Tetralogy of Fallot

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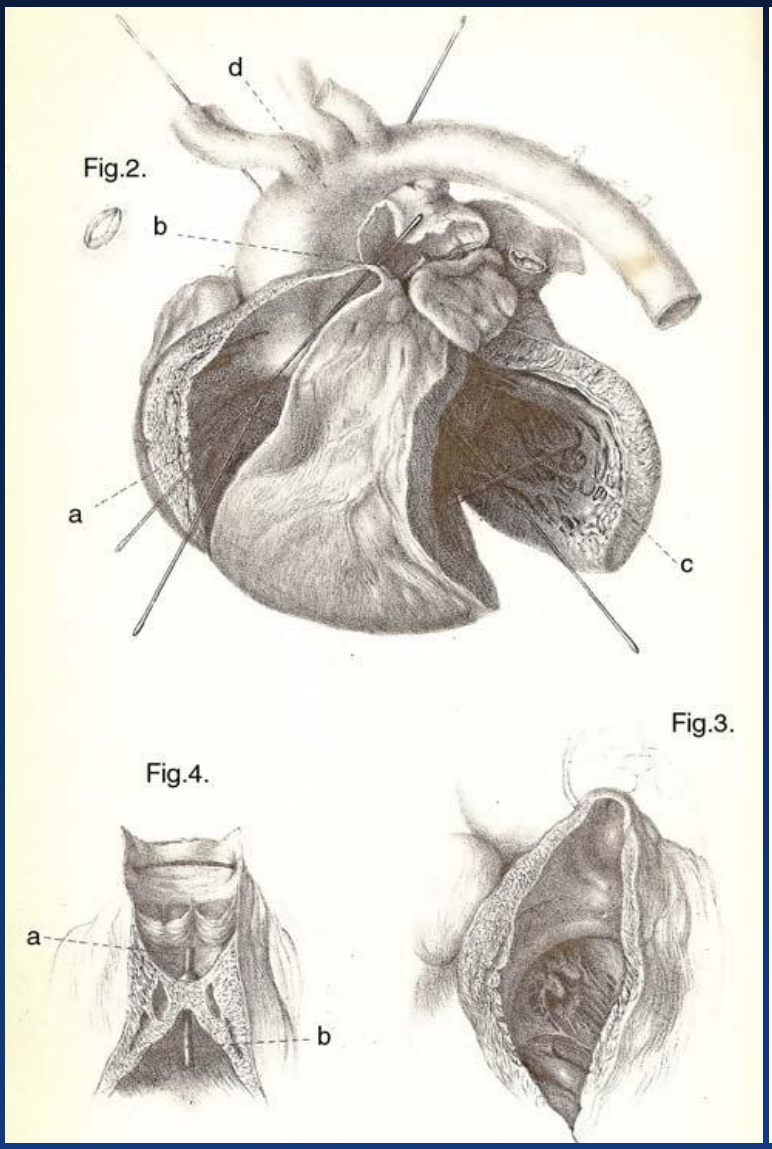
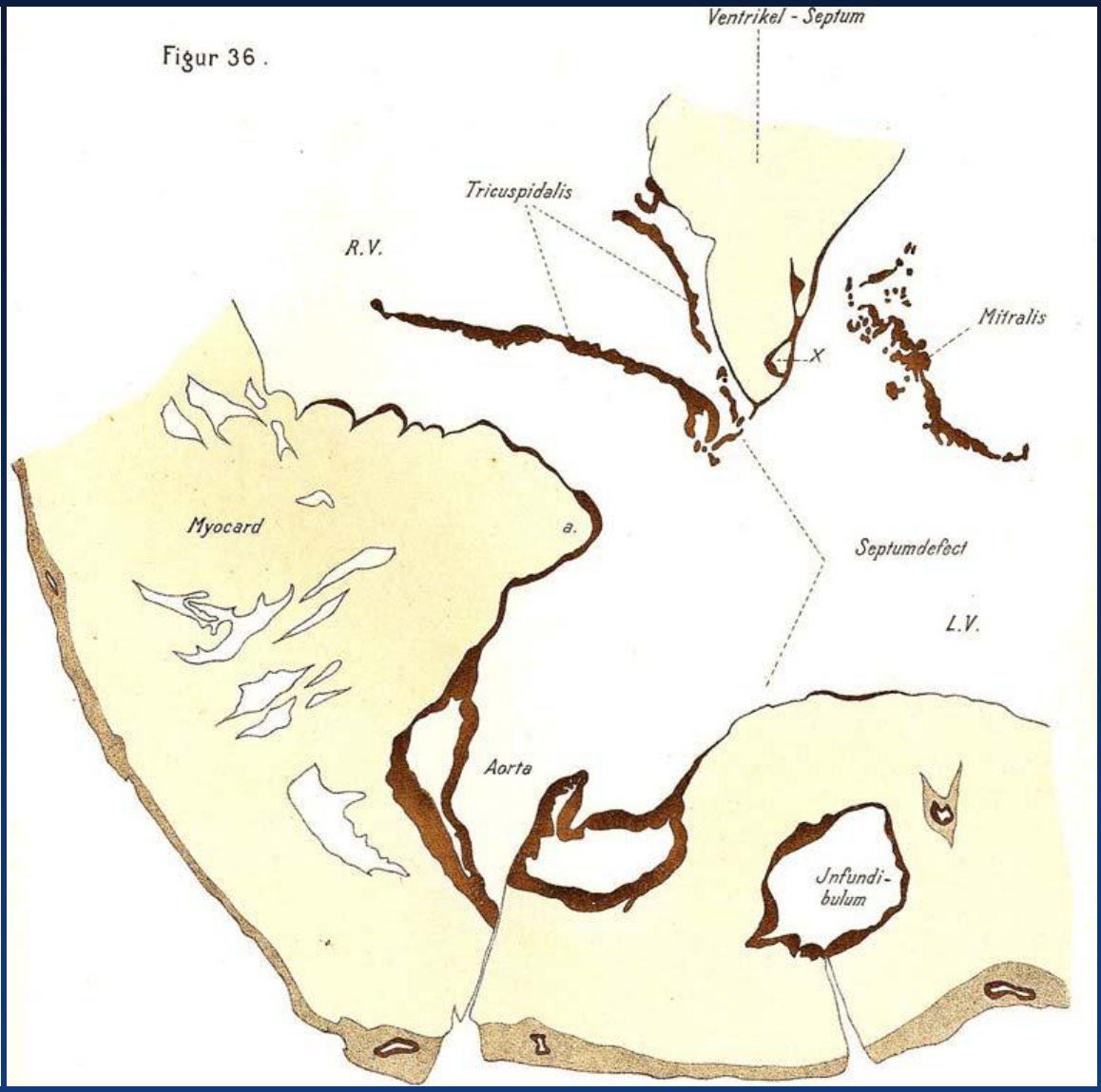
소아심장외과
박 천 수

Étienne-Louis Arthur Fallot

1. Pulmonary stenosis
2. Ventricular septal communication
3. Rightward deviation of the aorta's origin
4. Right ventricular hypertrophy

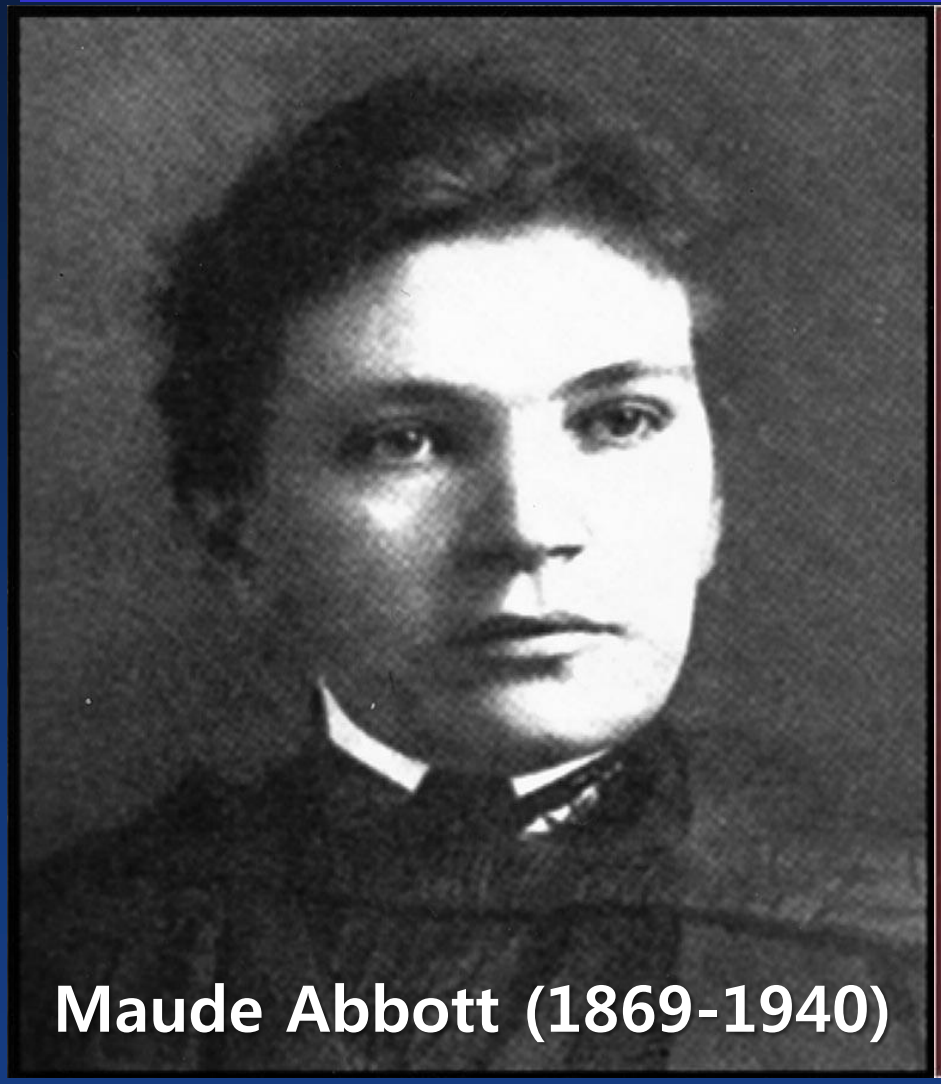
"Contribution a` l'anatomie pathologique de la maladie bleue"
Marseille Méd 1888



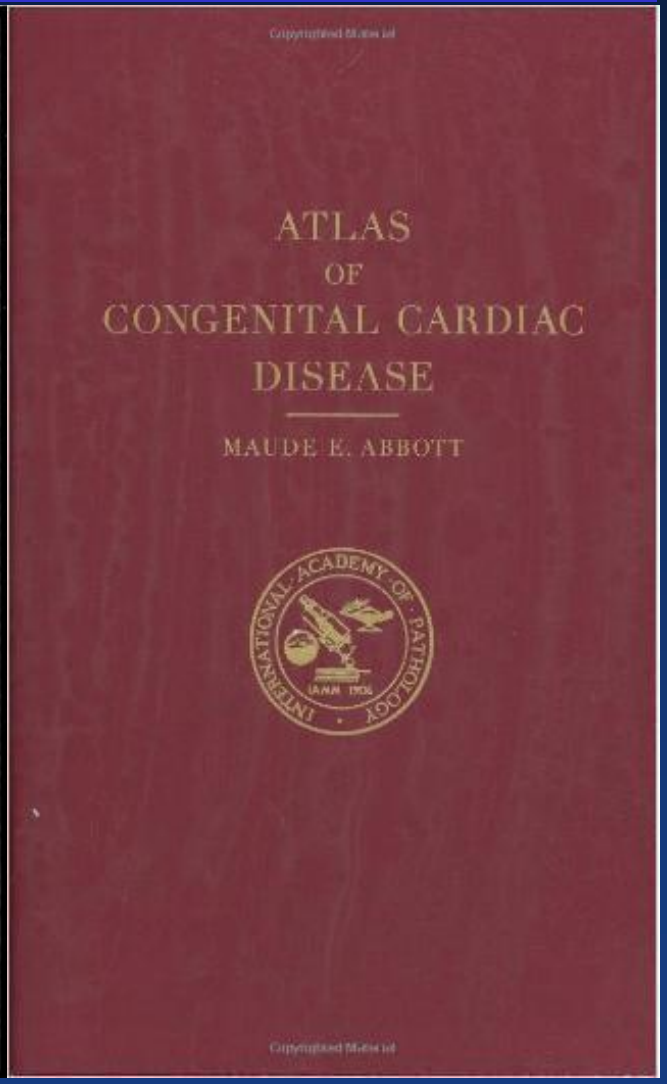


"Contribution a` l'anatomie pathologique de la maladie bleue"
Marseille Méd 1888

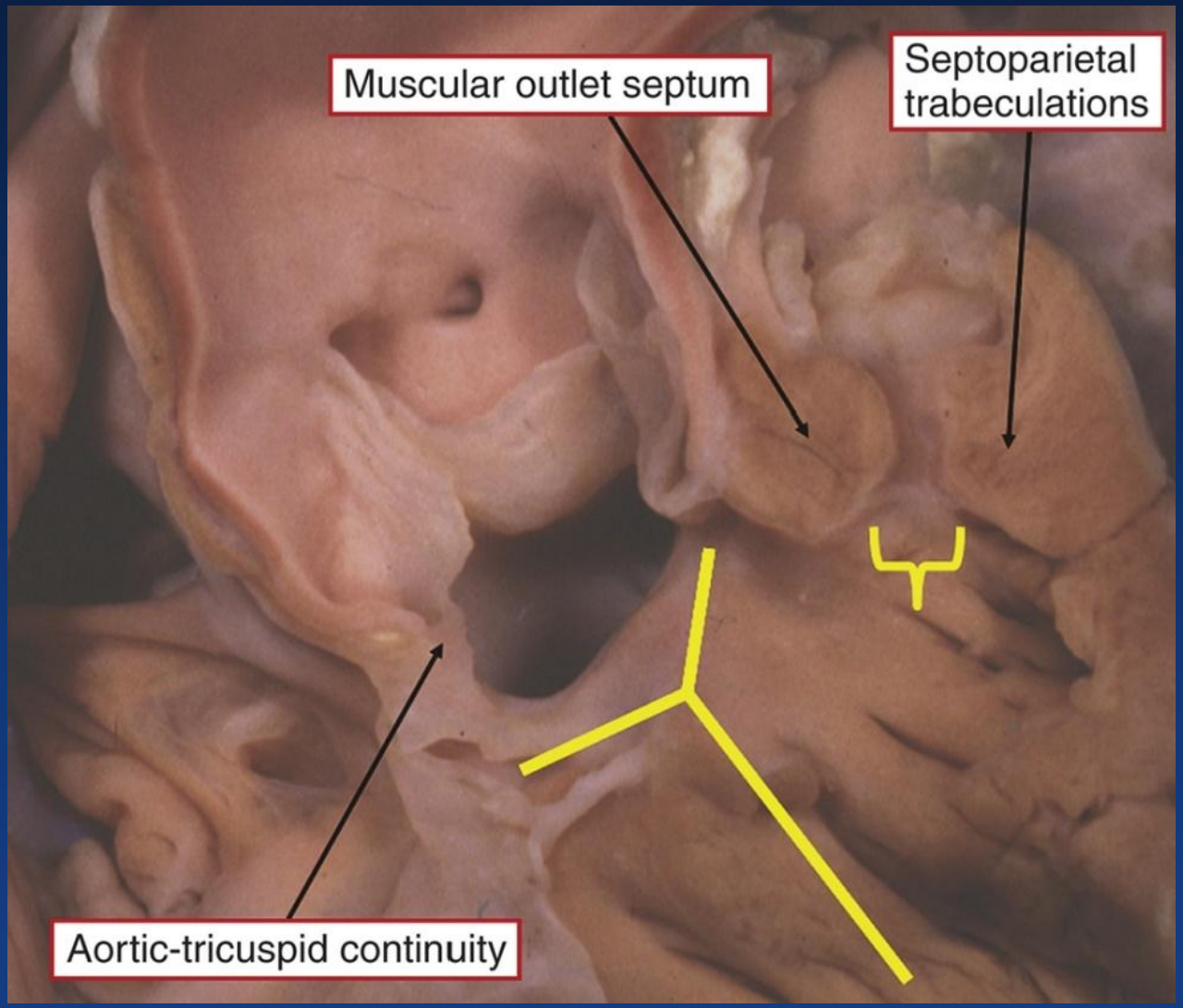
The Mother of Congenital Heart Disease



Maude Abbott (1869-1940)

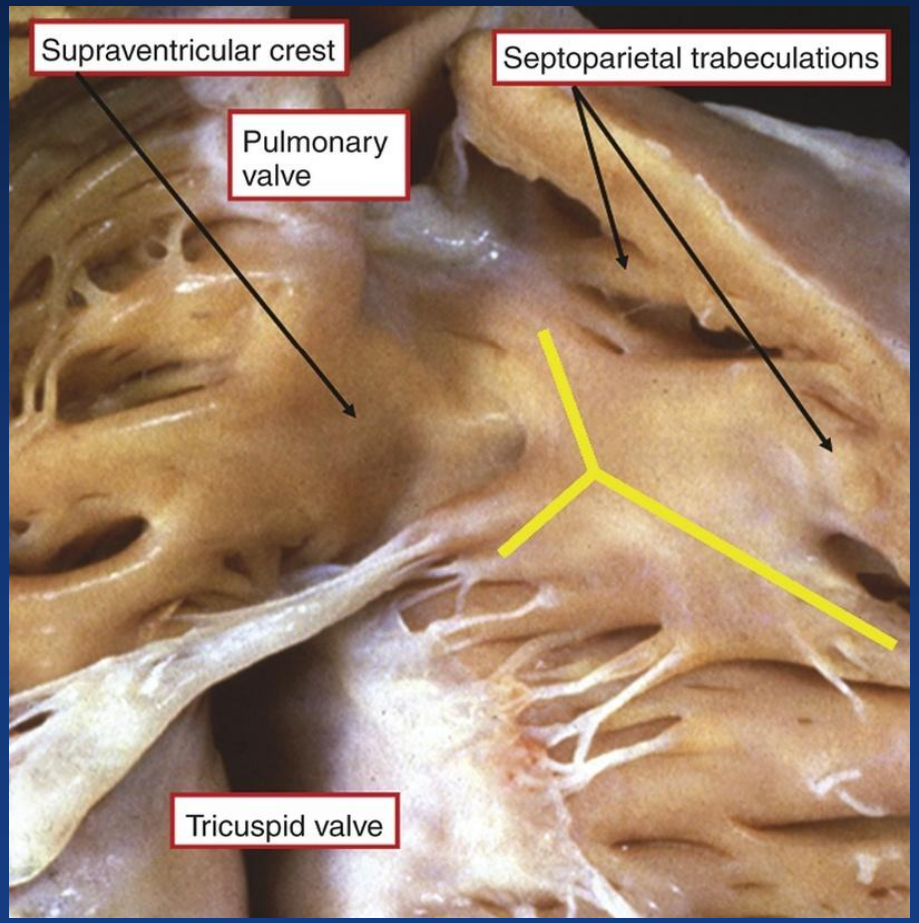


Key Anatomy


Muscular outlet septum
Septoparietal trabeculations
Aortic-tricuspid continuity

Key Anatomy

Normal



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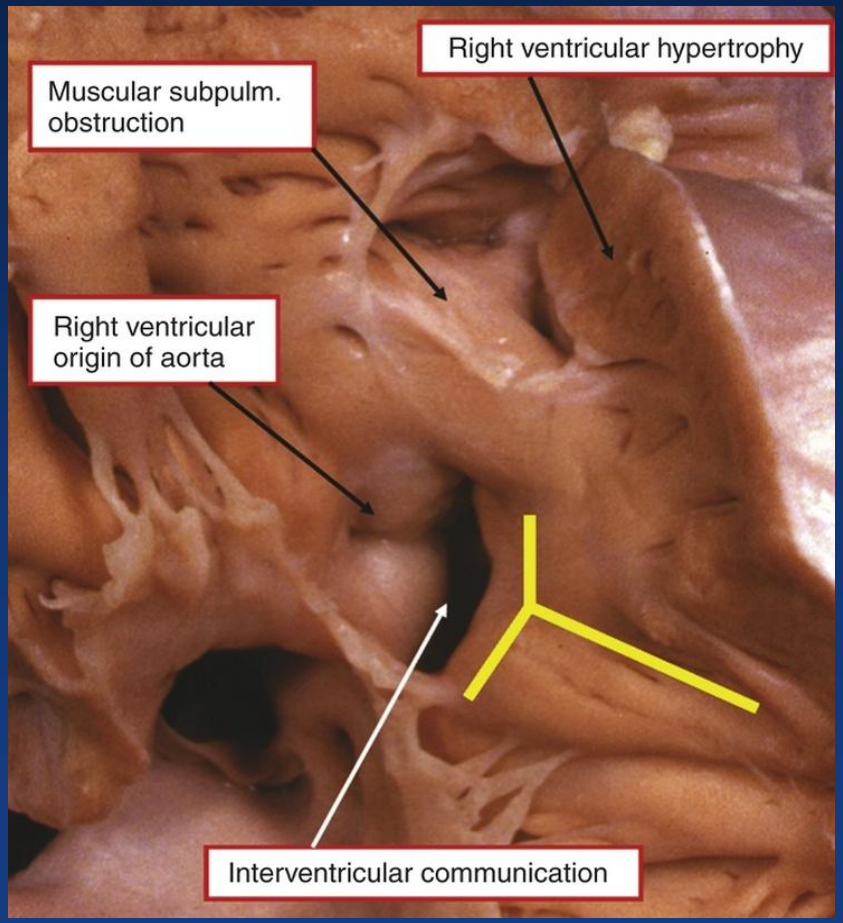
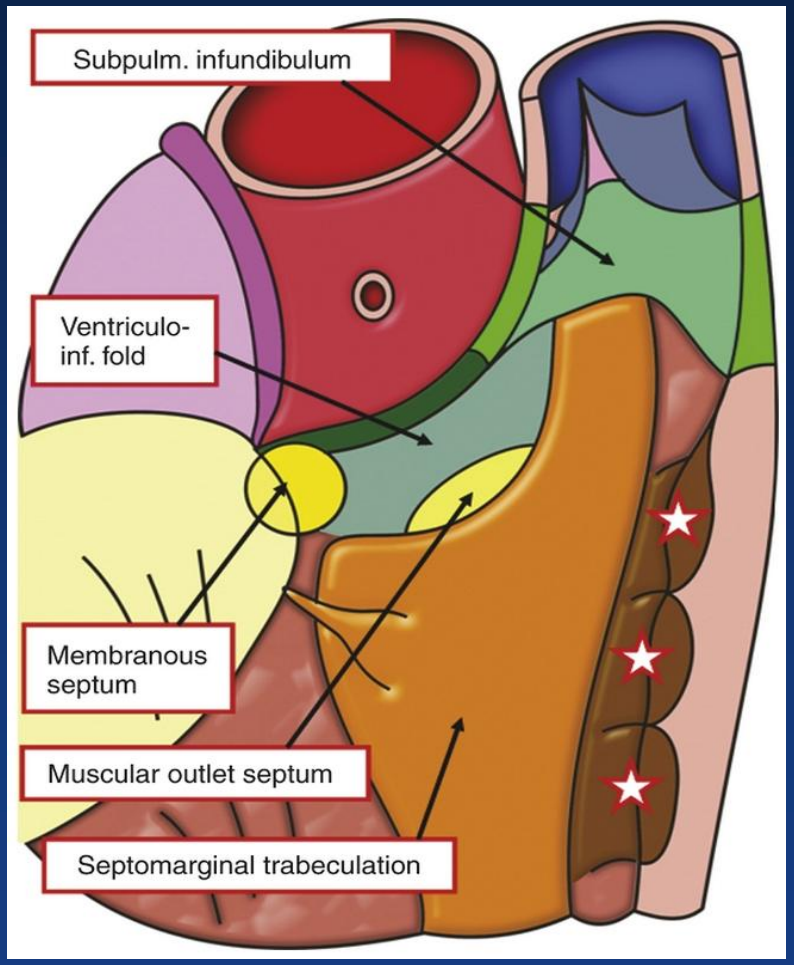


Key Anatomy

Normal



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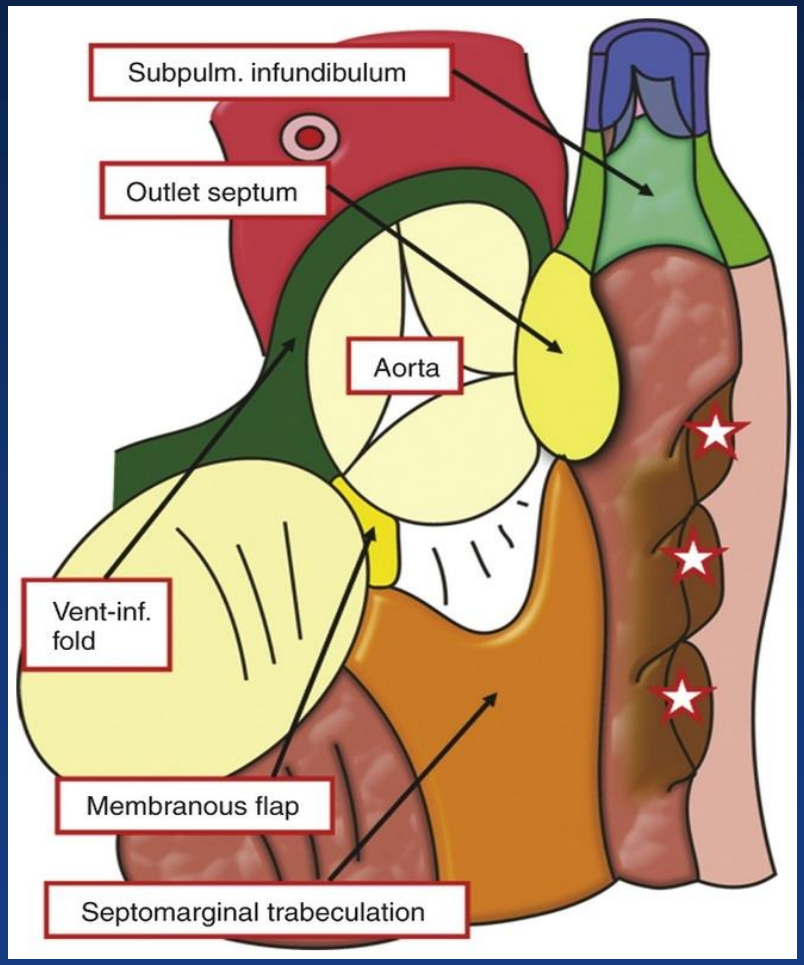


Clinical Presentation

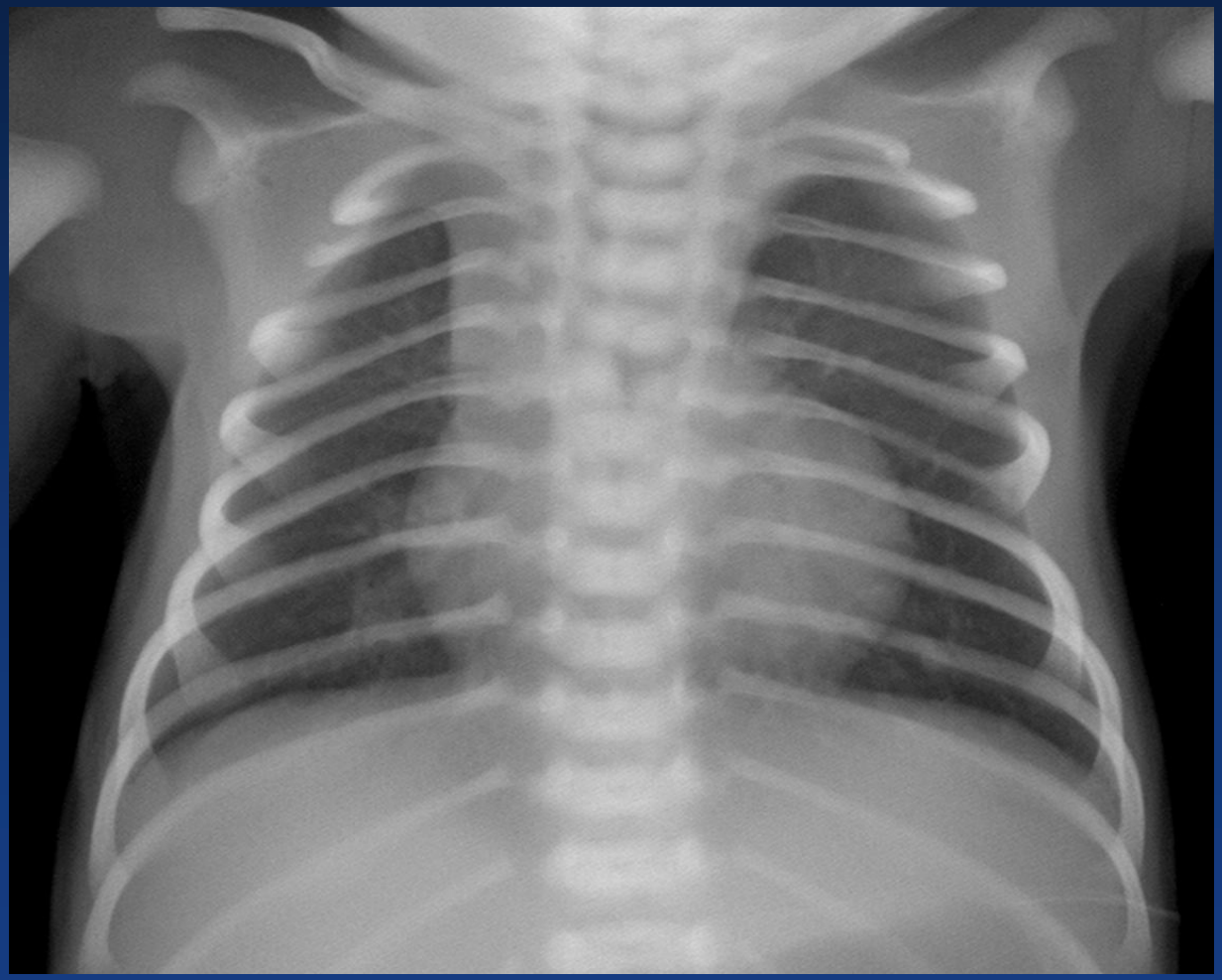
- Determining factors
  - The degree of RVOTO
  - Associated anomalies
    - PDA
    - MAPCA
- Cyanosis
  - Hypoxic spell: infundibular spasm
- Absent pulmonary valve
  - Respiratory symptom

Evaluation

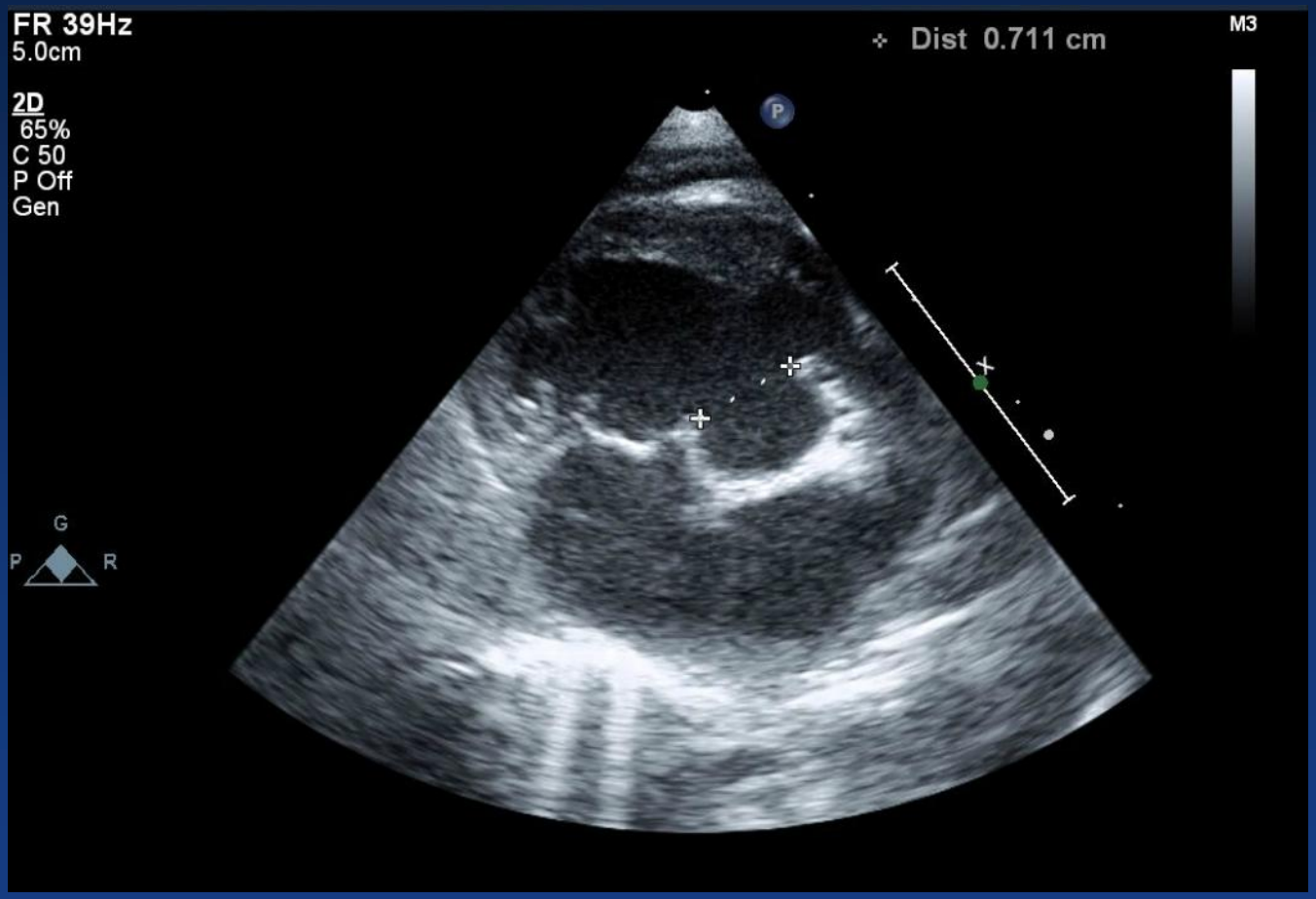
- CXR
- ECG: Not specific
- EchoCG: main W/U
- MRI: volume and flow quantification
- Cath and Angio: rarely performed
  - Mainly for intervention
- CT: increasingly used
  - Coronary anatomy, phasic RVOT morphology, PA morphology, ventricular volume

Evaluation

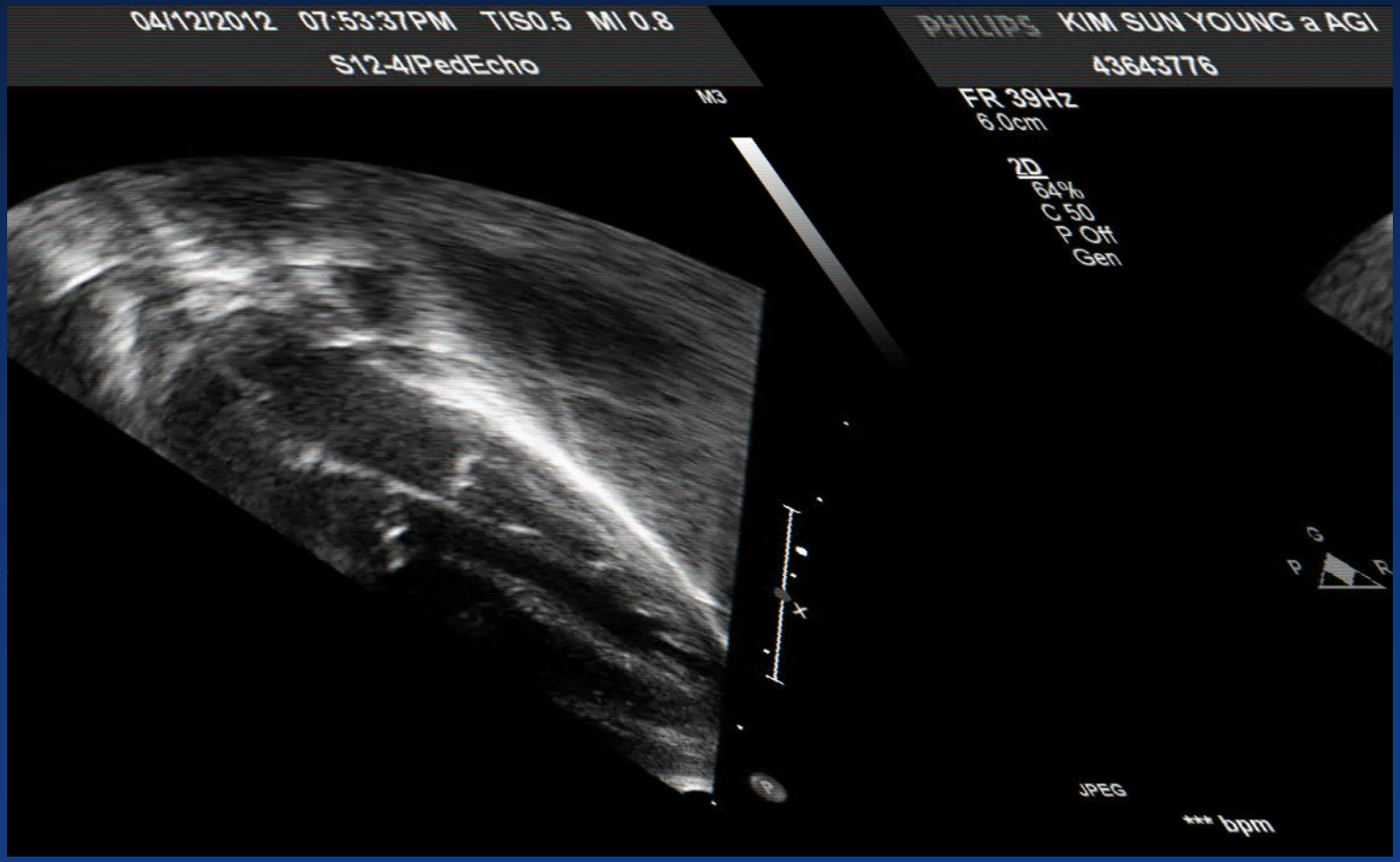
- CXR -



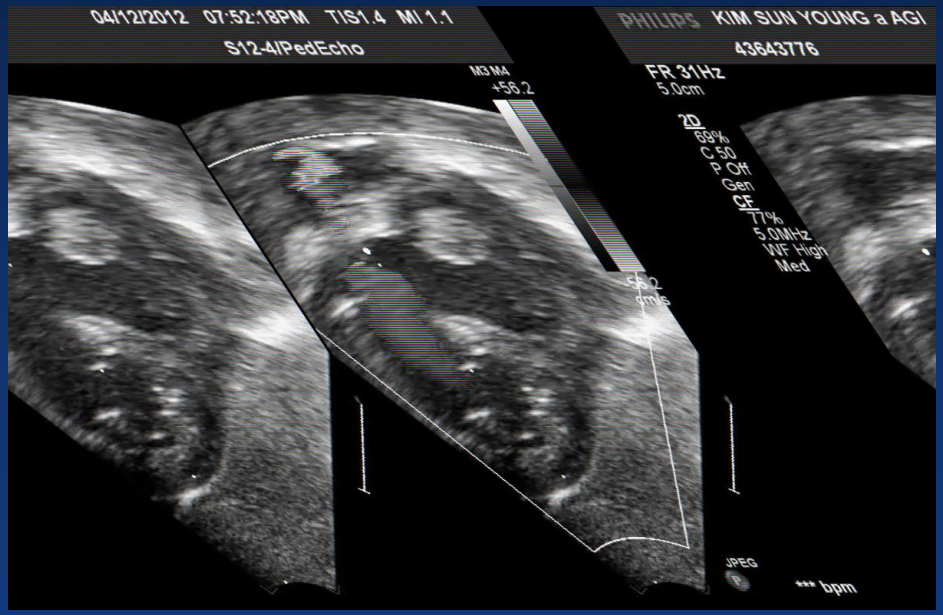
Evaluation
- EchoCG -

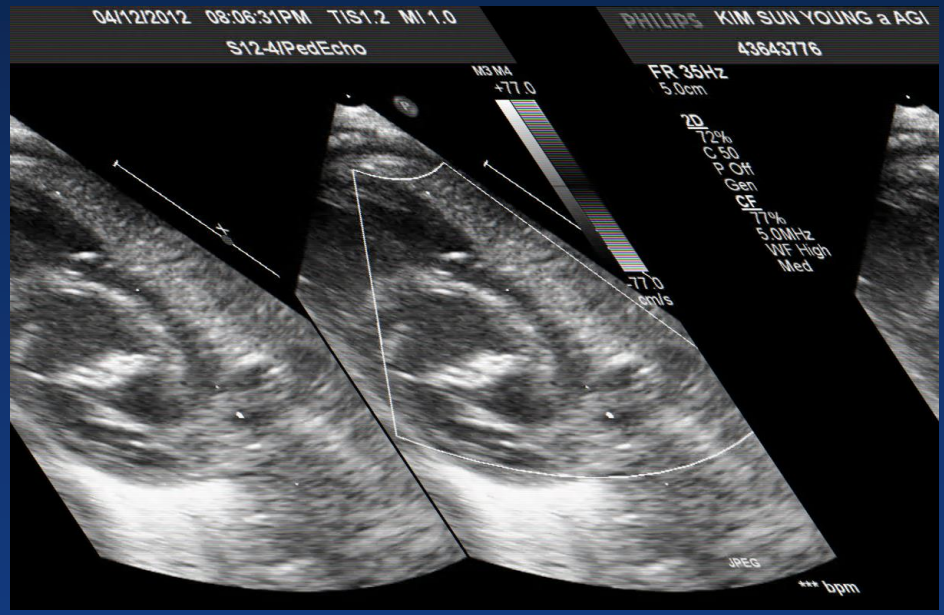


Evaluation
- EchoCG -



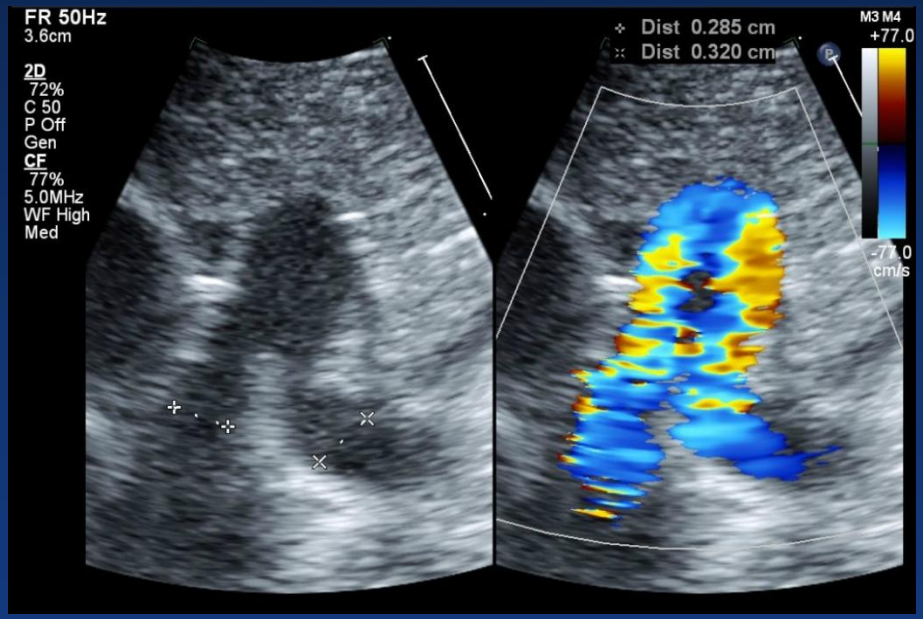
Evaluation
- EchoCG -

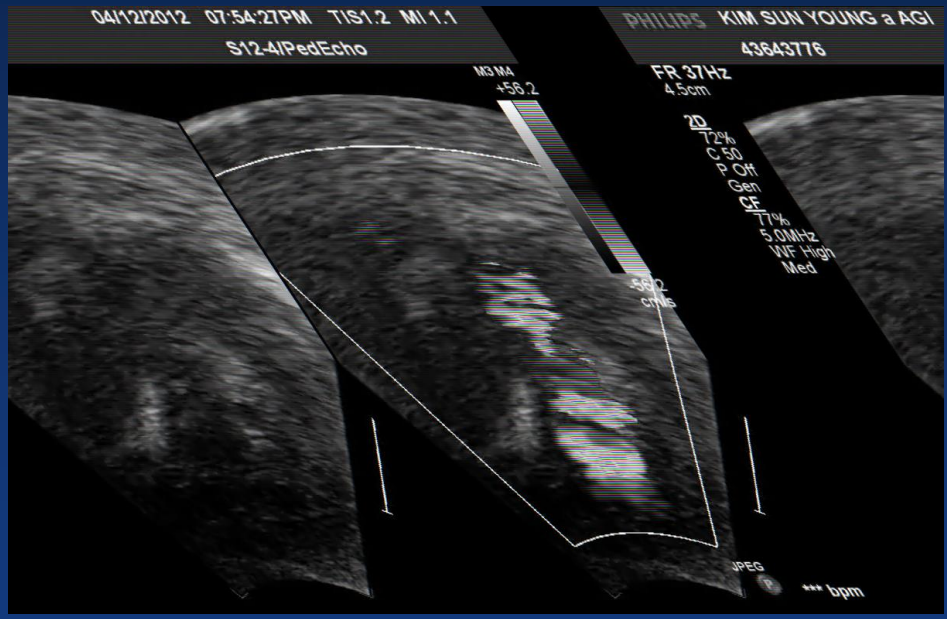




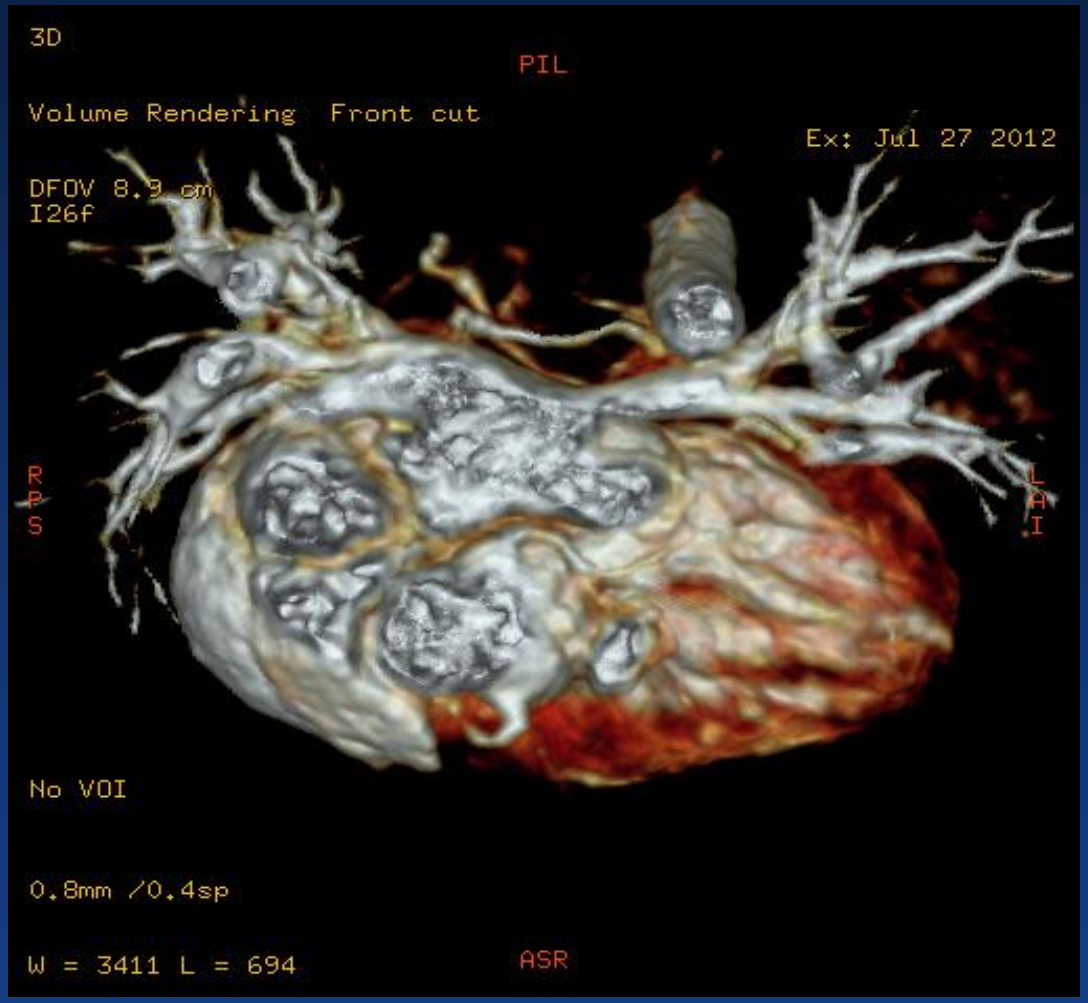
Evaluation

- EchoCG -

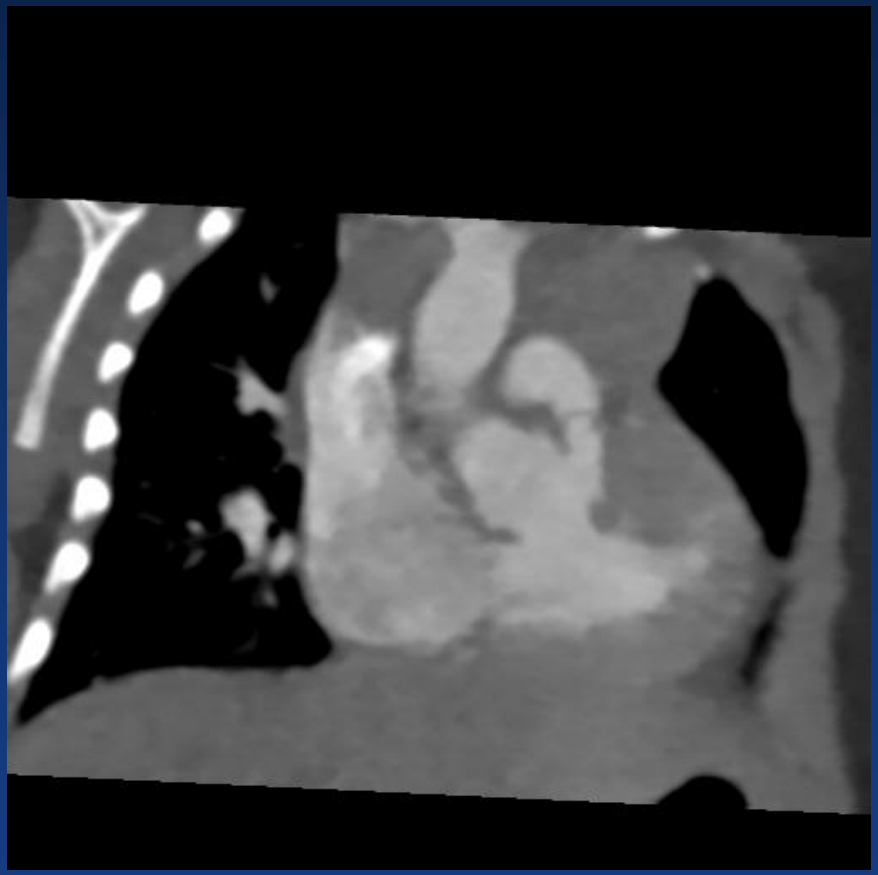


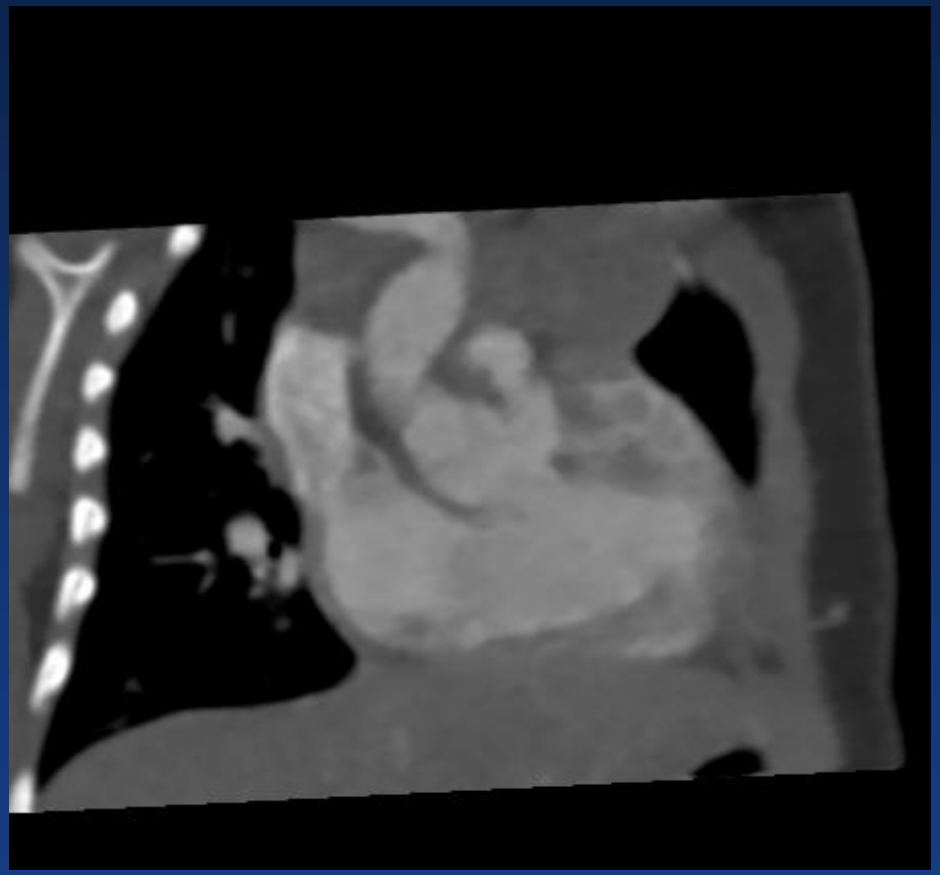


Evaluation
- CT -

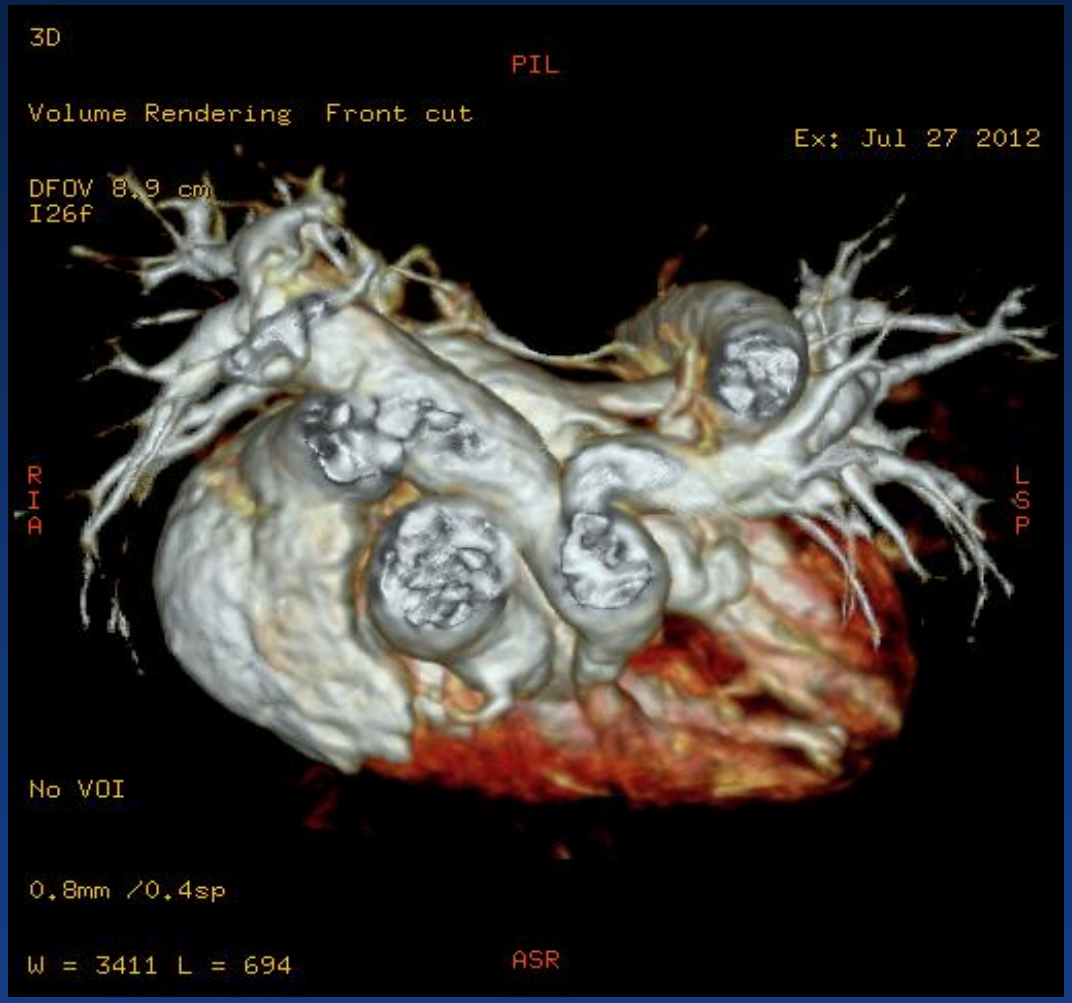


Evaluation
- CT -





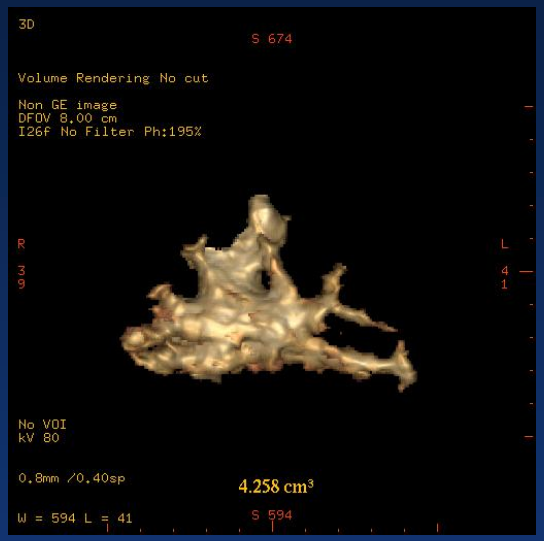
Evaluation
- CT -



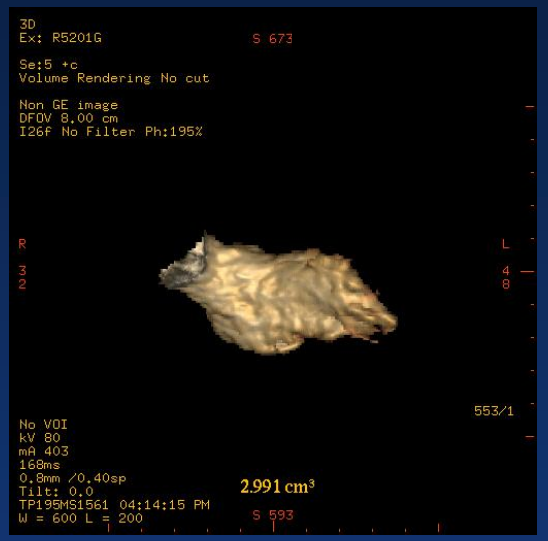
Evaluation

- CT -

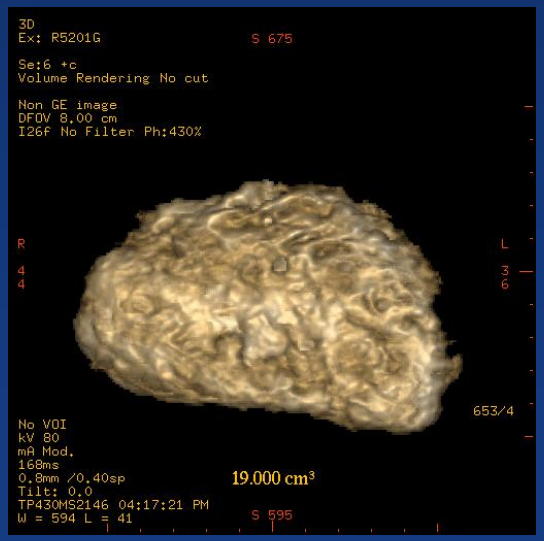
RV



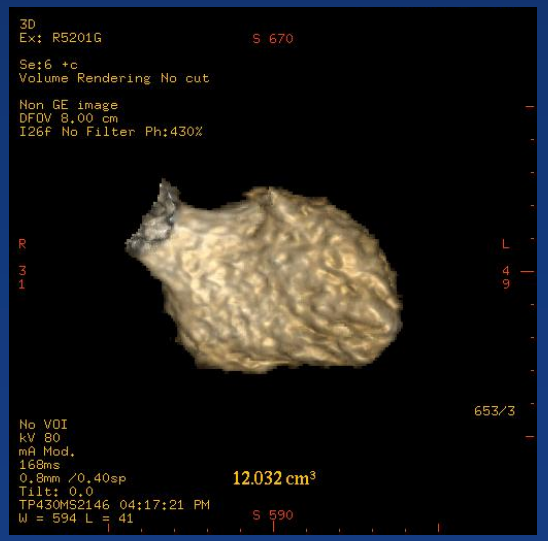
systole



LV



diastole



Palliative Surgery

- Systemic to pulmonary artery shunt
- Catheter intervention
  - Balloon pulmonary valvuloplasty
  - RVOT stent
- Surgical RVOT widening

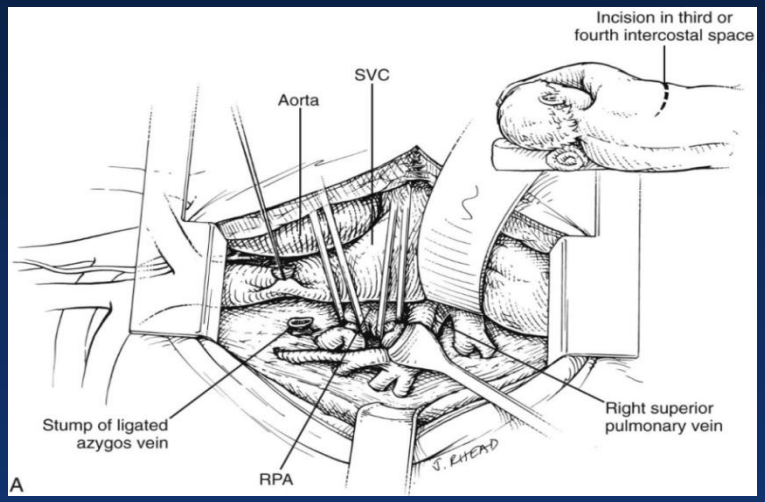
Palliative Surgery
- S-P Shunt -

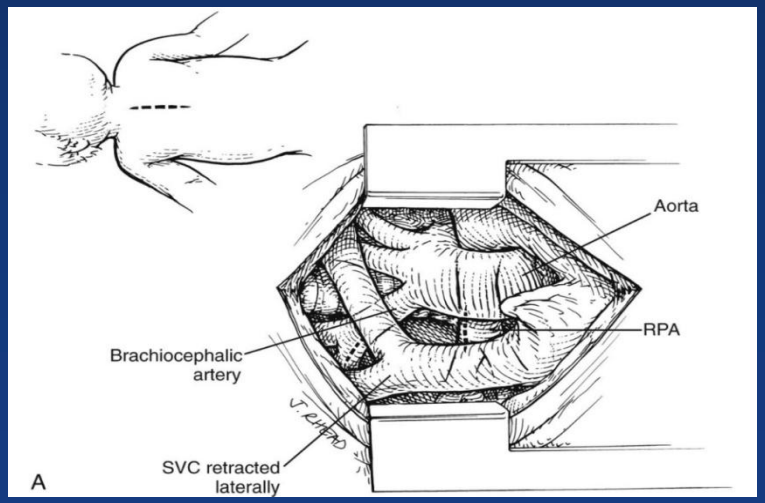
- Advantage
  - Avoidance of high risk OHS
  - Future growth of branch PA or annulus
- Disadvantage
  - Ventricular volume overloading
  - Persistent hypoxia
  - Shunt related complications or attrition

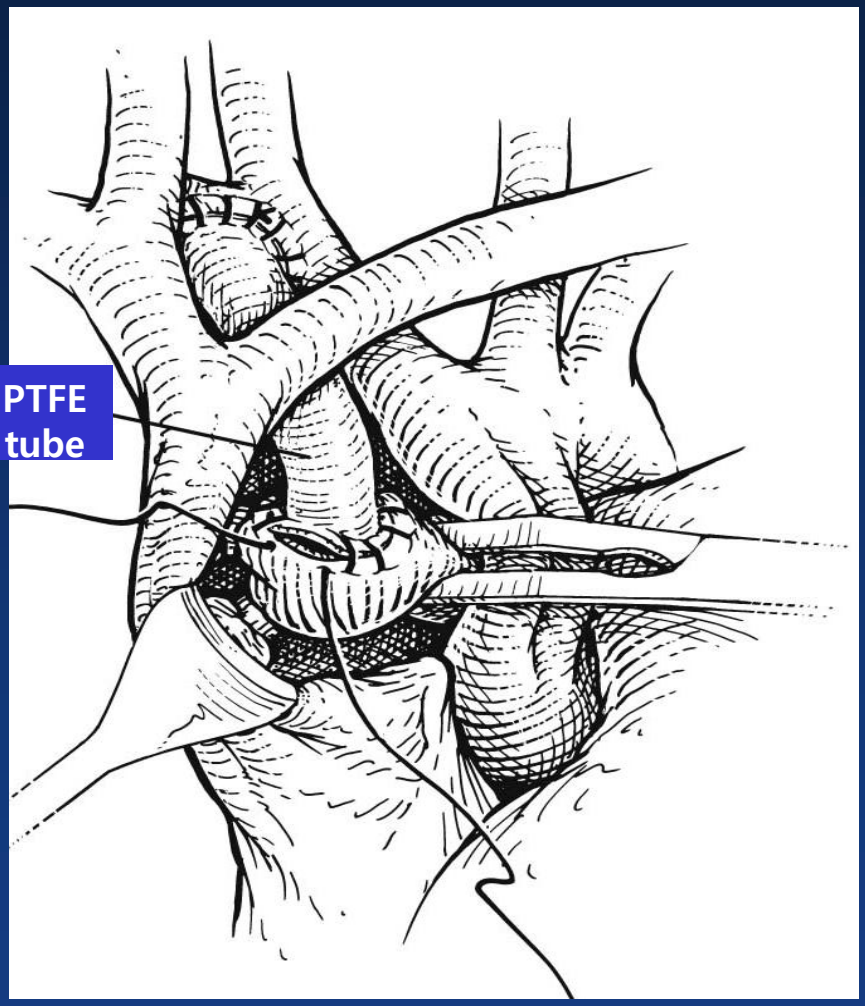
Palliative Surgery
- S-P Shunt -

- Arch
- Graft size
- Approach : central vs. thoracotomy
- Cardiopulmonary bypass
  - Additional procedures
  - Profound hypoxia

Palliative Surgery
- S-P Shunt -







Corrective Surgery
- Goal -

- Elimination of intracardiac shunt
- Alleviation of PS
- Minimizing PR

Corrective Surgery
- Optimal Timing -

- Symptomatic
  - Immediately
  - Neonate/ young infant

    → controversial !!
- Asymptomatic
  - Center specific
  - 3 ~ 6 months, upto 1 year

Cardiac surgery in infants with low birth weight is associated with increased mortality: Analysis of the Society of Thoracic Surgeons Congenital Heart Database

Christopher L. Curzon, DO,[a] Sarah Milford-Beland, MS,[b] Jennifer S. Li, MD, MHS,[a,b] Sean M. O'Brien, PhD,[b] Jeffrey Phillip Jacobs, MD,[c] Marshall Lewis Jacobs, MD,[d] Karl F. Welke, MD,[e] Andrew J. Lodge, MD,[f] Eric D. Peterson, MD, MPH,[b] and James Jaggers, MD[f]

J Thorac Cardiovasc Surg 2008;135:546-51

TABLE 3. Mortality data, diagnostic–procedure groups

	Mortality rate, overall (n = 3022)	Mortality rate, 1–2.5 kg (n = 517)	Mortality rate 2.5–4 kg (n = 2505)	Risk ratio (95% CI)	*P* value*
Tetralogy of Fallot					
Primary repair	6.1 (99)	5.3 (19)	6.3 (80)	0.84 (0.10–6.79)	.61
Shunt palliation	8.3 (97)	7.4 (27)	8.6 (70)	0.86 (0.19–4.02)	.58

Contemporary Patterns of Management of Tetralogy of Fallot: Data From The Society of Thoracic Surgeons Database

Hamad F. Al Habib, MBBS, Jeffrey Phillip Jacobs, MD, Constantine Mavroudis, MD, Christo I. Tchervenkov, MD, Sean M. O'Brien, PhD, Siamak Mohammadi, MD, and Marshall L. Jacobs, MD

Ann Thorac Surg 2010;90:813-820

Table 3. The Relationship Between Type of Procedure and Age at Operation

	0–30 Days No.	>30 Days–3 Months No.	>3–6 Months No.	>6 Months–1 Year No.	>1–2 Years No.	>2–18 Years No.	All Ages No.
TOF palliation							
Without previous cardiac operation	178	89	16	6	1	4	294
After previous cardiac operation	4	6	1	2	1	0	14
TOF repair							
Without previous cardiac operation	154	492	975	614	145	154	2534
After previous cardiac operation	8	10	47	111	37	4	217
All operations	344	597	1039	733	184	162	3059

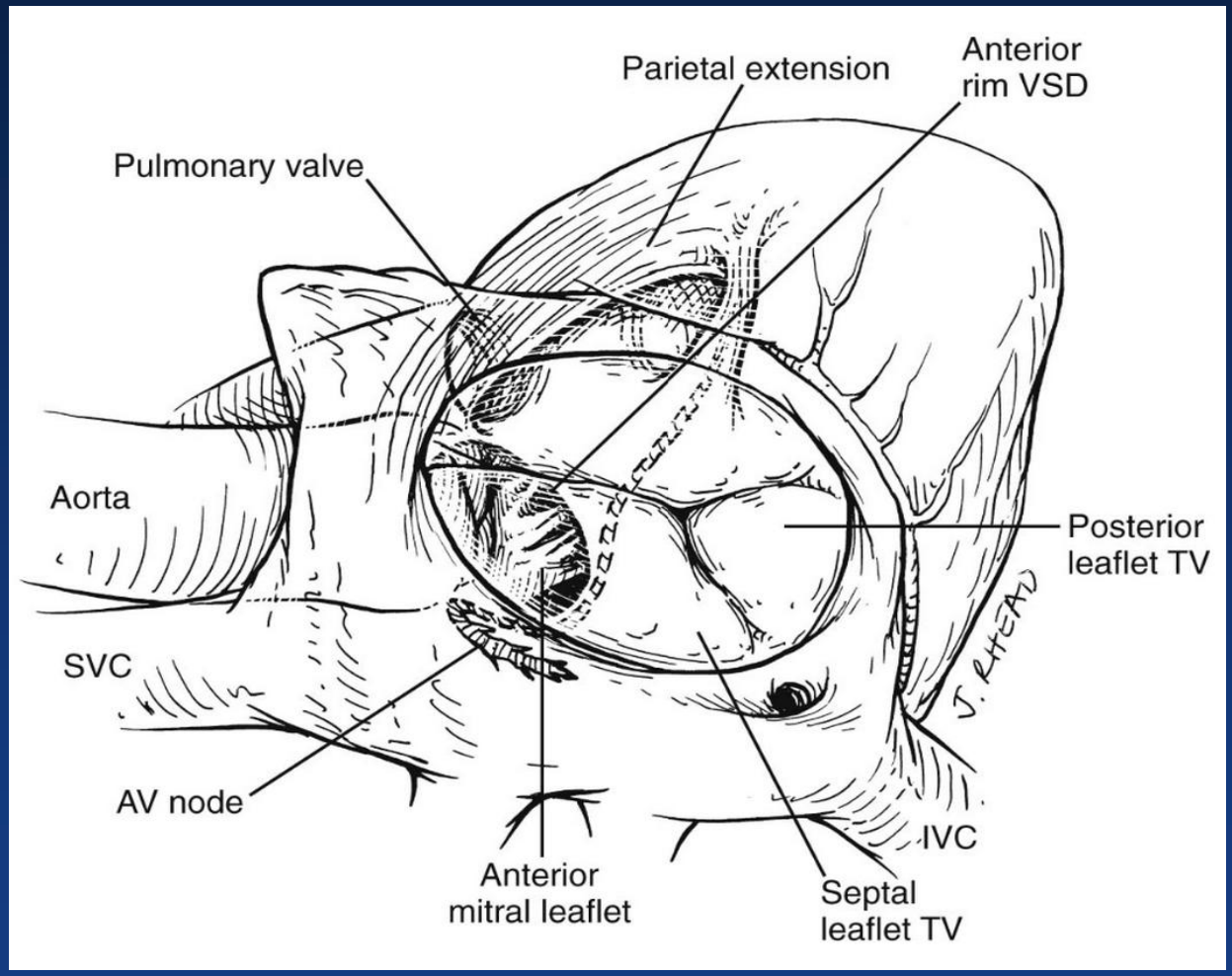
Corrective Surgery

- Trans-ventricular vs. -atrial VSD closure
- Transannular vs. Annulus preservation
  - PV annulus size (z-score)
  - PV morphology
  - Postbypass pRV/LV
- RVOT patching
- Branch PA widening

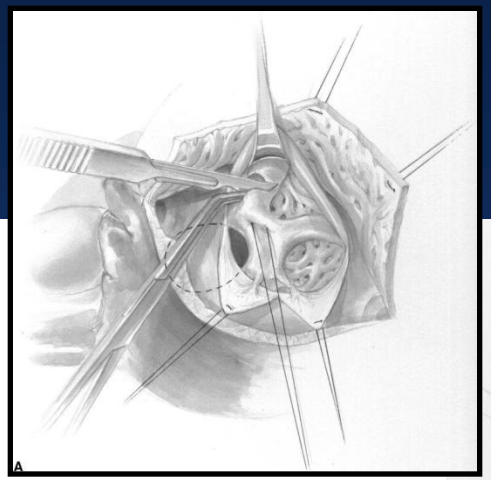
Corrective Surgery
- VSD Closure -

- Conduction axis
  - Same as simple VSD
- Malaligned outlet septum
  - Mobilization: division of parietal band
  - Anterosuperior margin
- Approach
  - Transatrial
  - Transventricular

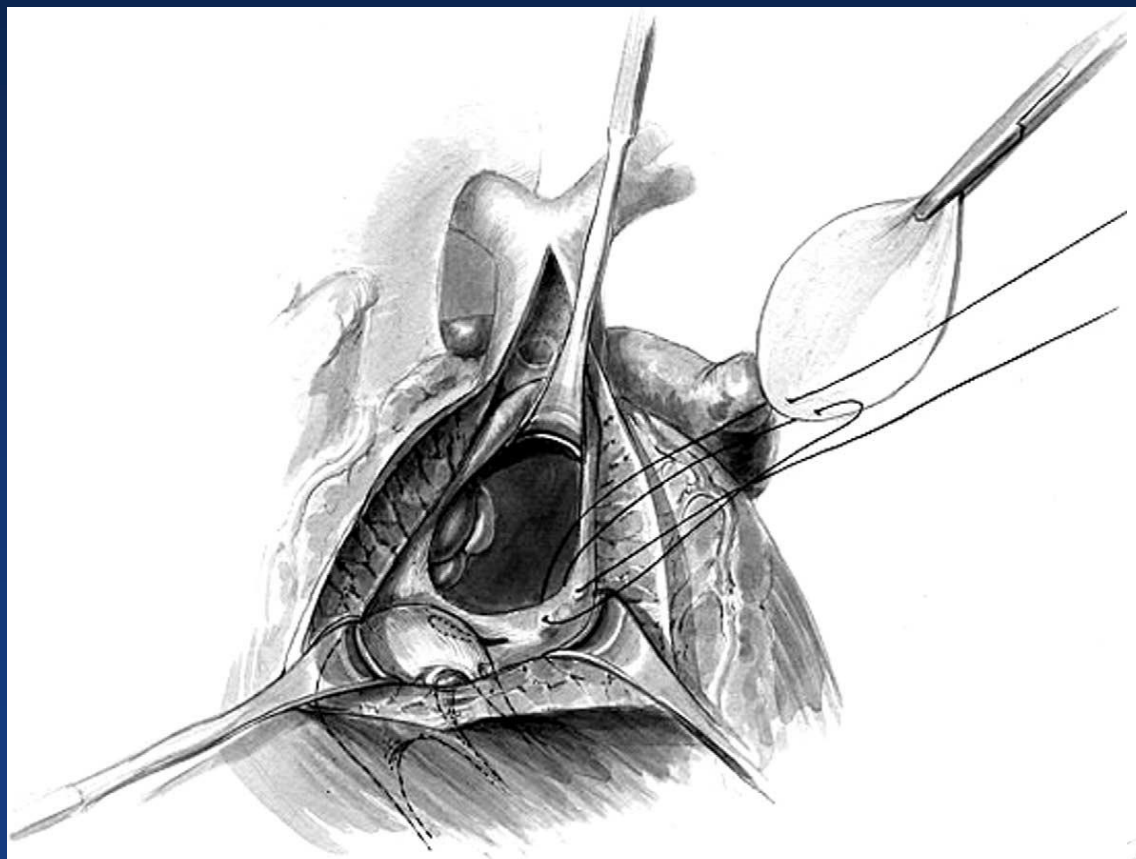
Corrective Surgery
- VSD Closure -

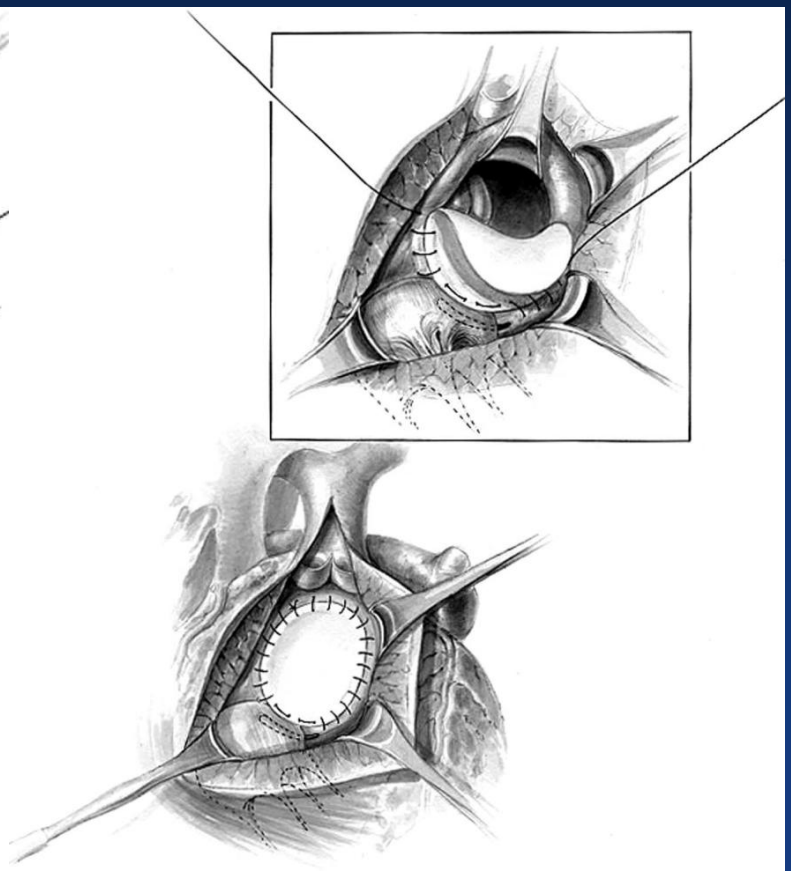


Corrective Surgery
- Transatrial VSD Closure -



Corrective Surgery
- Transventricular VSD Closure -

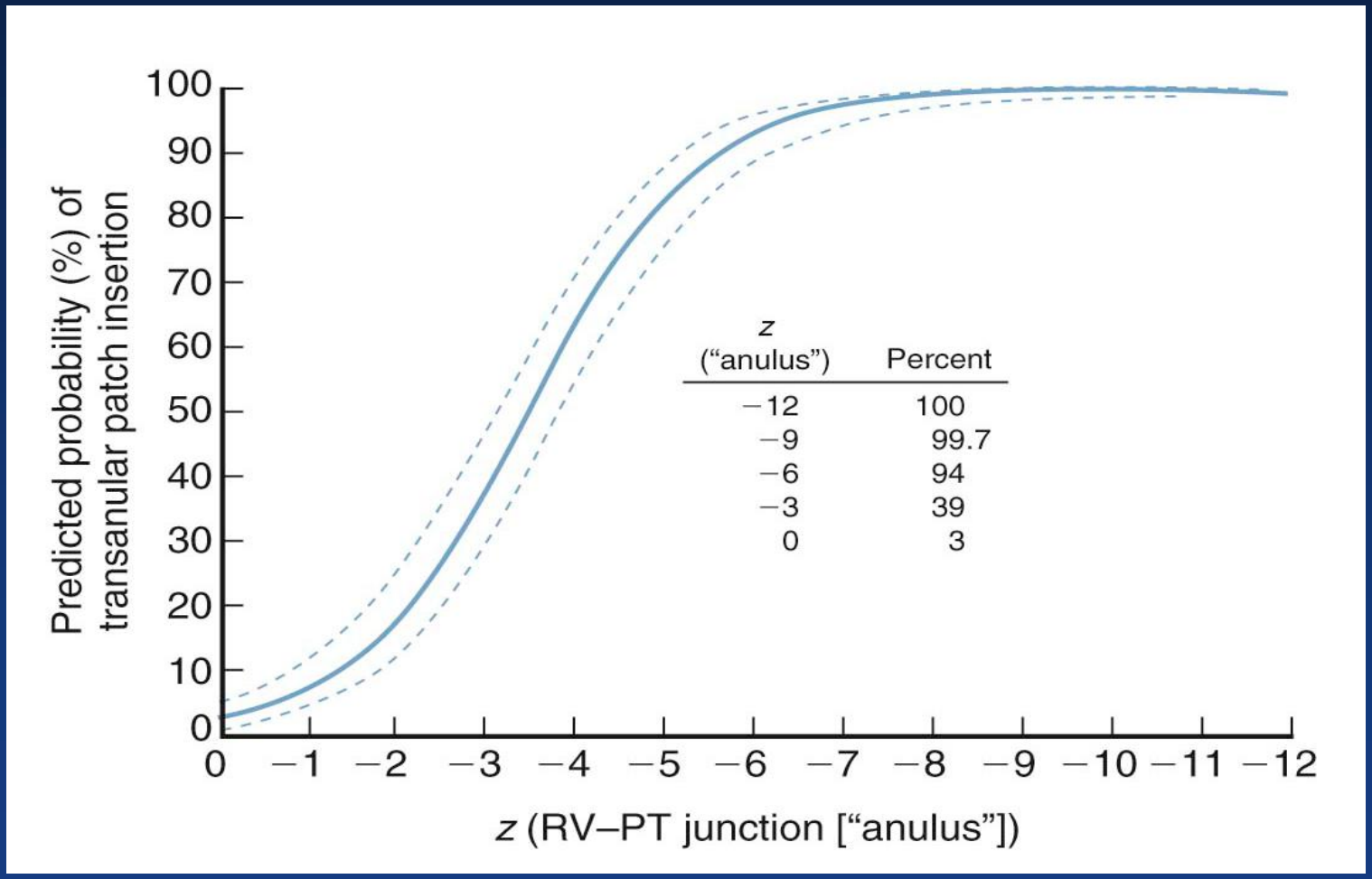




Corrective Surgery
- RVOT Relief -

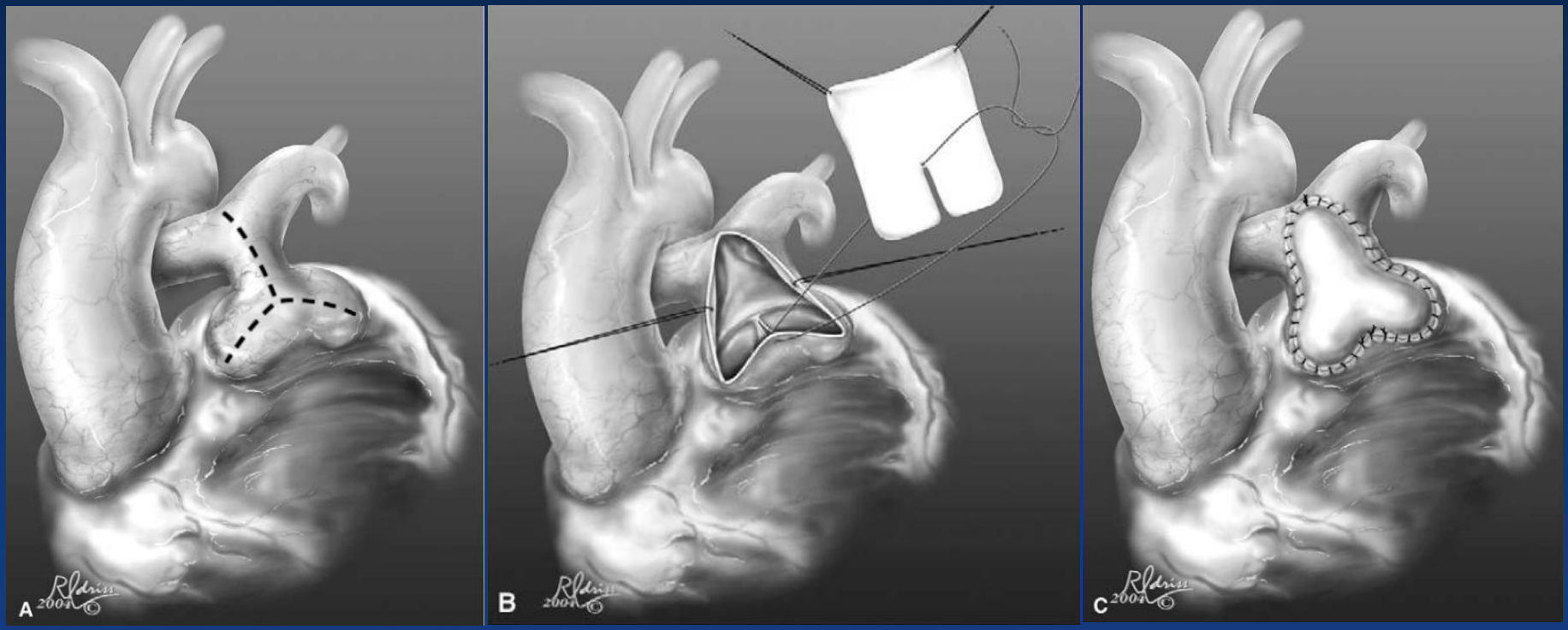
- PR ←→ PS
- Optimal PG through RVOT
  - pRV/LV <0.7
  - In neonate: RVP <50mmHg
- RVOT patching vs. m. resection
- Monocusp in transannular patch
- Major coronary artery crossing RVOT

Corrective Surgery
- RVOT Relief -

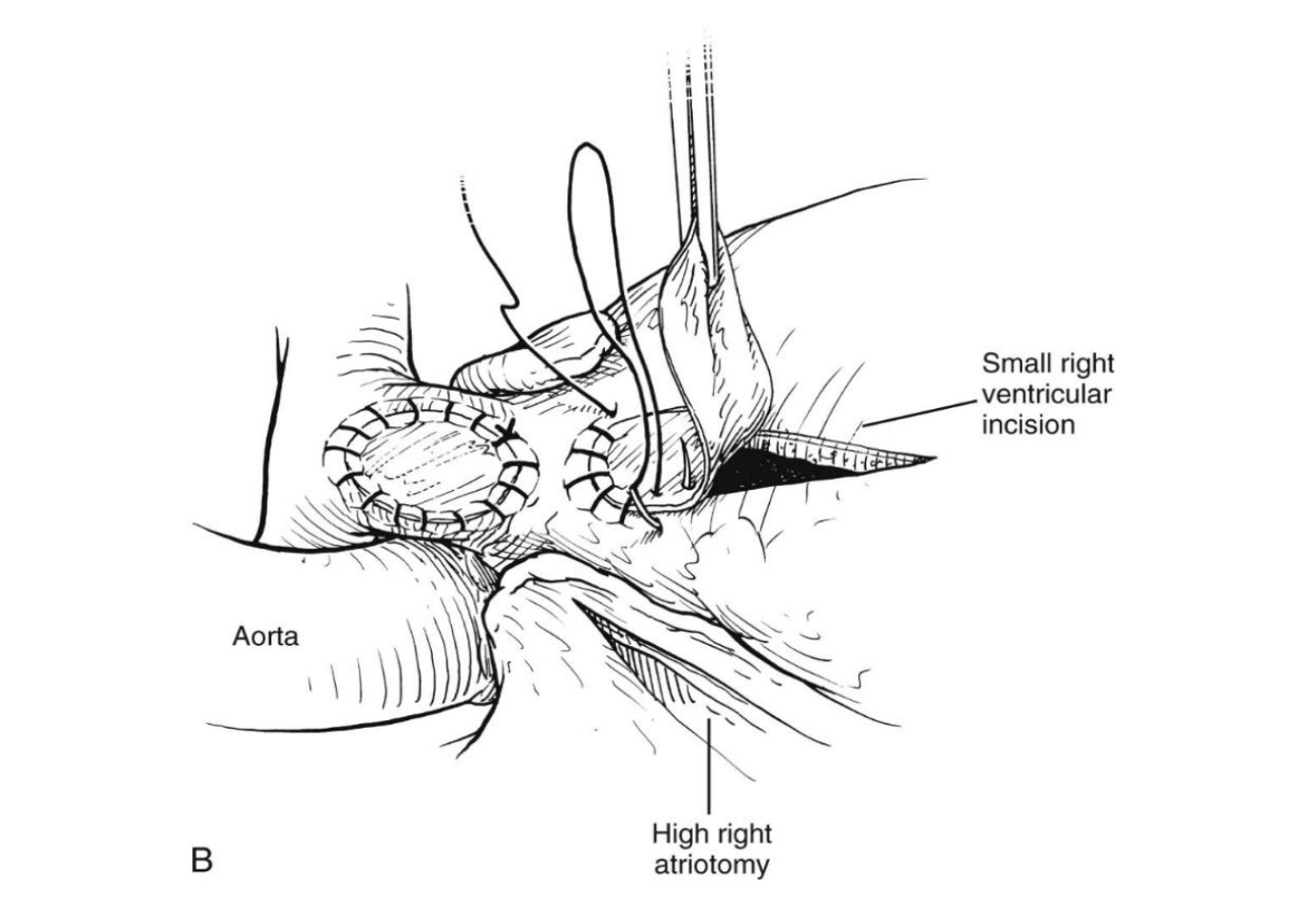


z (“anulus”)	Percent
−12	100
−9	99.7
−6	94
−3	39
0	3

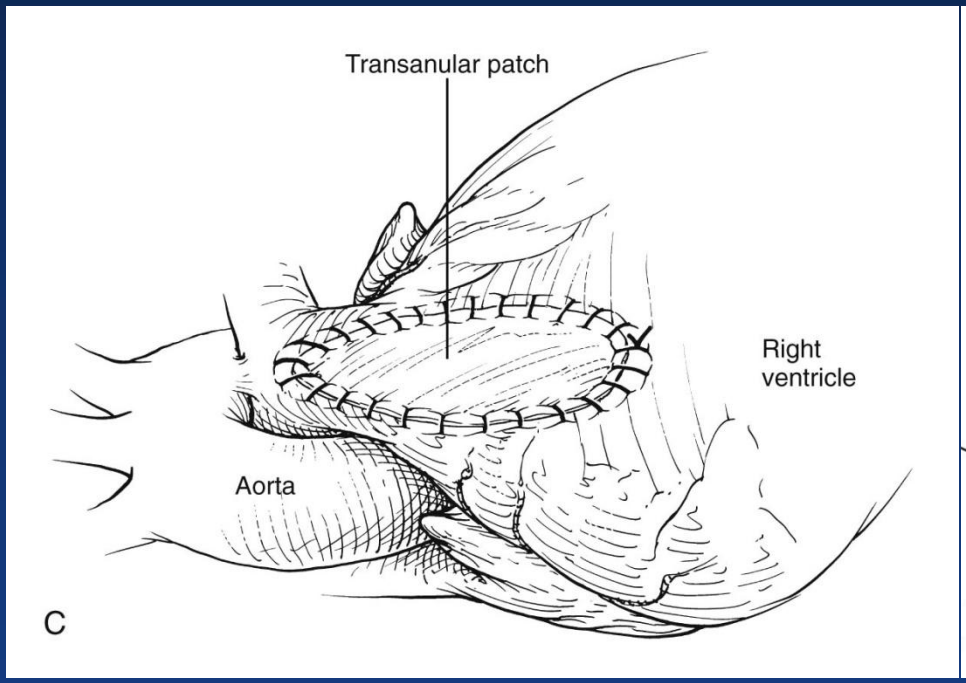
Corrective Surgery
- Annulus Preservation -

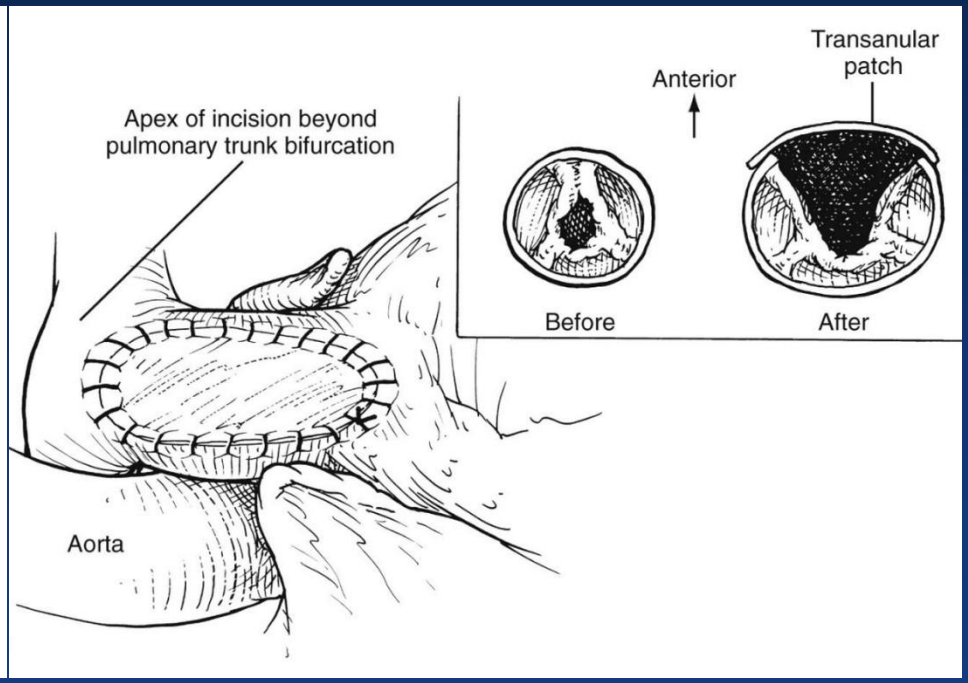


Corrective Surgery
- Annulus Preservation -



Corrective Surgery
- Transannular -





Corrective Surgery
- Transannular -

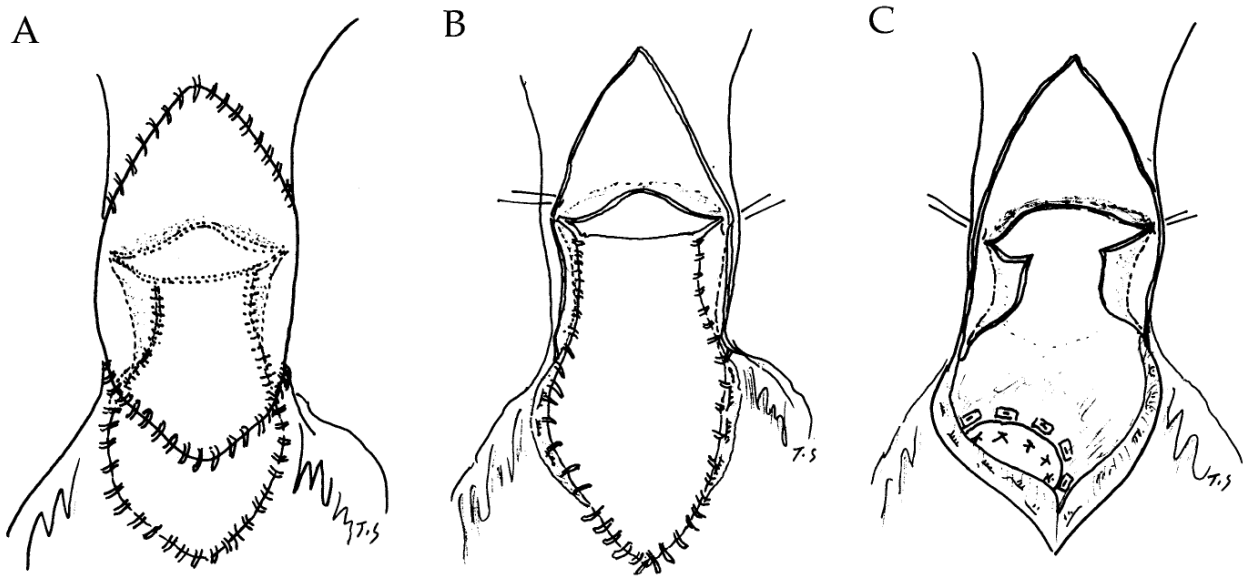
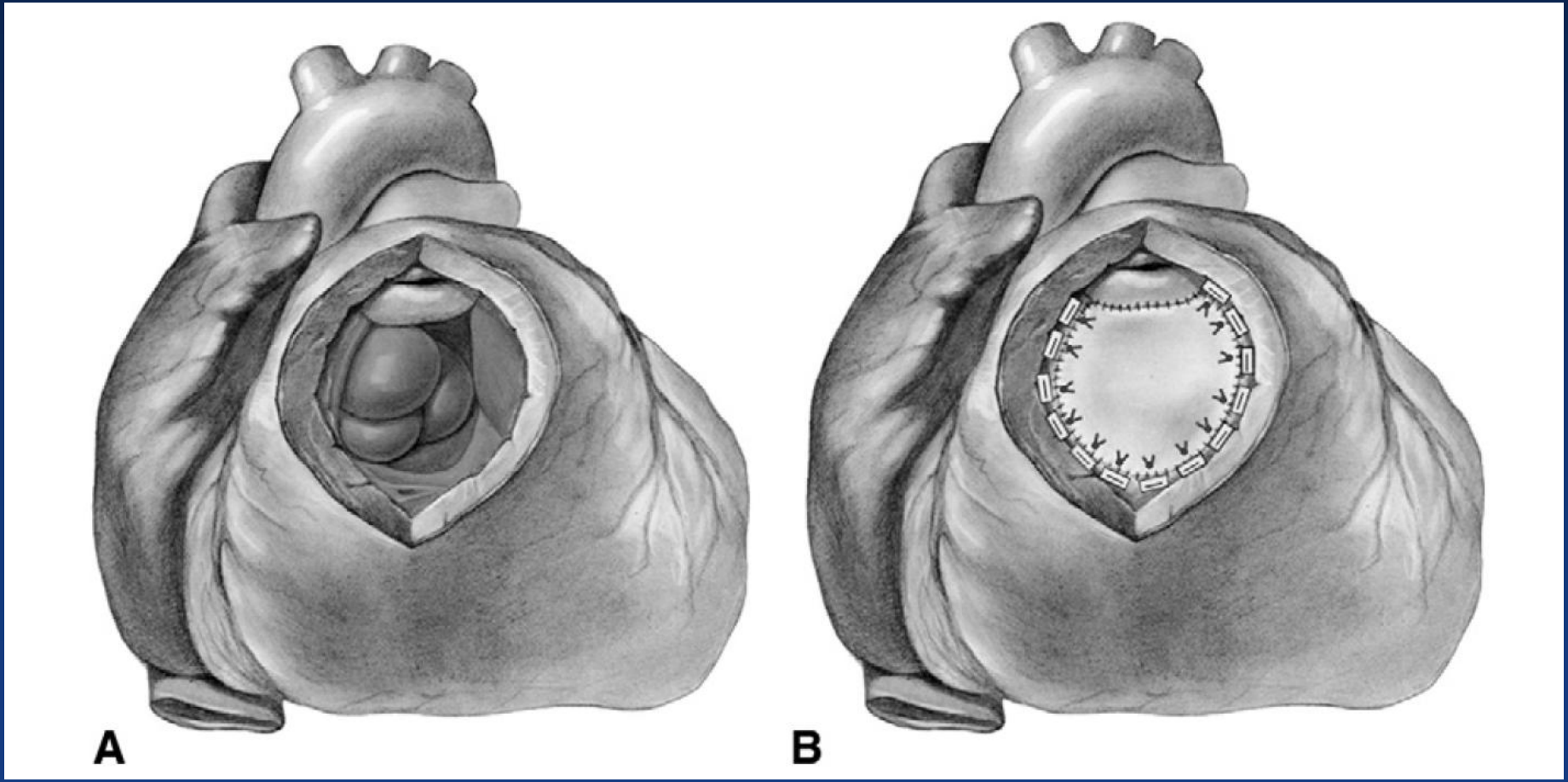
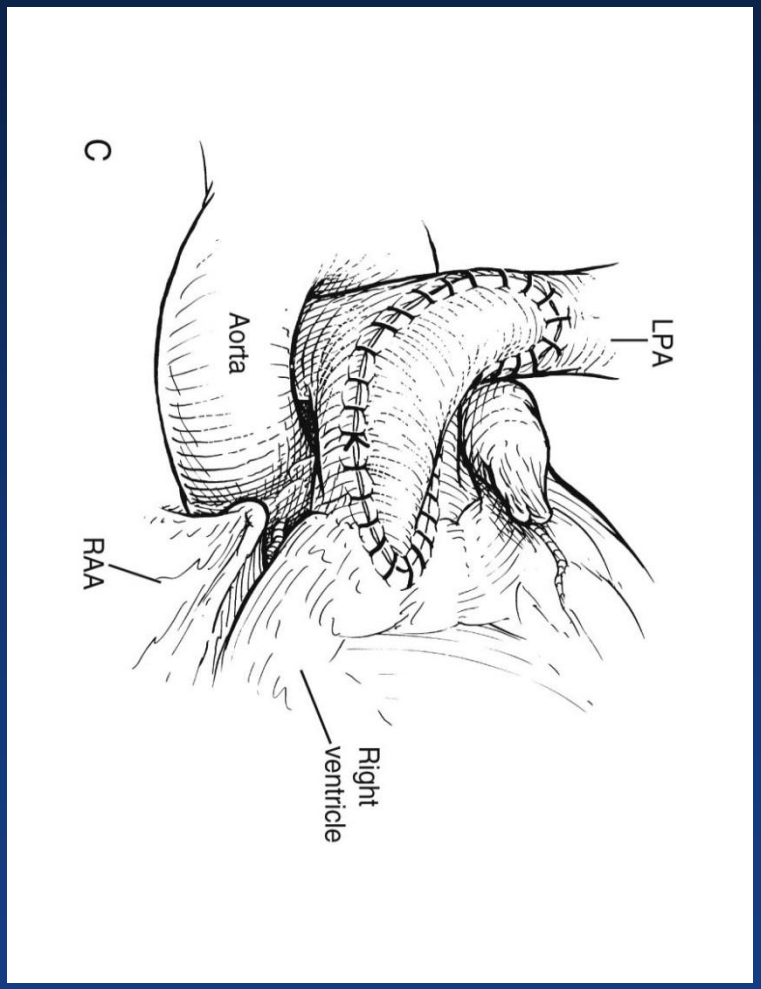


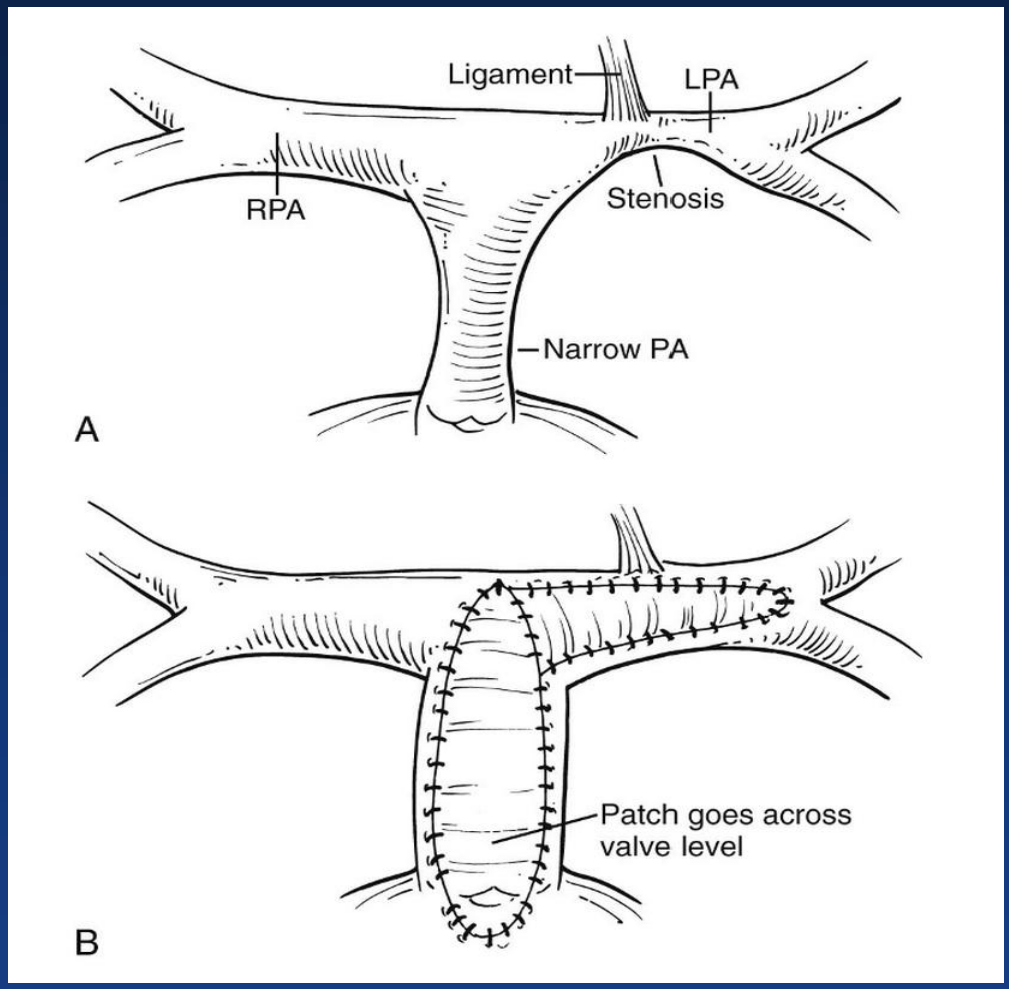
Fig 1. (A) The anterior pulmonic valve cusp is evenly divided after a small ventriculotomy is performed. (B) The rectangularly shaped patch is placed between the divided valve cusps, and the ventriculotomy incision was subsequently covered with the same patch. (C) The incision at the main pulmonary artery is then covered with a second ovoid glutaraldehyde-treated bovine pericardial patch, which is connected to the middle or lower part of the first patch used in valve repair, covering the ventriculotomy incision. A large anterior pulmonic valve cusp with a large sinus is created.

Corrective Surgery
- Infundibular Septal Deficiency -

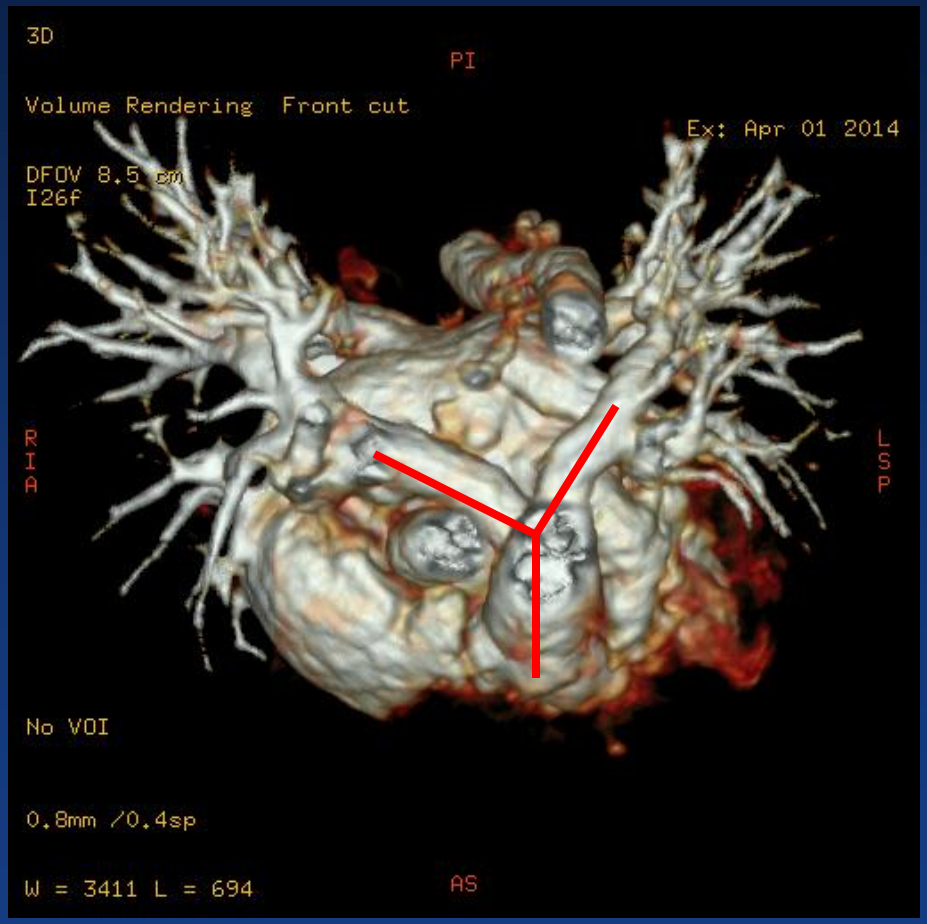


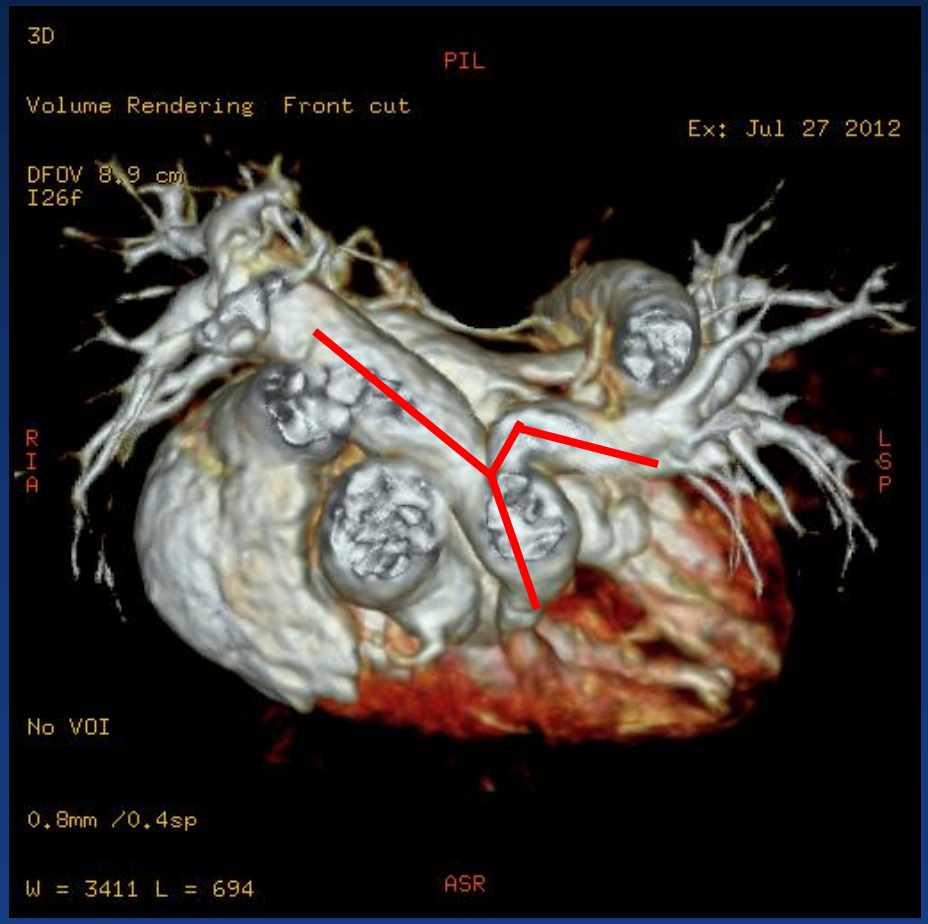
Corrective Surgery
- Branch PA Widening -





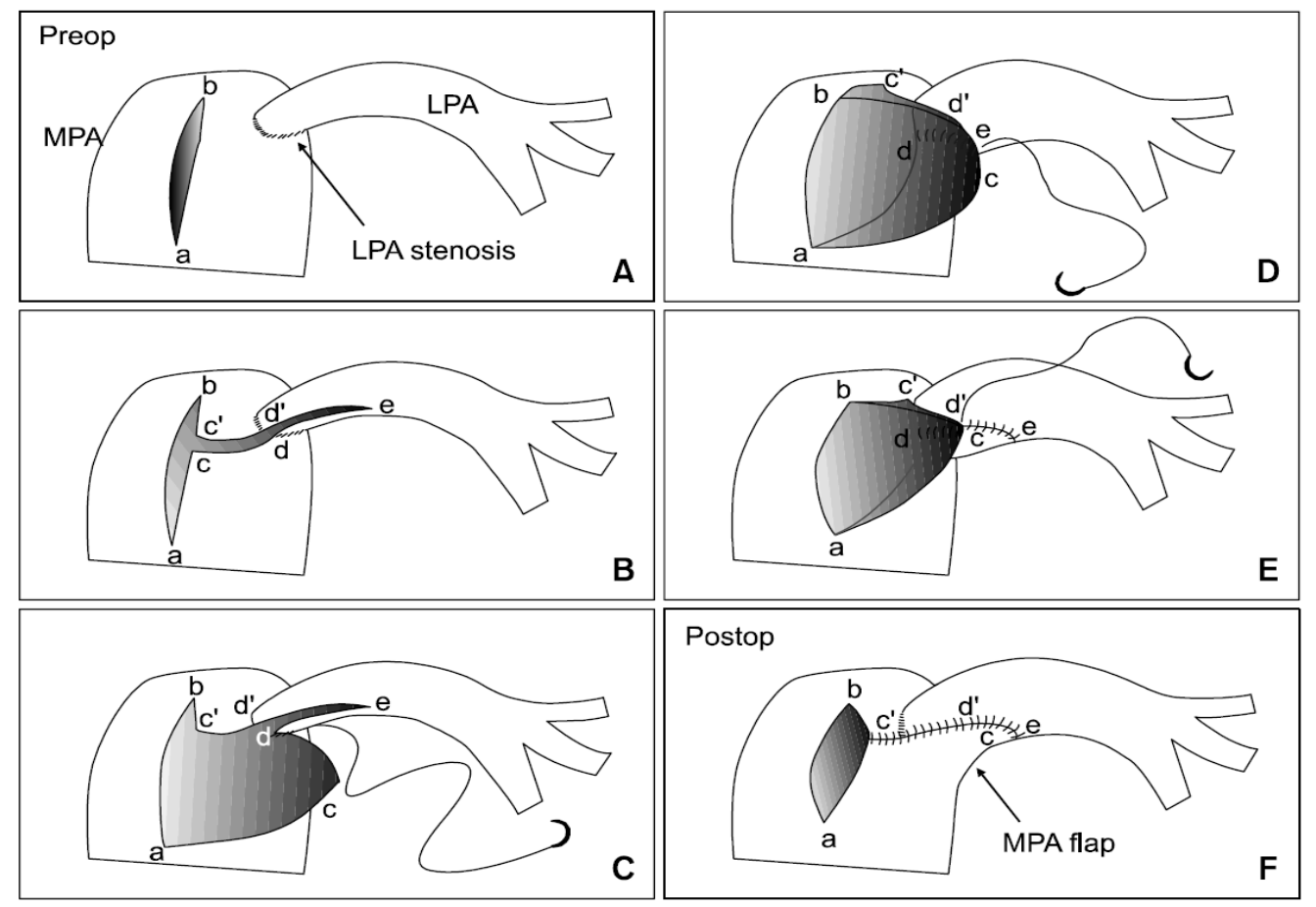
Corrective Surgery
- Branch PA Widening -



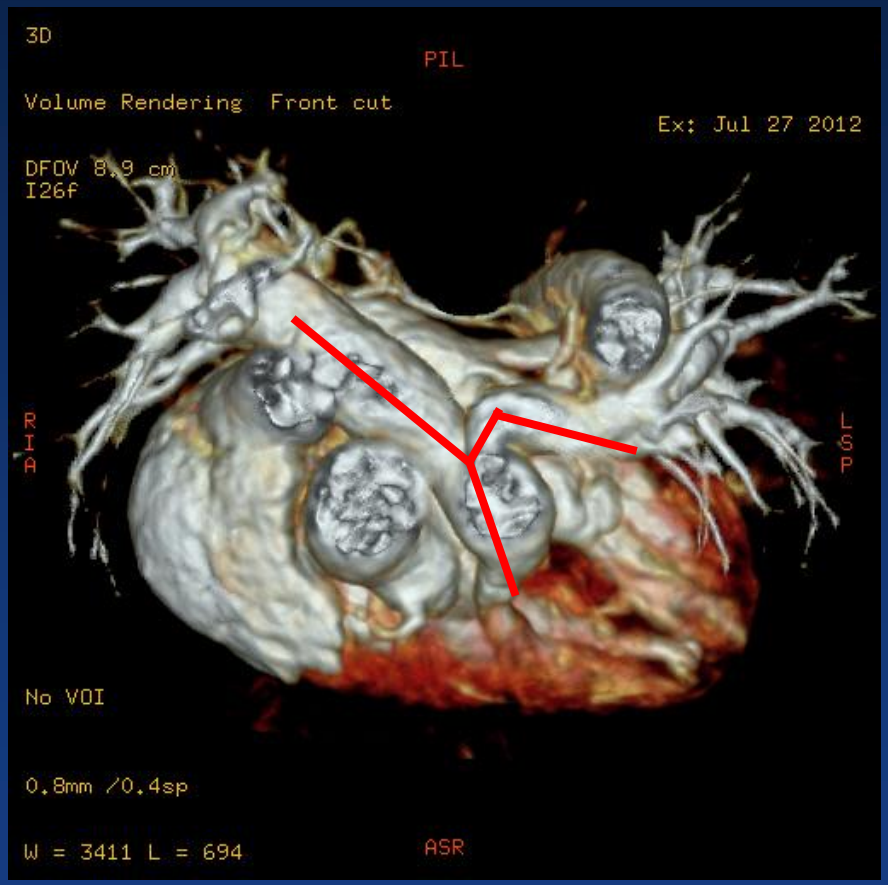


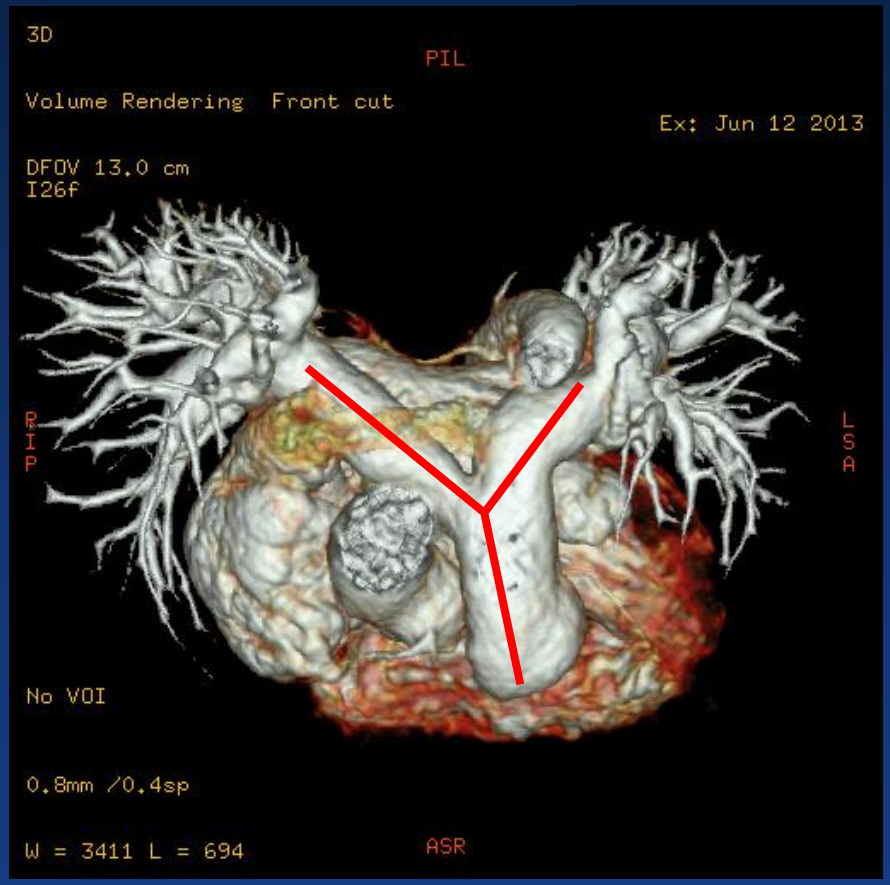
활로 4징 환자에서 자가 주폐동맥 플랩을 이용한 새로운 좌폐동맥 성형술

이창하* · 전양빈* · 이택연** · 이석기*** · 백만종**** · 김수철**** · 이영탁*****



Corrective Surgery
- Branch PA Widening -





Preop **Postop 10 mo**

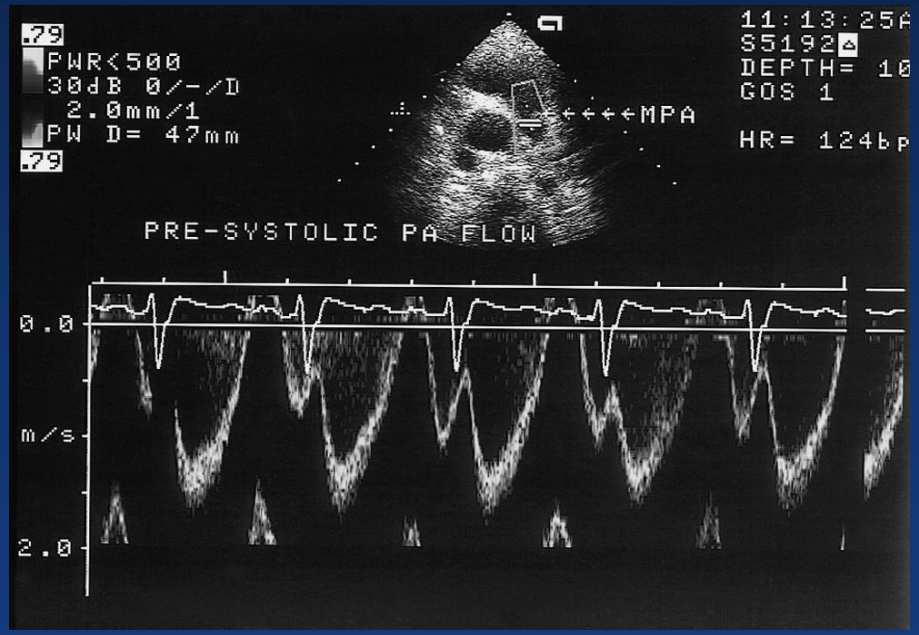
Postoperative Issues

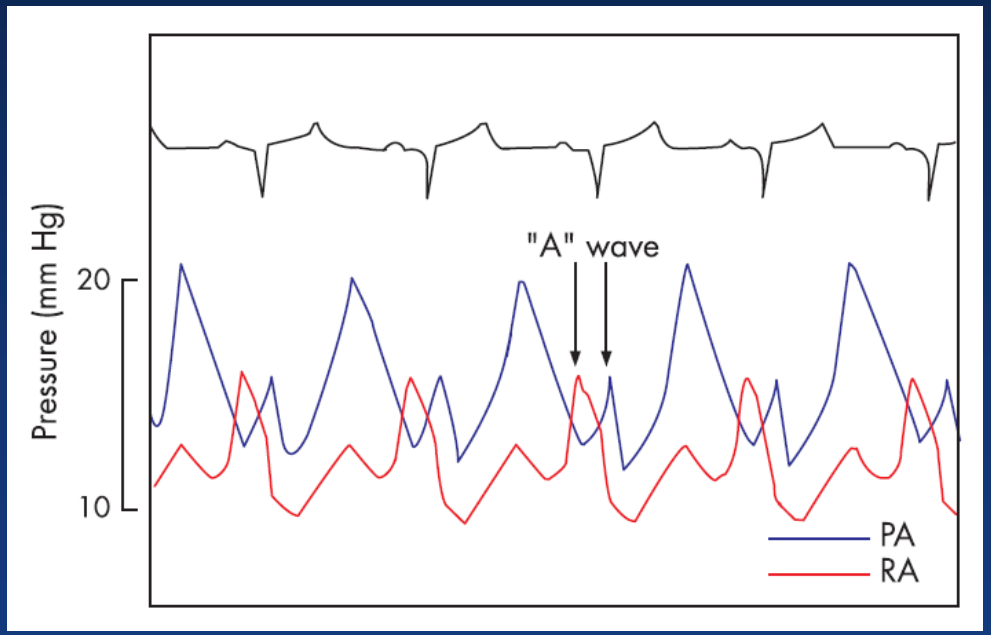
- RV dysfunction
- Residual lesions
  - Residual VSD: even small defect cannot be tolerated
  - Residual RVOTO
- Arrhythmia
- PR

Postoperative Issues
- RV Dysfunction -

- RV incision/ excision
- Inadequate myocardial protection
- Residual RVOTO
- Significant PR
- Residual VSD

Postoperative Issue
- Restrictive Physiology of RV -





"A" wave transmitted into the PA

Postoperative Issue
- Restrictive Physiology of RV -

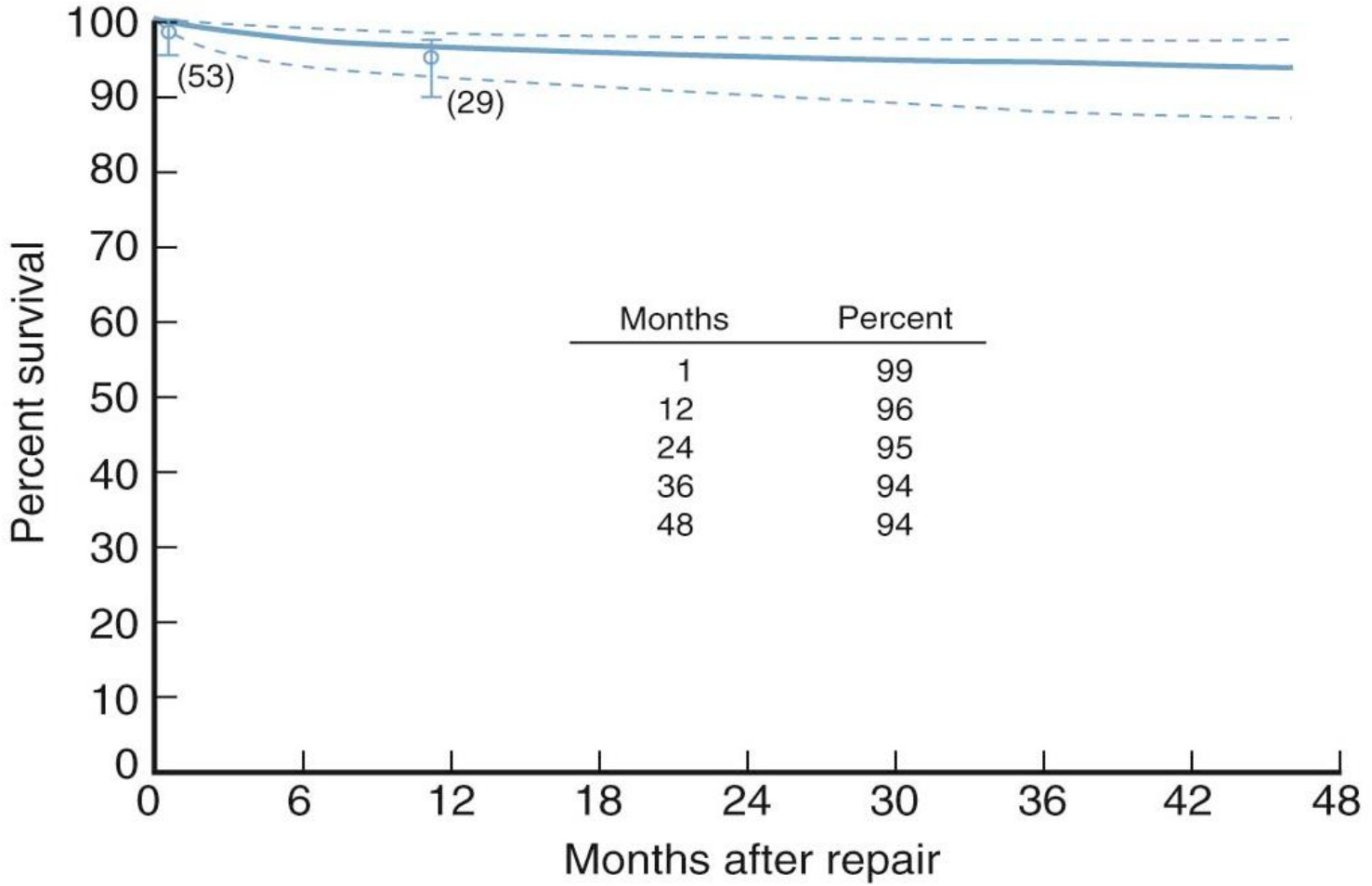
- Suboptimal cardiac output
- Elevated CVP
- Fluid collection : pleural/ peritoneal/ tissue (heart/lung/…)

→ Prolonged ICU care

Outcomes

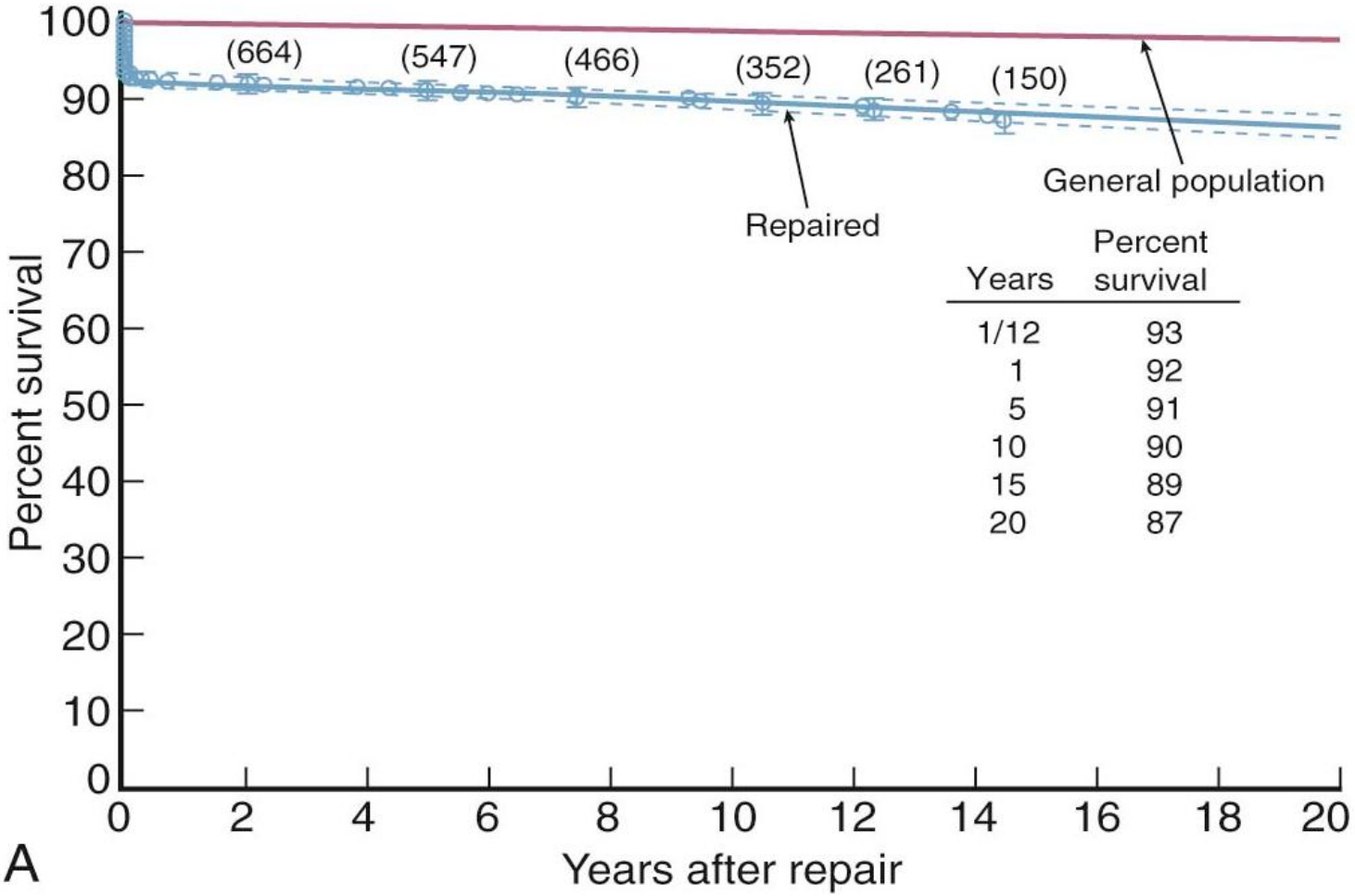
- Early mortality: 3~5%
  - Nearly zero percent in recent series
- Long-term survival
  - Excellent, but lower than general population
  - Mainly related to RV

Outcomes



Months	Percent
1	99
12	96
24	95
36	94
48	94

Outcomes


100
90
80
70
60
50
40
30
20
10
0
Percent survival
(664)
(547)
(466)
(352)
(261)
(150)
Repaired
General population
Years
Percent survival
1/12 93
1 92
5 91
10 90
15 89
20 87
0 2 4 6 8 10 12 14 16 18 20
Years after repair
A

Late Problems

- Branch PA stenosis
- PR
- RV dilatation and dysfunction
  → LV dysfunction
- Arrhythmia

Late Problems
- Optimal Timing of PVR -

Pediatric Cardiac Surgery Annual

Indications and Timing of Pulmonary Valve Replacement After Tetralogy of Fallot Repair

Tal Geva

Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2006;9:11-22

Late Problems
- Optimal Timing of PVR -

- Repaired TOF or similar physiology with moderate or severe pulmonary regurgitation (regurgitation fraction ≥25% measured by CMR) and two or more of the following criteria:

1. RV end-diastolic volume index ≥160 mL/m^2 (Z score >5)
2. RV end-systolic volume index ≥70 mL/m^2
3. LV end-diastolic volume index ≤65 mL/m^2
4. RV ejection fraction ≤45%
5. RVOT aneurysm
6. Clinical criteria: exercise intolerance, symptoms and signs of heart failure, cardiac medications, syncope, sustained ventricular tachycardia.

Outcomes of Pulmonary Valve Replacement in 170 Patients With Chronic Pulmonary Regurgitation After Relief of Right Ventricular Outflow Tract Obstruction

Implications for Optimal Timing of Pulmonary Valve Replacement

Cheul Lee, MD,* Yang Min Kim, MD,† Chang-Ha Lee, MD,* Jae Gun Kwak, MD,* Chun Soo Park, MD,* Jin Young Song, MD,‡ Woo-Sup Shim, MD,‡ Eun Young Choi, MD,‡ Sang Yun Lee, MD,‡ Jae Suk Baek, MD‡

Bucheon, Korea

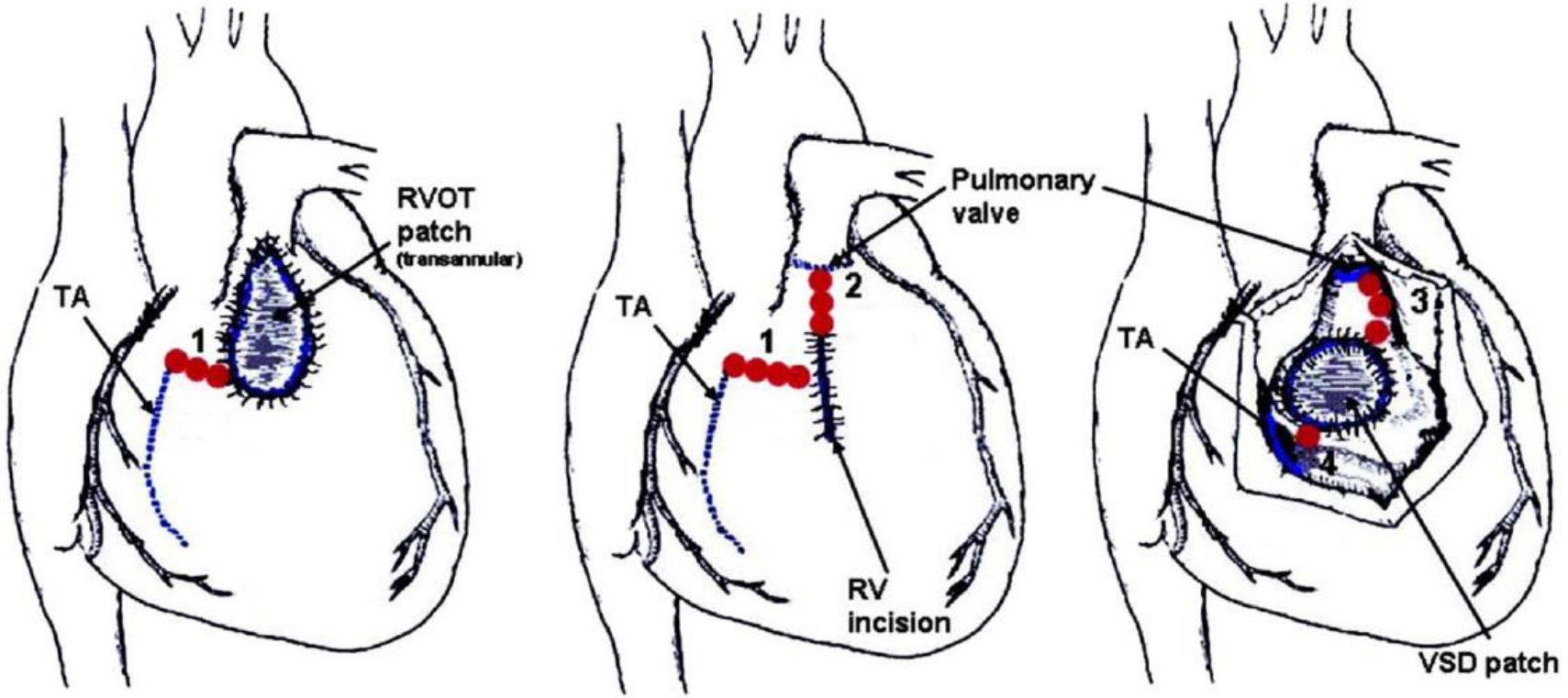
J Am Coll Cardiol 2012;60:1005-14

Midterm outcomes of PVR in patients with chronic PR were acceptable. PVR should be considered before RV EDVI exceeds 163 ml/m^2 or RV ESVI exceeds 80 ml/m^2, with more attention to RV ESVI. (J Am Coll Cardiol 2012;60:1005–14)

Late Problems
- Arrhythmia -

- Sustained arrhythmia: upto 30%
- Atrial arrhythmia: m/c
  - Intra-atrial re-entrant tachycardia
  - Atrial fibrillation
- Ventricular tachycardia: single m/c

Late Problems
- Arrhythmia -



Summary

- Early outcome is satisfactory
- Late outcome is suboptimal
- Every effort is needed to reduce the late event, when perfoming initial corrective surgery.

COMMENTARY

Tetralogy of Fallot repair: Ready for a new paradigm

Tal Geva, MD

J Thorac Cardiovasc Surg 2012;143:1305-6

The ongoing quest for refinement of valve-sparing RVOT reconstruction and the unresolved debate as to the optimal balance between residual PS and PR highlight the urgent need for a new paradigm in the surgical management of TOF. Although small incremental improvements have an

COMMENTARY

Tetralogy of Fallot repair: Ready for a new paradigm

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important role, it is now necessary to develop a bioengineered pulmonary valve substitute, as well as a contracting RVOT patch with biomechanical properties that are as close as possible to those of the myocardium. These biotechno-