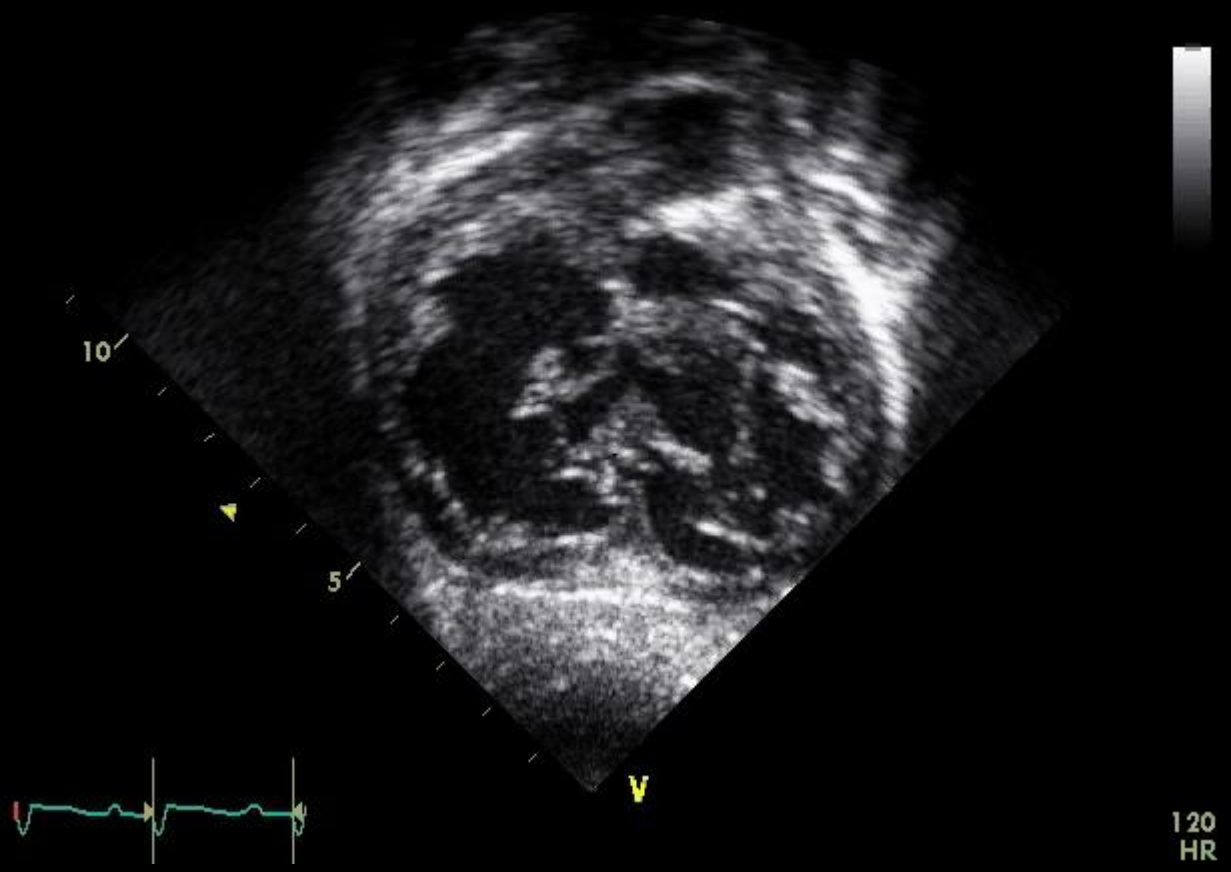
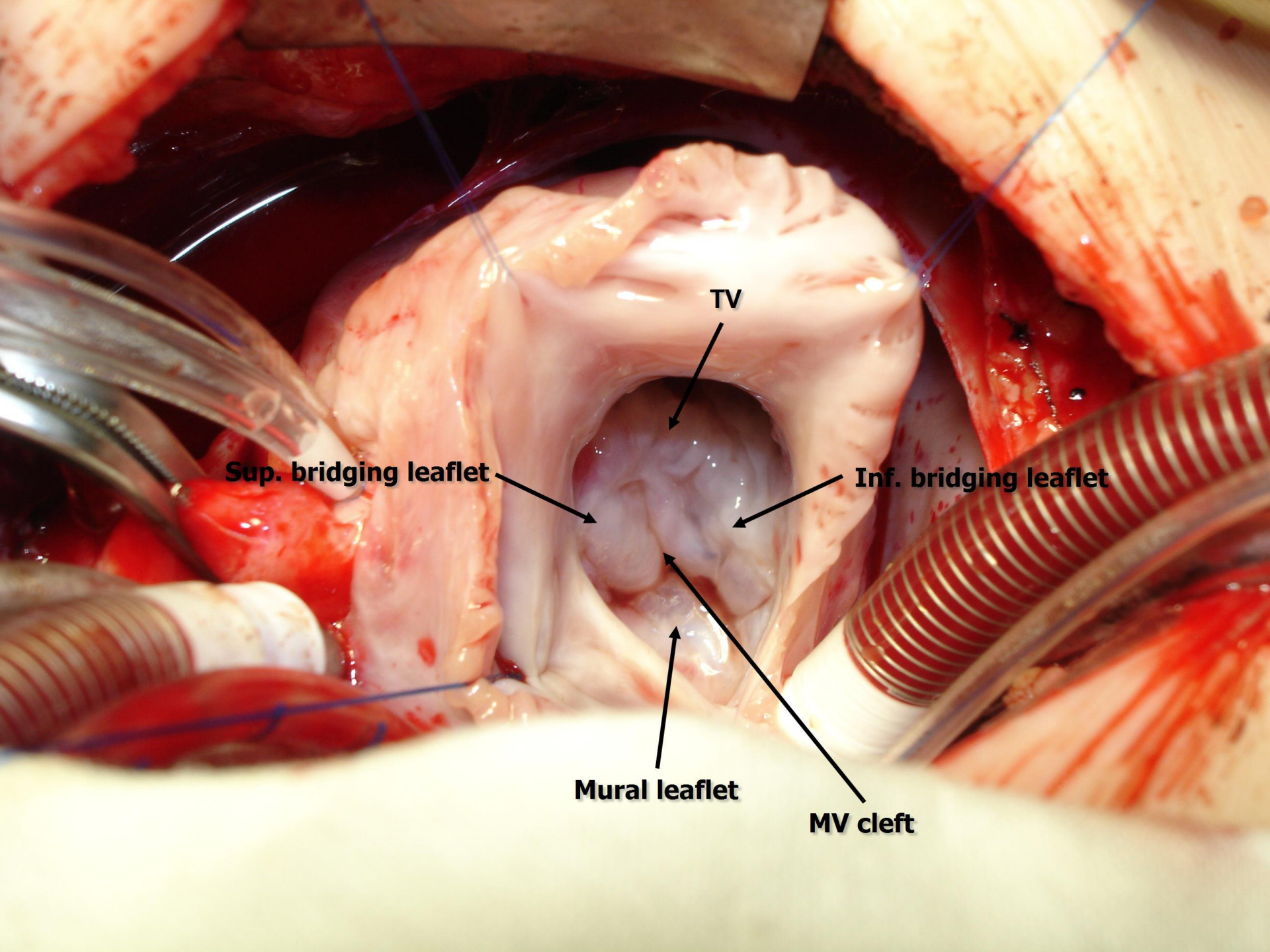
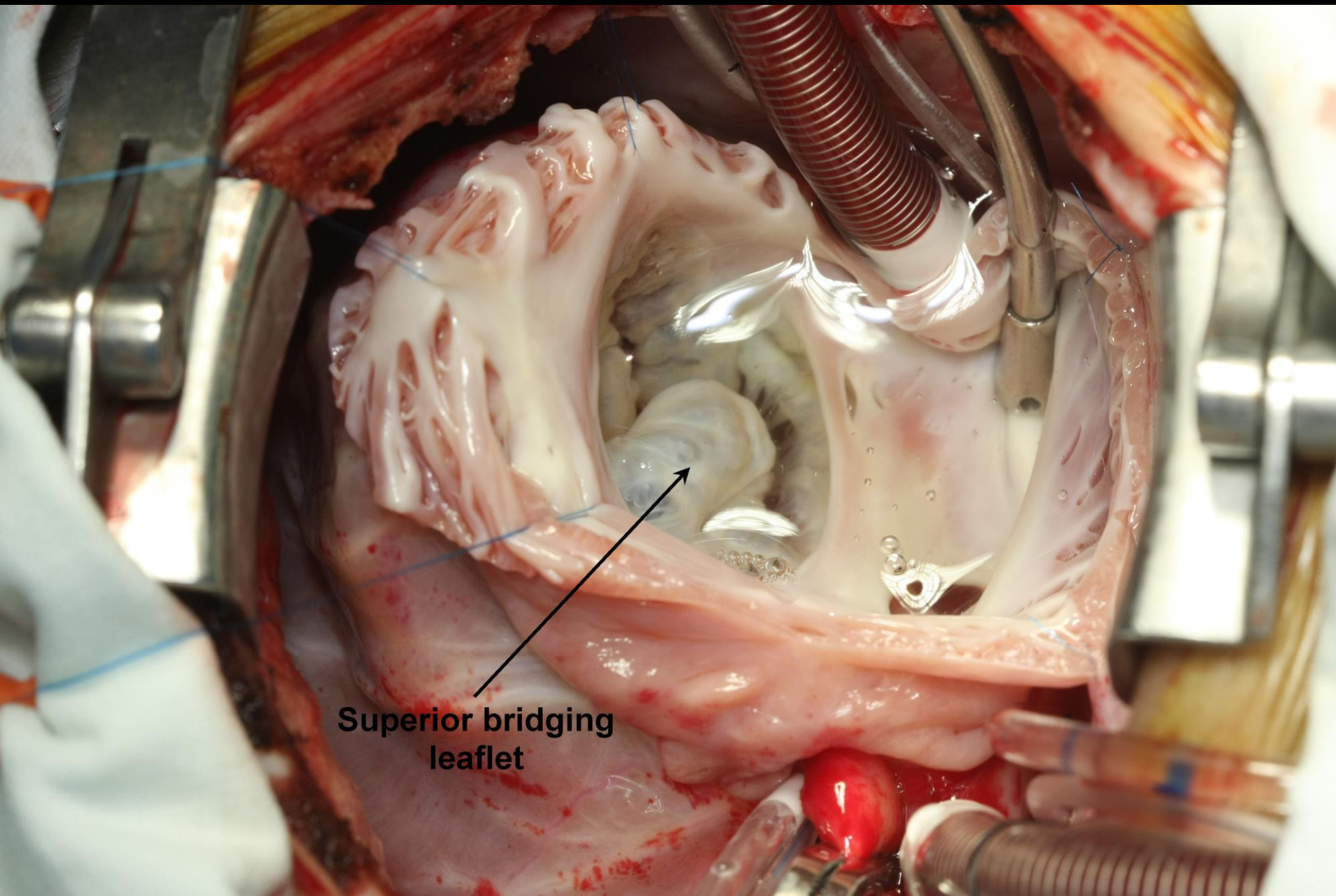
Common Atrioventricular Junction

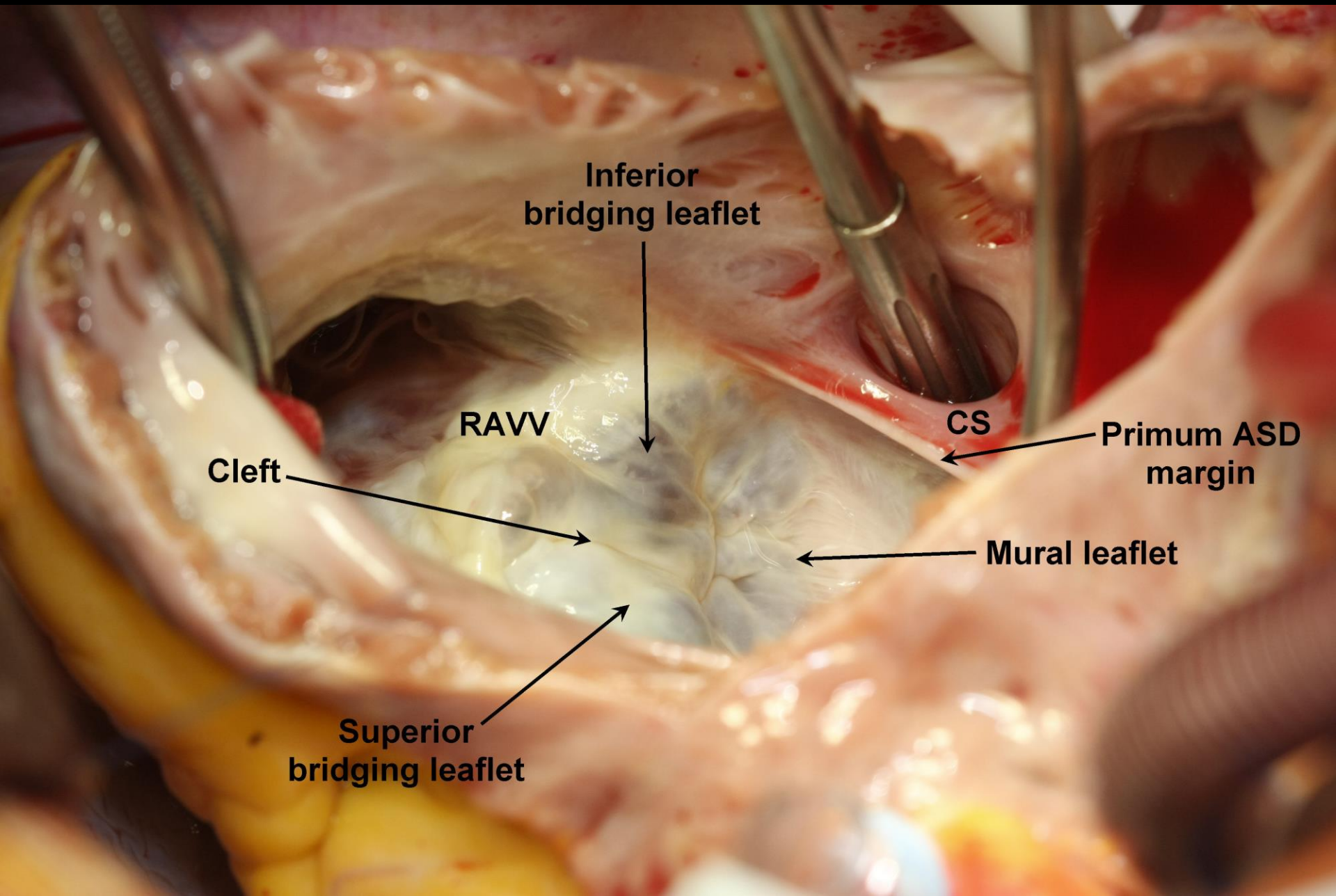



TV
Sup. bridging leaflet
Inf. bridging leaflet
Mural leaflet
MV cleft

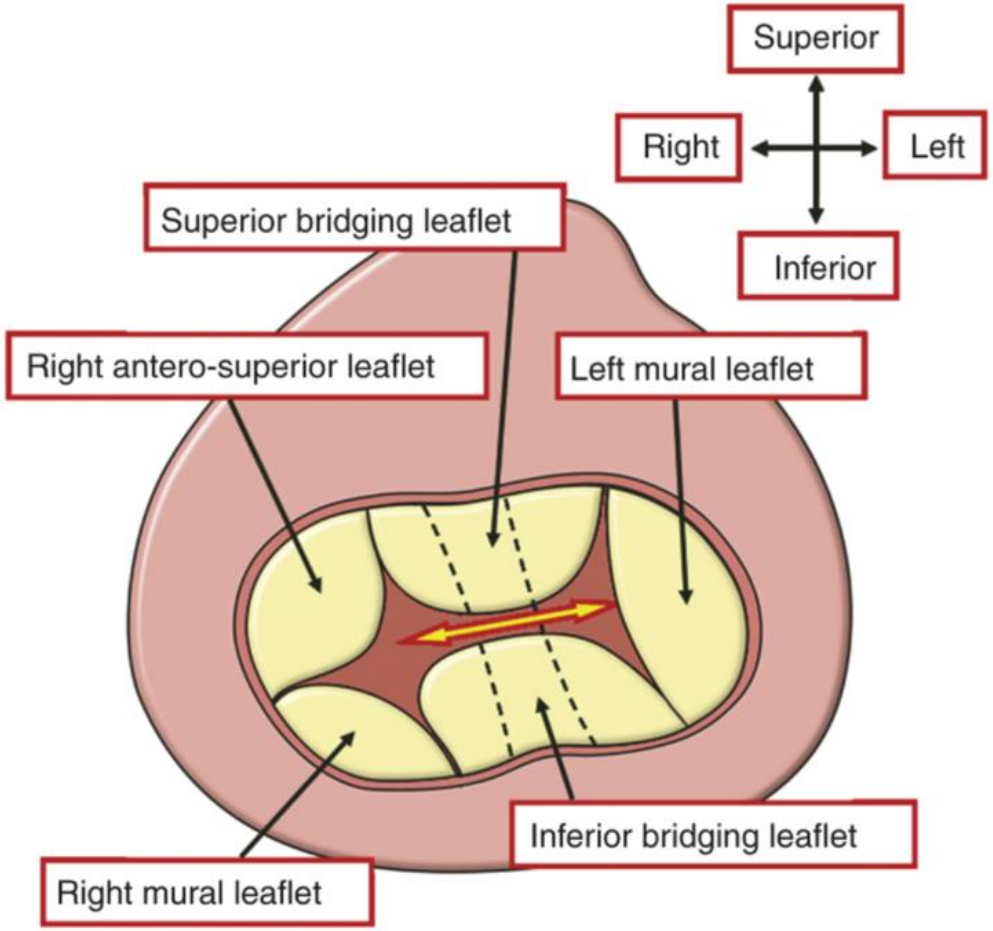
Common Atrioventricular Junction



Common Atrioventricular Junction

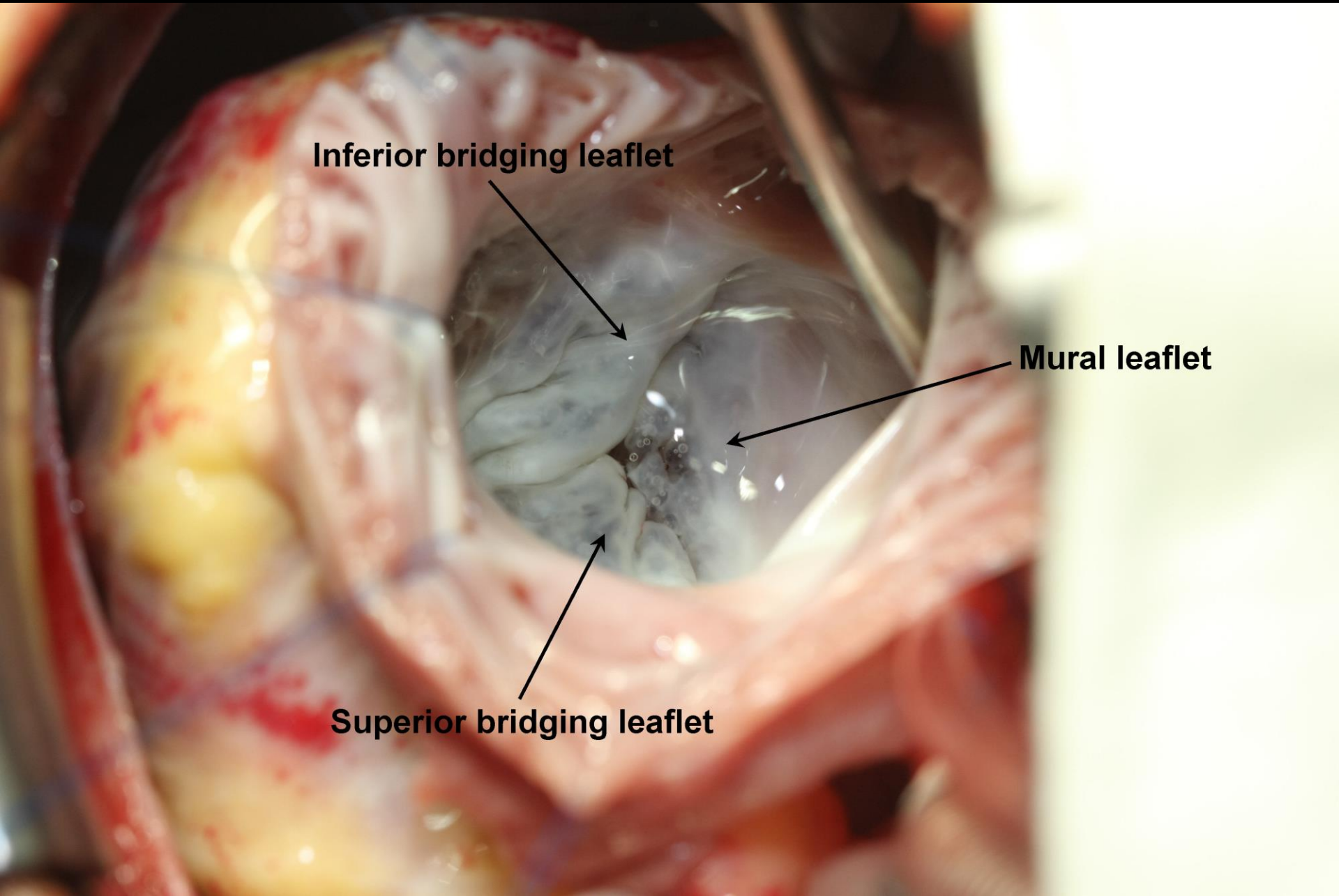


Atrioventricular Valve Leaflets

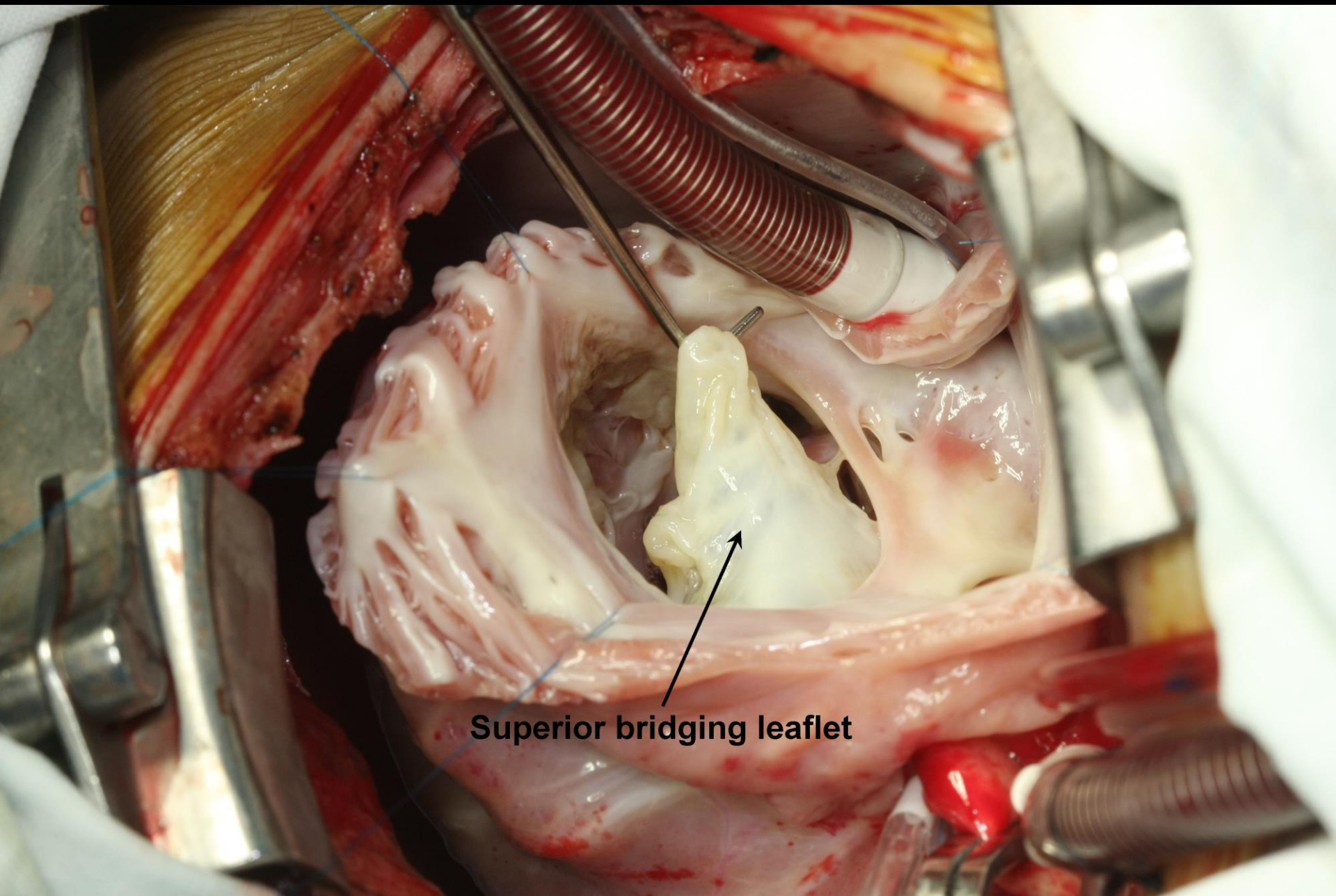


Anderson RH, et al. Paediatric Cardiology. 3rd ed.

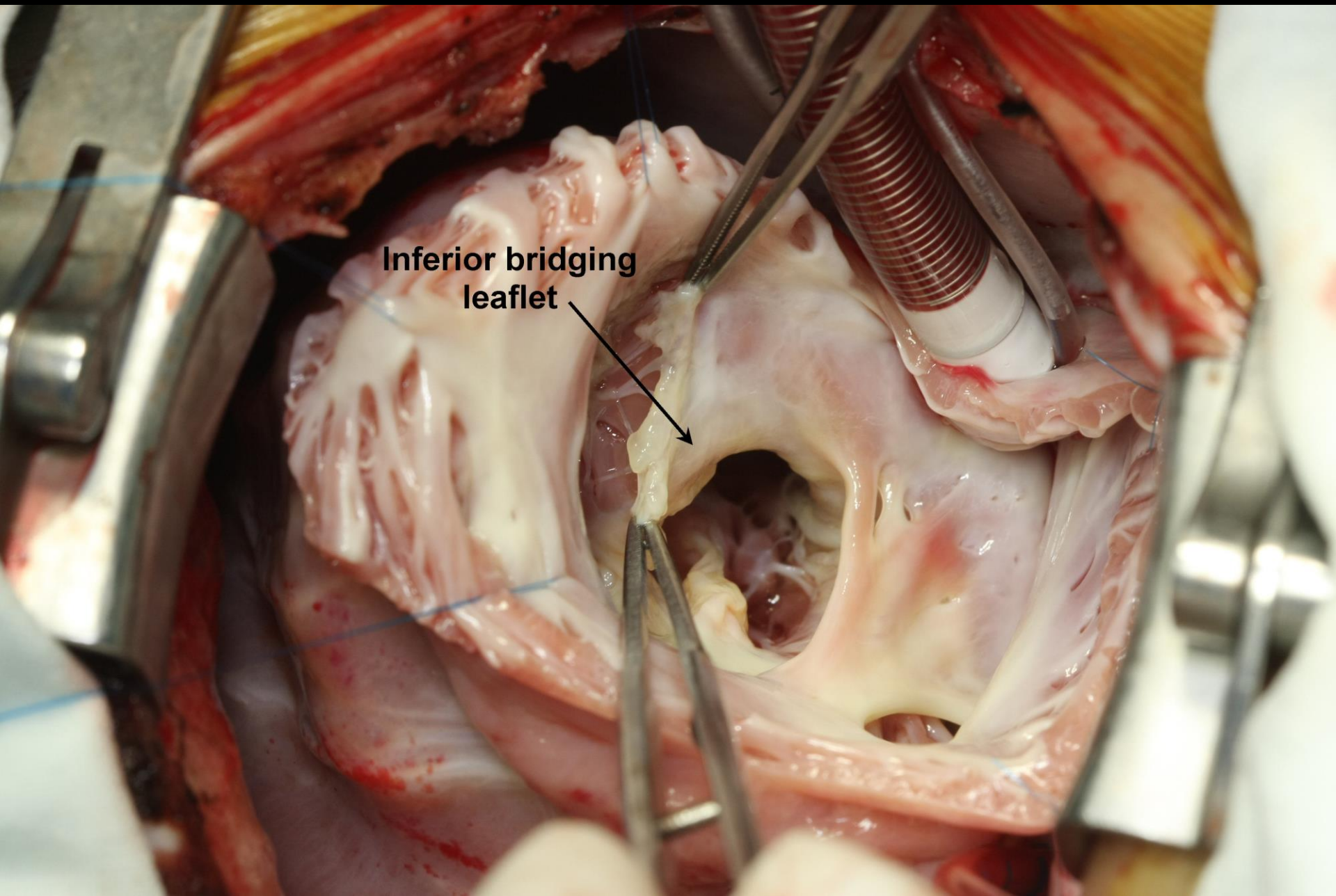
Left Atrioventricular Valve



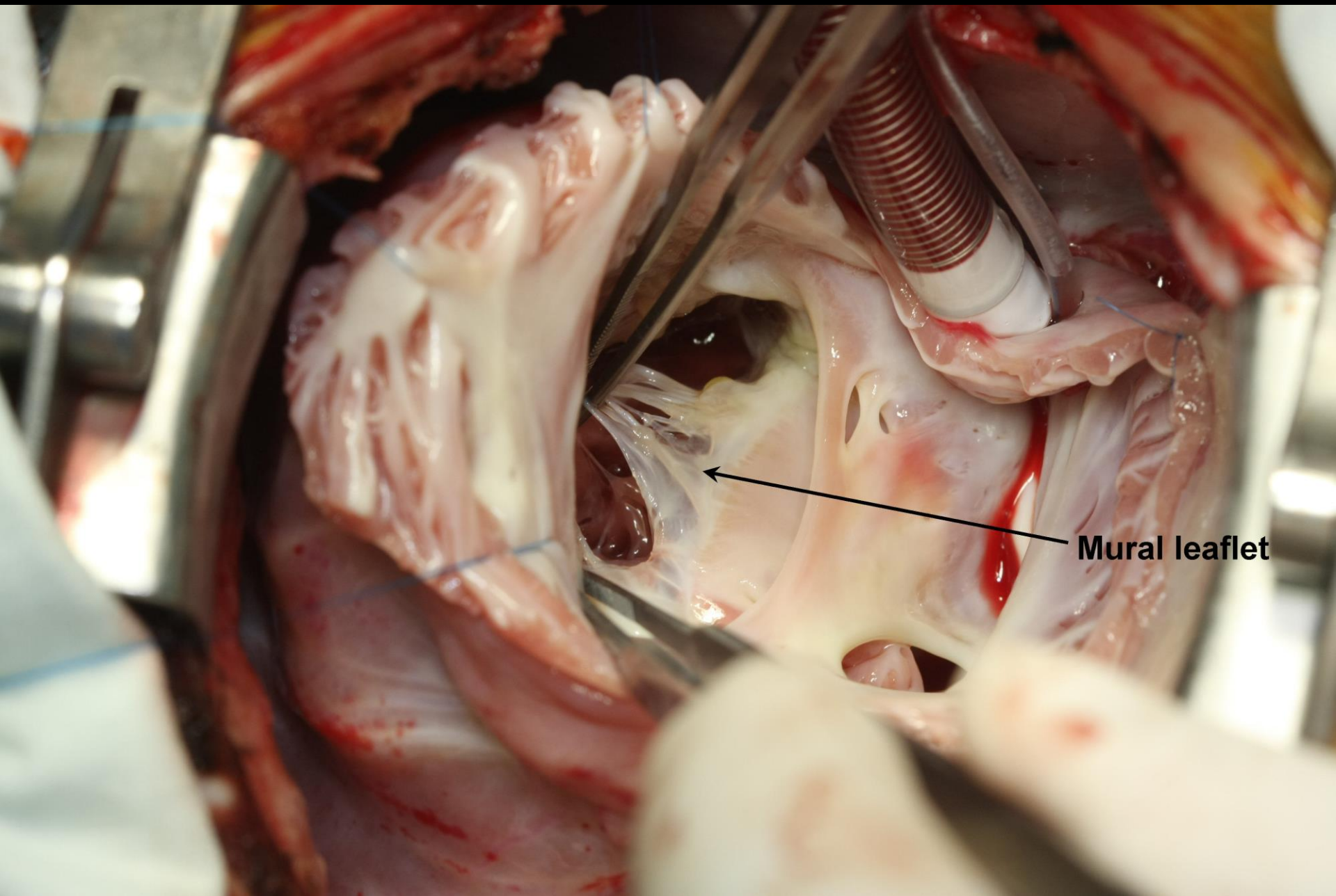
Superior Bridging Leaflet



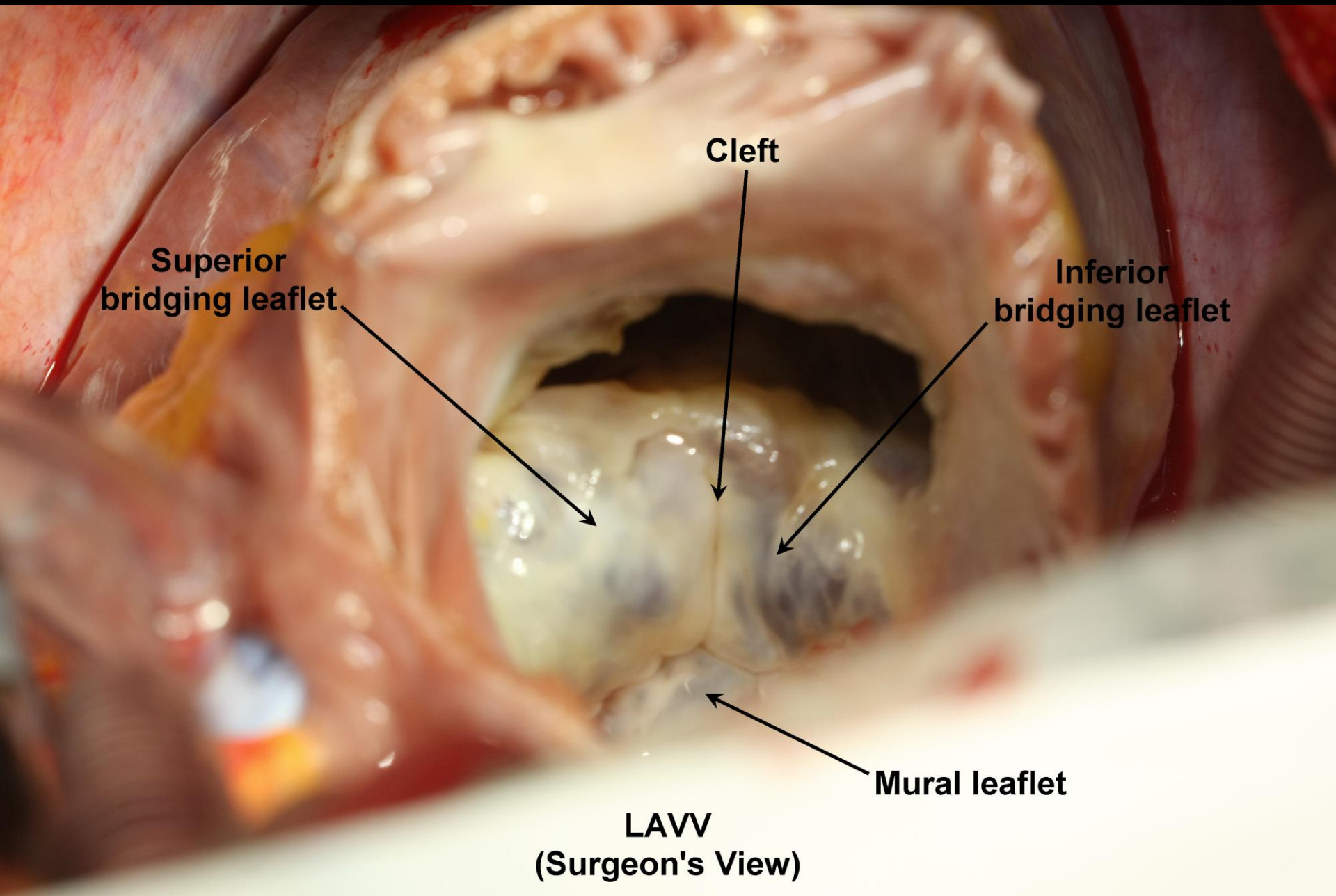
Inferior Bridging Leaflet



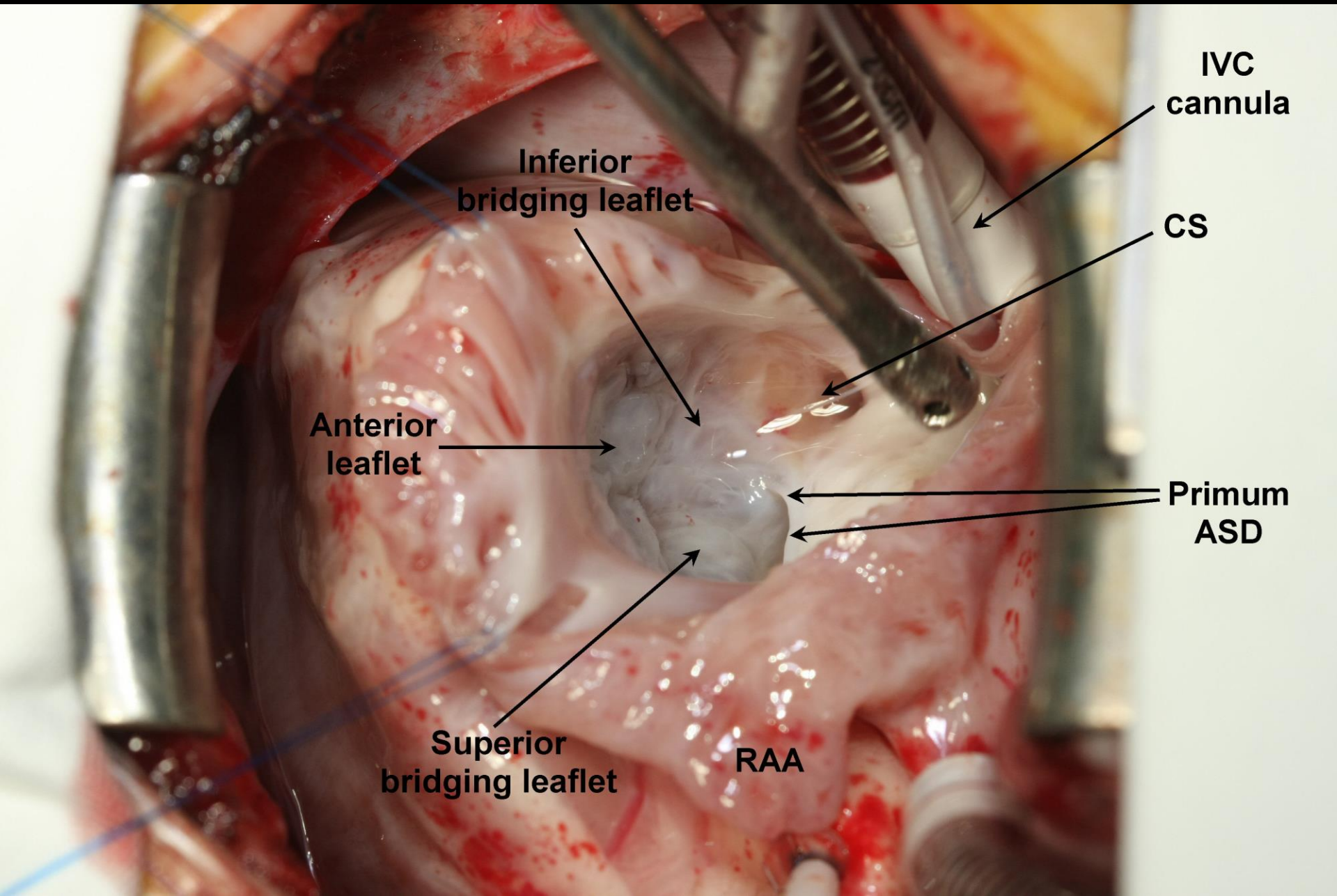
Left Mural Leaflet



Left Atrioventricular Valve



Right Atrioventricular Valve



Congenital Heart Surgery Nomenclature and Database Project

- Partial (incomplete) AVSD

  = separate AV valve orifices + primum ASD

- Intermediate (transitional) AVSD

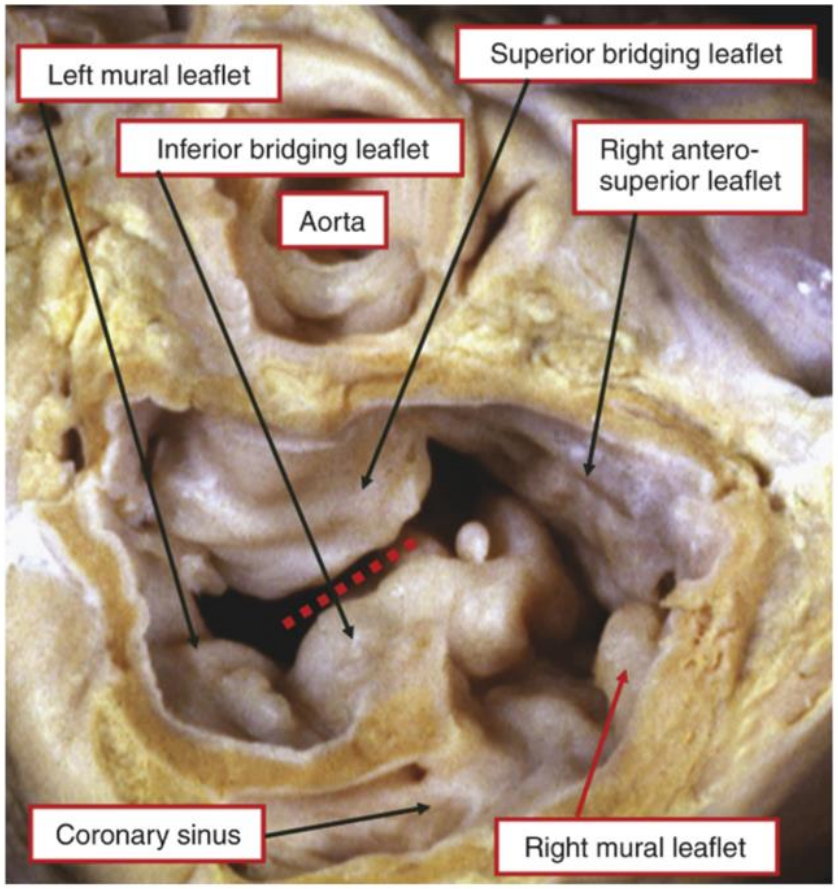
  = separate AV valve orifices + primum ASD + restrictive VSD

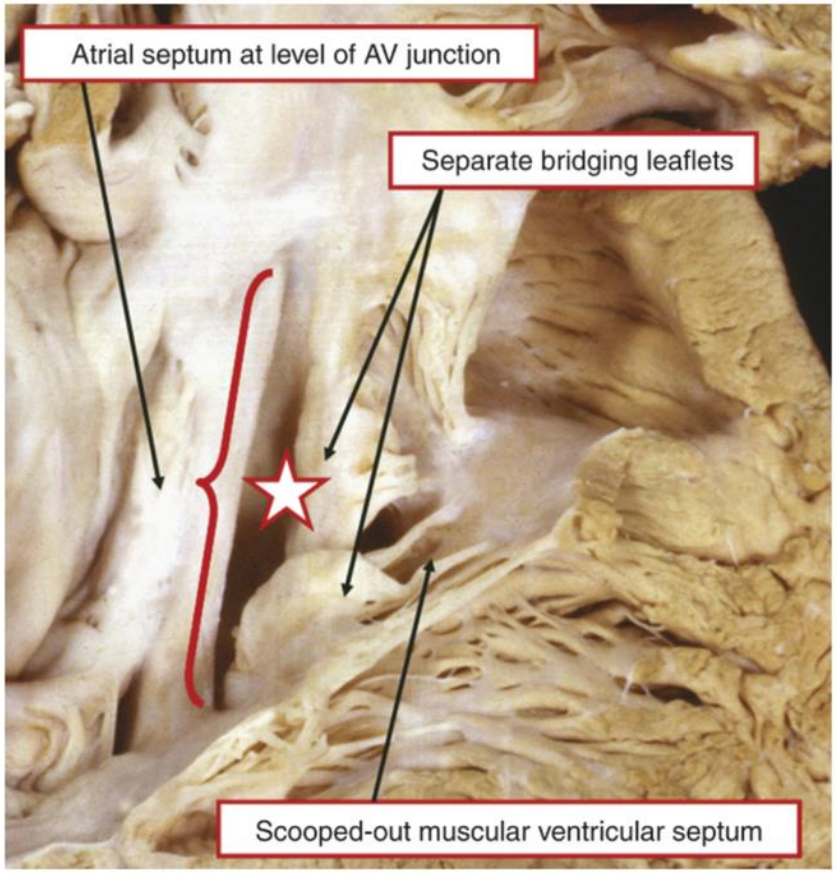
- Complete AVSD

  = common AV valve orifice + primum ASD + nonrestrictive VSD

Ann Thorac Surg. 2000;69:S36-43.

Complete AVSD





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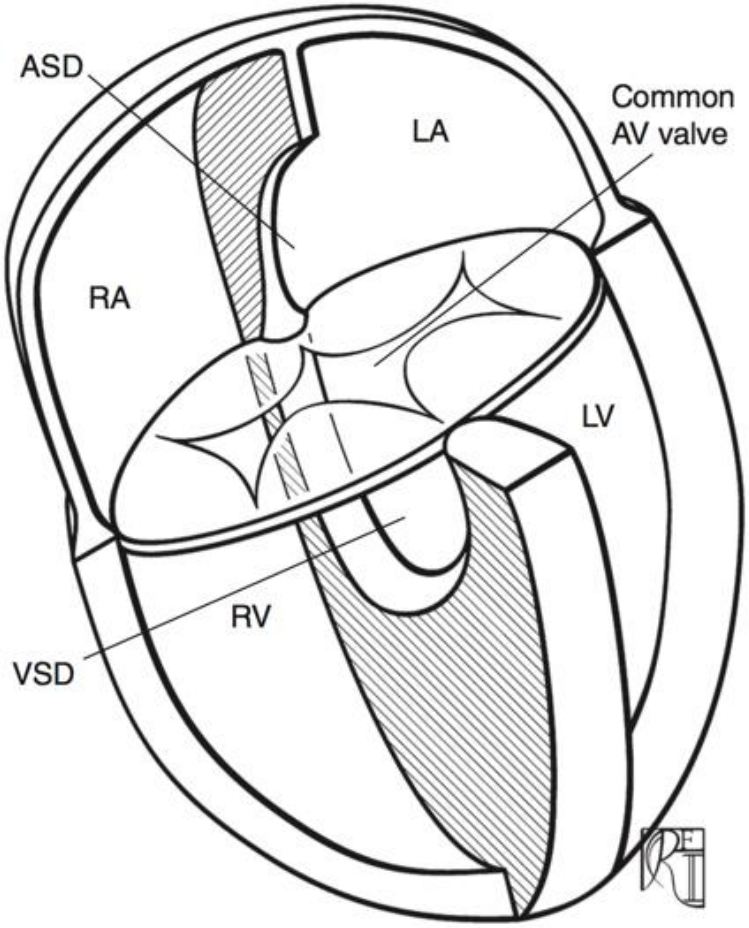
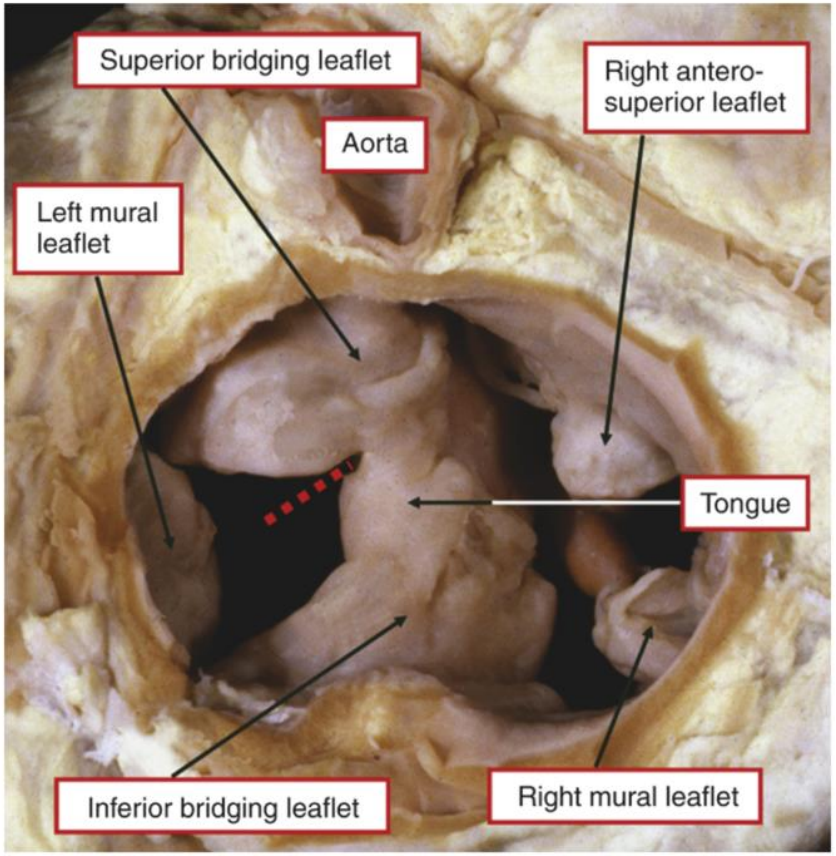
Complete AVSD

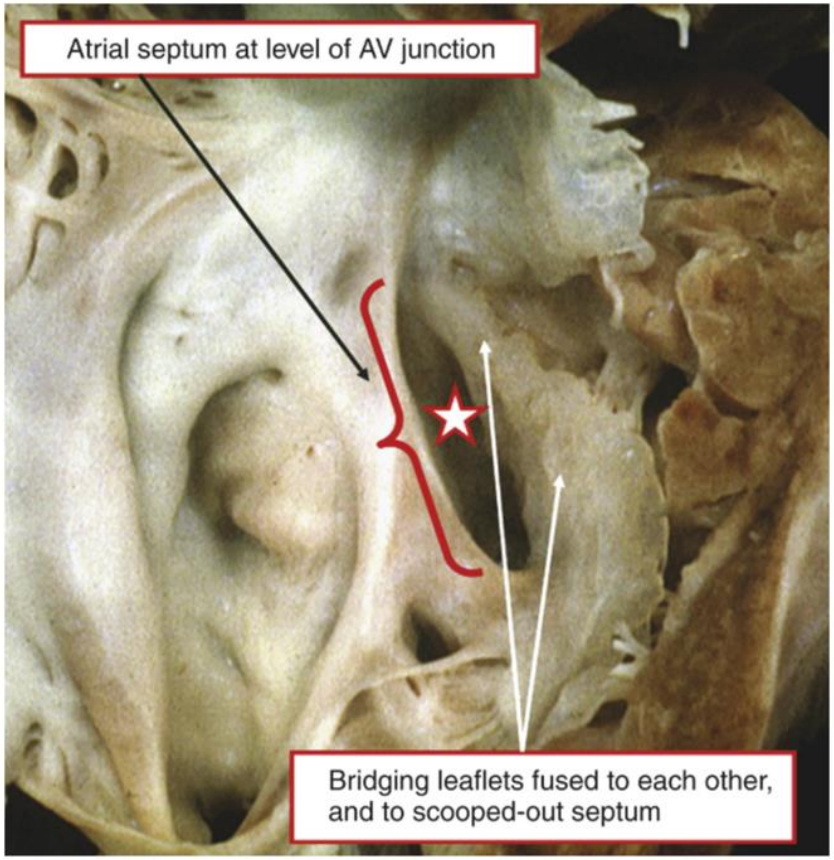


Mavroudis C, et al. Pediatric Cardiac Surgery. 4th ed.

Complete AVSD

Partial AVSD





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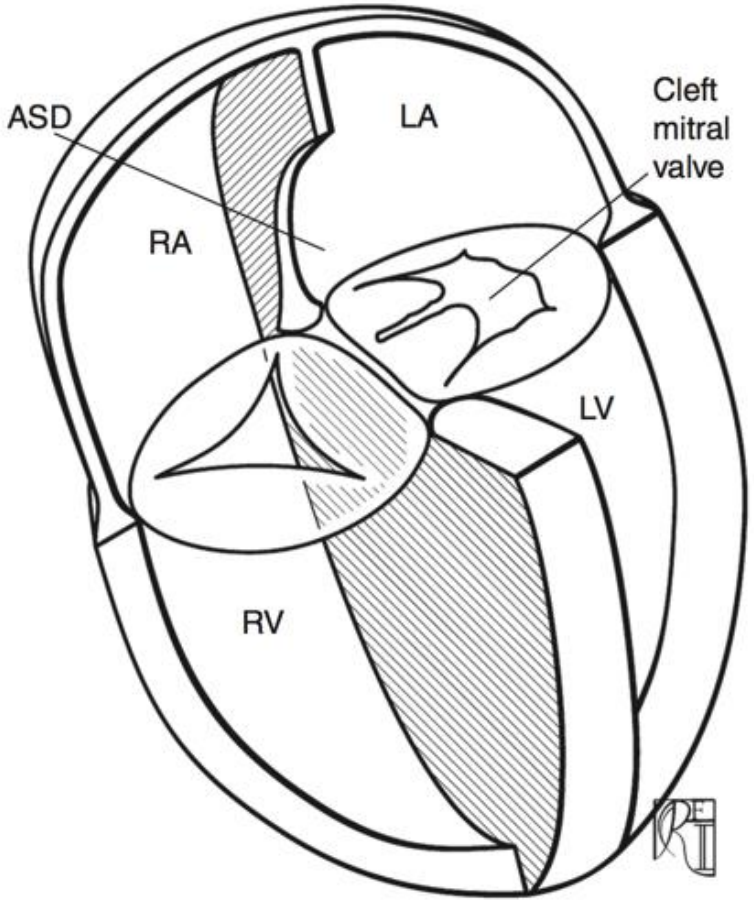
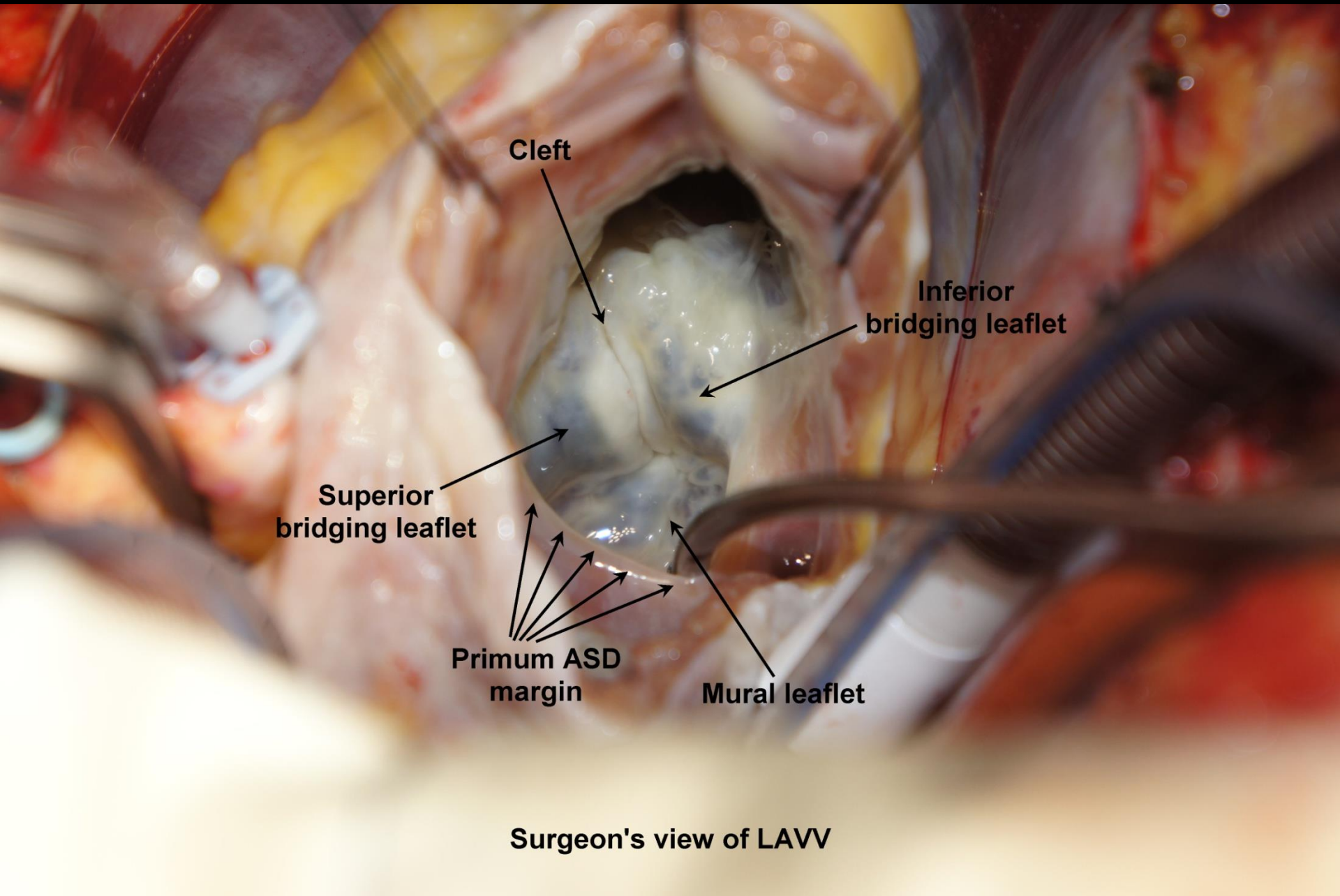
Partial AVSD



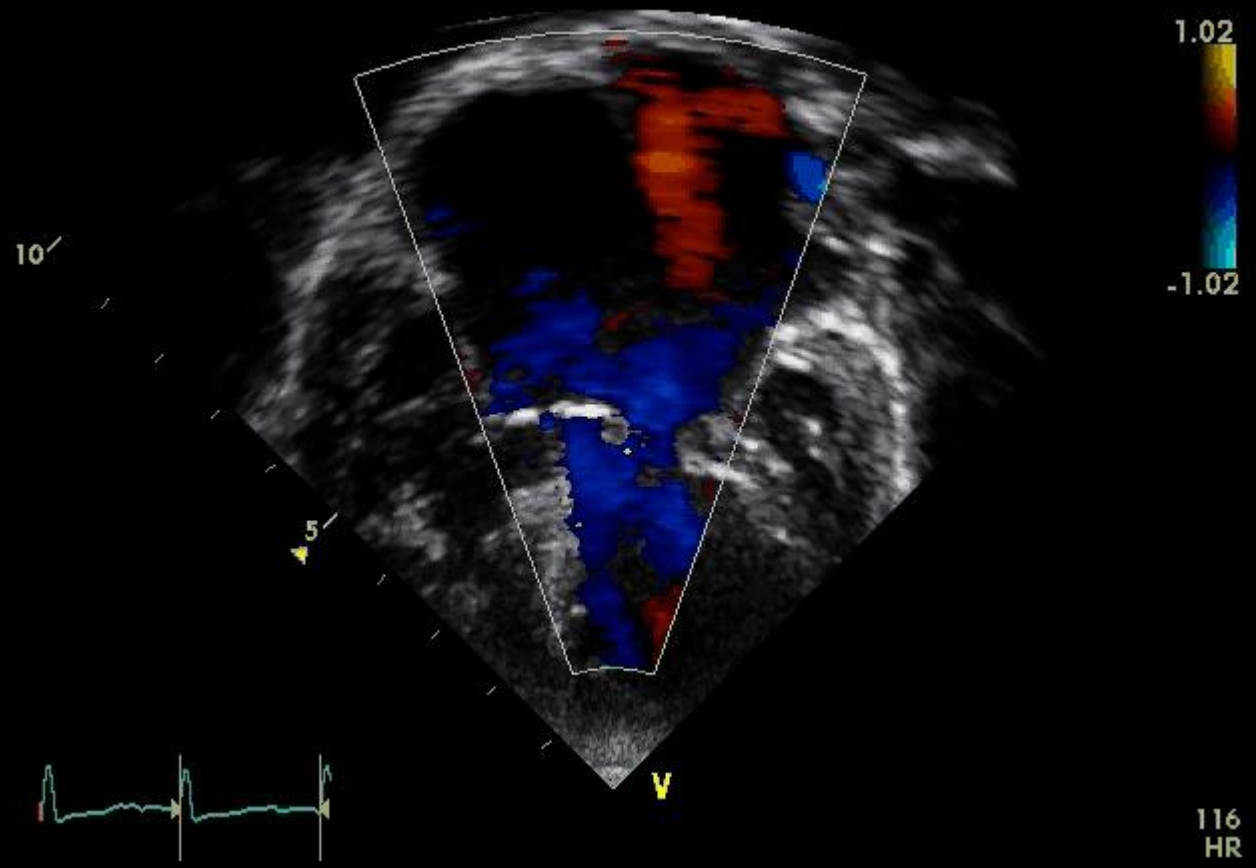
Mavroudis C, et al. Pediatric Cardiac Surgery. 4th ed.

Partial AVSD

Left Atrioventricular Valve Cleft

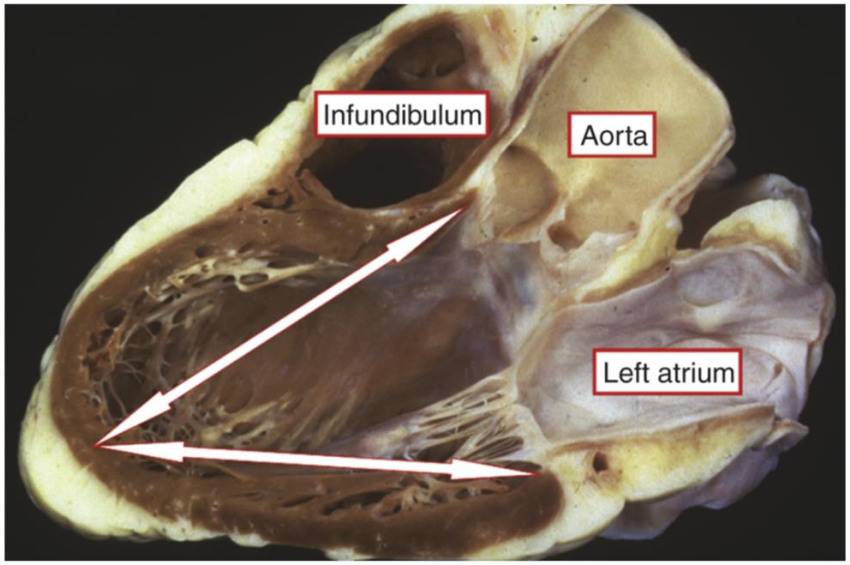


Atrioventricular Valve Regurgitation

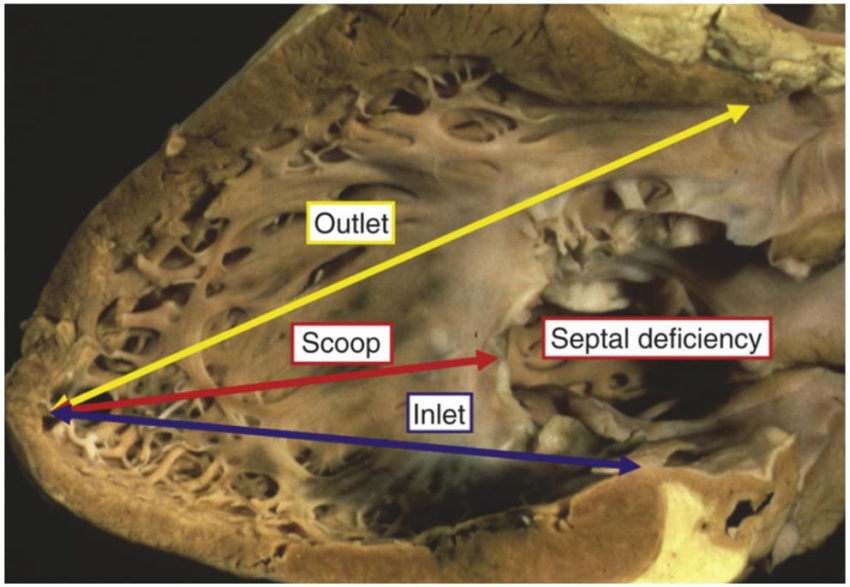


Inlet and Outlet Dimensions of the LV

Normal Heart

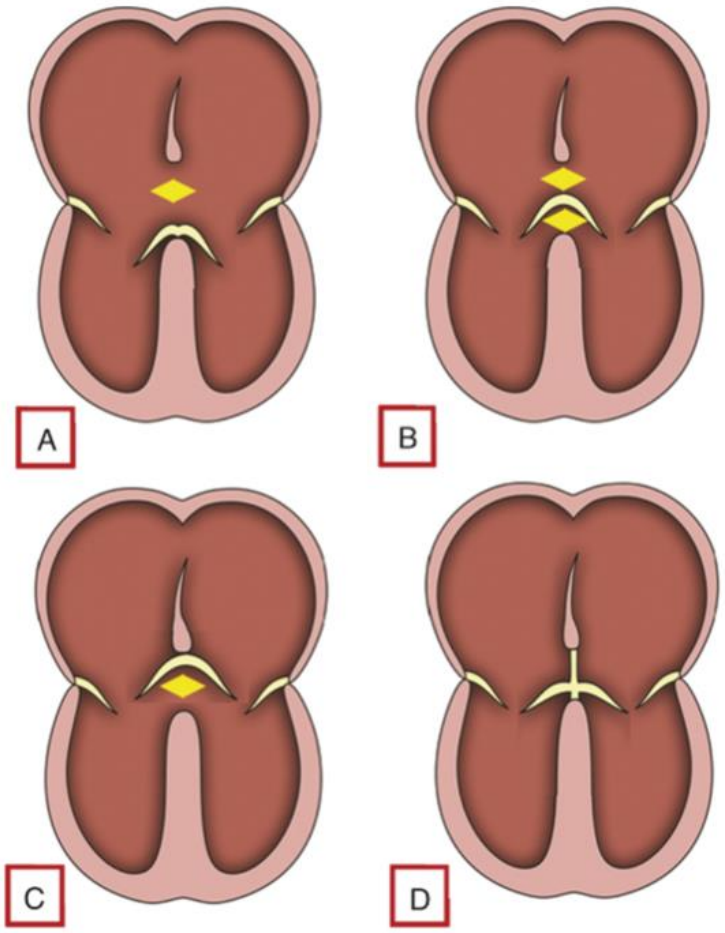


Atrioventricular Septal Defect



Anderson RH, et al. Paediatric Cardiology. 3rd ed.

Potential for Shunting



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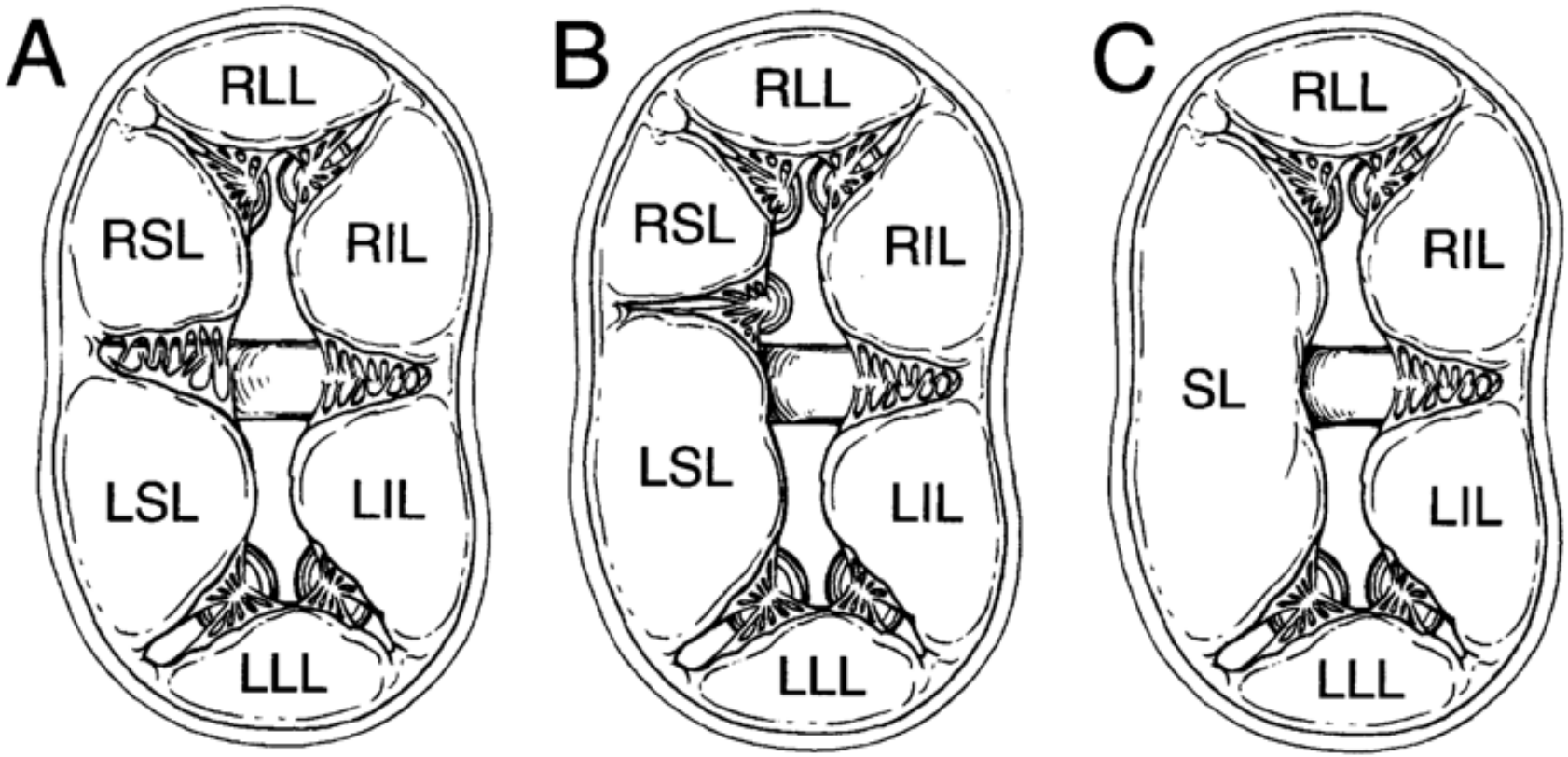
Rastelli Classification



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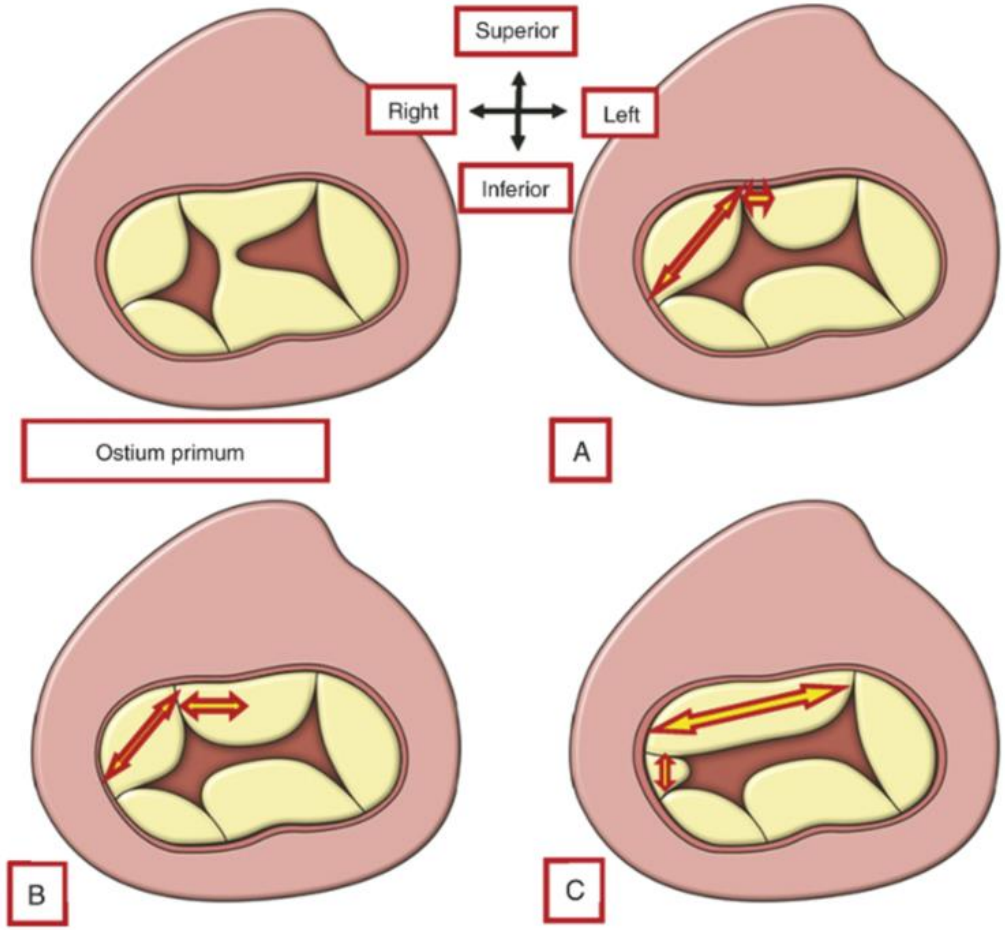
Rastelli Classification



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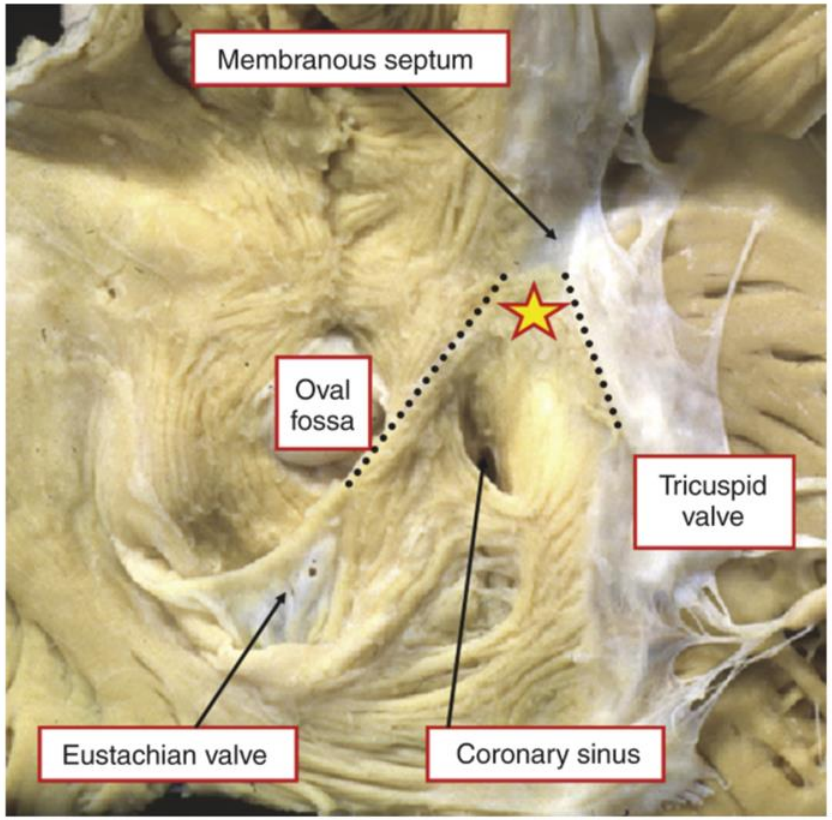
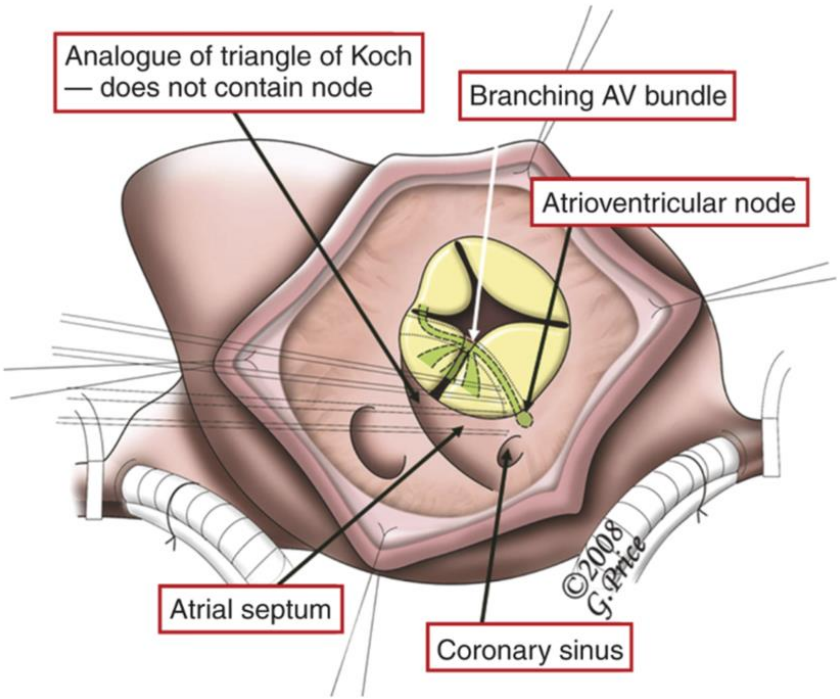
Location of the AV Node

Normal Heart

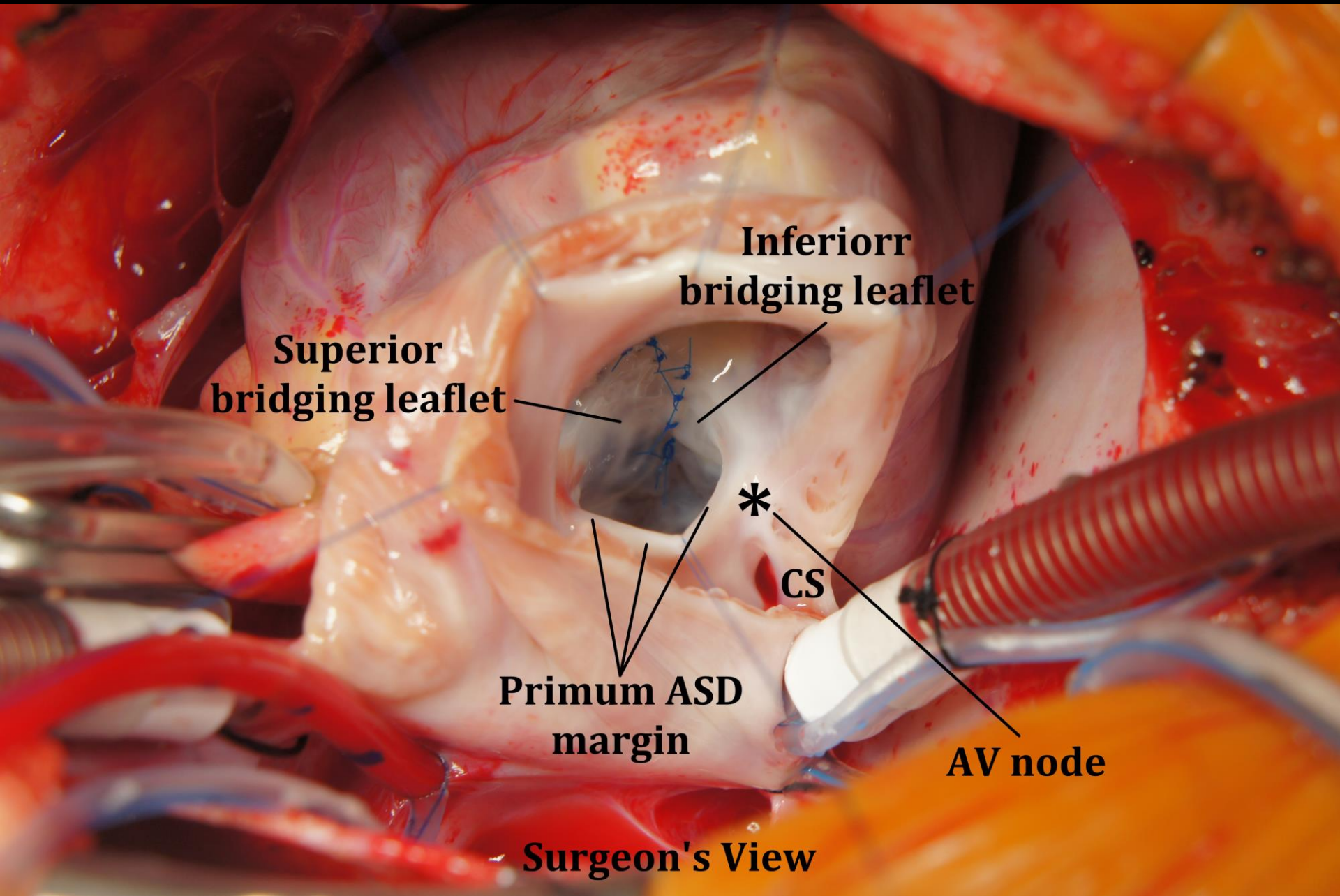


Atrioventricular Septal Defect



Anderson RH, et al. Paediatric Cardiology. 3rd ed.

Location of the AV Node in AVSD



Associated Cardiac Anomalies

- Patent ductus arteriosus
- Tetralogy of Fallot
- Completely unroofed coronary sinus with left SVC
- Heterotaxia
- Double outlet right ventricle
- Additional VSDs
- Total anomalous pulmonary venous connection
- Left ventricular outflow tract obstruction
- Transposition of the great arteries
- Etc.

Down Syndrome

- Rare in patients with partial AVSD
- Common in patients with complete AVSD (about 75%)
- More frequent advanced pulmonary vascular disease

Pathophysiology

- Left-to-right shunting is present unless severe pulmonary vascular disease has developed or important right ventricular outflow tract obstruction coexists.
- Partial AVSD: similar to that of an isolated ASD
- Complete AVSD
  * Large left-to-right shunt
  * Important elevation of PVR after age 6 to 12 months
- AV valve regurgitation: ventricular volume overload

AV Valve Regurgitation

- About 10-15% of patients with partial AVSD have important AV valve regurgitation.
- About 35% of patients with complete AVSD have important AV valve regurgitation.
- AV valve regurgitation may be more common in older patients with complete AVSD.

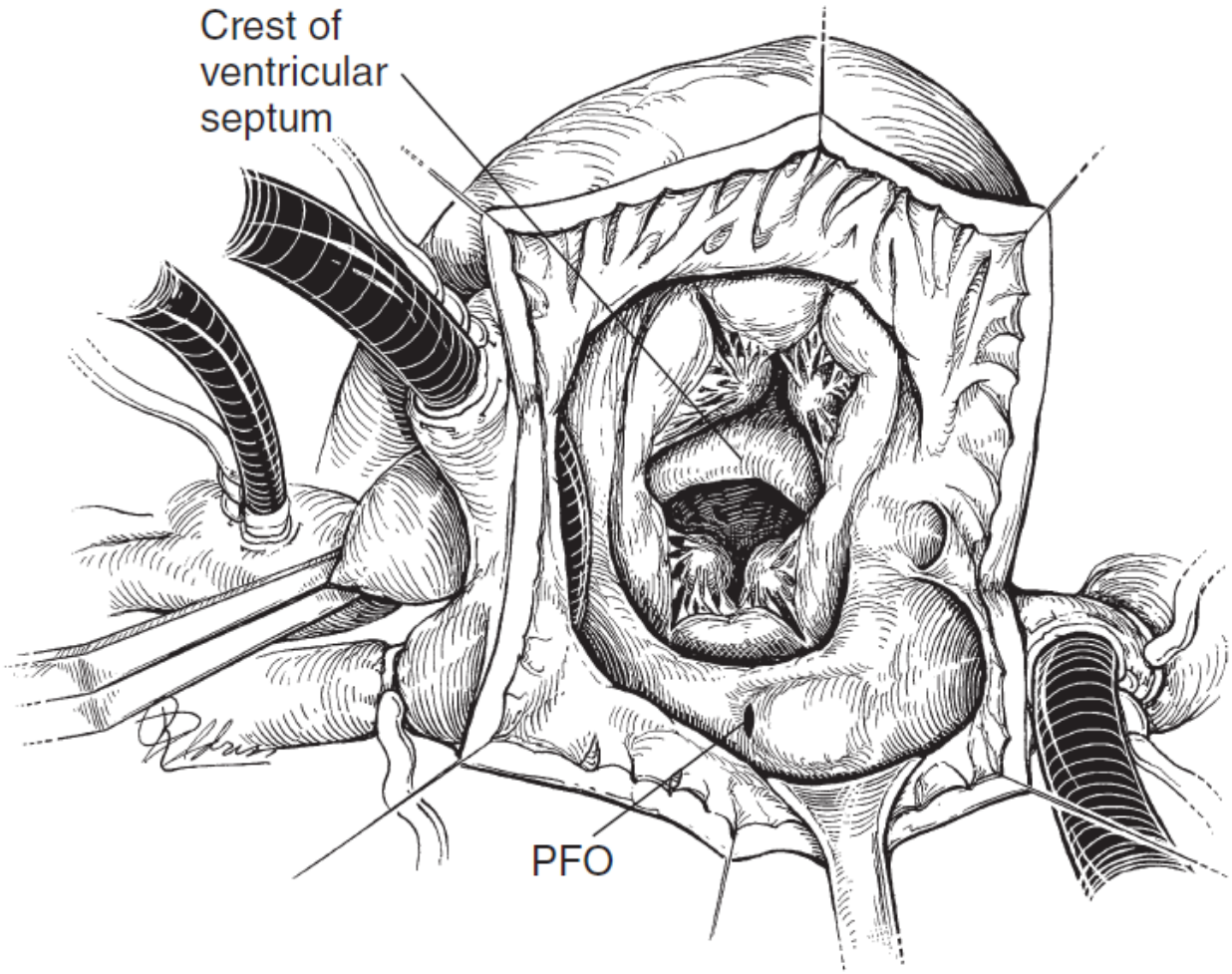
Timing of Surgery

- Asymptomatic partial AVSD: 1-3 years of age
- Partial AVSD with significant AVVR: earlier repair
- Complete AVSD: 3-6 months of age
- Symptomatic complete AVSD: earlier repair
- Role of pulmonary artery banding?

Goals of Surgery

1. Closing the interatrial communication
2. Closing the interventricular communication
3. Avoiding damage to the AV node and bundle of His
4. Maintaining or creating two competent, nonstenotic AV valves

Exposure

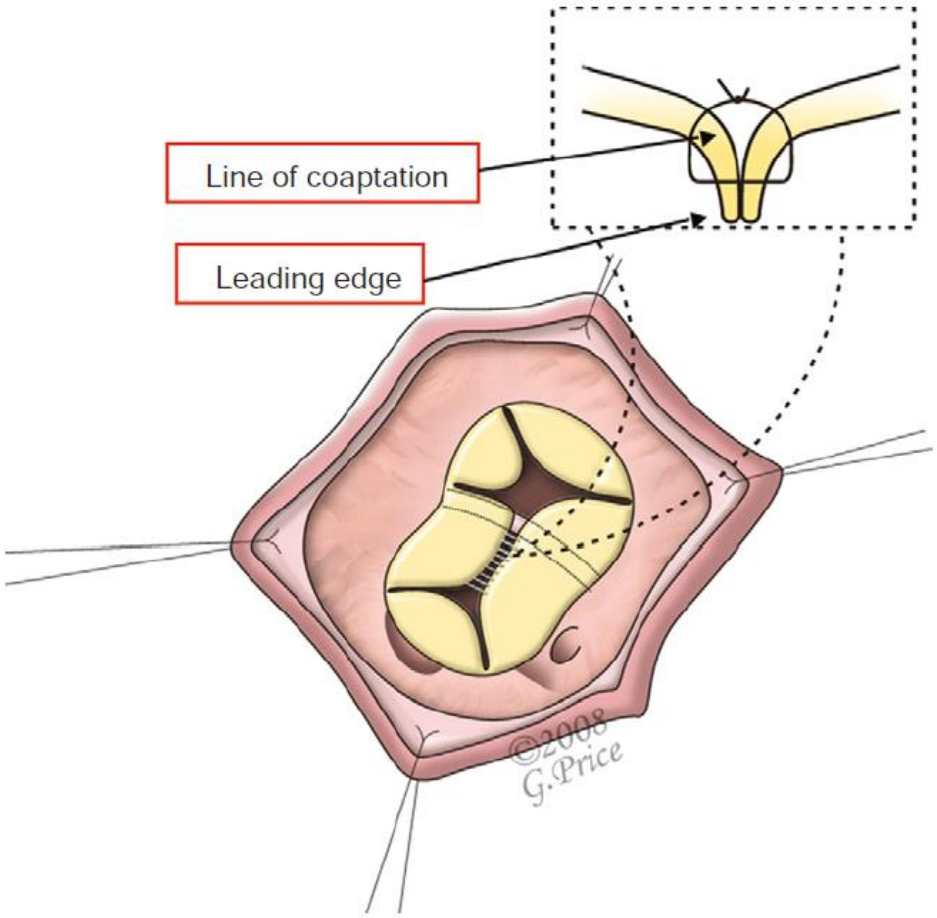


Mavroudis C, et al. Pediatric Cardiac Surgery. 4th ed.

Left AV Valve Cleft Repair

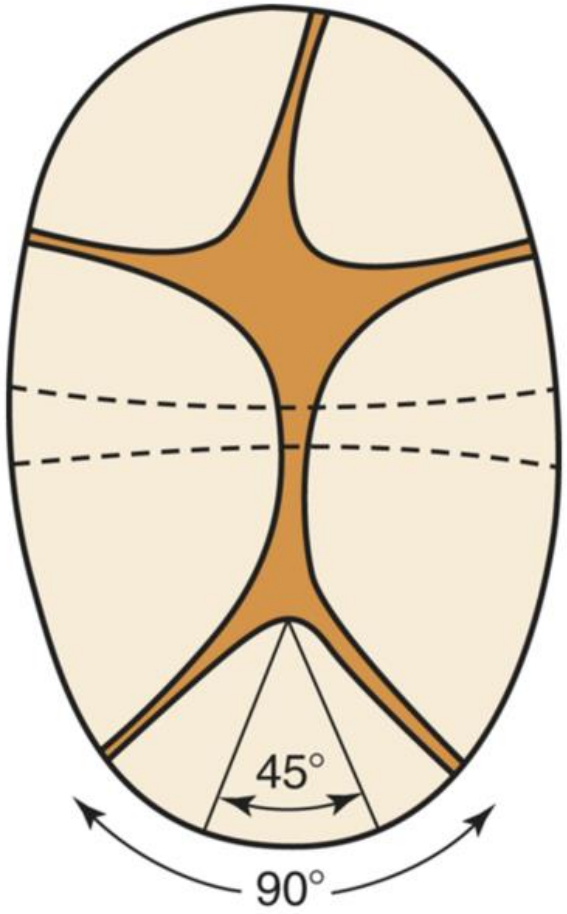
- Routine cleft repair is a current standard.
- The extent of cleft repair is determined by the position of the papillary muscles and the size of the left mural leaflet.
- When the papillary muscles are close together or a single papillary muscle is present, complete cleft repair can result in significant stenosis.

Left AV Valve Cleft Repair



Anderson RH, et al. Paediatric Cardiology. 3rd ed.

Angular Size of the Left Mural Leaflet



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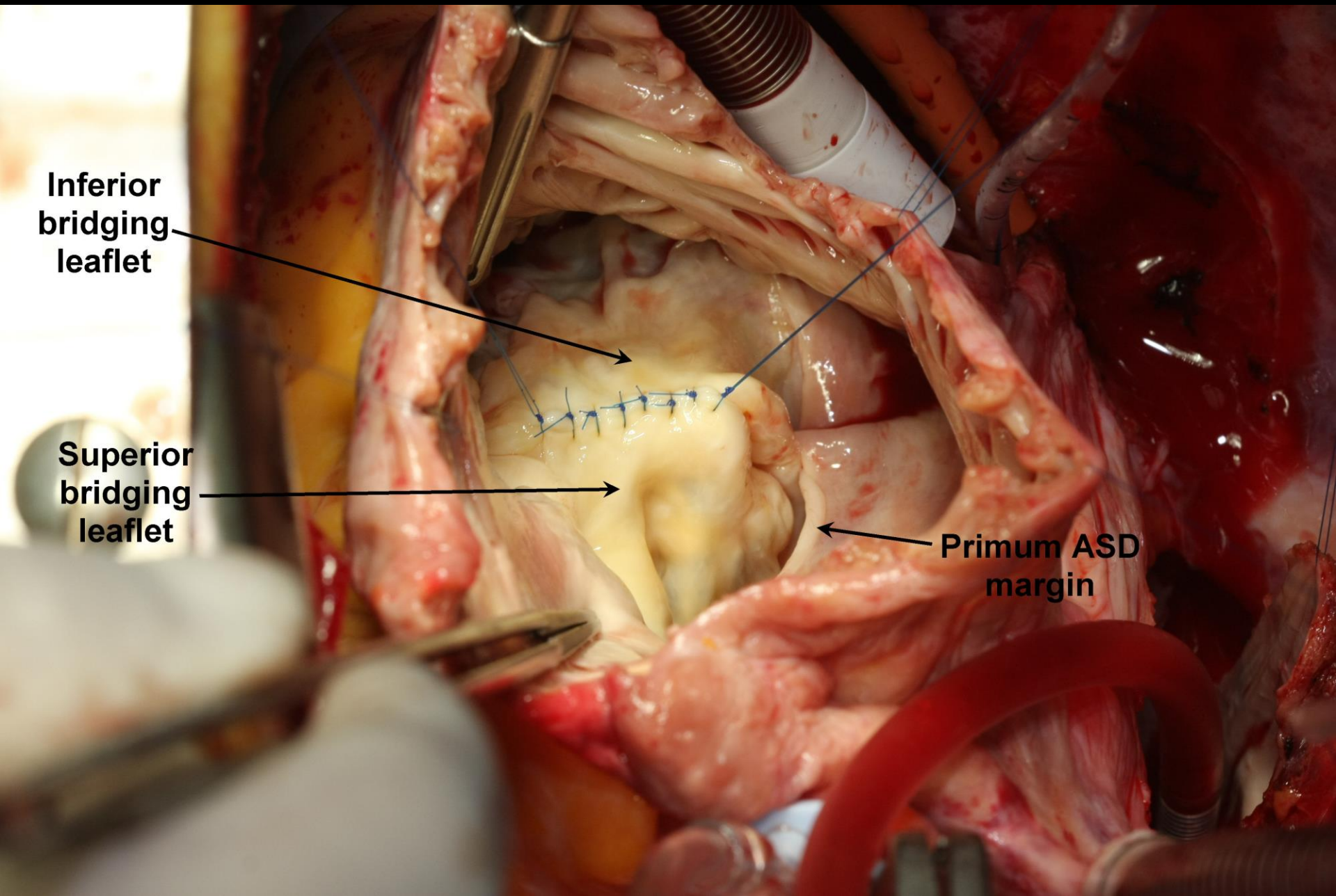
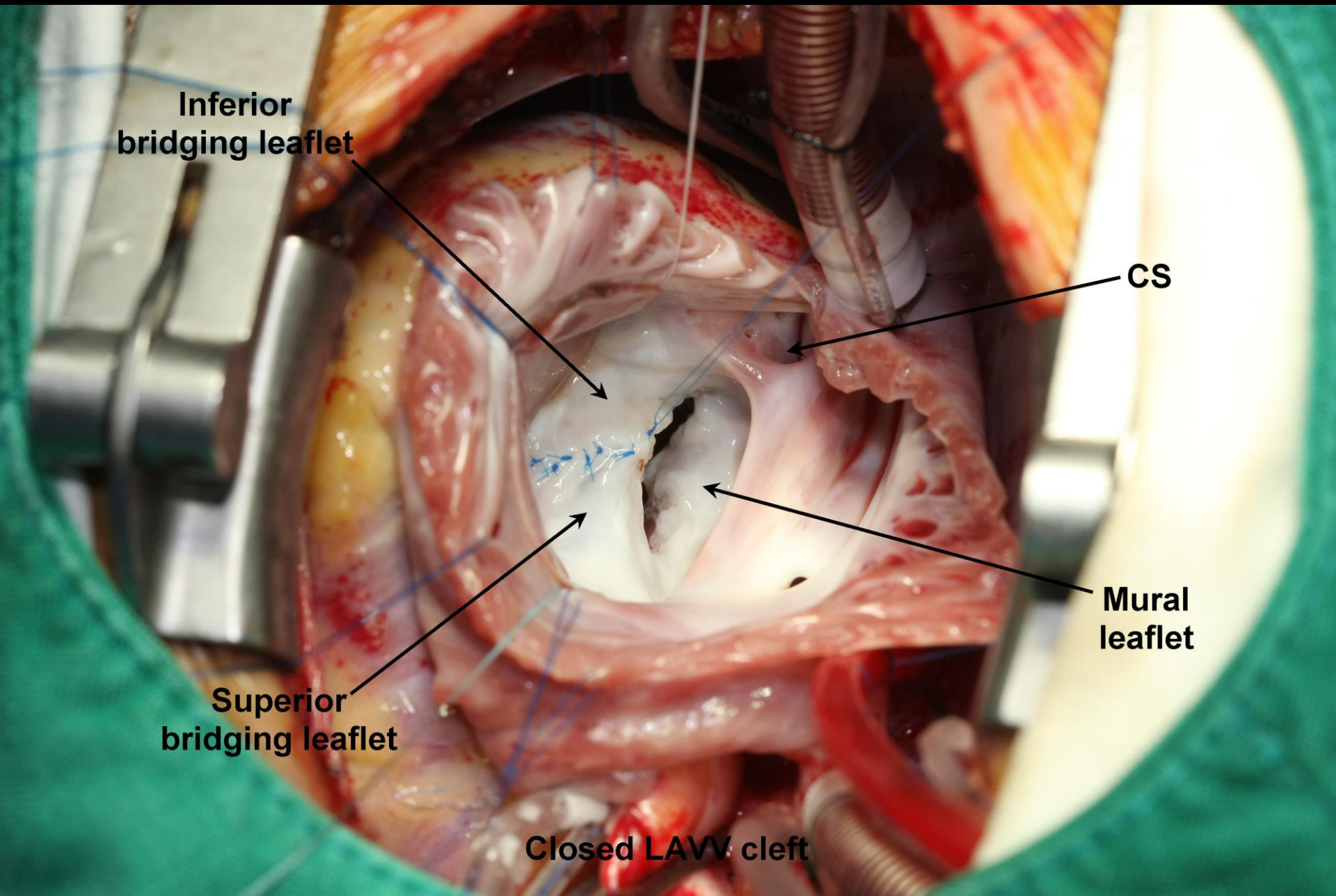
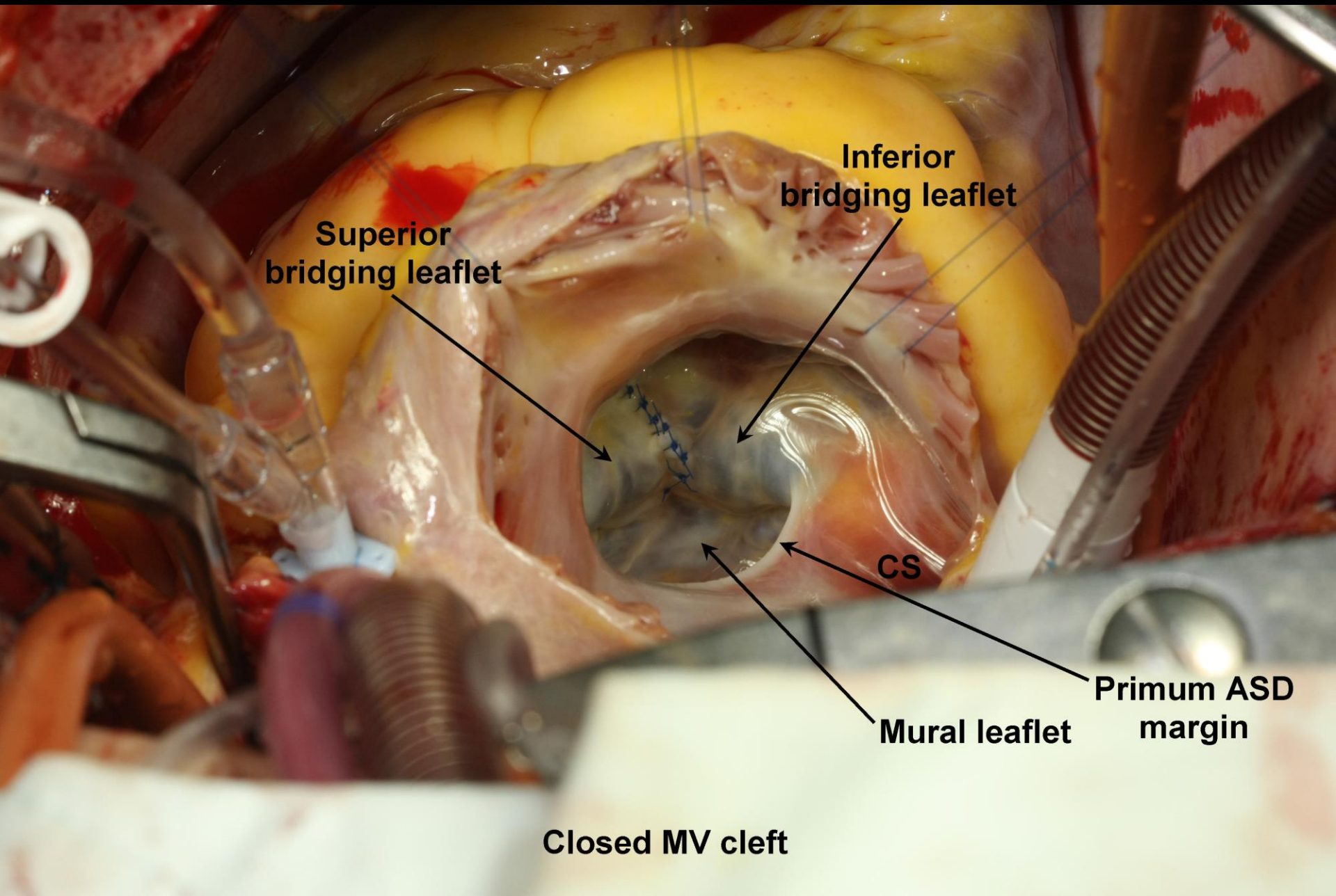
Repair of the Left AV Valve Cleft



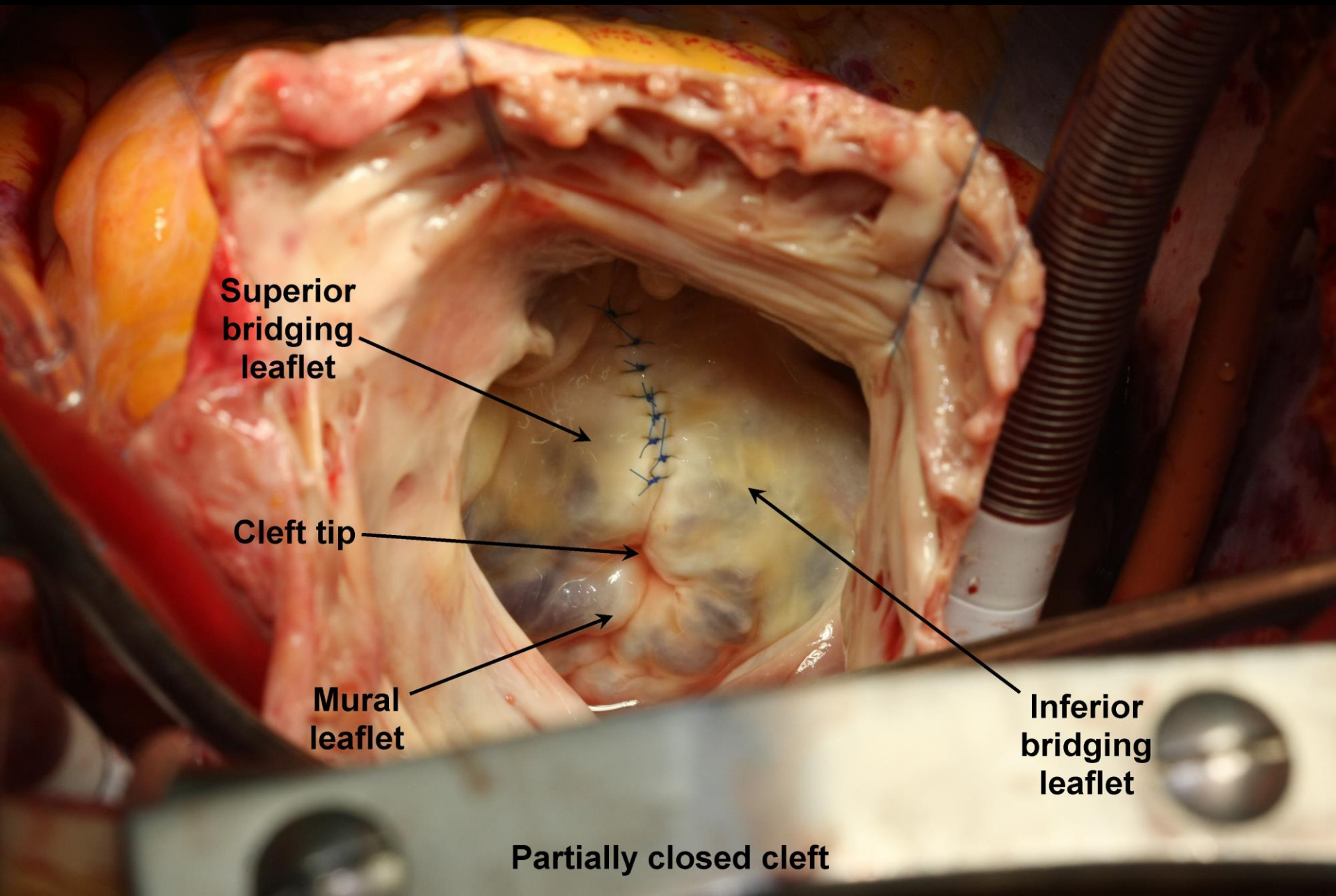
Completely Repaired Left AV Valve Cleft



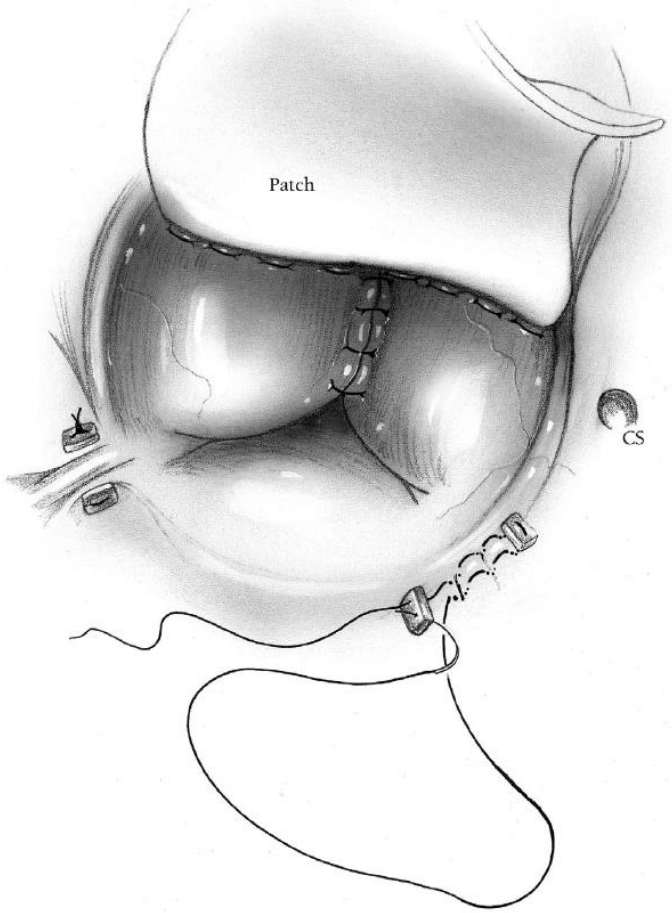
Completely Repaired Left AV Valve Cleft



Partially Repaired Left AV Valve Cleft

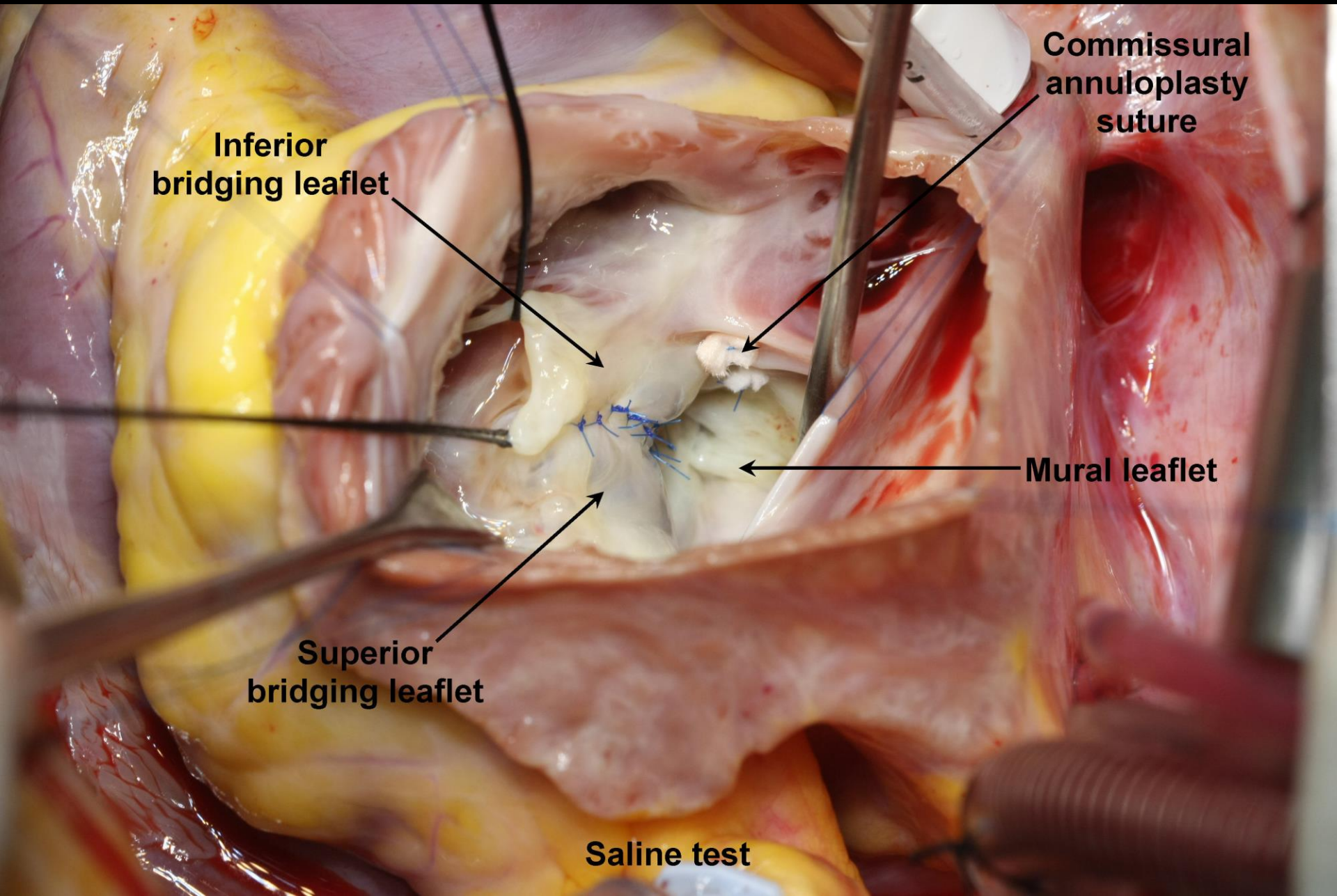


Left AV Valve Annuloplasty

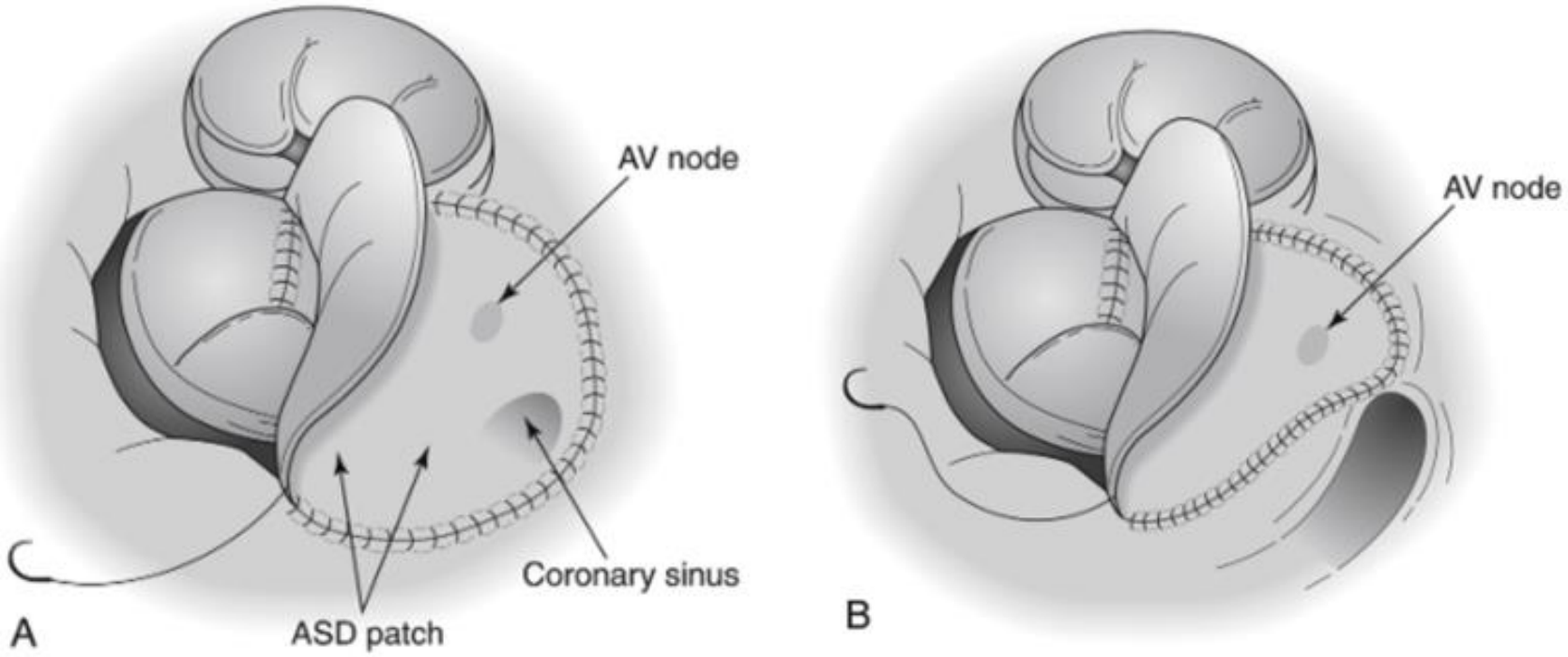


Oper Tech Thorac Cardiovasc Surg. 2004;9:221-32.

Left AV Valve Annuloplasty

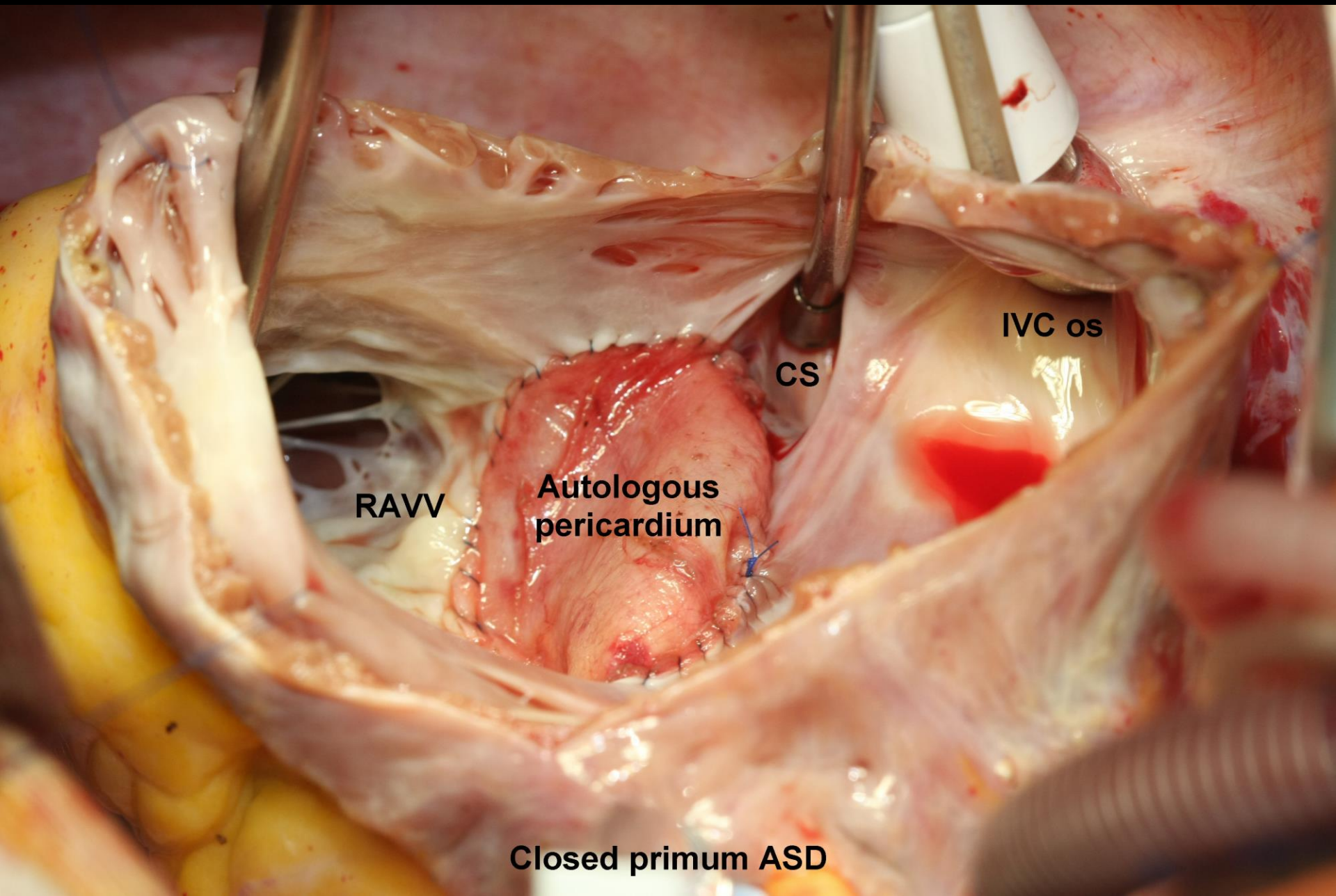


Closure of the Primum ASD



Selke FW, et al. Sabiston & Spencer Surgery of the Chest. 8th ed.

Closed Primum ASD



Repair Techniques for Complete AVSD

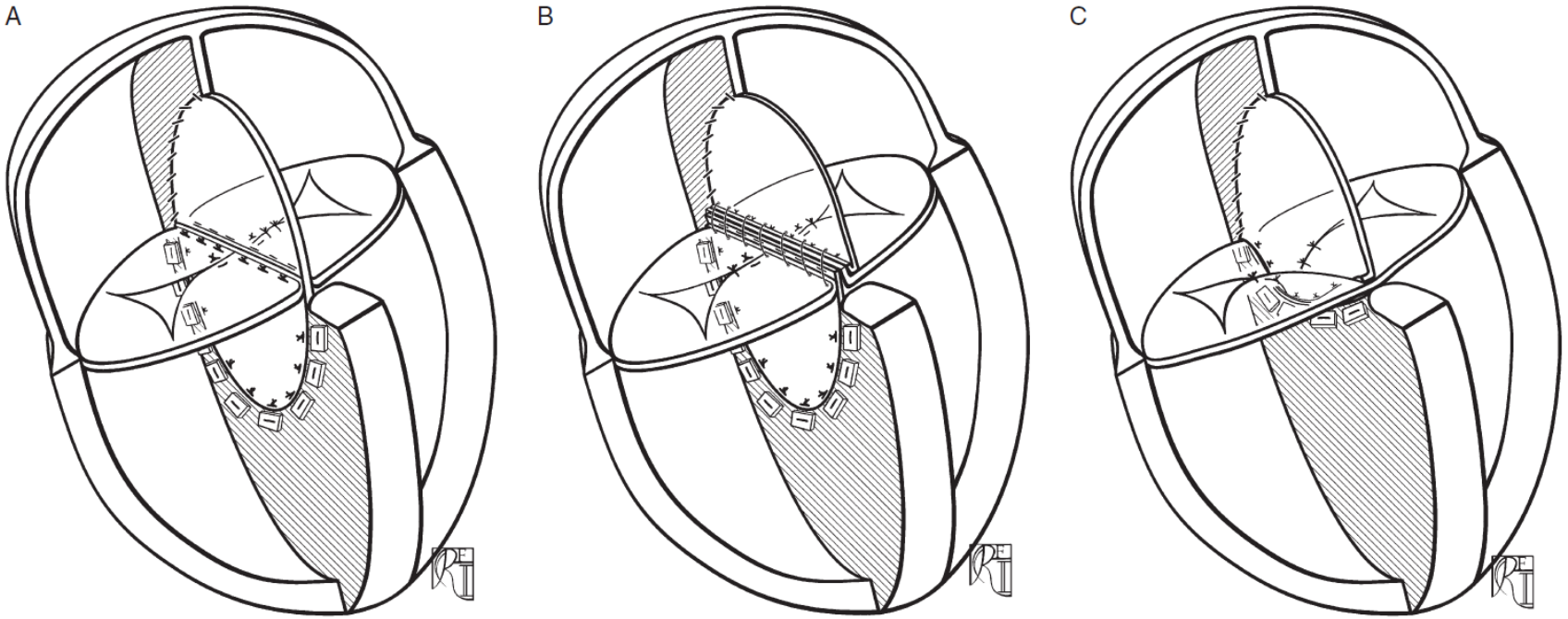
- One-patch technique
- Two-patch technique
- Modified one-patch technique

Repair Techniques for Complete AVSD

Two-patch

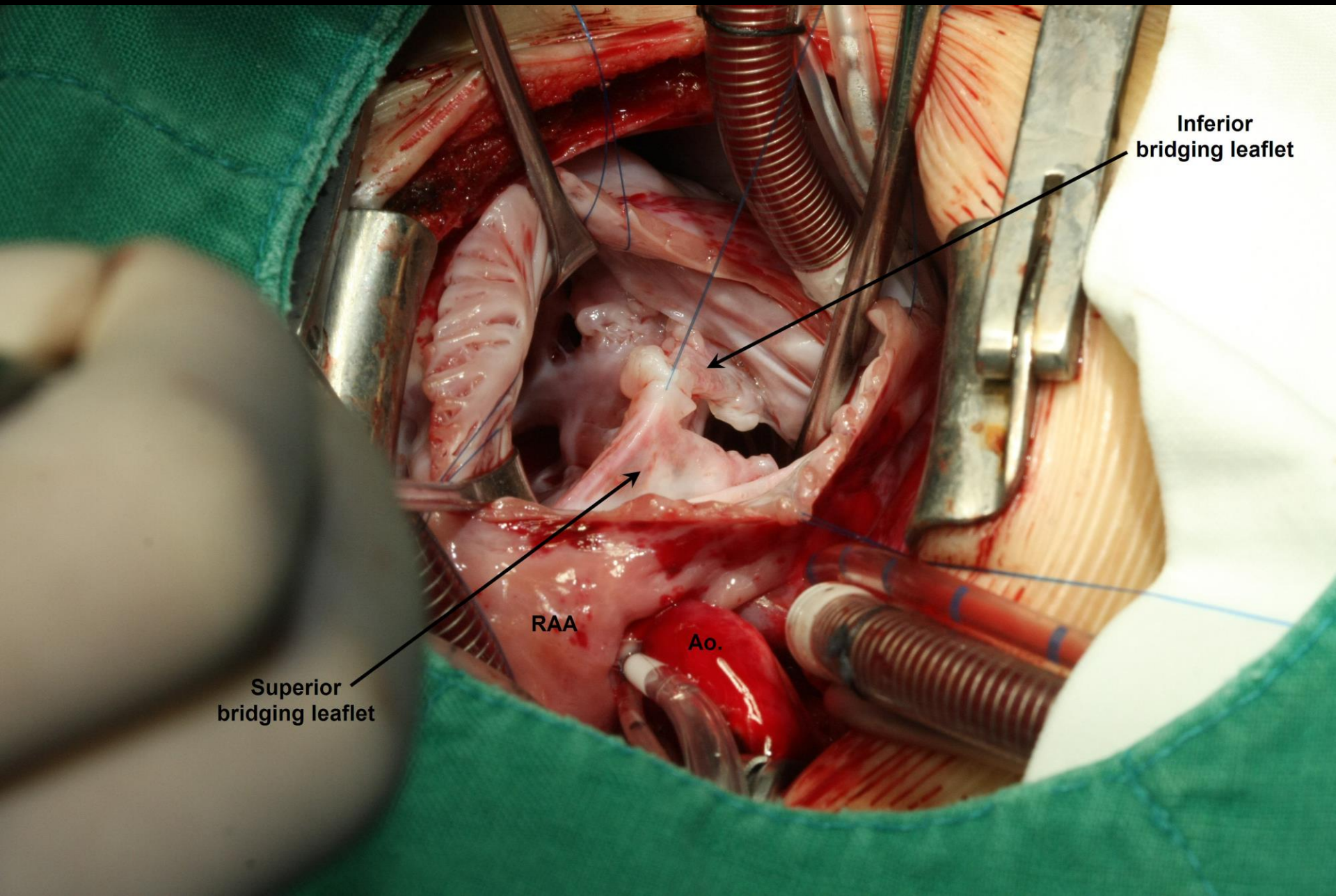
One-patch

Modified one-patch

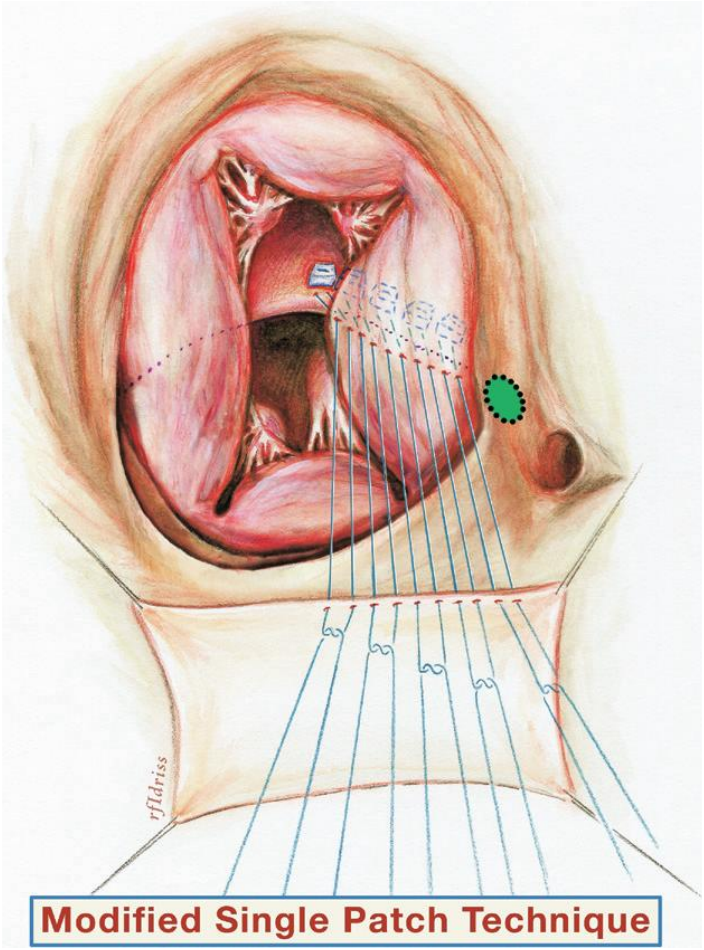


Mavroudis C, et al. Pediatric Cardiac Surgery. 4th ed.

Approximation of the Zone of Apposition

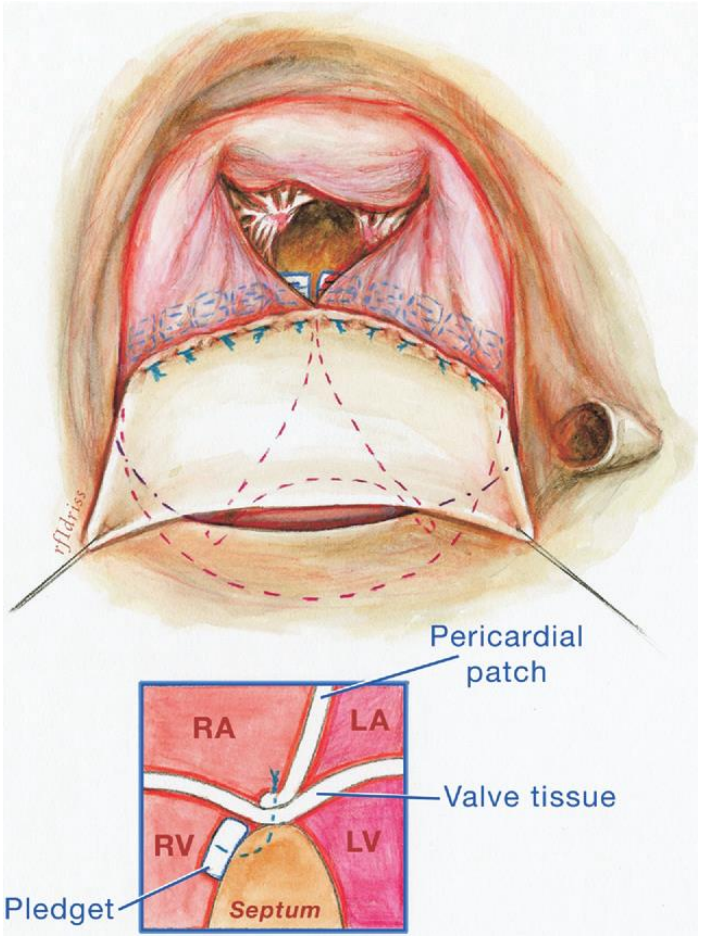


Modified One-Patch Technique



Ann Thorac Surg. 2007;84:2038-46.

Modified One-Patch Technique



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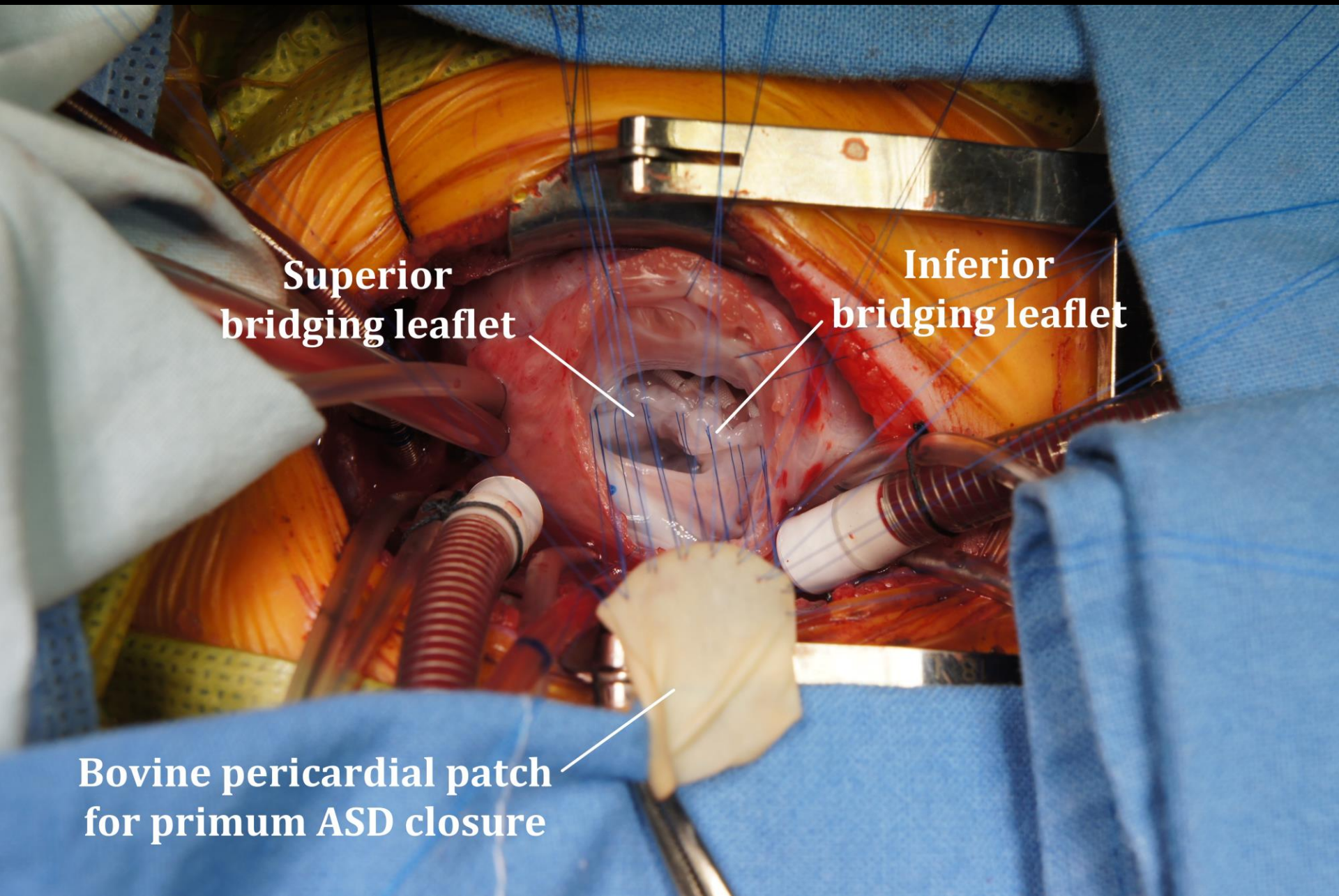
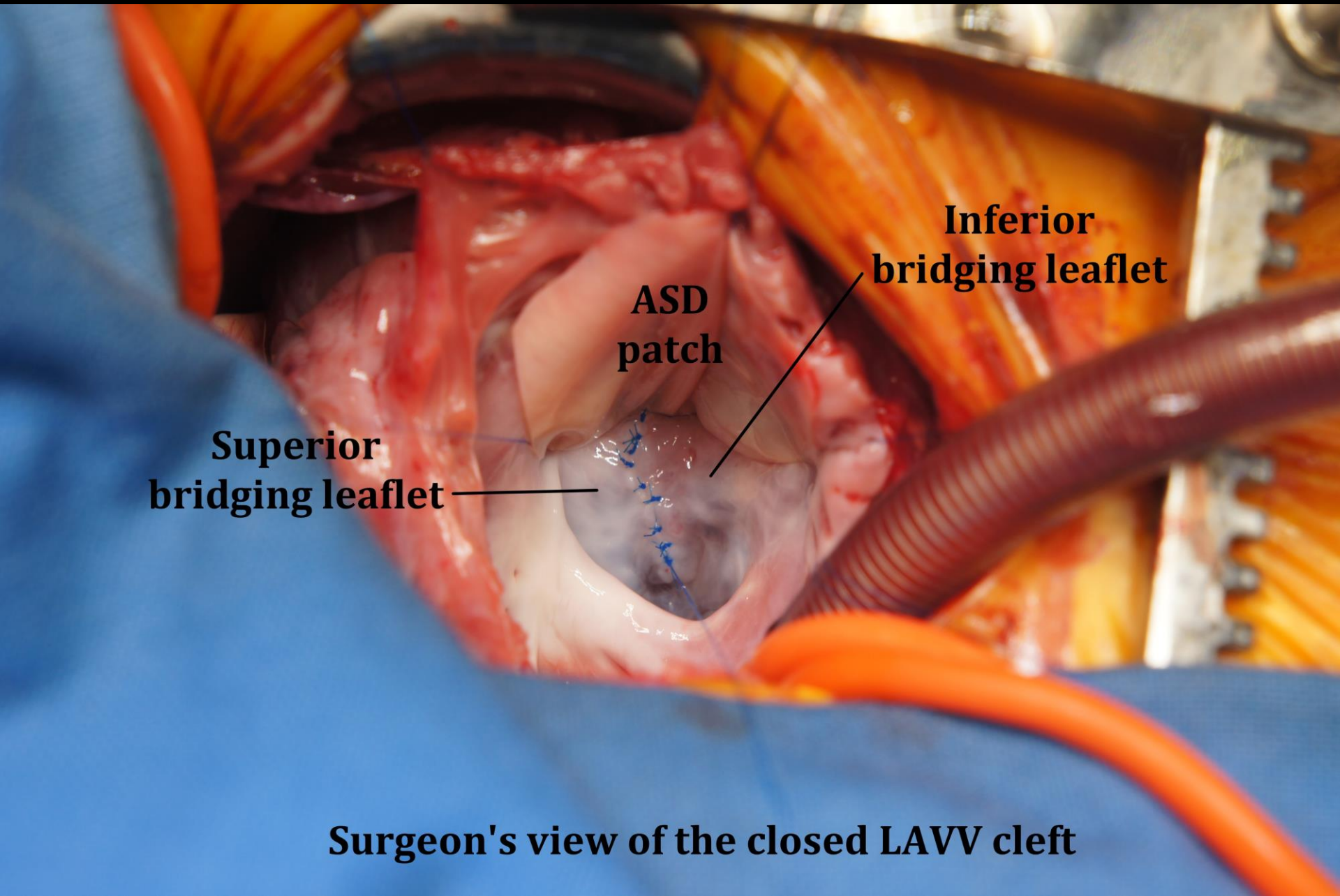
Modified One-Patch Technique



Modified One-Patch Technique



Surgeon's view of the closed LAVV cleft

Contemporary outcomes of complete atrioventricular septal defect repair: Analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database

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Objective: Contemporary outcomes data for complete atrioventricular septal defect (CAVSD) repair are limited. We sought to describe early outcomes of CAVSD repair across a large multicenter cohort, and explore potential associations with patient characteristics, including age, weight, and genetic syndromes.

Methods: Patients in the Society of Thoracic Surgeons Congenital Heart Surgery Database having repair of CAVSD (2008-2011) were included. Preoperative, operative, and outcomes data were described. Univariate associations between patient factors and outcomes were described.

Results: Of 2399 patients (101 centers), 78.4% had Down syndrome. Median age at surgery was 4.6 months (interquartile range, 3.5-6.1 months), with 11.8% (n = 284) aged ≤2.5 months. Median weight at surgery was 5.0 kg (interquartile range, 4.3-5.8 kg) with 6.3% (n = 151) < 3.5 kg. Pulmonary artery band removal at CAVSD repair was performed in 122 patients (4.6%). Major complications occurred in 9.8%, including permanent pacemaker implantation in 2.7%. Median postoperative length of stay (PLOS) was 8 days (interquartile range, 5-14 days). Overall hospital mortality was 3.0%. Weight < 3.5 kg and age ≤ 2.5 months were associated with higher mortality, longer PLOS, and increased frequency of major complications. Patients with Down syndrome had lower rates of mortality and morbidities than other patients; PLOS was similar.

Conclusions: In a contemporary multicenter cohort, most patients with CAVSD have repair early in the first year of life. Prior pulmonary artery band is rare. Hospital mortality is generally low, although patients at extremes of low weight and younger age have worse outcomes. Mortality and major complication rates are lower in patients with Down syndrome. (J Thorac Cardiovasc Surg 2014;148:2526-31)

Early Outcomes of Complete AVSD Repair
STS Congenital Heart Surgery Database Study

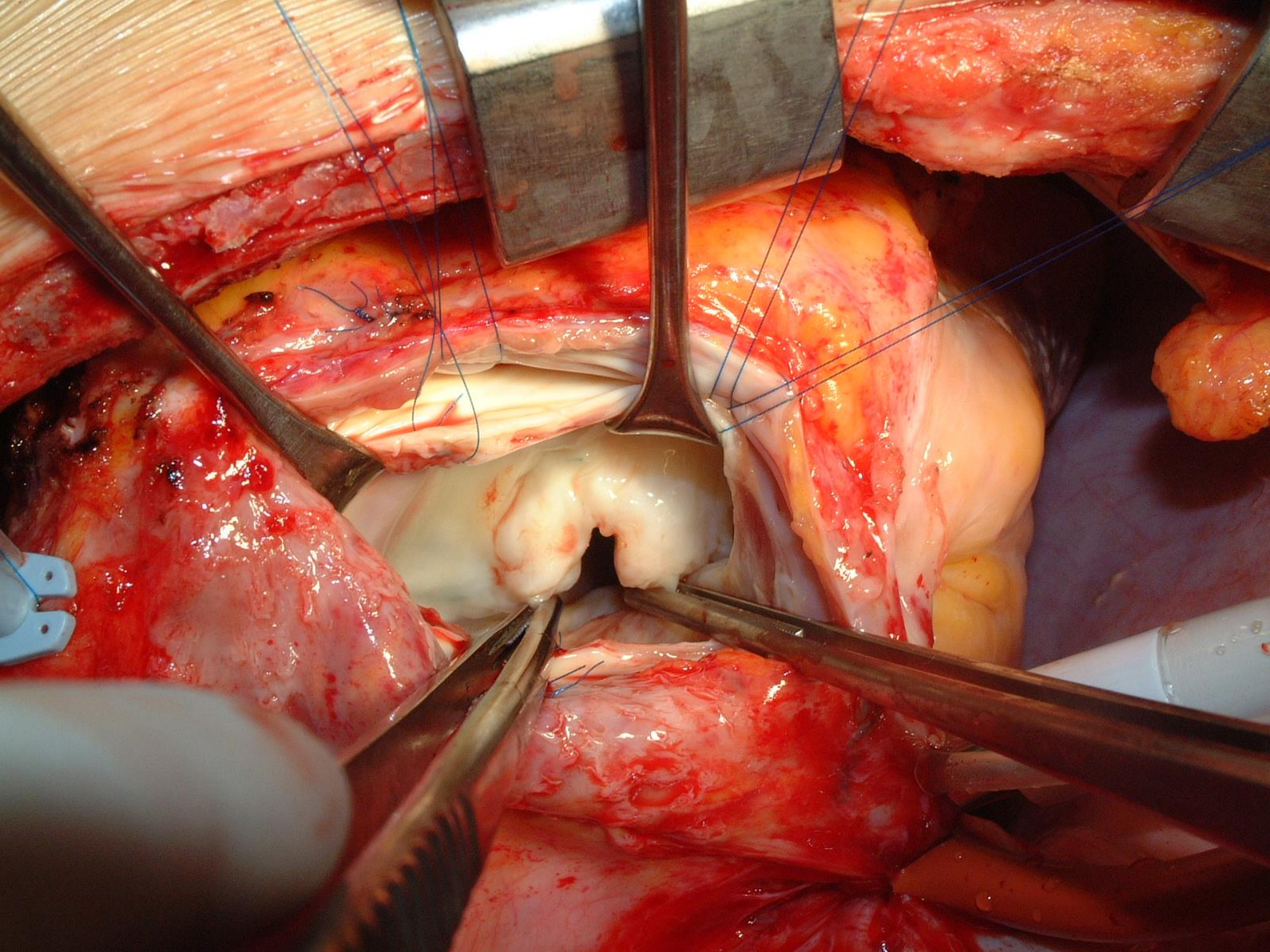
- Down syndrome: 78%
- Medina age at surgery: 4.6 months
- Prior pulmonary artery banding: 4.6%
- Major complications: 9.8% (pacemaker 2.7%)
- Hospital mortality: 3.0%
- Risk factors for mortality and complications:
  weight < 3.5 kg, age < 2.5 months
- Down syndrome:
  lower rate of mortality and morbidities

J Thorac Cardiovasc Surg. 2014;148:2026-31.

Late Reoperation

- The most common cause of late reoperation after repair of AVSD is left AV valve regurgitation.

- The reoperation rate for left AV valve regurgitation is approximately 10%.



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