전공의 연수강좌
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Tetralogy of Fallot

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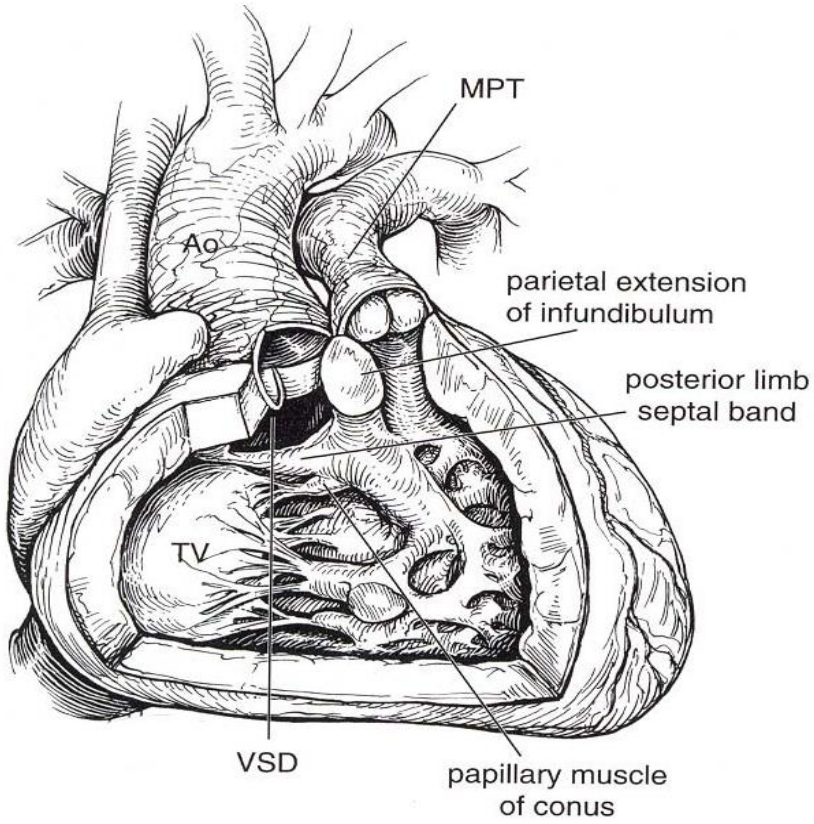
Definition

- Classic theory
  - Unequal sprial septation of conotruncus

- Van Praagh's theory
  - Underdevelopment of RV infundibulum with **anterior & leftward displacement (malalignment) of infundibular (conal, outlet) septum**



Anterior and leftward displacement of the infundibular (conal) septum

- ROVTO & RVH
- Large VSD
- Overriding of aorta

Incidence & Associated anomaly

- 3/10000 live birth
- 7~10% of CHD & the most common among the cyanotic defect (50%)
- M>F
- Genetic defect in 28%
  - Chromosome 22q11.2 defect
  - Trisomy 21
  - VACTERL (Vertebral anomaly, Imperforate anus, Cardiac anomaly, TEF, Esophageal atresia, Renal anomaly, Limb defect)

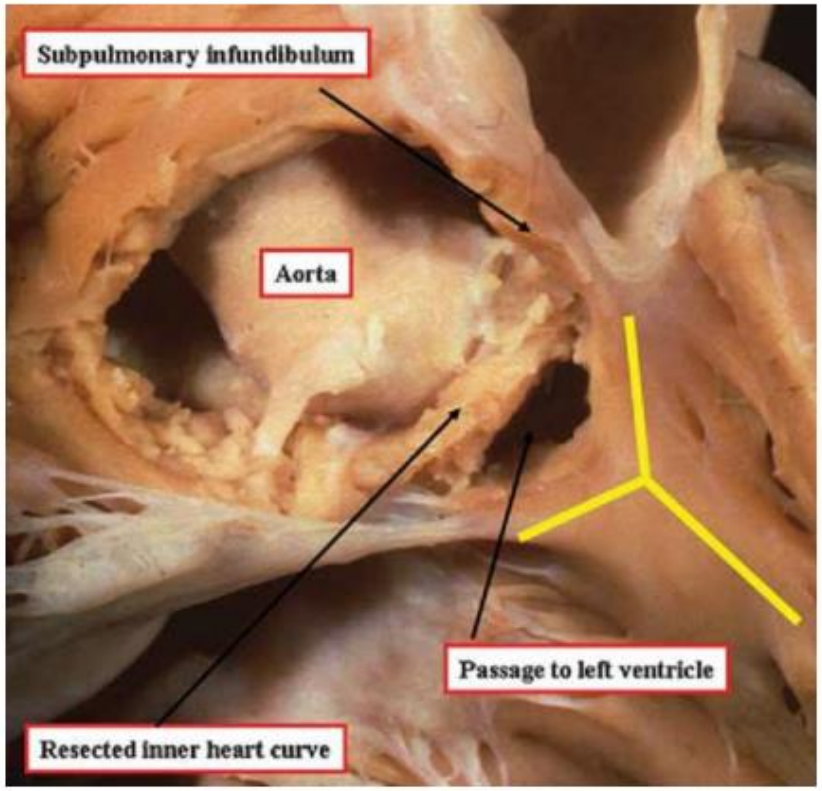
Clinical features

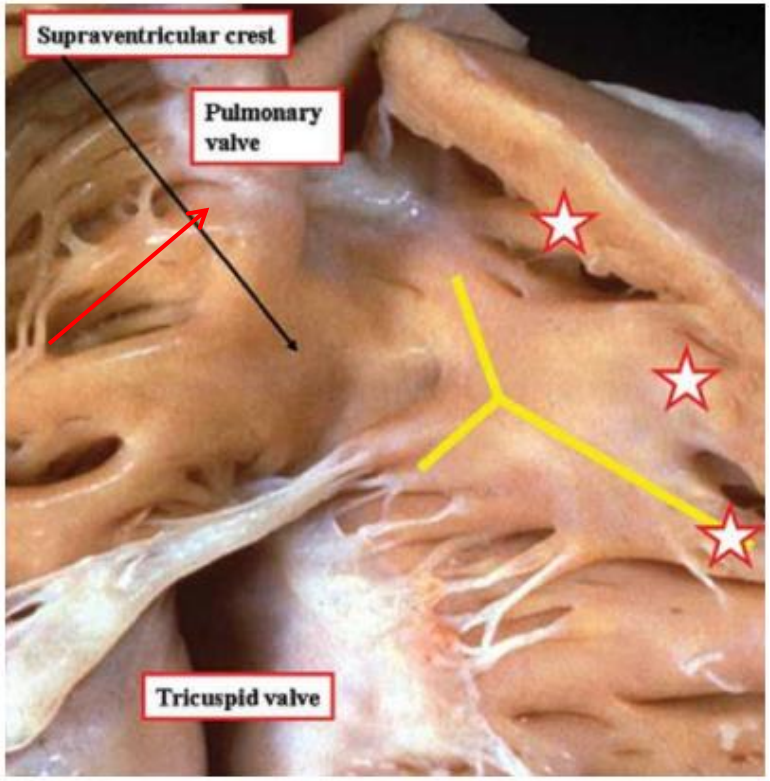
- Severity of RVOTO
  - Degrees of cyanosis
- Mild RVOTO
  - Predominant L to R shunt → CHF (pink TOF)

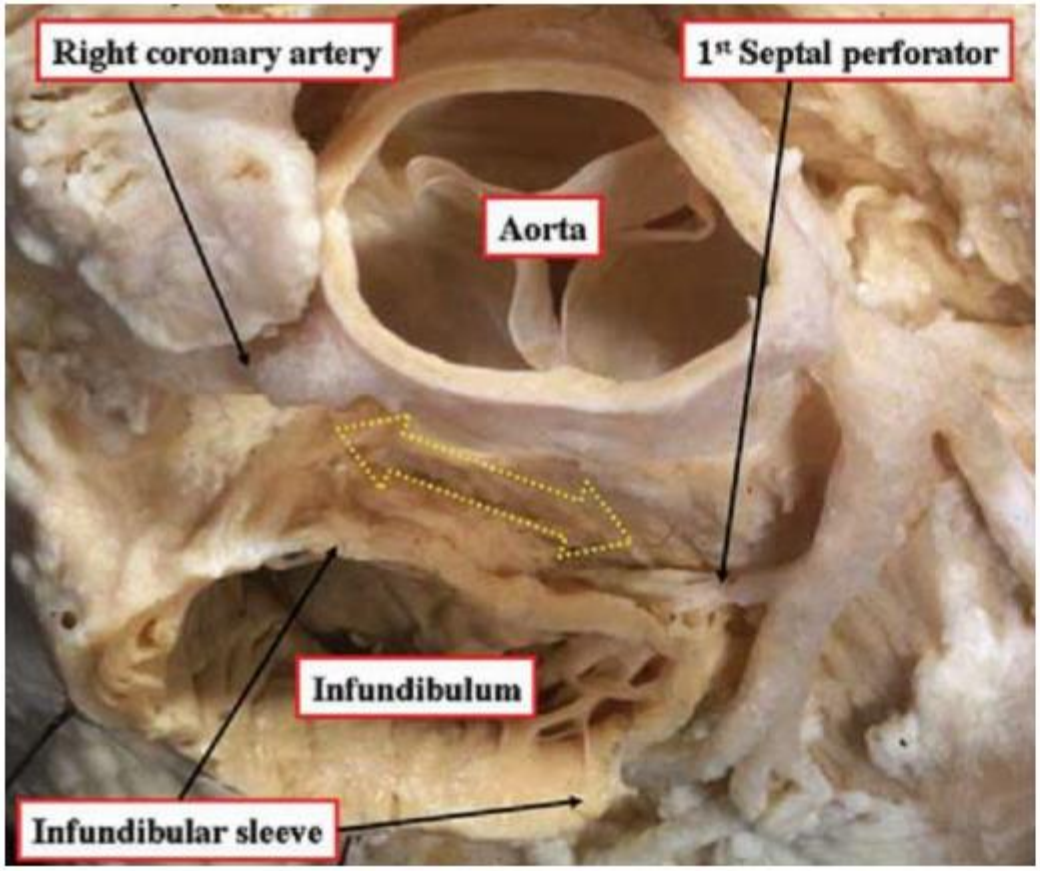
- Most, commonly, cyanosis is mild at birth and gradually progresses with age
  - increasing hypertrophy of the RV infundibulum
- Mechanism of "cyanotic spell" initiation
  - Reduction of cardiac afterload or preload and tachycardia (dehydration, viral infection, vasodilatation by medication, etc.)

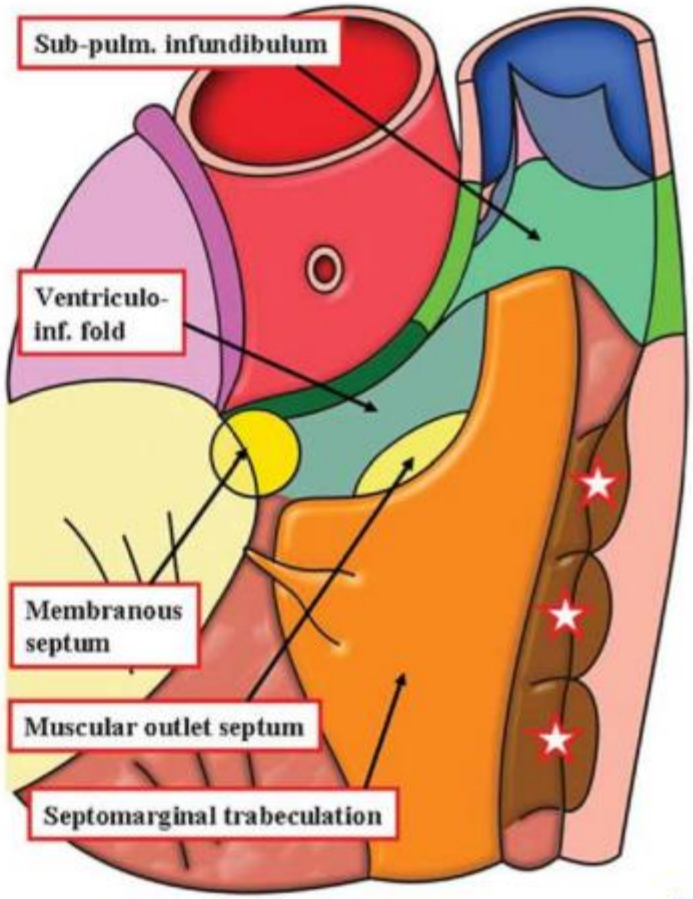
Normal Heart

Crista supraventricularis (Supraventricular crest)
Muscular area separating the attachments of the TV and PV in the roof of the RV

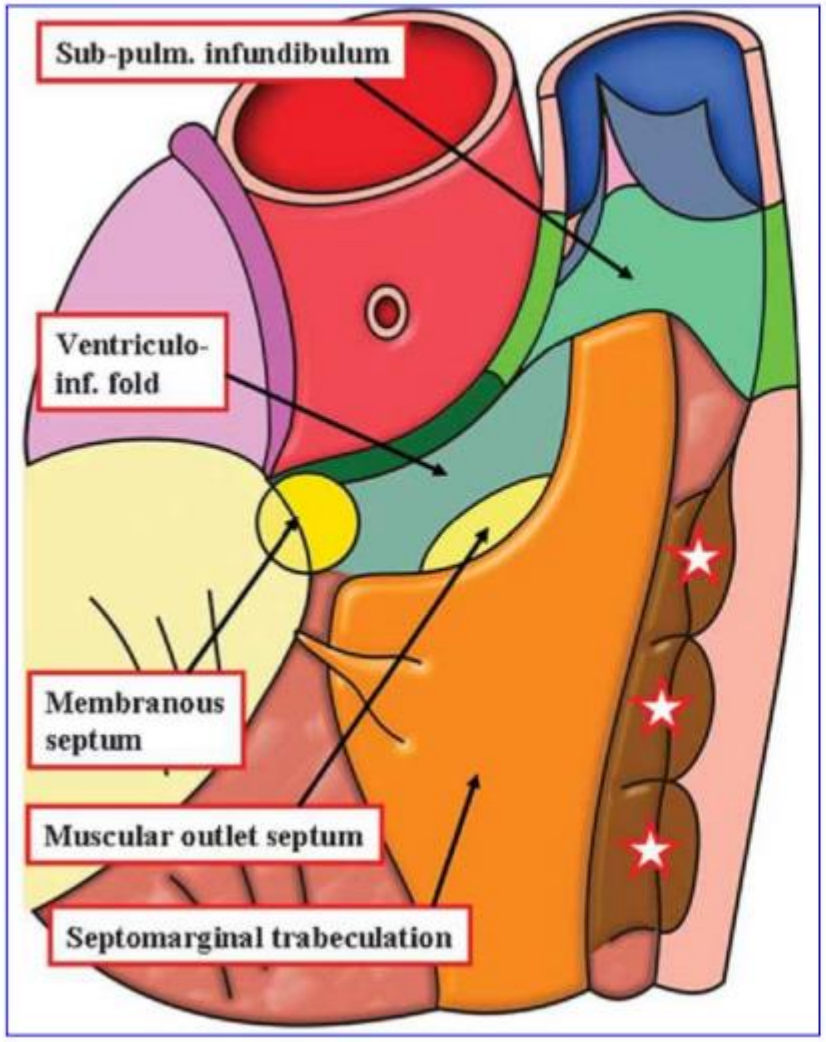




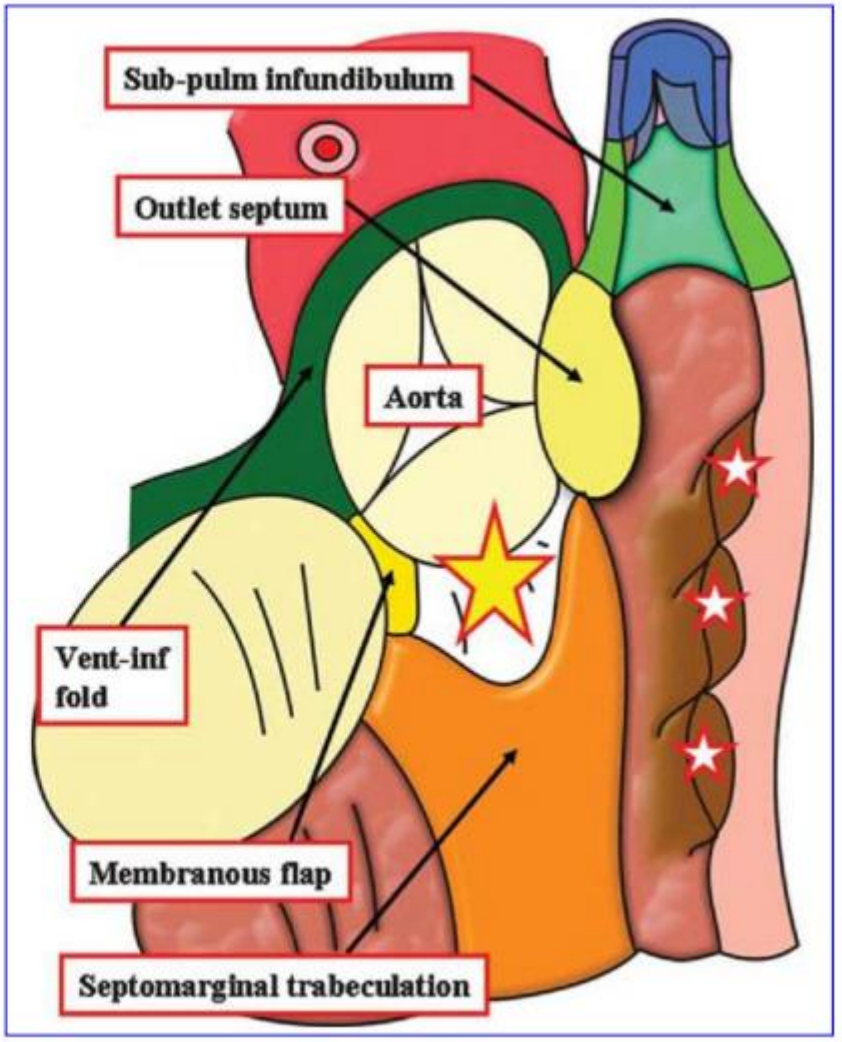

Right coronary artery
1st Septal perforator
Aorta
Infundibulum
Infundibular sleeve


Sub-pulm. infundibulum
Ventriculo-inf. fold
Membranous septum
Muscular outlet septum
Septomarginal trabeculation

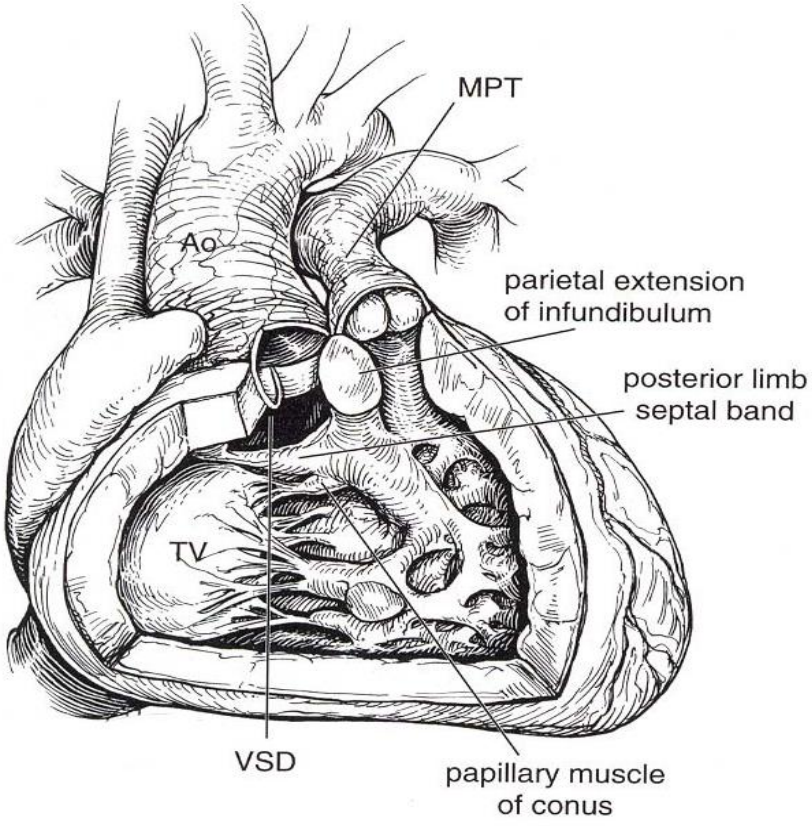
Normal

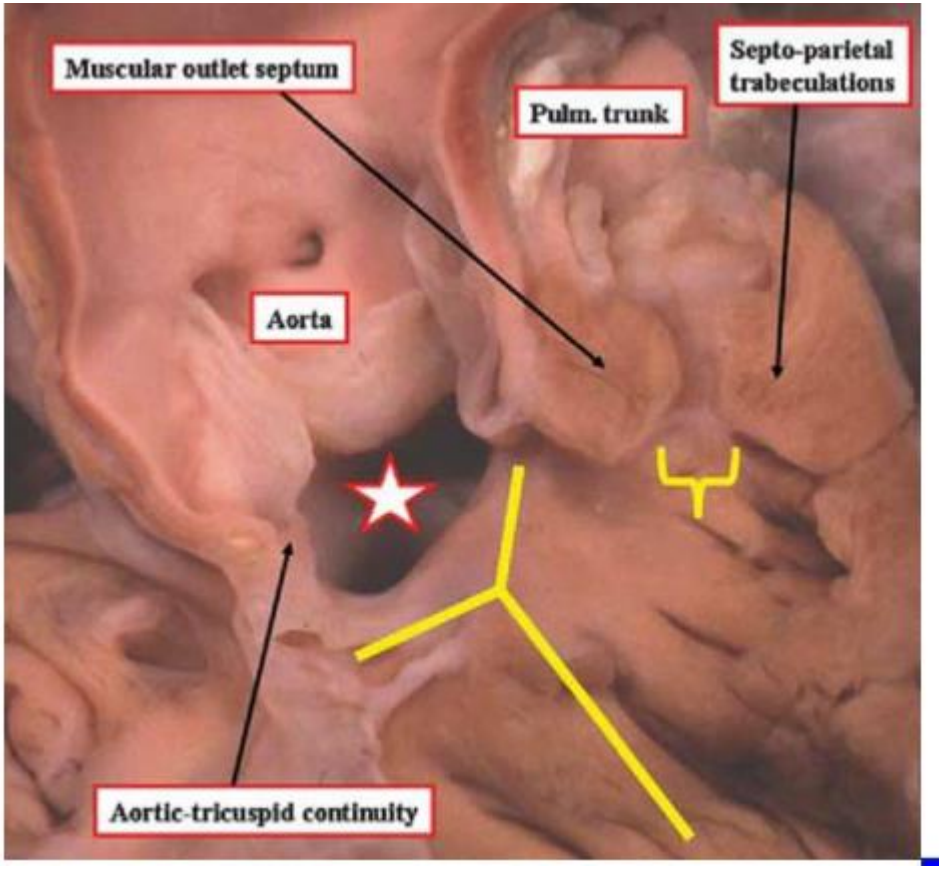

Sub-pulm. infundibulum
Ventriculo-inf. fold
Membranous septum
Muscular outlet septum
Septomarginal trabeculation

TOF


Sub-pulm infundibulum
Outlet septum
Aorta
Vent-inf fold
Membranous flap
Septomarginal trabeculation

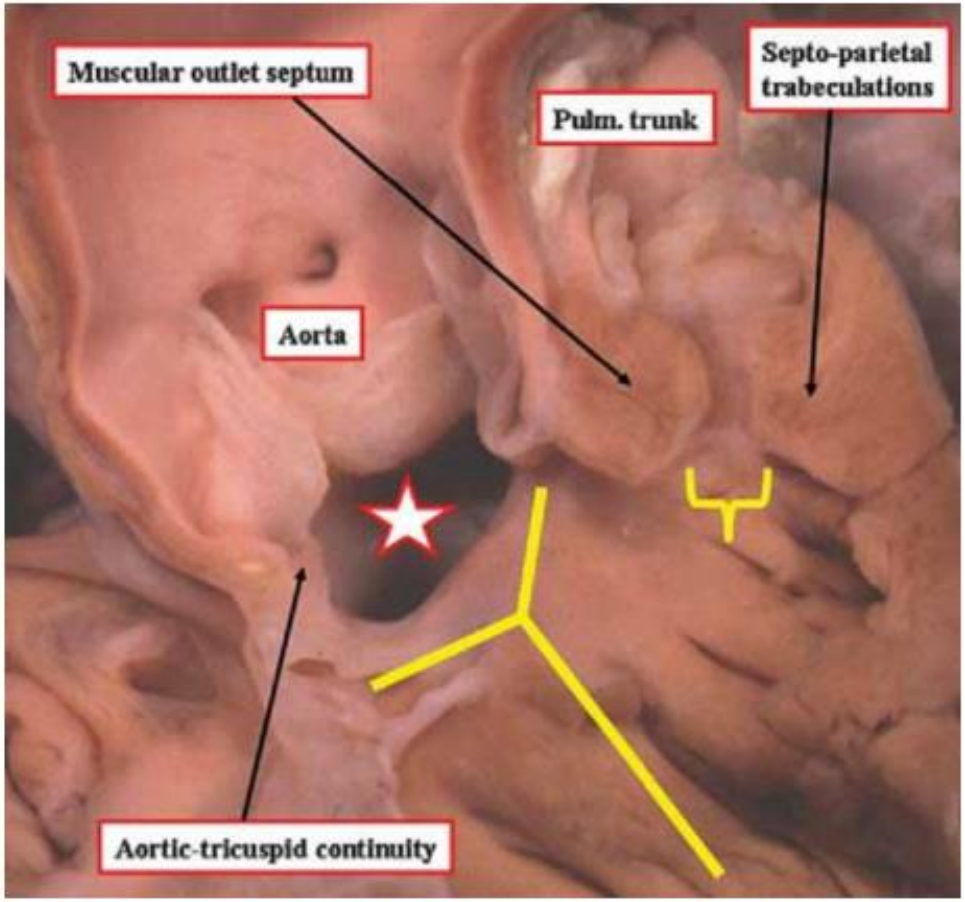
TOF

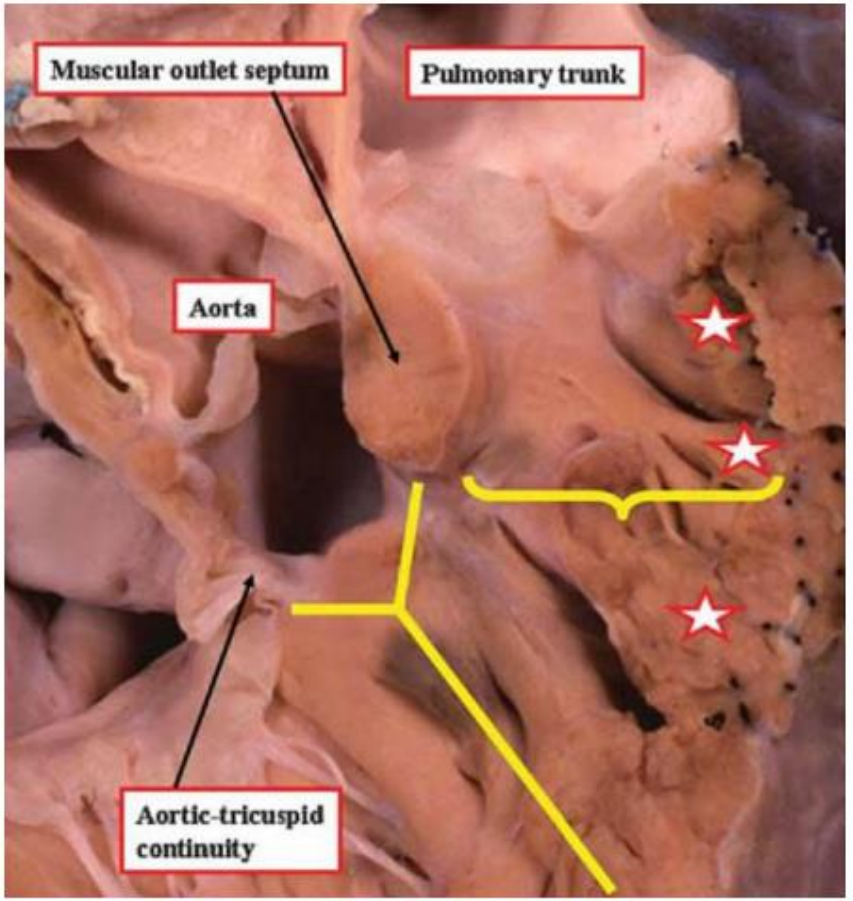

MPT
Ao
parietal extension
of infundibulum
posterior limb
septal band
TV
VSD
papillary muscle
of conus


Muscular outlet septum
Pulm. trunk
Septo-parietal
trabeculations
Aorta
Aortic-tricuspid continuity

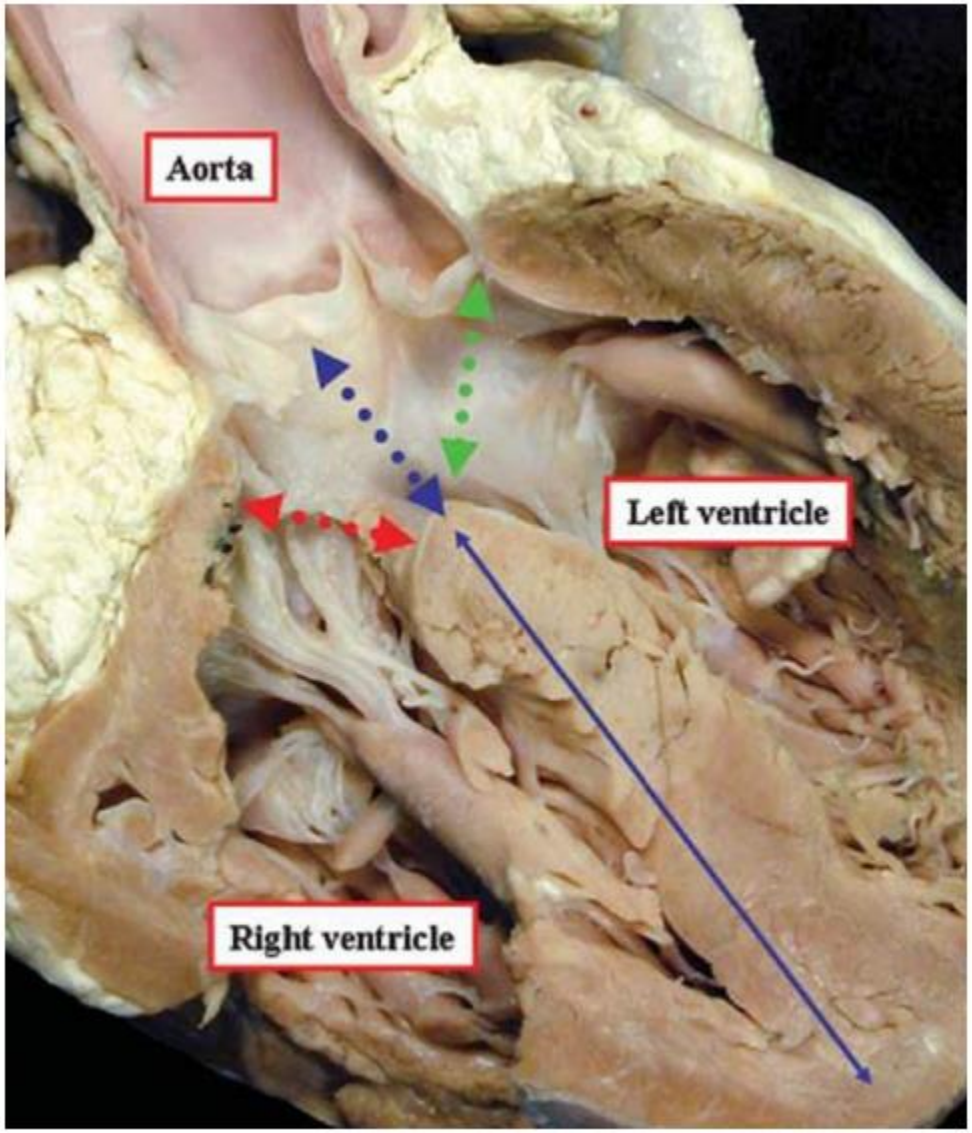
TOF

Anteriorly malaligned VSD
(Eisenmeger defect)





Overriding of the aortic valve

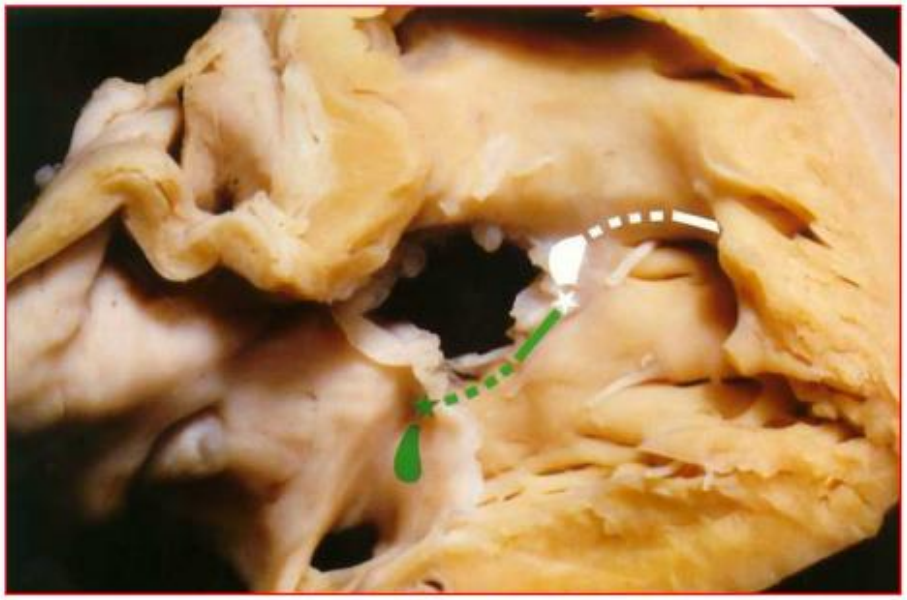


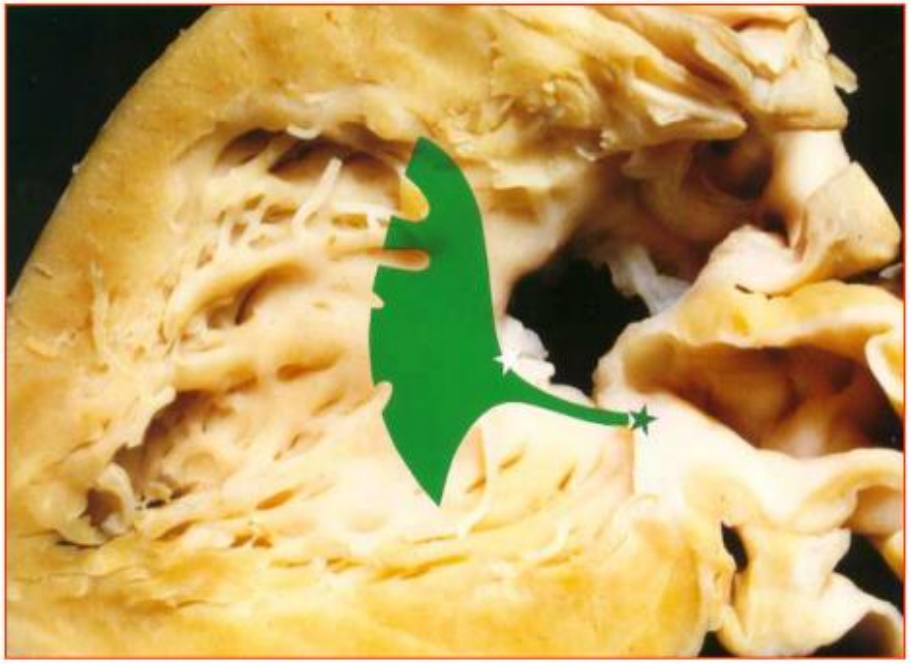
Pulmonary valve and annulus

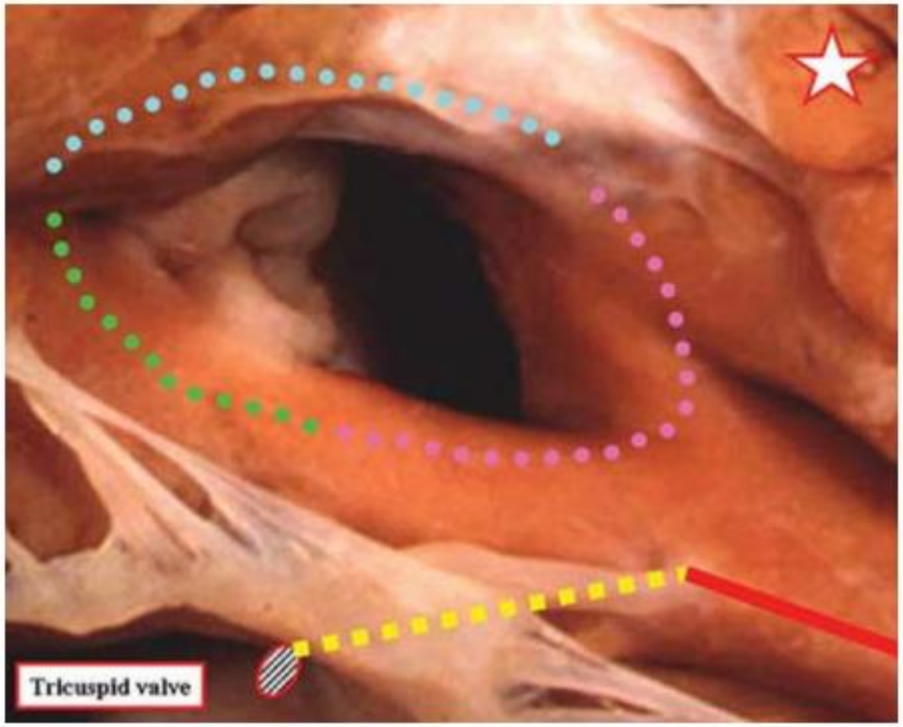
- Stenosis in 75%
- Leaflets
  - Thickened, tethered to the PA
  - Bicuspid in 75%
  - Vertical or horizontal position

- MPA & branch Pas
  - Usually somewhat diffusely small, often short
  - Narrowest portion of MPA is often at STJ
  - Branch PA stenosis in 10%
    - LPA os

- VSD
  - Large anteriorly malaligned
  - 25%: VIF extends to the posterior limb of TSM
    - Muscle bar beneath TV (MO)
  - Additional VSDs in 3~15%
- Conduction system
  - SA and AV node: normal in location






Tricuspid valve

- Coronary artery
  - 5%, LAD from RCA, dual LAD
  - Very occasionally, RCA from single LCA, LCA from single RCA
  - Crossing over RVOT, rarely in the myocardium
- Other anatomic features
  - PA: 7%
  - Absence of PV leaflet: 5%
  - 25%, right aortic arch

- Outpatient management
  - Relieving hypoxemia and preventing hypoxic spell
  - Beta-blocker : propranolol
    - Not reliable and should rarely be used
  - Protect from viral infections

Indications and timing of surgery

- Symptoms
  - PG dependent neonate
  - Worsening cyanosis
  - Cyanotic spell

Symptomatic neonates or young infants with TOF

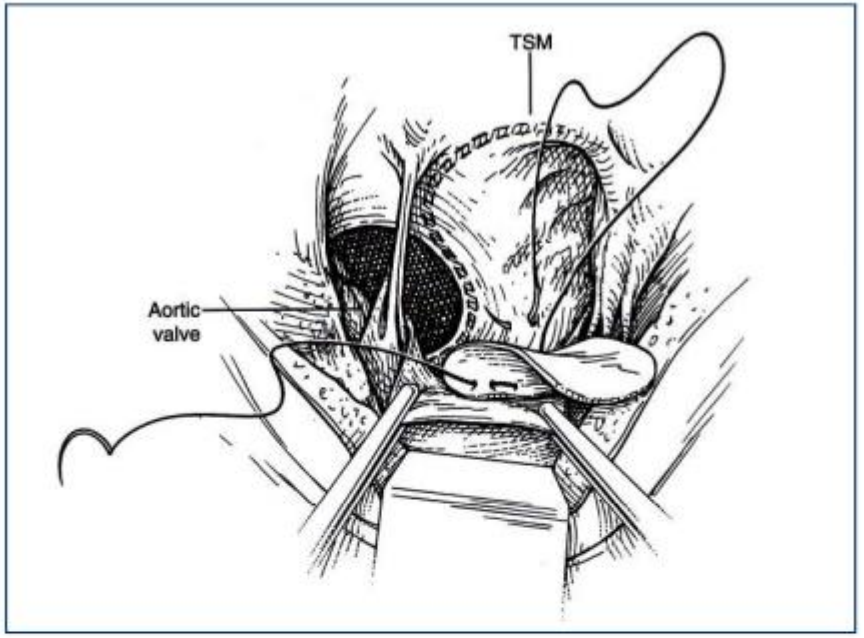
- Shunt vs Early primary repair
- Controversies in “Early primary repair”
  - **Pro**
    - Low operative mortality of early primary repair
    - Avoidance harmful effect of shunt operation and late repair
    - RVH
  - **Con**
    - Neonatal myocardium may be less capable of handling of RV volume load

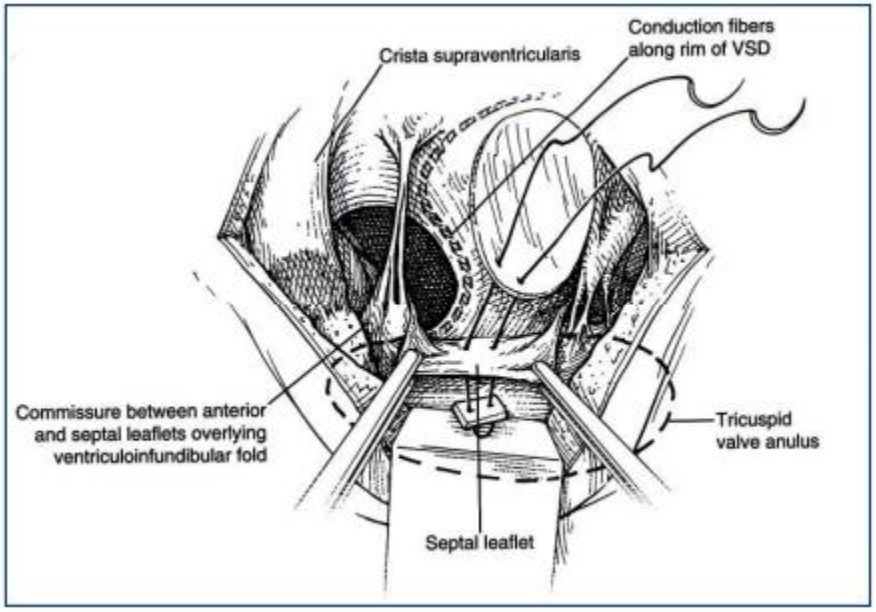
- Potential disadvantages of staged approach
  - Long-standing pressure overload of RV
  - Persistent cyanosis
    - Cardiomyocytic degeneration and interstitial fibrosis
    - Myocardial dysfunction and ventricular arrhythmia

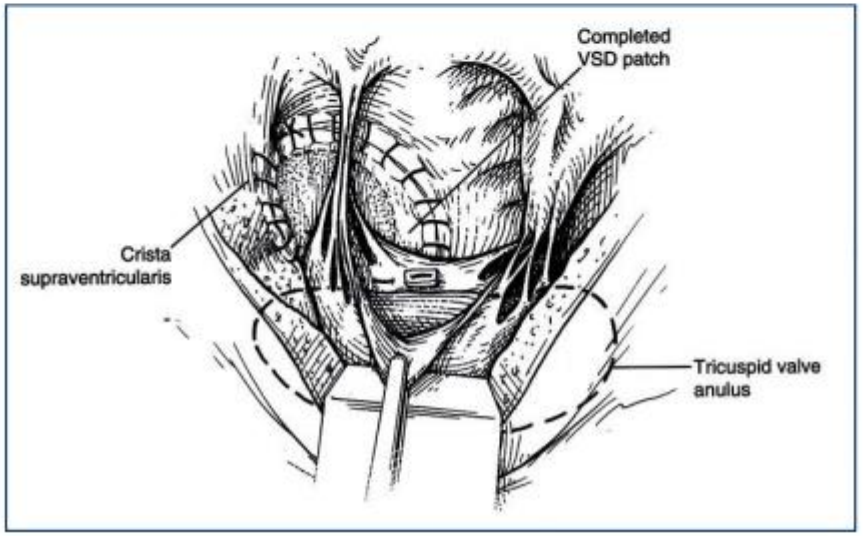
- Potential disadvantage of early primary repair
  - Frequent need of transannular patch
  - Adverse effects of early bypass surgery on the neonatal brain
  - Often complicated and lengthy postoperative recovery in small infants

Surgical Management

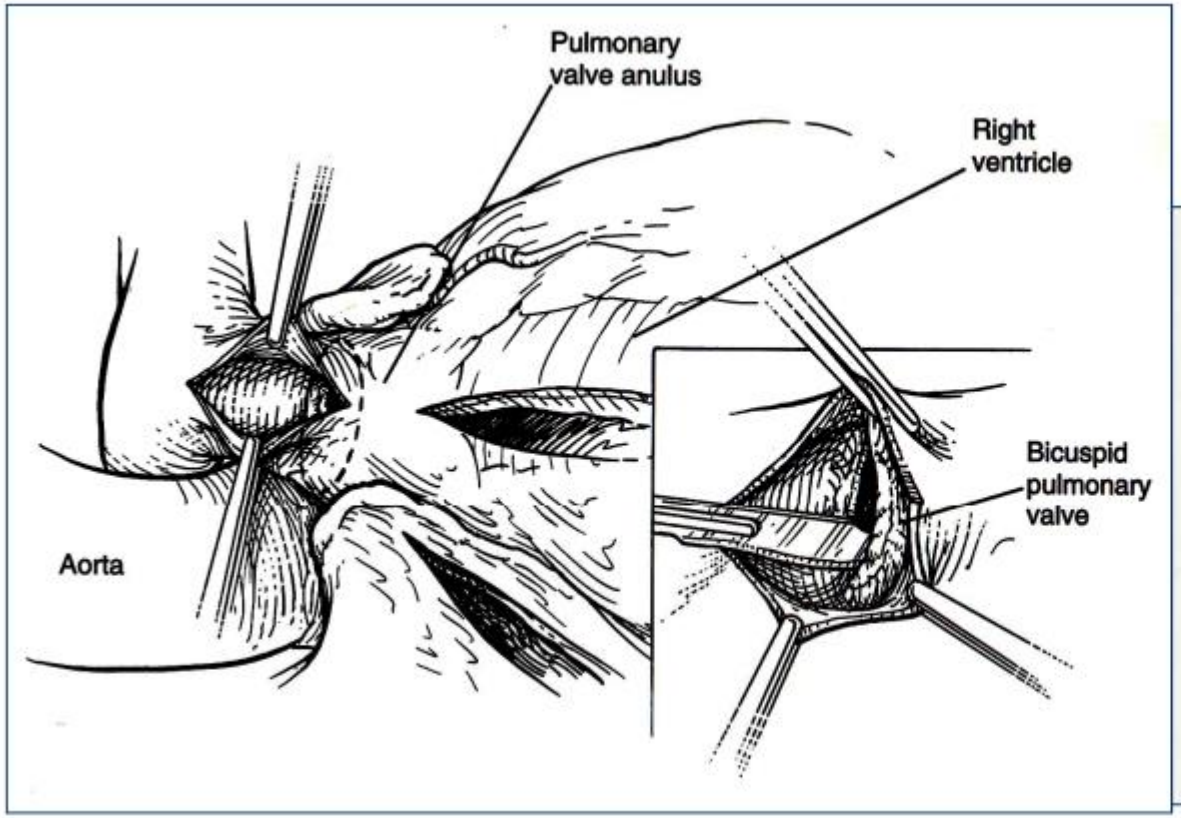
VSD closure

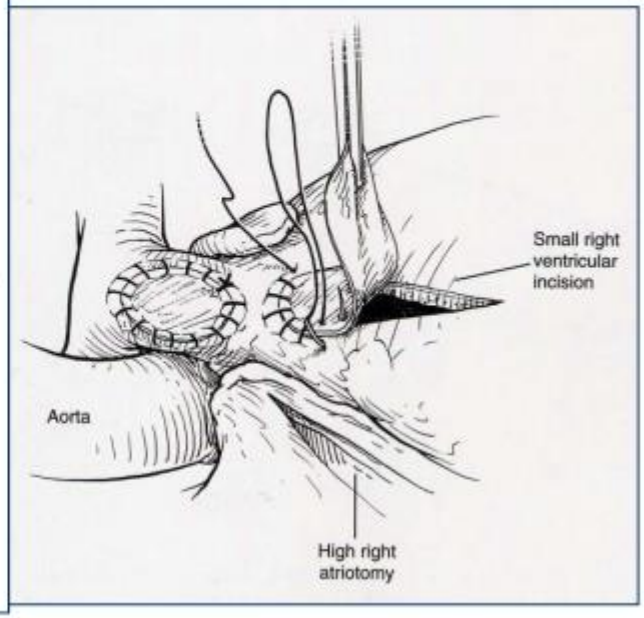




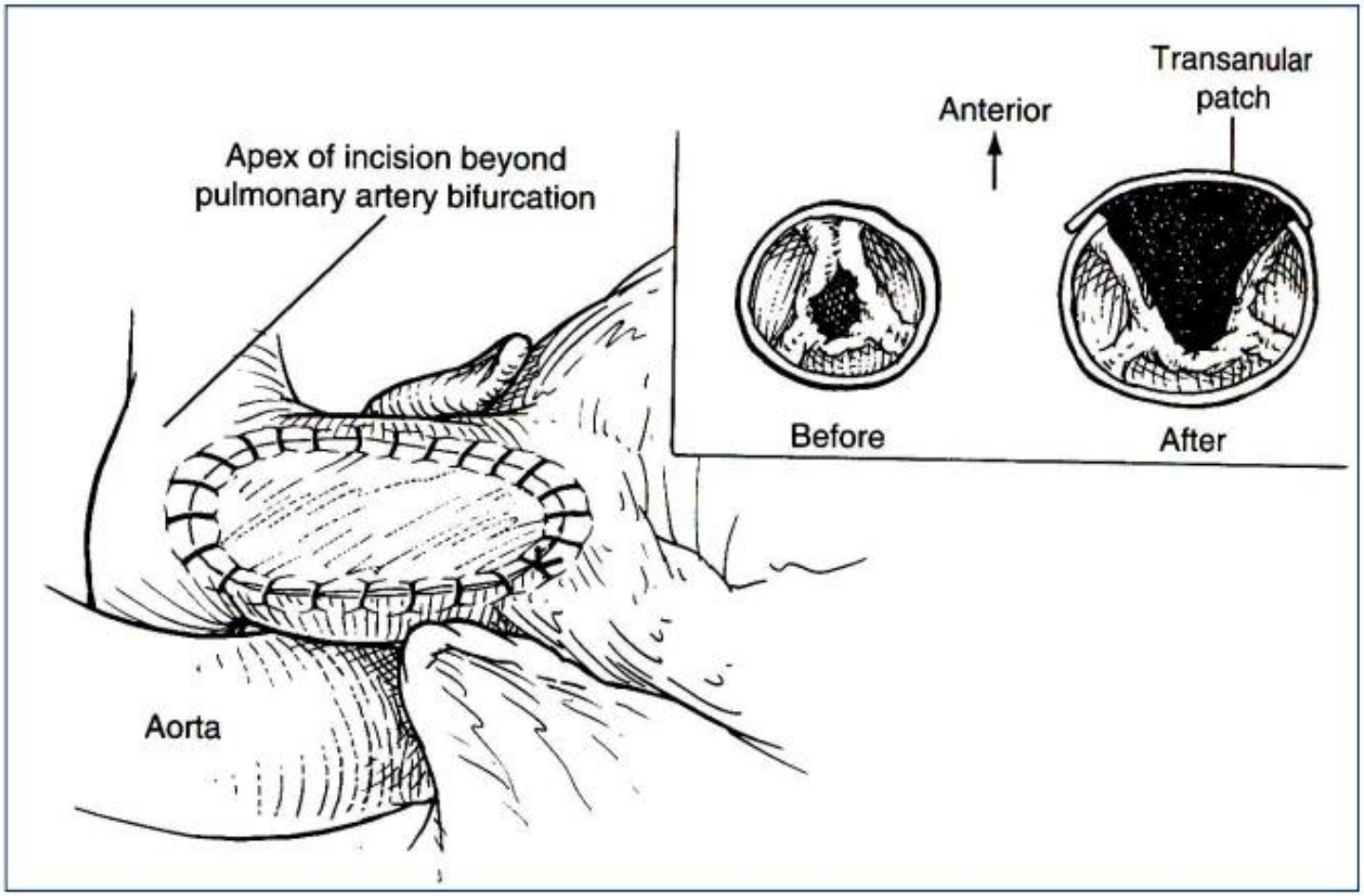


Pulmonary valvotomy





Transannular patch



Transannular patch

Criteria for preservation of pulmonary valve ?

z-valve of pulmonary annulus > -3

diameter of pulmonary annulus (mm) > 0.8mm/kg of body weight

Intraperative relaxed heart, PV annulus > (1-2mm or 0mm ?) + mean PV annulus

Postrepair RV/LV > 0.7

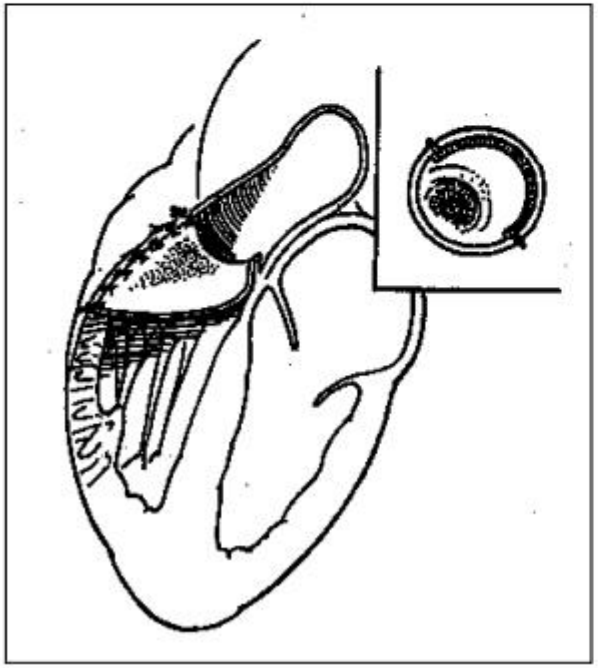
If TAP has not been placed, TAP should be considered.

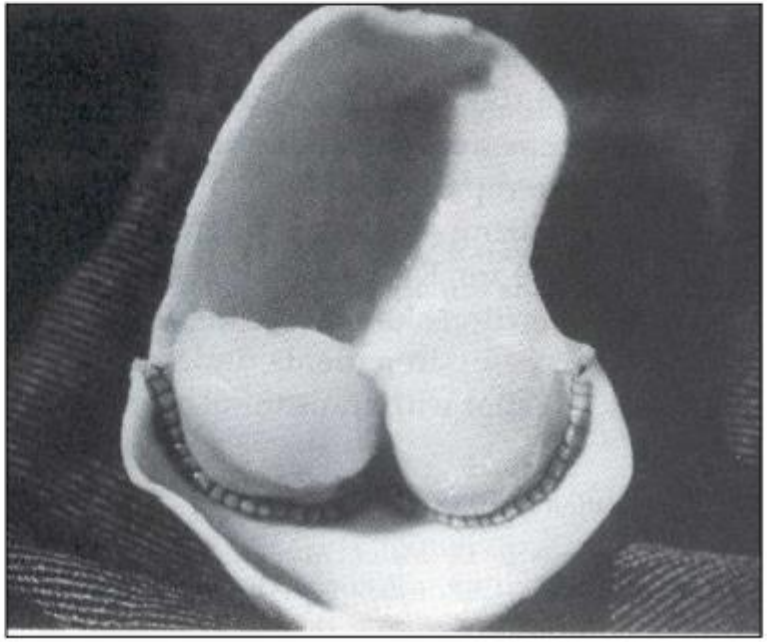
If TAP has been place, other causes must be considered.

- branch PA stenosis
- hypoplasia of peripheral PAs
- residual VSD
- residual infundibular obstruction

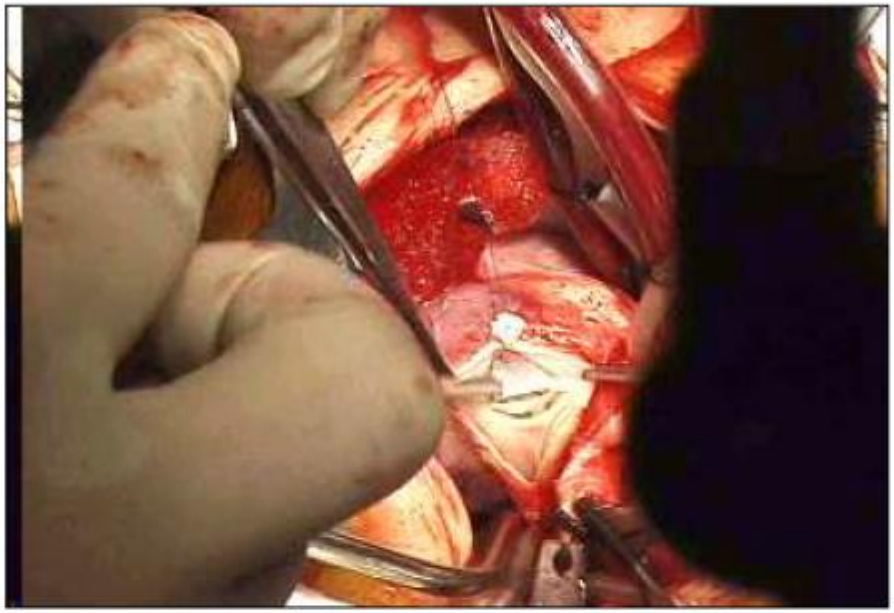
- Often elevation of RV pressure results from dynamic RVOTO.
- Ultra-short acting β-blocker (esmolol) can help in intraoperative differentiation.

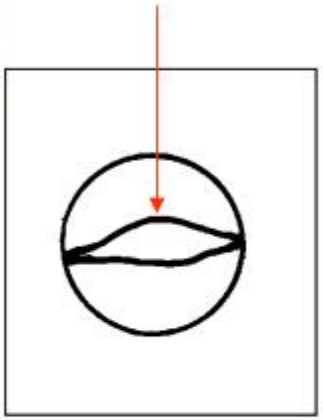
Monocups implantation



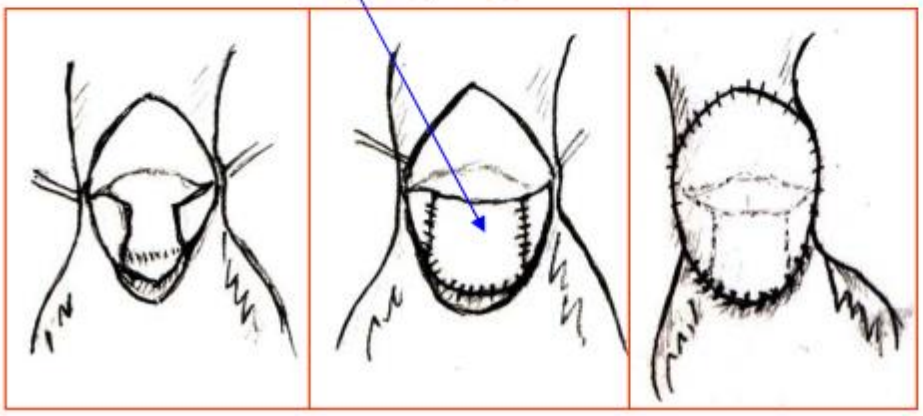


PV annular enlargement with valve reconstruction

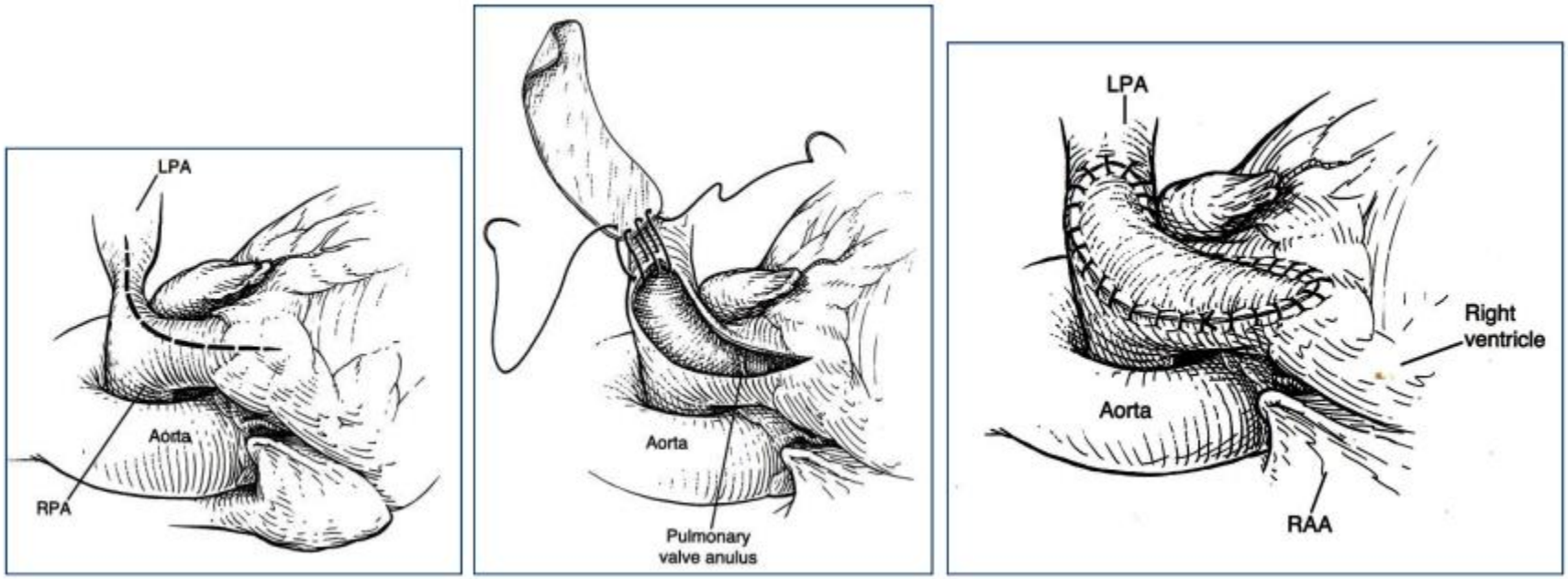




Gore-Tex membrane or autologous pericardium

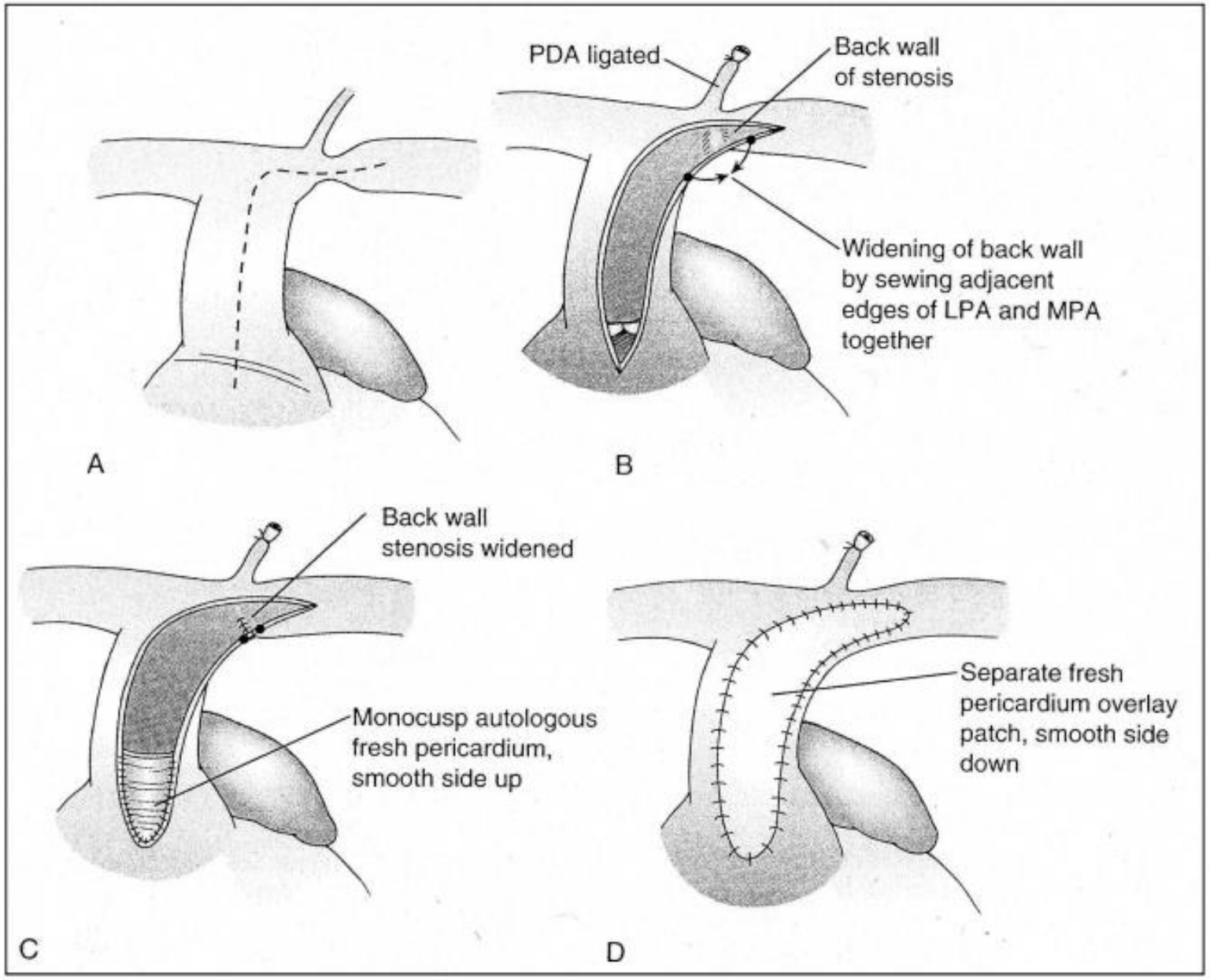


PA angioplasty



Causes of postoperative LPA stenosis

- Inadequate enlargement
- Aneurysmal dilatation of RVOT patch
- Kinking


PDA ligated
Back wall of stenosis
Widening of back wall by sewing adjacent edges of LPA and MPA together
A
B
Back wall stenosis widened
Monocusp autologous fresh pericardium, smooth side up
Separate fresh pericardium overlay patch, smooth side down
C
D

Acute angle of LPA with or without stenosis

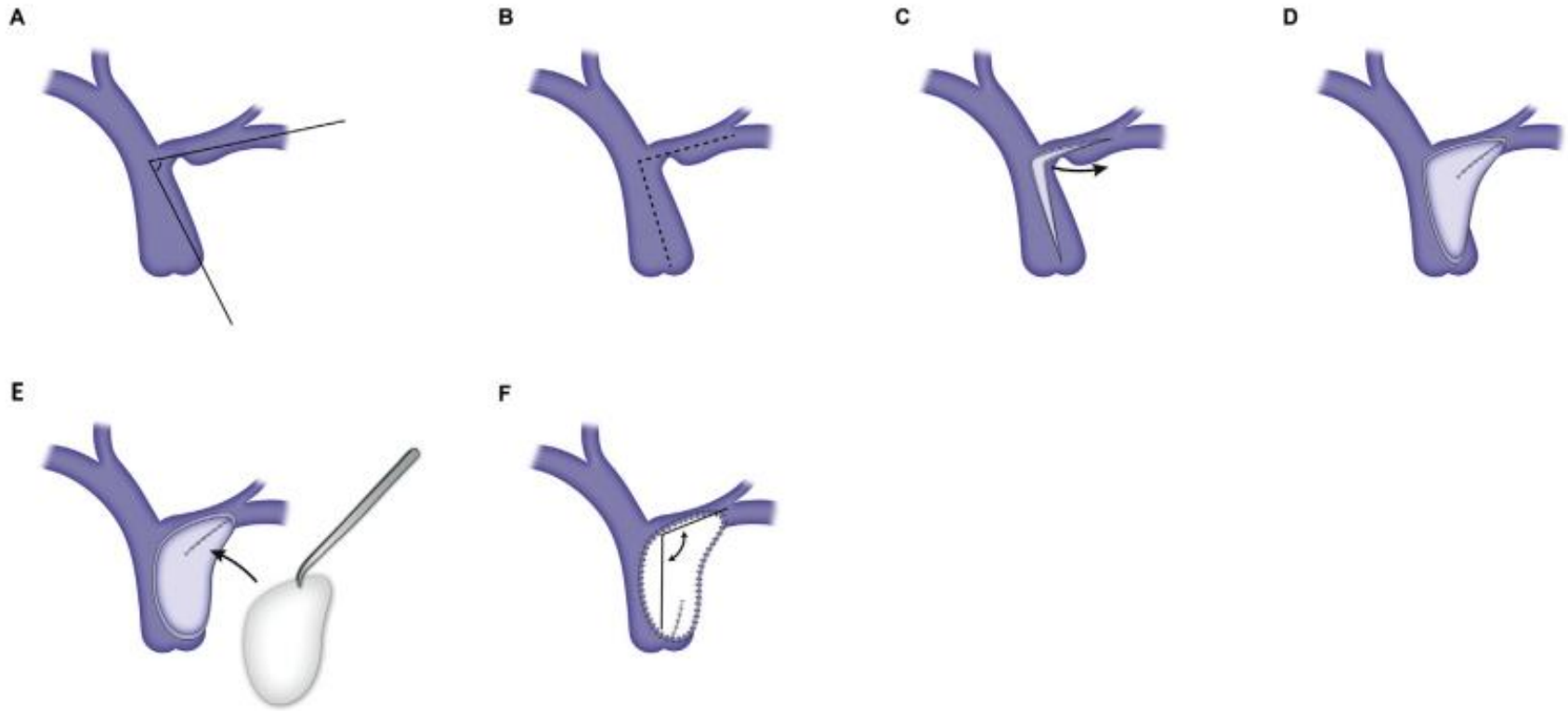


Fig 1. Schematic drawing of a left pulmonary artery (LPA) acute-angle correction angioplasty. (A–B) The main pulmonary artery (MPA)

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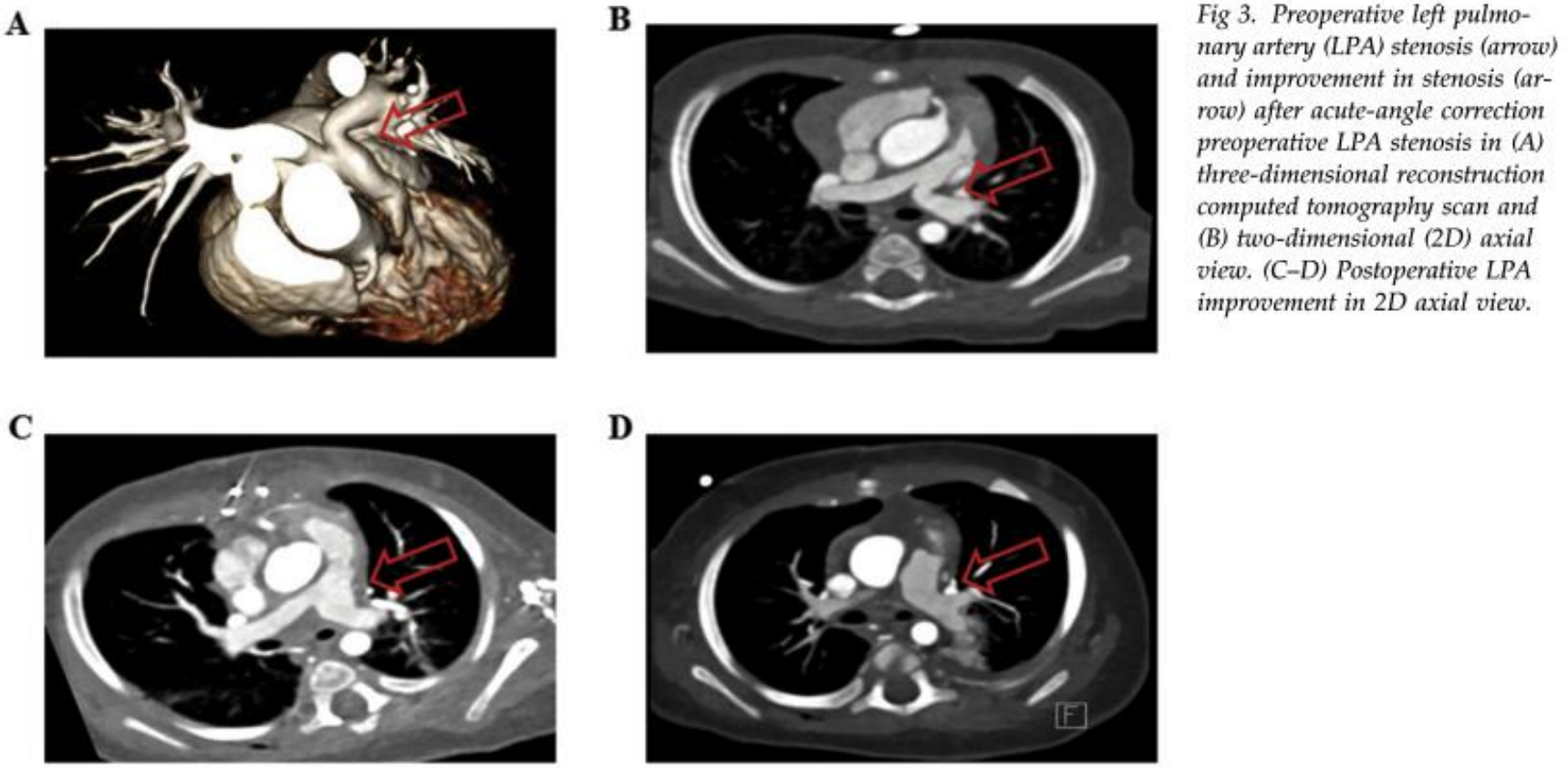

Fig 3. Preoperative left pulmonary artery (LPA) stenosis (arrow) and improvement in stenosis (arrow) after acute-angle correction preoperative LPA stenosis in (A) three-dimensional reconstruction computed tomography scan and (B) two-dimensional (2D) axial view. (C–D) Postoperative LPA improvement in 2D axial view.

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Special topics in surgical management

- Hypoplastic PA
  - McGoon ratio <1.2
  - Nakata index <70
  - Uncommon in the patients with TOF with PS
  - Hypoplasia is most likely to be a result of underperfusion of the PAs
  - Prompt enlargement can be expected when pressure and flow are restored
  - Intraop RV pr
    - ASD creation or large perforation in the VSD patch

- Use of monocusp valve

Functions transiently at best

Bigras et al. no significant differences in the degree of early postoprative PR or in clinical outcomes (JTCS 1966;112:33)

Gundry et al. 16 of 19 patients had competent monocusp valves immediately after operation, but only one of 7 patients had a competent valve by 24 months postoperatively (JTCS 1994;107:908)

If extensive reconstruction for the branch pulmonary arteries is required or if there is distal disease of the pulmonary vasculature, inclusion of a monocusp in the repair may improve hemodynamics in the immediate postoperative state.

Surgical Results of Monocusp Implantation with Transannular Patch Angioplasty in Tetralogy of Fallot Repair

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Department of Thoracic and Cardiovascular Surgery, Kyungpook National University Hospital, Kyungpook National University School of Medicine

Background: Monocusp reconstruction with a transannular patch (TAP) results in early improvement because it relieves residual volume hypertension during the immediate postoperative period. However, few reports have assessed the long-term surgical outcomes of this procedure. The purpose of the present study was to evaluate the mid-term surgical outcomes of tetralogy of Fallot (TOF) repair using monocusp reconstruction with a TAP. **Methods:** Between March 2000 and March 2009, 36 patients with a TOF received a TAP. A TAP with monocusp reconstruction (group I) was used in 25 patients and a TAP without monocusp reconstruction (group II) was used in 11 patients. We evaluated hemodynamic parameters using echocardiography during the follow-up period in both groups. **Results:** At the most recent follow-up echocardiography (mean follow-up, 8.2 years), the mean pulmonary valve velocities of the patients in group I and group II were 2.1±1.0 m/sec and 0.9±0.9 m/sec, respectively (p=0.001). Although the incidence of grade 3–4 pulmonary regurgitation (PR) was not significantly different between the two groups (group I: 16 patients, 64.0%; group II: 7 patients, 70.0%; p=0.735) during the follow-up period, the interval between the treatment and the incidence of PR aggravation was longer in group I than in group II (group I: 6.5±3.4 years; group II: 3.8±2.2 years; p=0.037). **Conclusion:** Monocusp reconstruction with a TAP prolonged the interval between the initial treatment and grade 3–4 PR aggravation. Patients who received a TAP with monocusp reconstruction to repair TOF were not to progress to pulmonary stenosis during the follow-up period as those who received a TAP without monocusp reconstruction.

Key words: 1. Tetralogy of Fallot
2. Transannular patch
3. Monocusp reconstruction

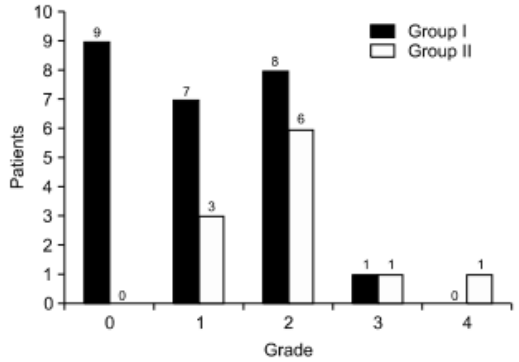


Fig. 1. Pulmonary regurgitation grade during the immediate postoperative period. No significant difference was found between group I and group II in immediate follow-up echocardiography (p=0.108).

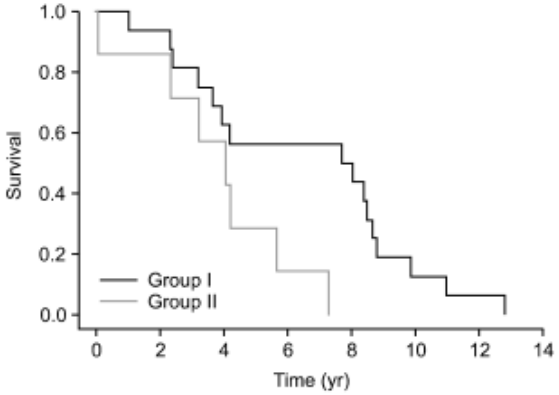
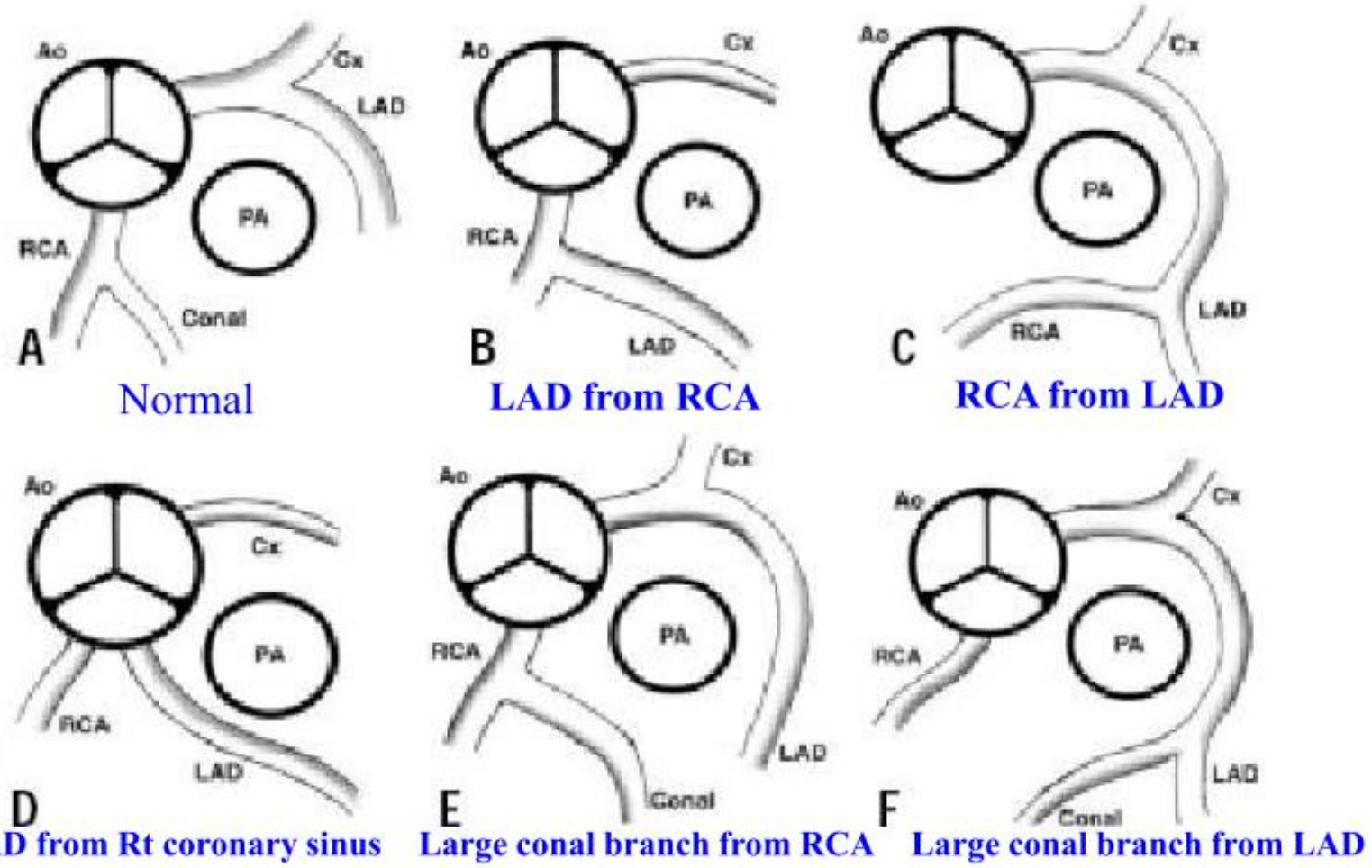
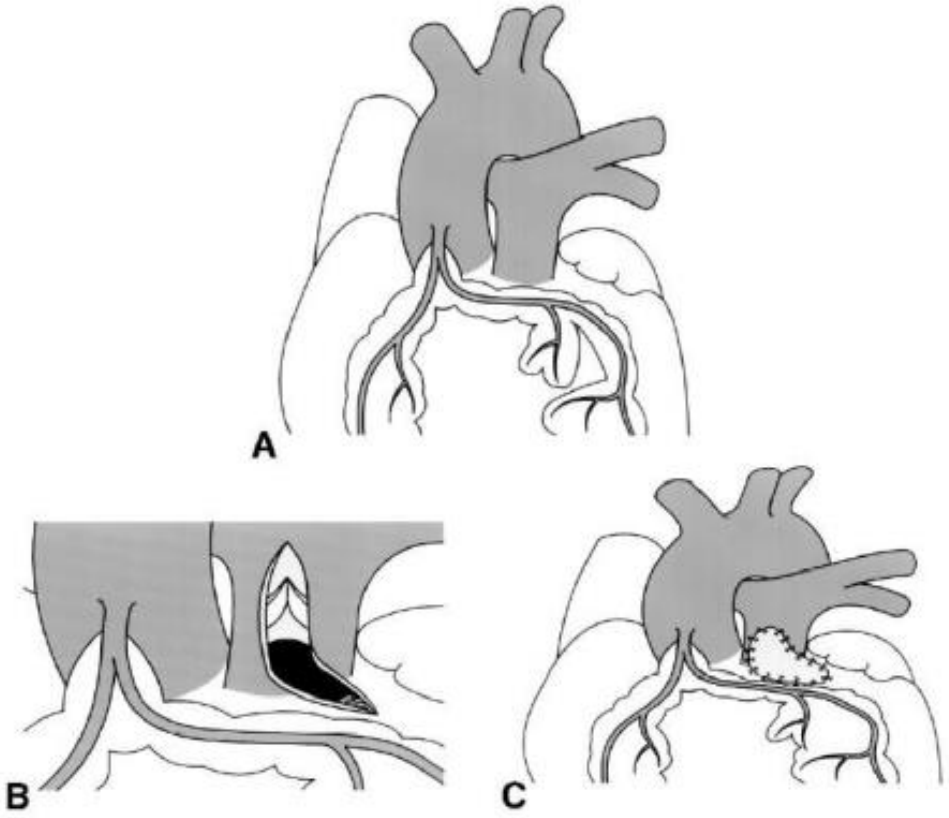


Fig. 3. Comparison of the extent to which patients in each group were free from the aggravation of PR. More patients in group I were free from grade 3–4 PR than in group II. PR, pulmonary regurgitation. Group I: 3 years, 81.3%±9.8%; 5 years, 56.3%±12.4%; 10 years, 43.8%±12.4%; group II: 3 years, 71.4%±17.1%; 5 years, 42.9%±18.7%; p=0.025.

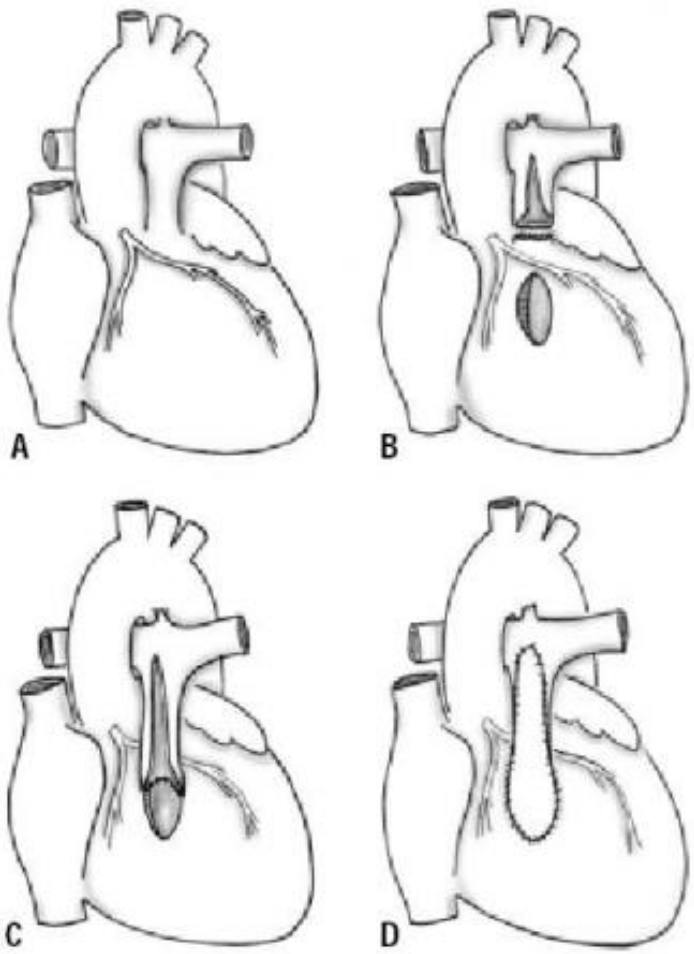
- Anomalous coronary artery crossing RVOT



- Surgical management
  - Transatrial and transpulmonary approach
  - Incision through MPA and PA annulus and to left of anomalous LAD
  - RVOT patching under the anomalous coronary artery
  - Conduit reconstruction
  - Translocation of the PA to a distal ventriculotomy
  - Proximally based PA flap-double outlet technique

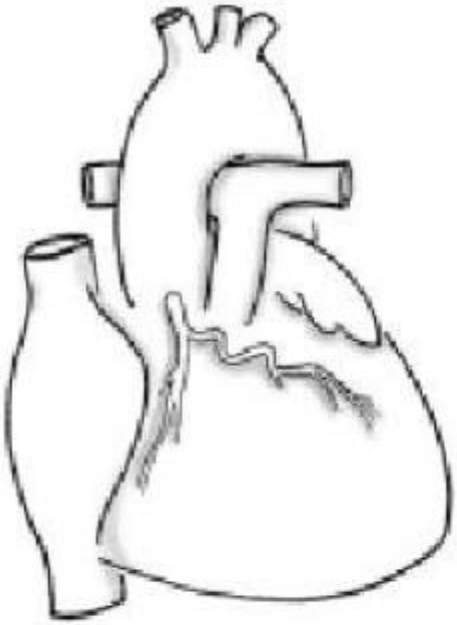


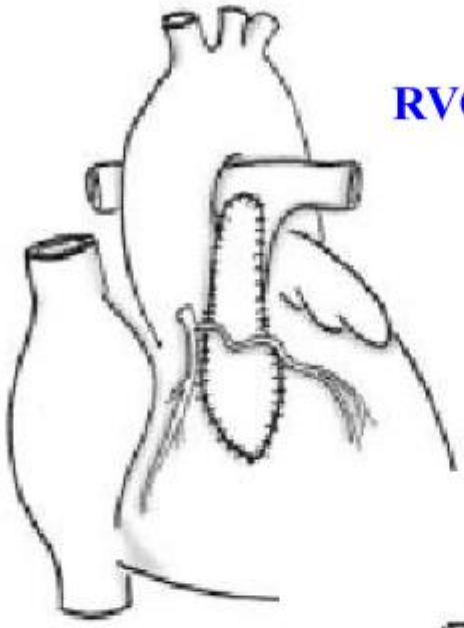
Incision to left of anomalous LAD

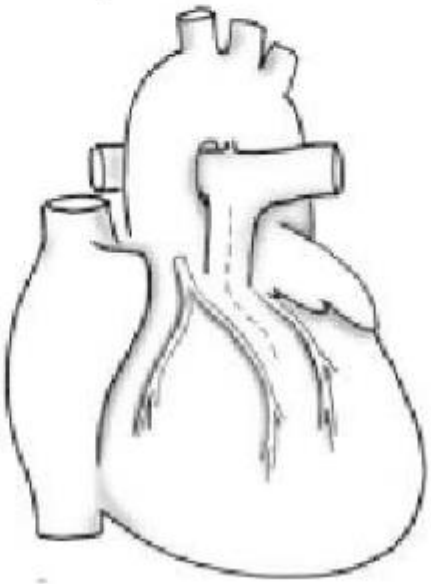


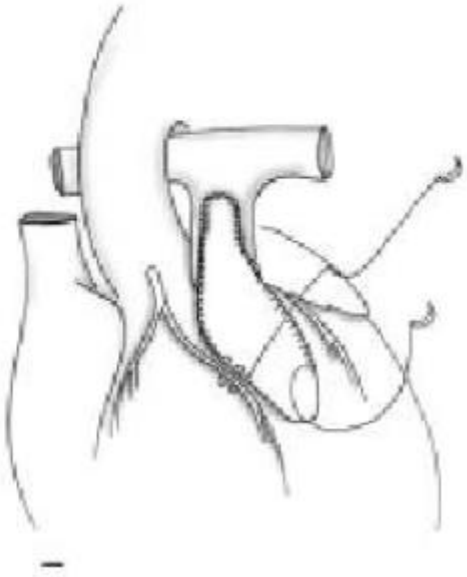
Translocation of MPA to a ventriculotomy

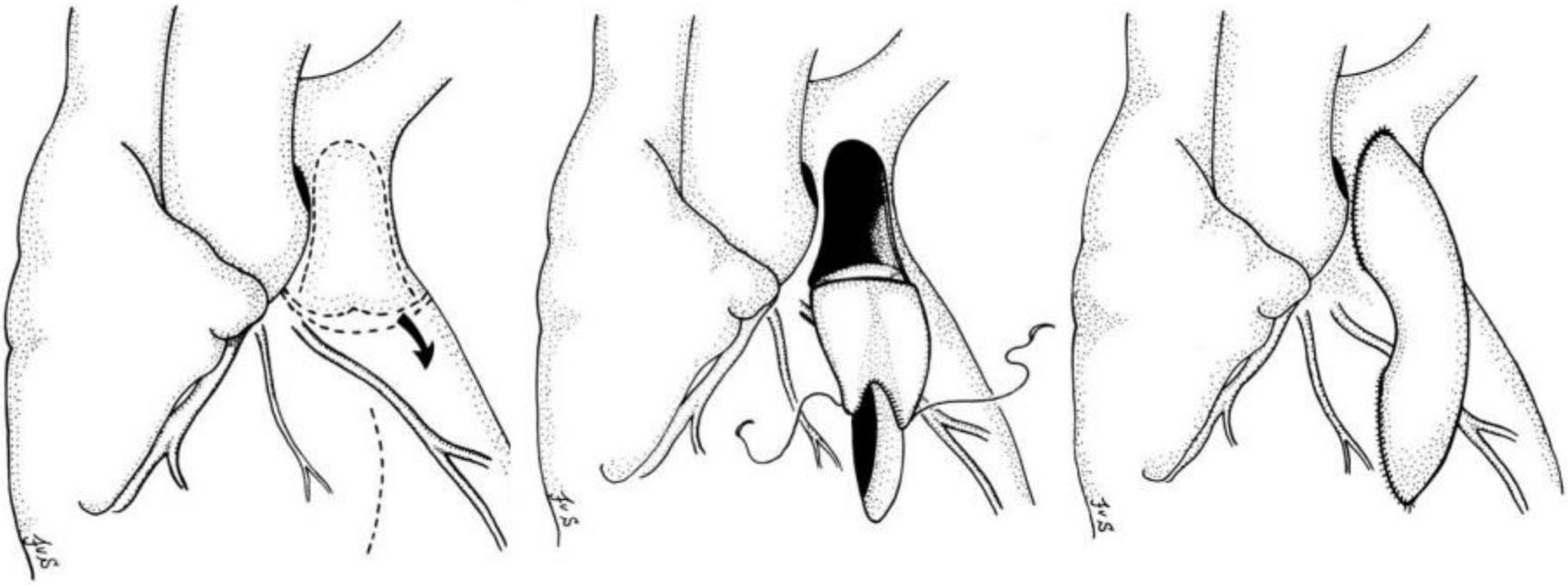
RVOT patching under the anomalous CA











Pulmonary artery flap - double outlet technique

Results of reoperation

- RVOTO
- PR
- Residual VSD

- Main causes of cardiac death in long-term survivors
  - Sudden death (Arrhythmia)
  - HF

- PVR
  - 2% of patients at 10-years FU and 12% after 20 years
  - Risk factors
    - TAP (large RVOT patch)
    - Peripheral PA stenosis
    - Residual VSD
    - Duration after TOF repair
    - Early repair or late repair?

- Indication for PVR
  - Symptomatic (decreased exercise tolerance, decreased functional class)
  - Progressive RV enlargement and RV dysfunction
  - LV dysfunction by RV
  - Increasing TR
  - Ventricular or supraventricular arrhythmia
  - RVEDV > 150 ml/㎡

- Valve choice at PVR
  - Bioprosthesis (bovine vs porcine)
  - Mechanical valve
    - When anticoagulation is required
      - Mechanical valve at aortic or mitral position
      - Afib
    - Repaid degeneration of previous tissue valve
    - Previous multiple operation

Arrhythmias

- Follow more than 20 years after TOF
  - 2~4% for Afib/Flutter
  - 3~4% for sustained VT
  - 2~4% for sudden cardiac death
- Holter monitoring
  - 19% for sustained VT
  - 23% for Afib/Flutter
- Risk factors
  - Elevated RV volume and pressure
  - Decreased RV or LV EF
  - PR
  - RVOT aneurysm