전공의 연수강좌
2025.09.25

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Tetralogy of Fallot

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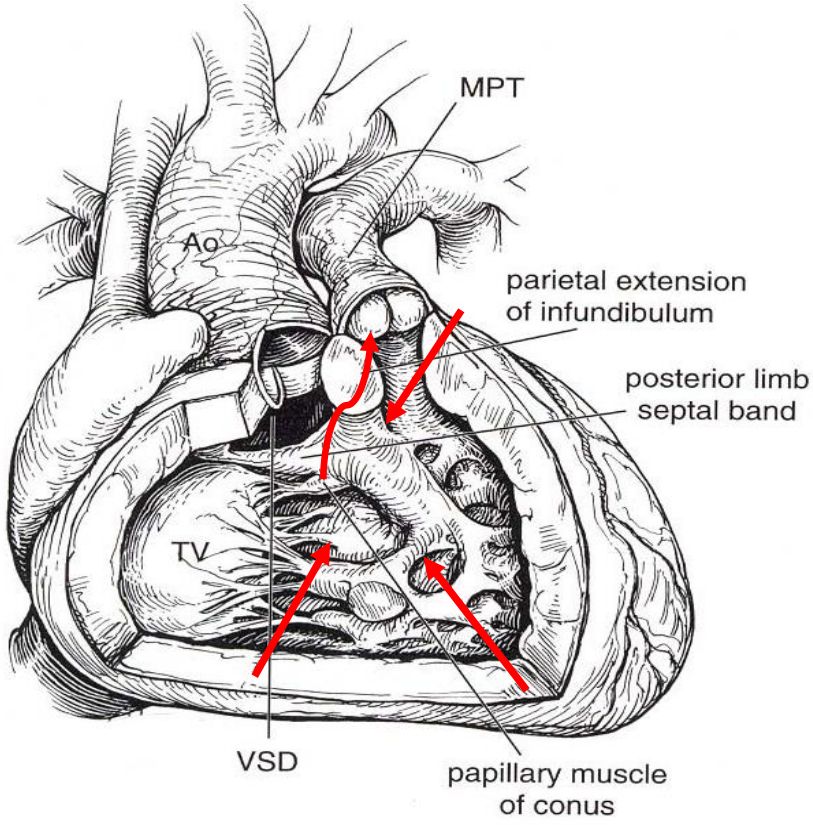
Definition

- Classic theory
  - Unequal spirial septation of conotruncus

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

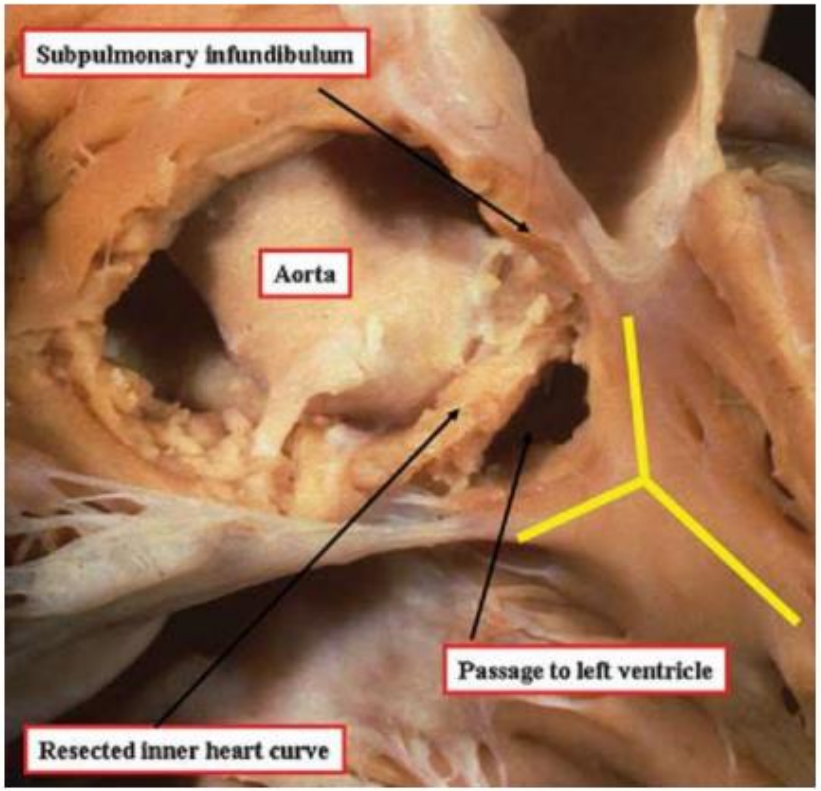


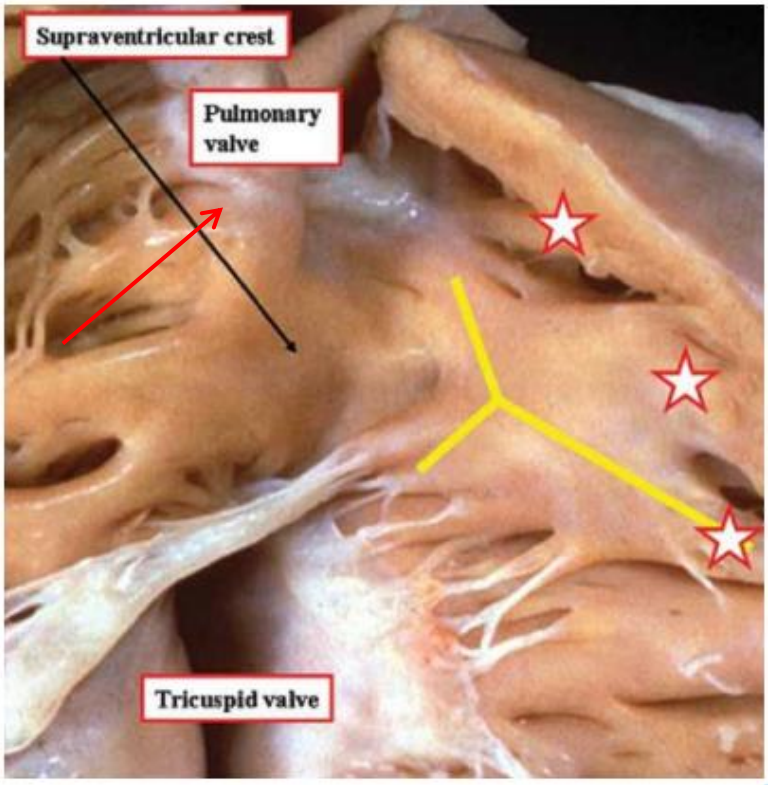
Anterior and leftward displacement of the infundibular (conal) septum

- ROVTO & RVH
- Large VSD
- Overriding of aorta

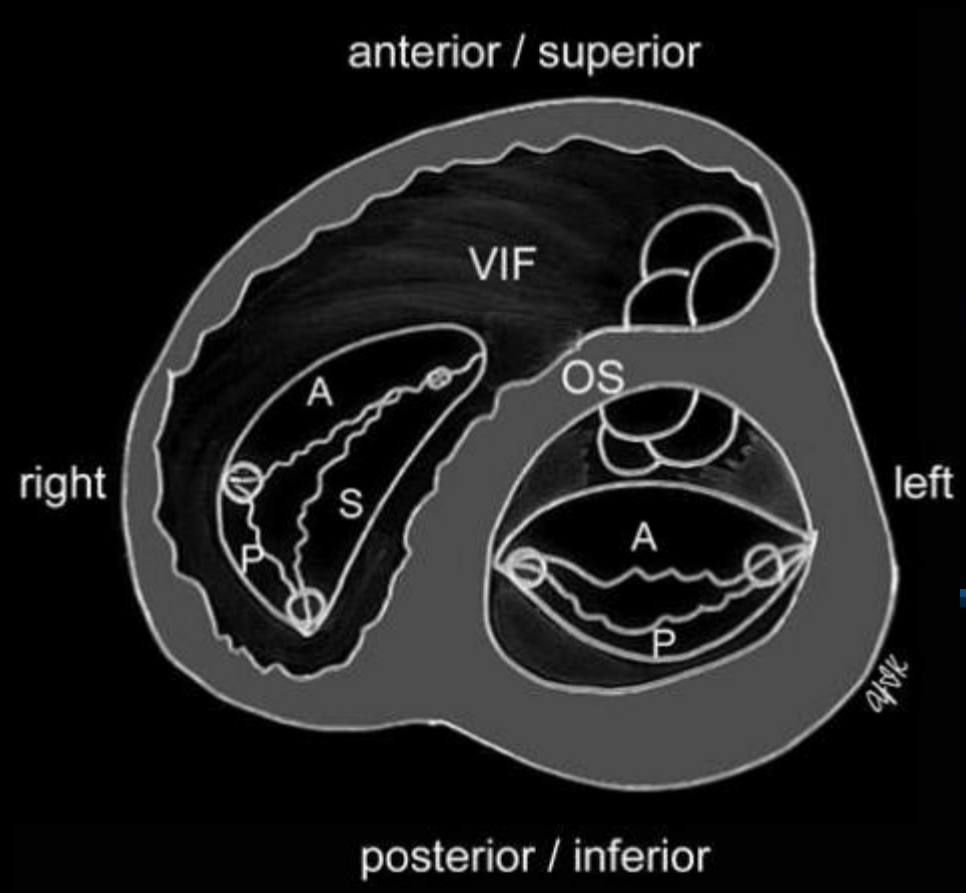
Normal Heart

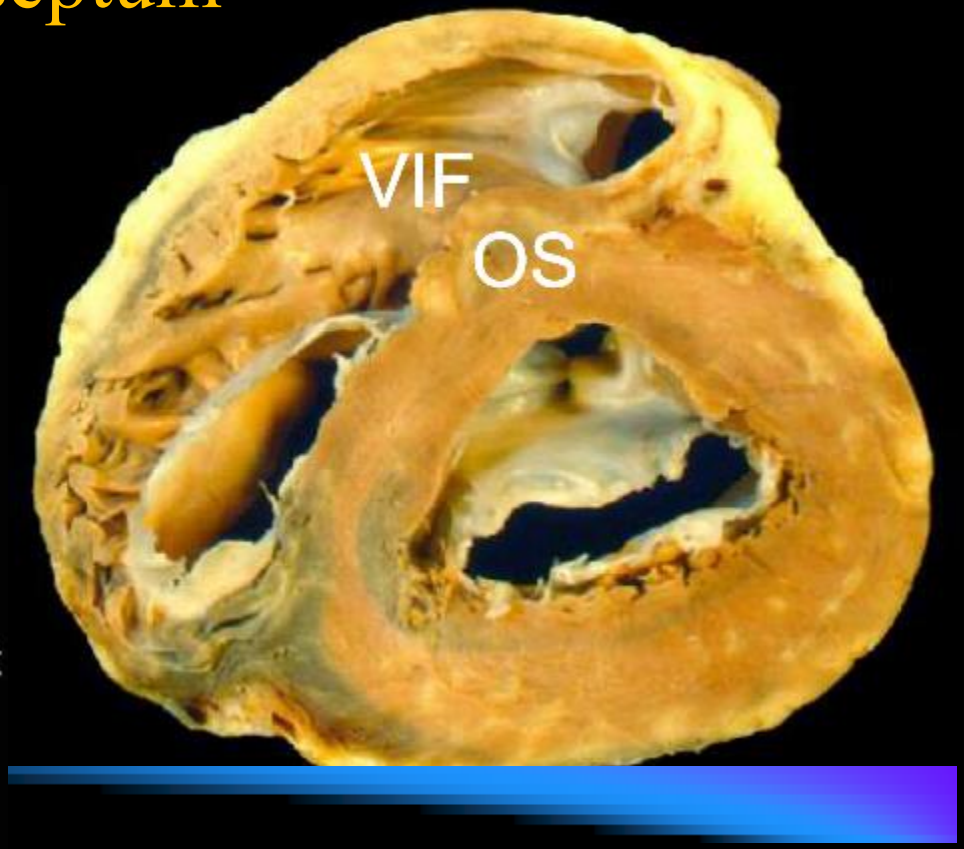
Crista supraventricularis (Supraventricular crest)
Muscular area separating the attachments of the TV and PV in the roof of the RV

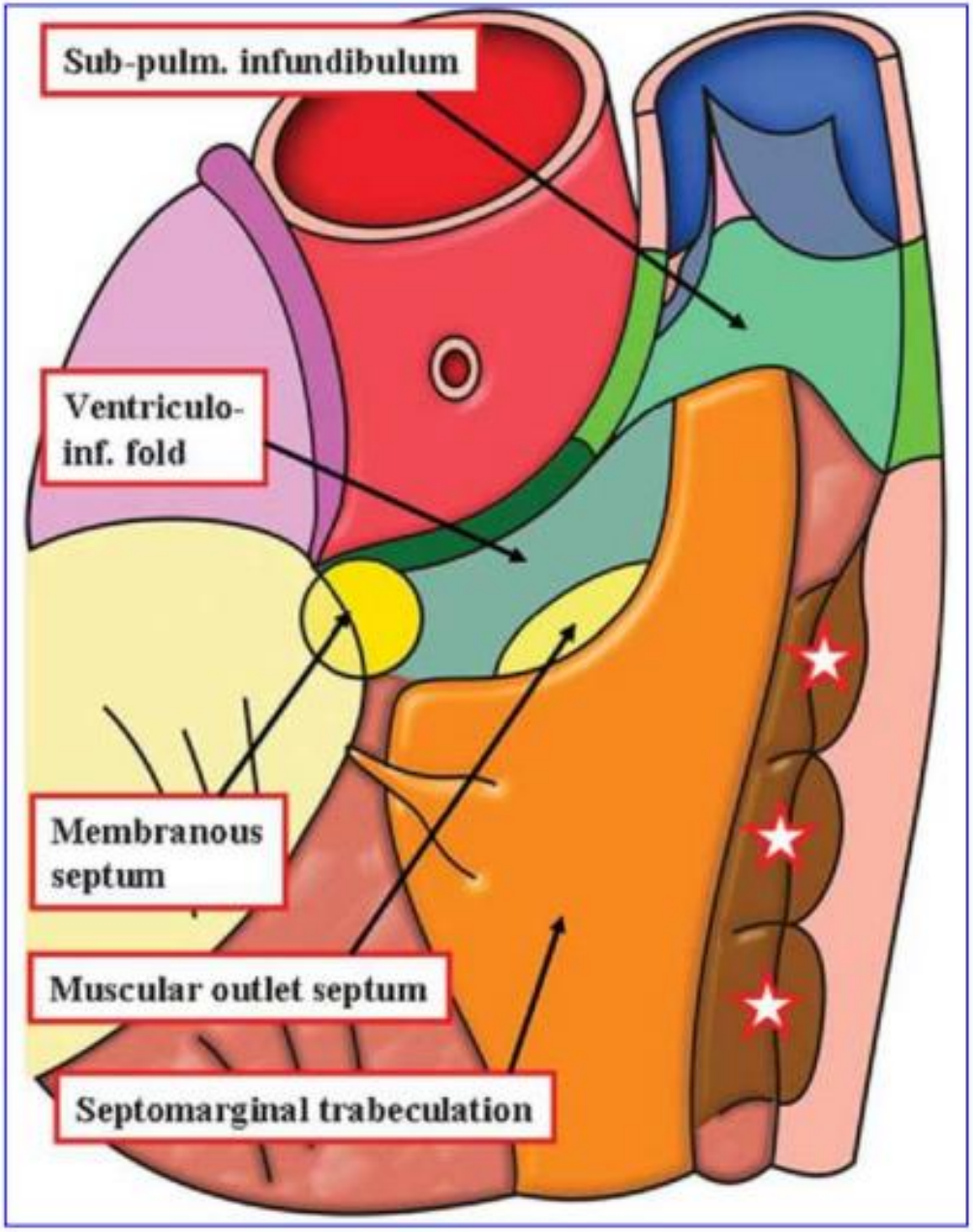




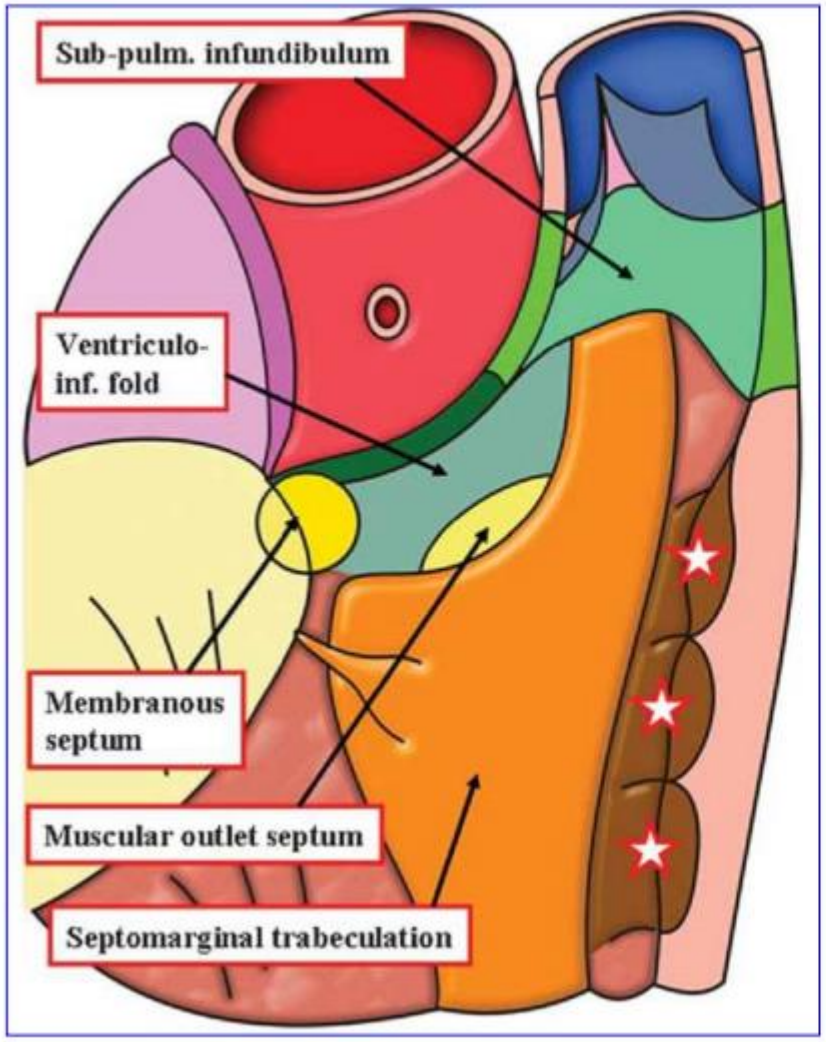
Crista supraventricularis: ventriculoinfundibular fold (VIF) + outlet or infundubular septum



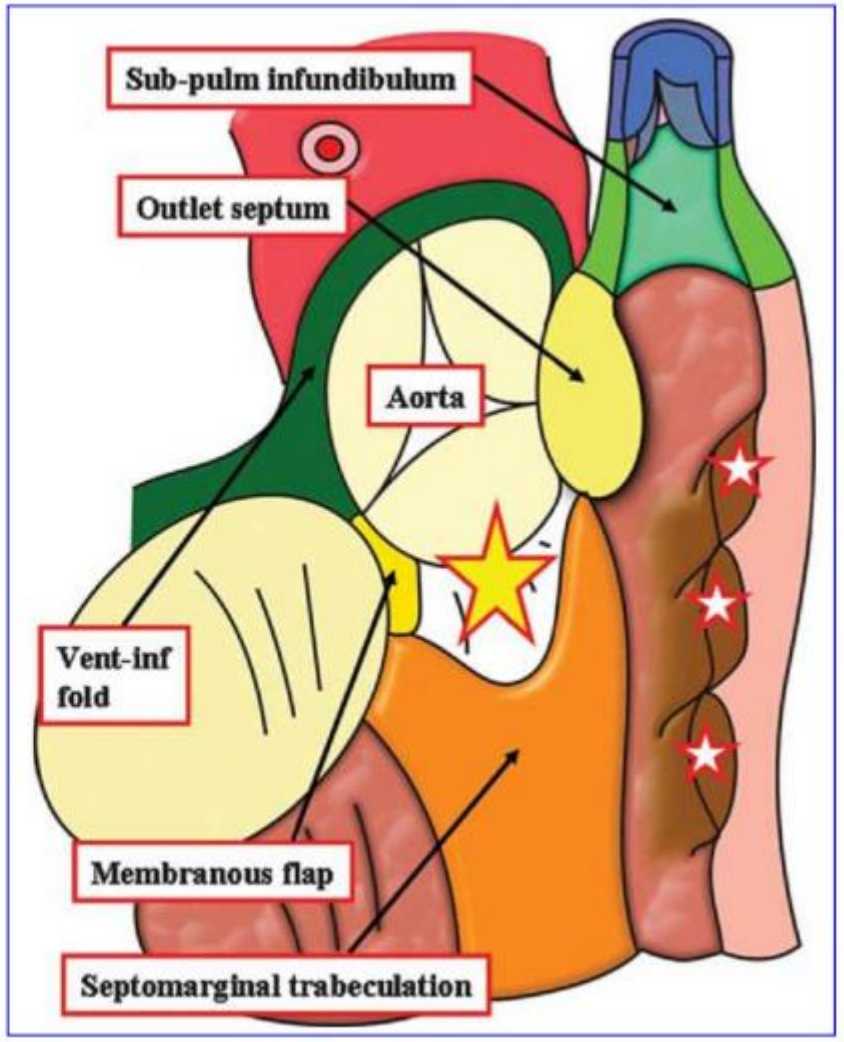



Sub-pulm. infundibulum
Ventriculo-inf. fold
Membranous septum
Muscular outlet septum
Septomarginal trabeculation

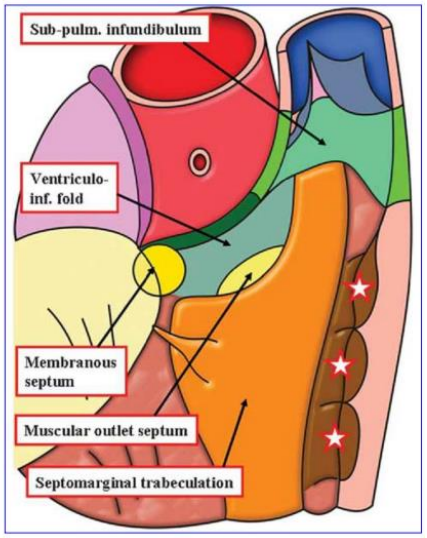
Normal


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

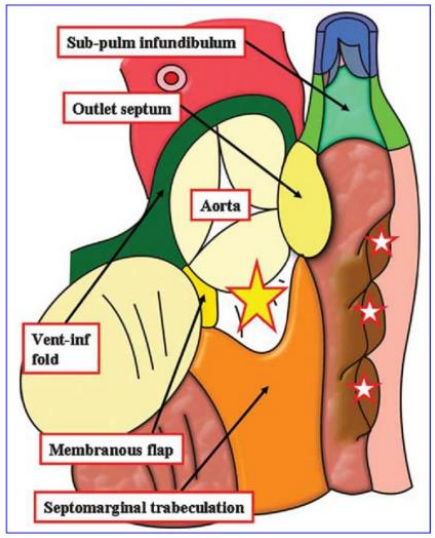
TOF


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

Normal



TOF

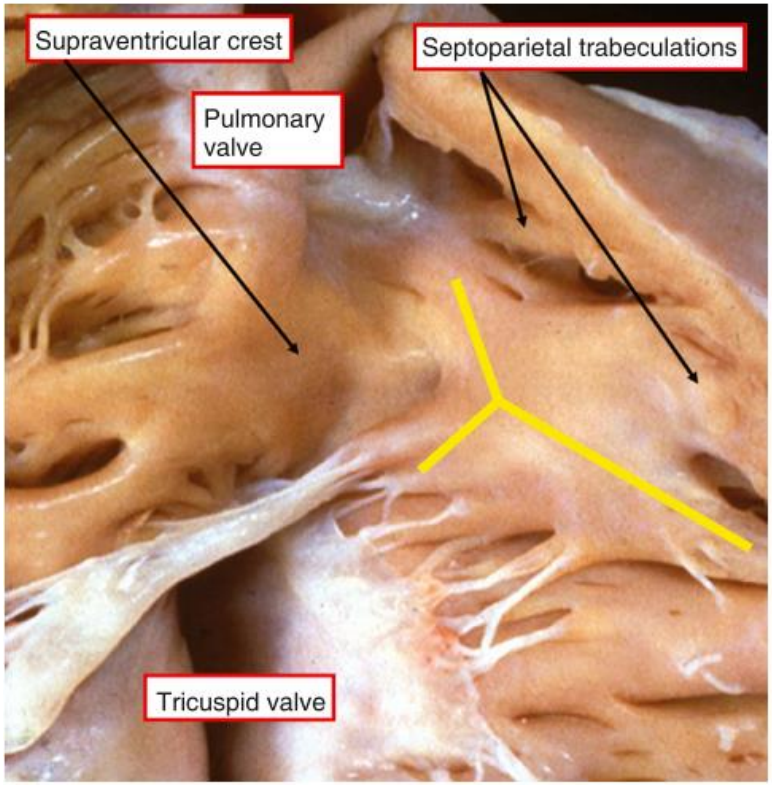


 # Muscular outlet septum:

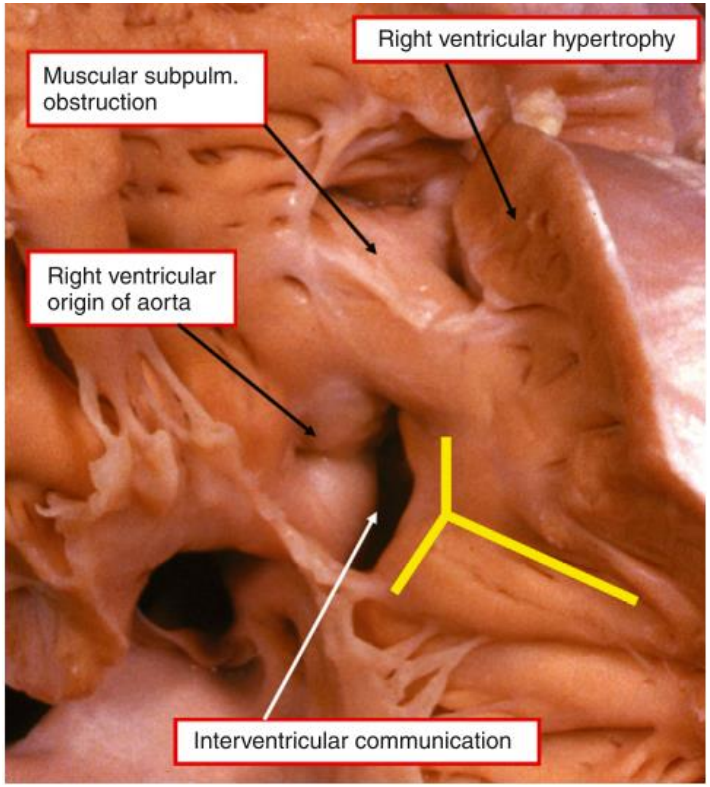
- Insignificant structure, inserted and buried between the limbs of SMT
- Not possible to distinguish
- **VIF** (part of Supraventricular crest) -> support the subpulmonary infundibulum

- **Antero-cephalad deviation** of the insertion of the **muscular outlet septum** relatively to the limbs of the septomaginal trabeculation
- Supraventricular crest divorced one from the other, **Muscular outlet septum**, rather than VIF, **support the narrowed subpulmonary infundibulum**
- **VSD**: situated between TSM limb and VIF

Normal


Supraventricular crest
Septoparietal trabeculations
Pulmonary valve
Tricuspid valve

TOF


Right ventricular hypertrophy
Muscular subpulm. obstruction
Right ventricular origin of aorta
Interventricular communication

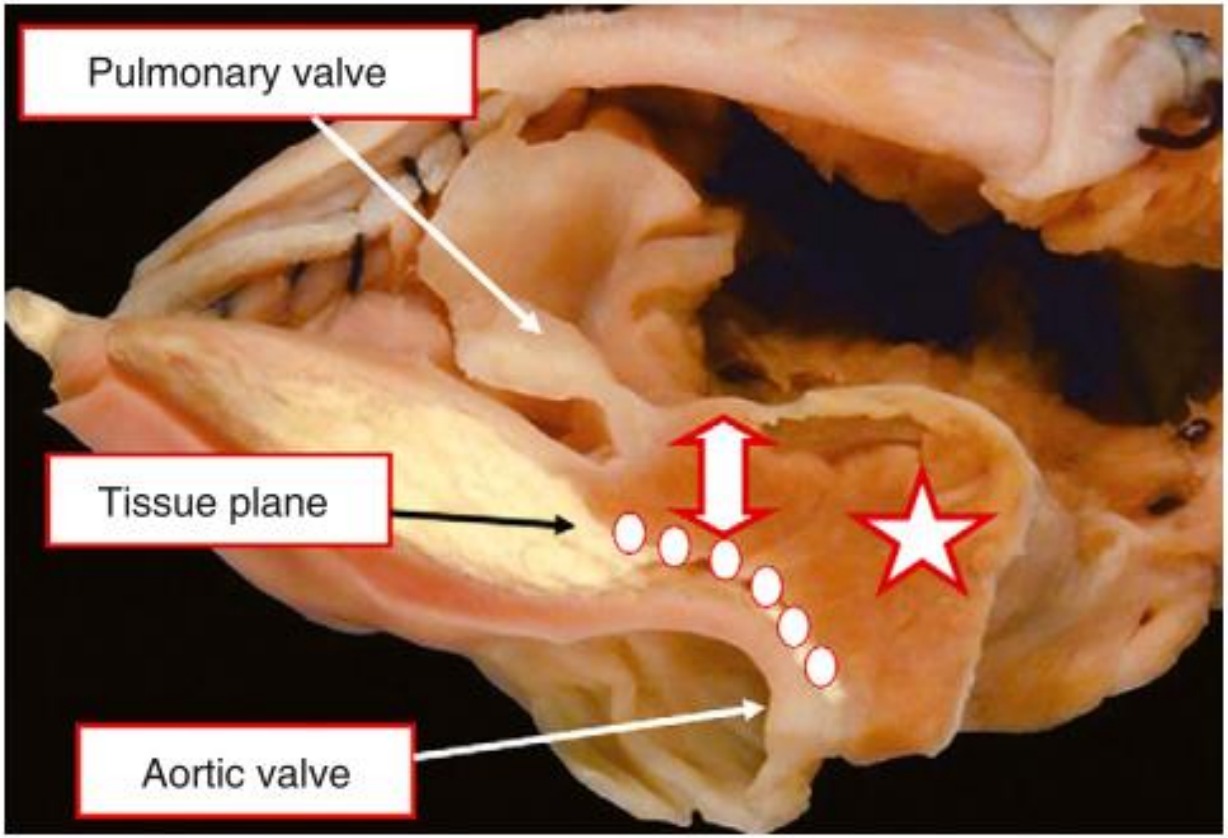
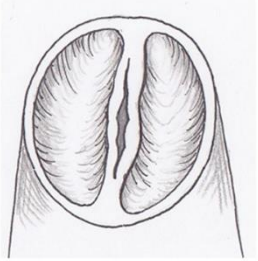
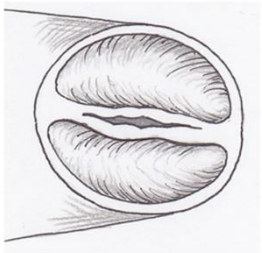


Figure 36-5 The adjacent parts of the subaortic and subpulmonary outlets have been removed from a heart with tetralogy of Fallot. The section shows how the narrowed subpulmonary infundibulum is made up of the outlet septum (*star*) and the free-standing infundibular sleeve (*double-headed arrow*). Note the tissue plane (*dots*) between the infundibulum and the aortic root.

Pulmonary valve and annulus

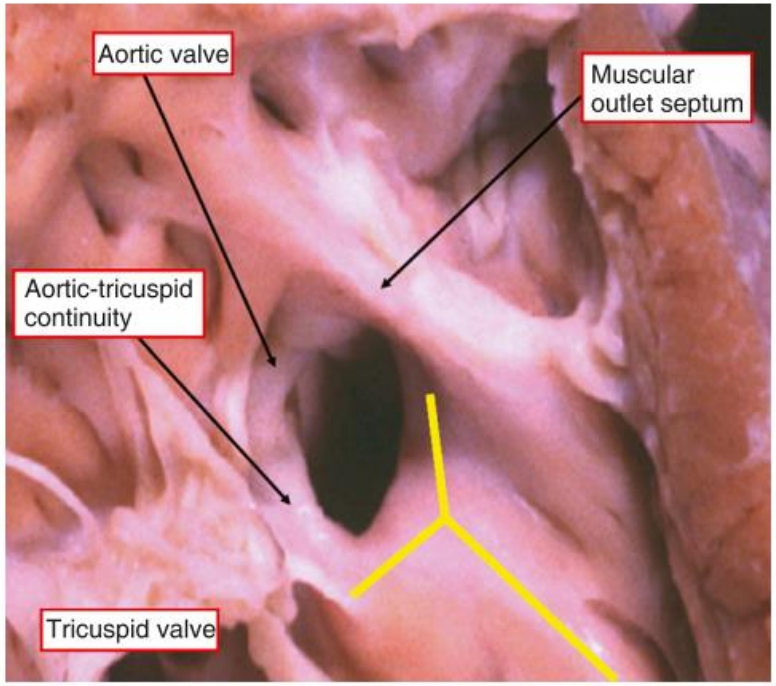
- Stenosis in 75%
- Leaflets
  - Thickened, tethered to the PA
  - Bicuspid in 75%
    - Monocusp, tricusp
  - Vertical or horizontal position



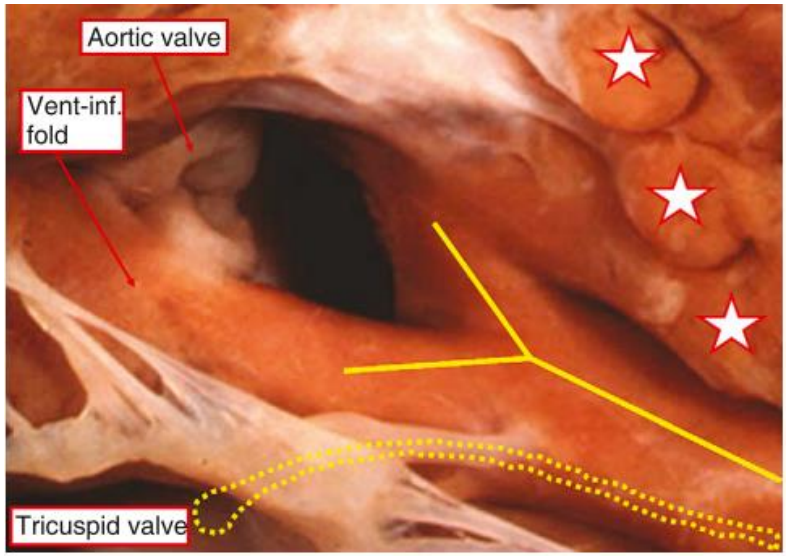


- MPA & branch PAs
  - Usually somewhat diffusely small, often short
  - Narrowest portion of **MPA** is often at STJ
  - Branch PA stenosis in 10%
    - **LPA os**

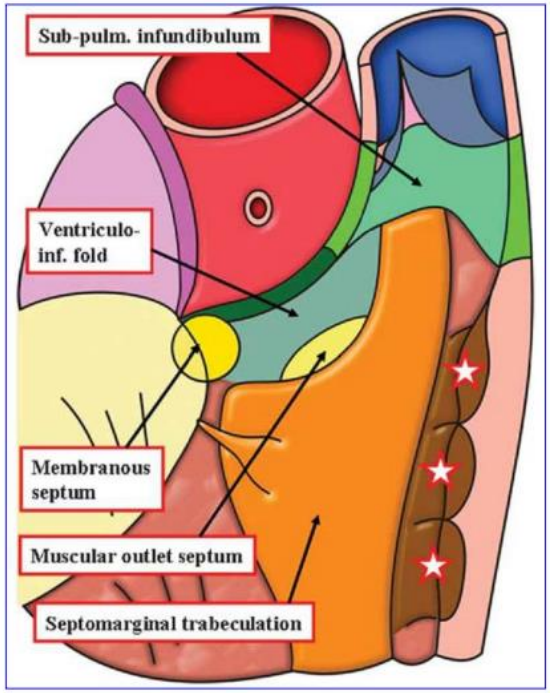
- **VSD**
  - Large anteriorly malaligned
  - 25%: VIF extends to the posterior limb of TSM
    - Muscle bar beneath TV (MO)
  - Additional VSDs in 3~15%

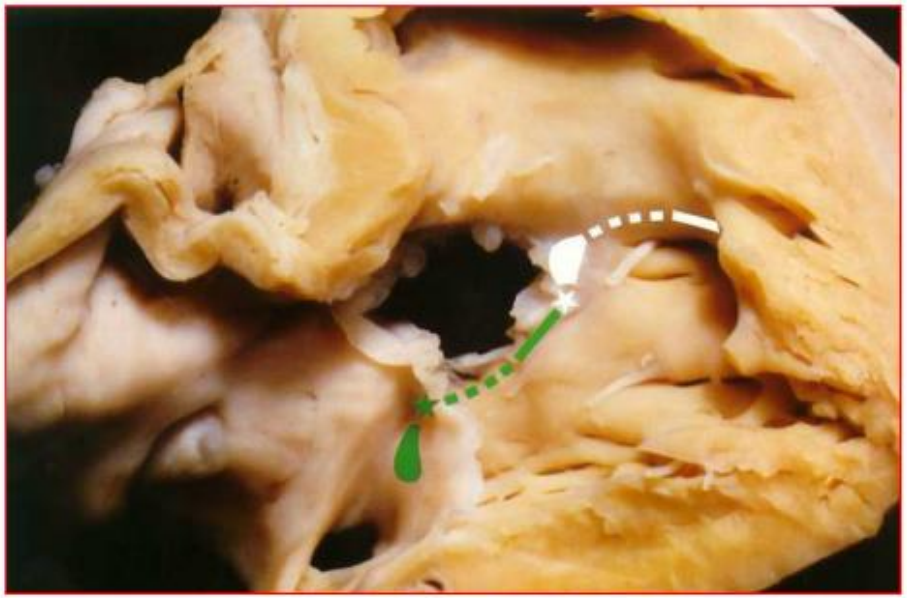

Aortic valve
Muscular outlet septum
Aortic-tricuspid continuity
Tricuspid valve

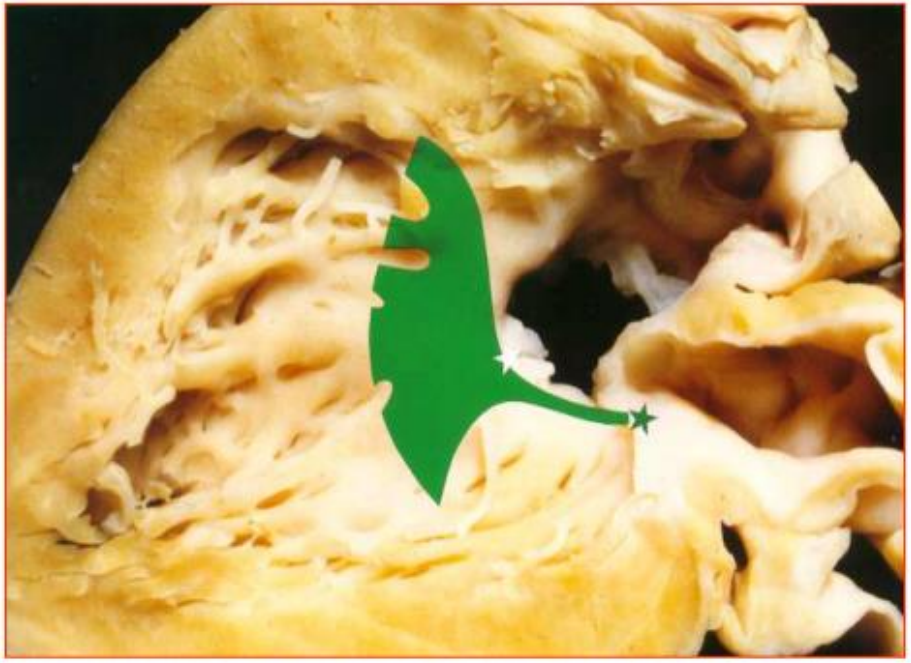
70~80%


Aortic valve
Vent-inf. fold
Tricuspid valve

20%

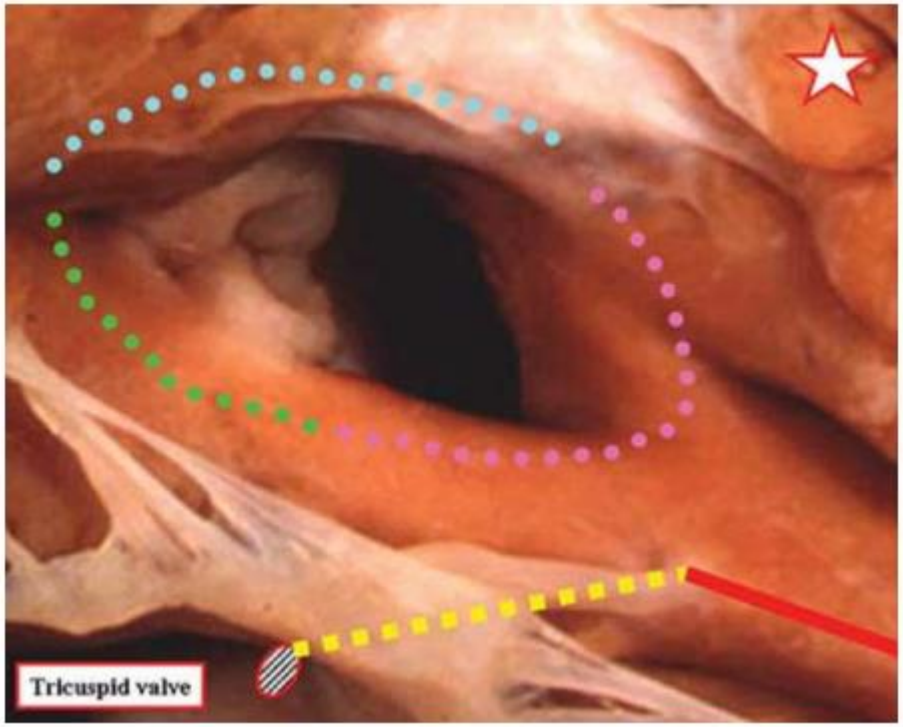

Sub-pulm. infundibulum
Ventriculo-inf. fold
Membranous septum
Muscular outlet septum
Septomarginal trabeculation





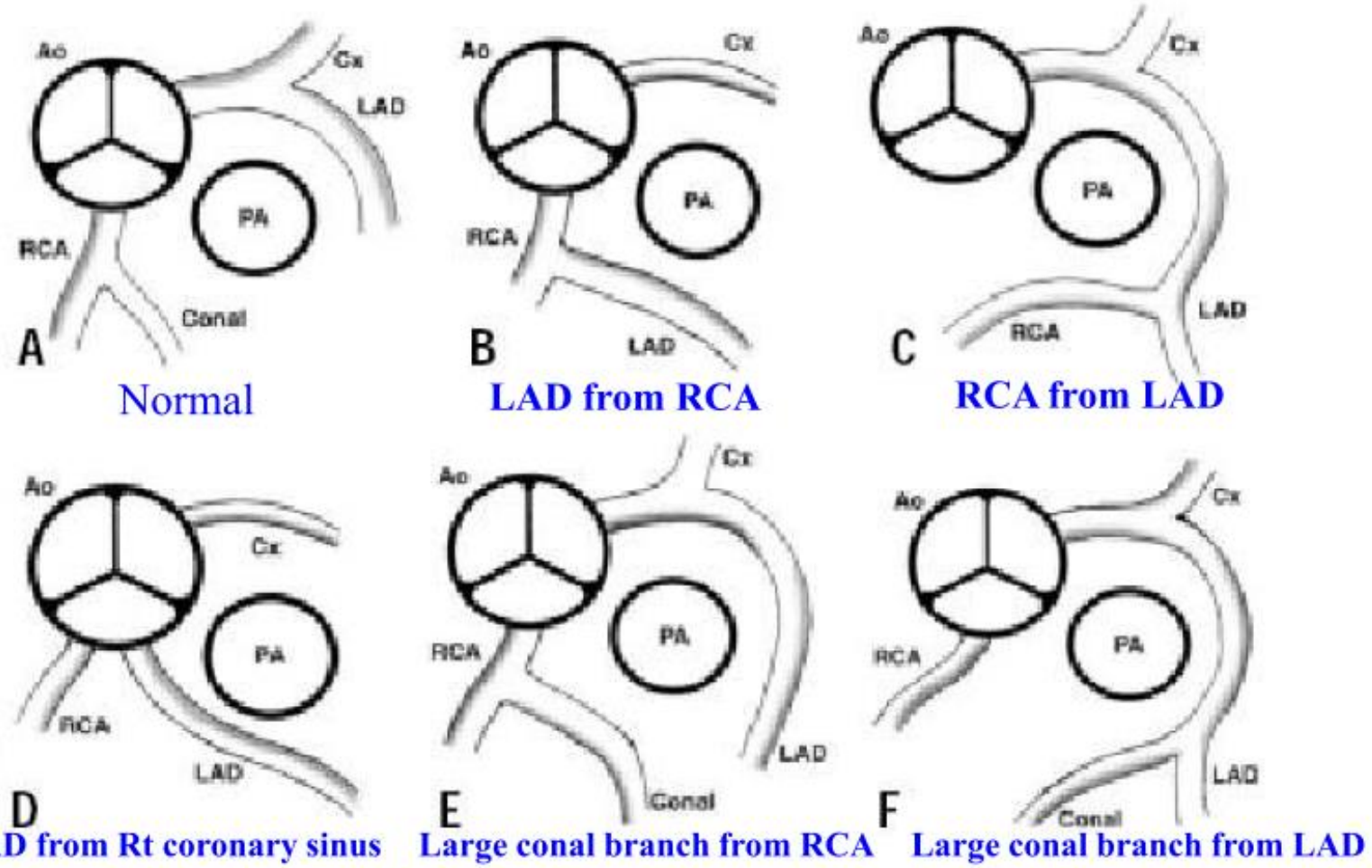
Conduction system

SA and AV node: normal in location



- 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
  - 25%, right aortic arch

- Anomalous coronary artery crossing RVOT



Indications and timing of surgery

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

- 6~12mo

Symptomatic neonates or young infants with TOF

- Shunt vs Early primary repair

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

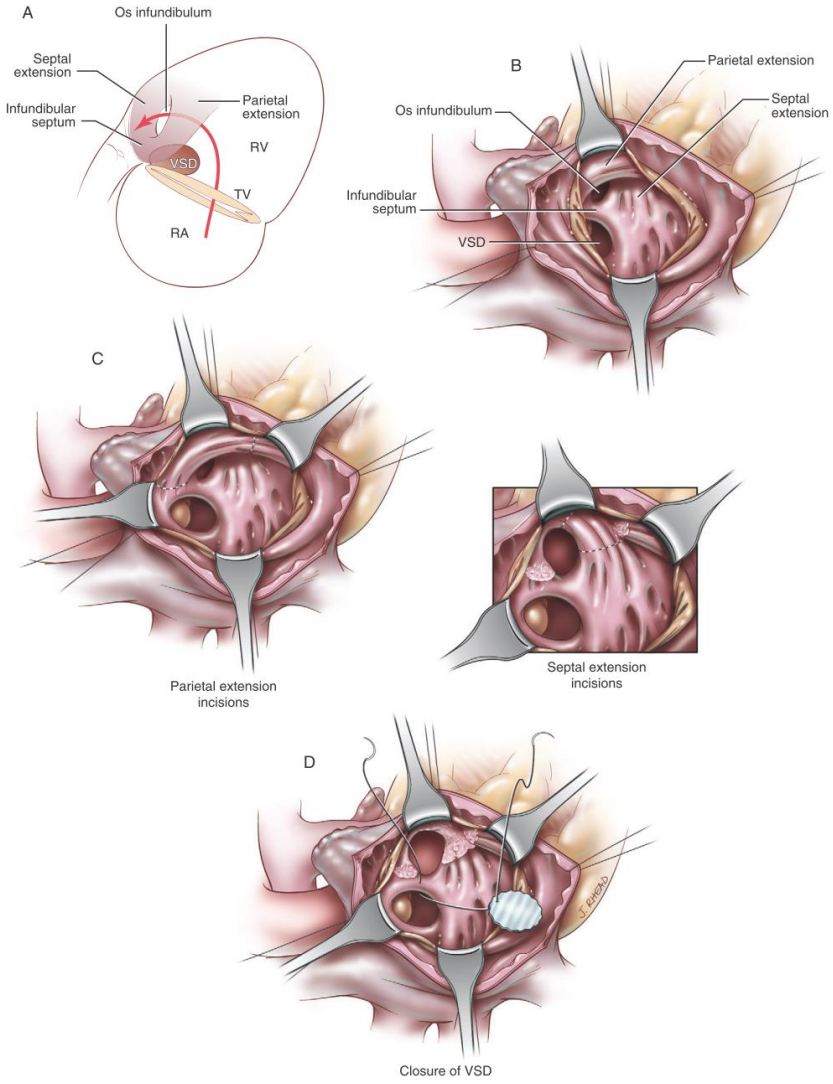
Symptomatic neonates or young infants with TOF

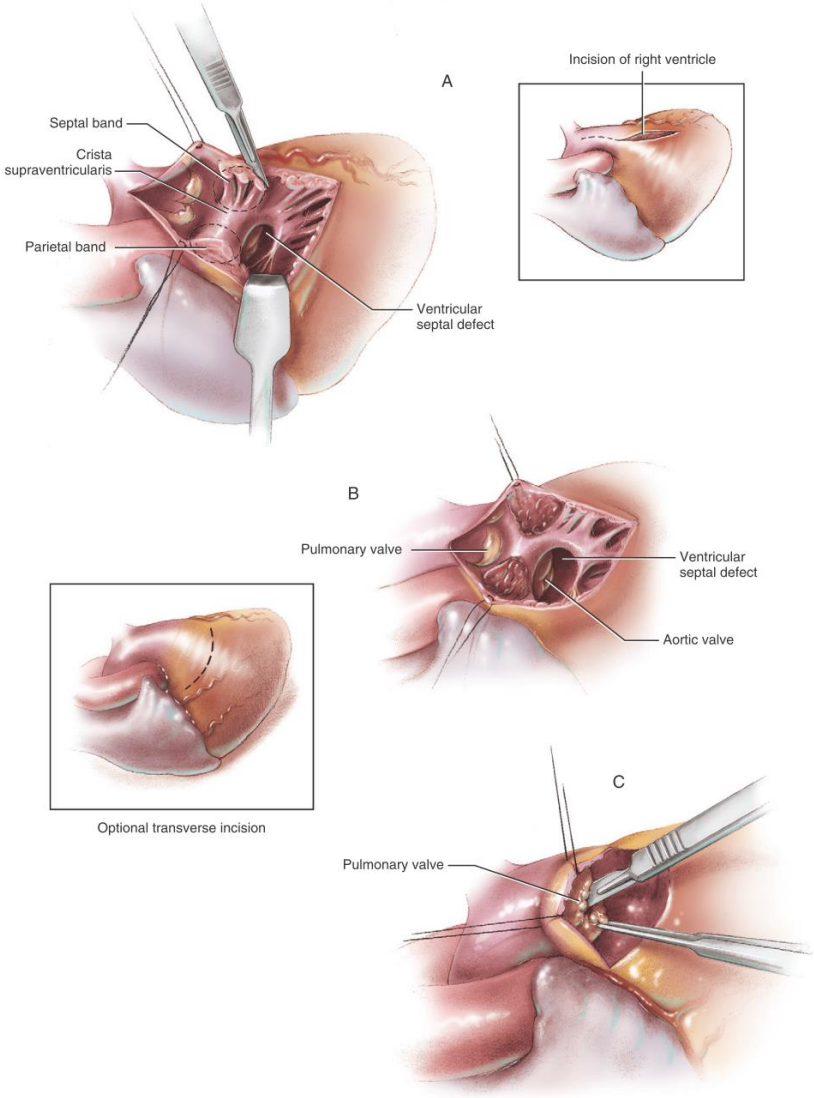
- Shunt vs Early primary repair

- **Potential disadvantage of early primary repair**
  - Frequent need of transannular patch
  - Neonatal myocardium may be **less capable of handling of RV volume load**
  - Adverse effects of **early bypass surgery** on the **neonatal brain**
  - Often complicated and lengthy postoperative recovery in small infants

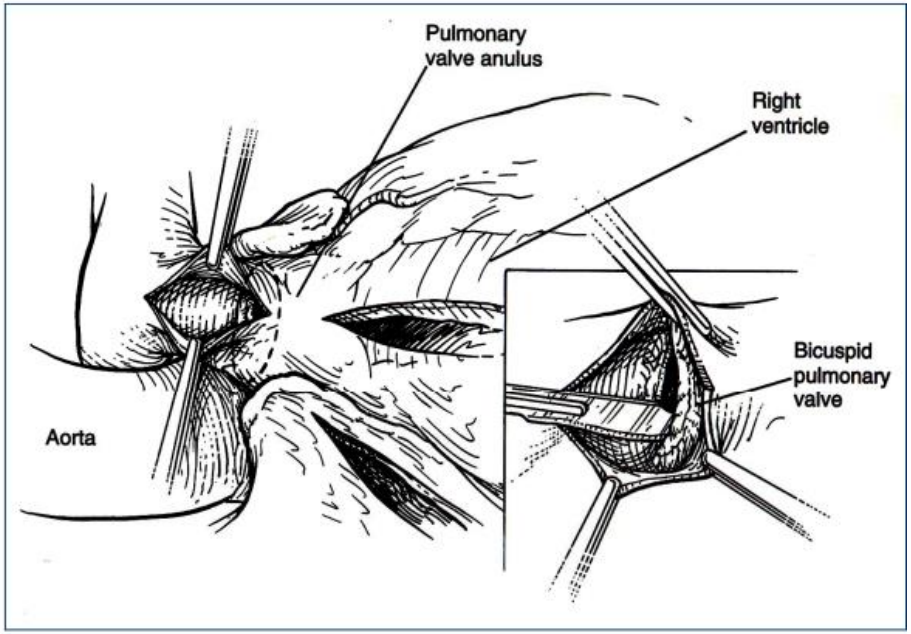
Surgical Management

VSD closure & RV muscle resection


A
Os infundibulum
Septal extension
Infundibular septum
Parietal extension
RV
VSD
TV
RA
B
Parietal extension
Os infundibulum
Septal extension
Infundibular septum
VSD
C
Parietal extension incisions
Septal extension incisions
D
Closure of VSD


Septal band
Crista supraventricularis
Parietal band
Ventricular septal defect
A
Incision of right ventricle
B
Pulmonary valve
Ventricular septal defect
Aortic valve
Optional transverse incision
C
Pulmonary valve

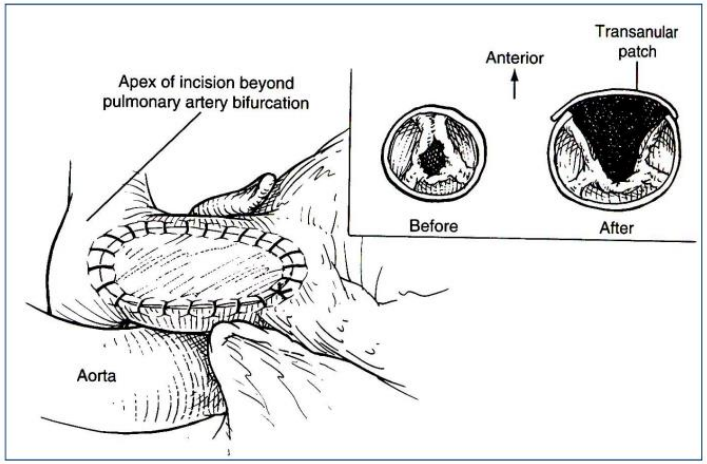
Pulmonary valvotomy



- **Commissurotomy**
- **Commissural mobilization** by excising the web-like structure around the commissures
- **Shaving** of the lumpy valve leaflet

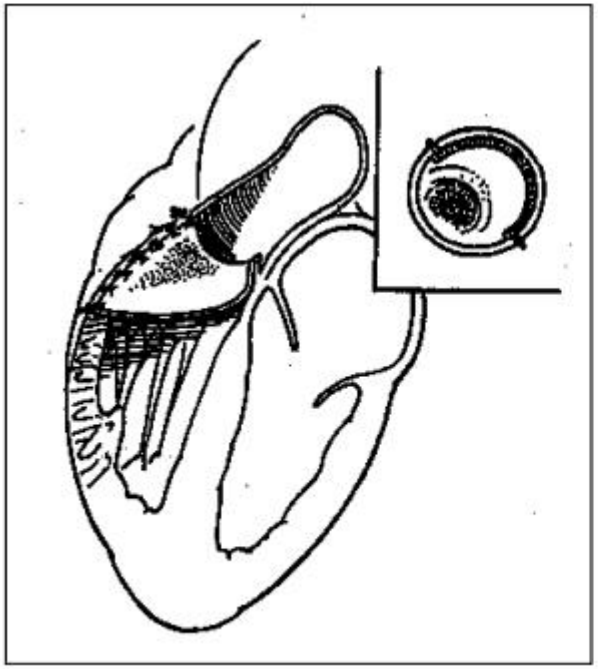
Transannular patch

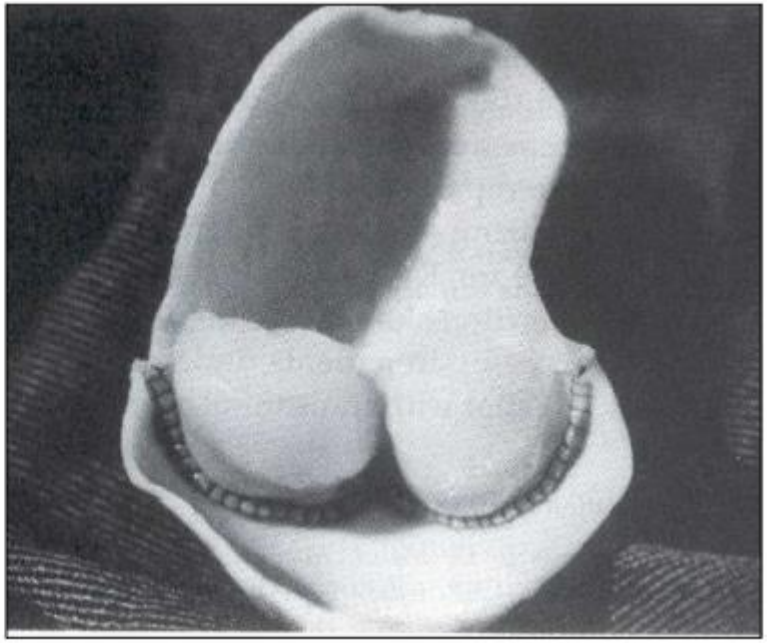
- Criteria for PV preservation
  - Z-value of PA >-3
  - Diameter of PA (mm) >0.8 mm/kg



- Post-repair RV/LV >0.7
  - If TAP has not been placed, TAP should be considered
  - If TAP has been place
    - Branch PA stenosis
    - Hypoplasia of peripheral PAs
    - Residual VSD
    - Residual infundibular stenosis

Monocups implantation





- Use of monocusp valve

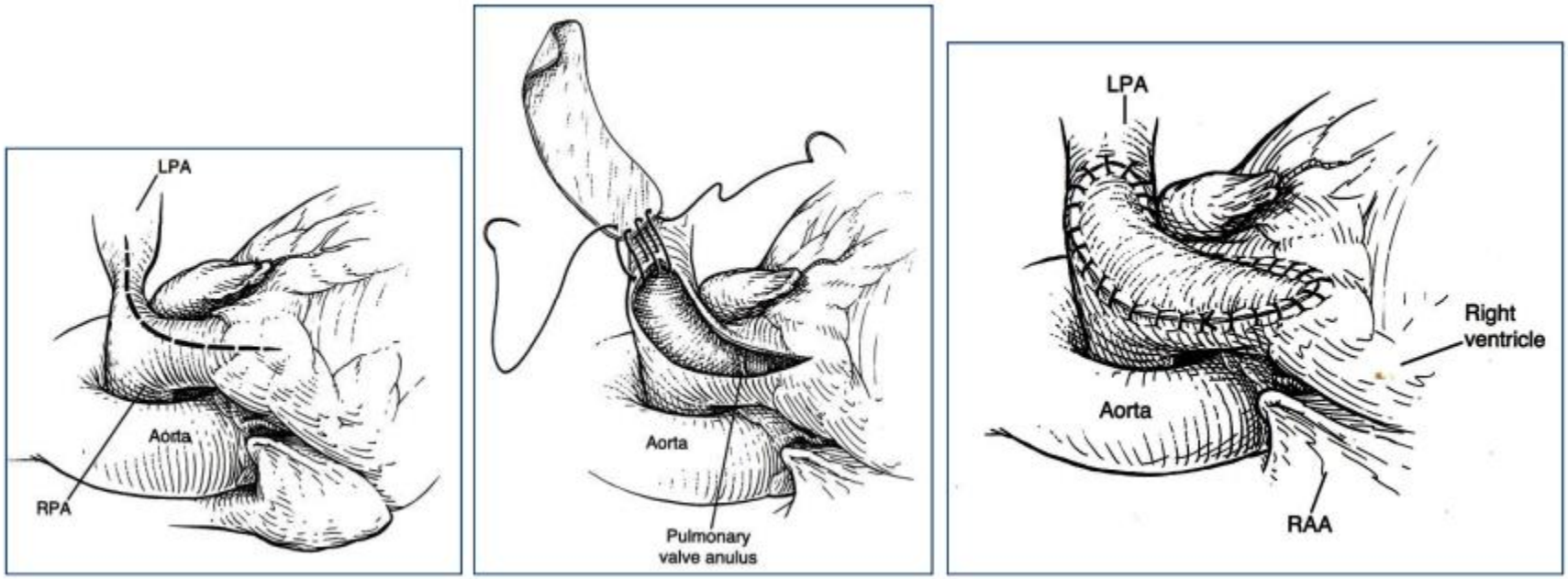
Functions transiently at best

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.

PA angioplasty



Causes of postoperative LPA stenosis

- Inadequate enlargement
- Aneurysmal dilatation of RVOT patch
- Kinking

Acute angle of LPA with or without stenosis

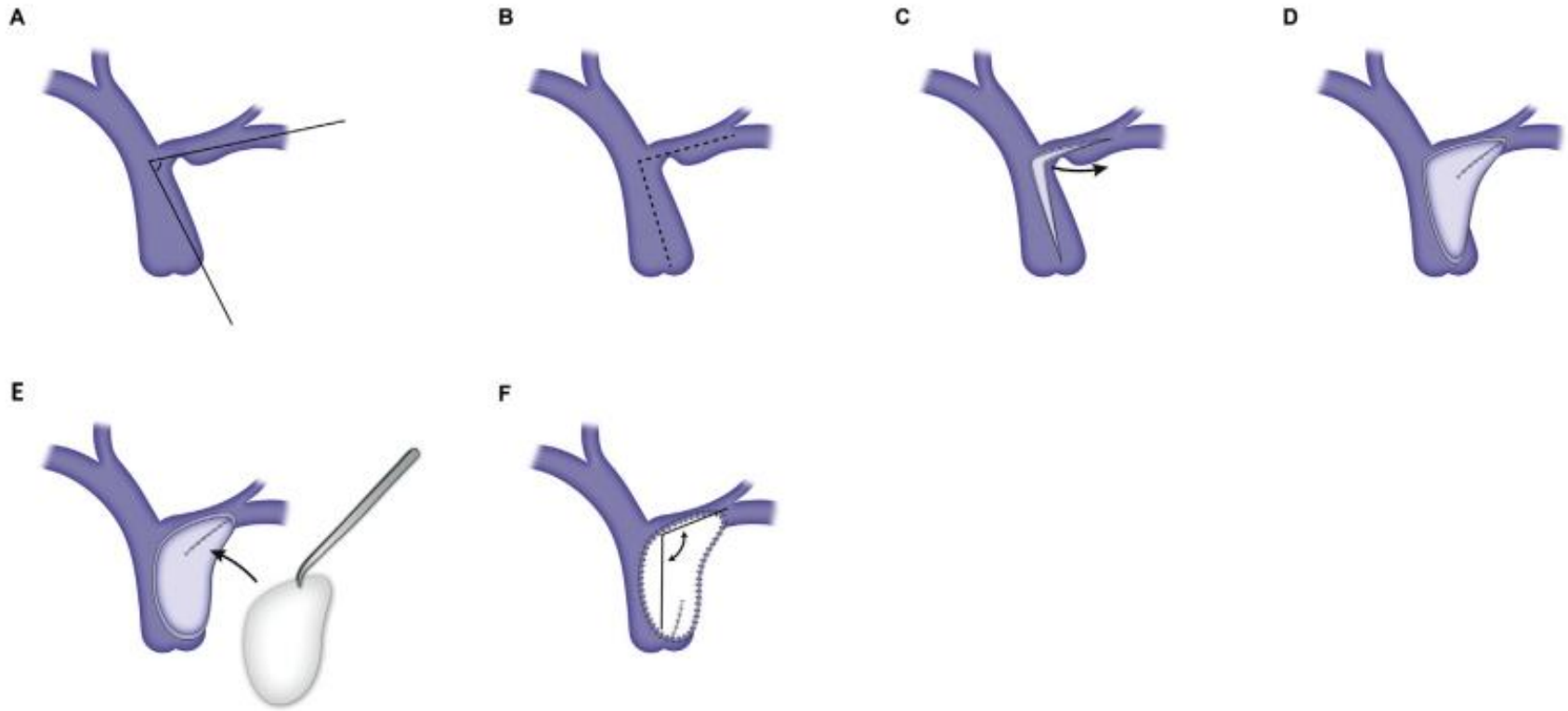


Fig 1. Schematic drawing of a left pulmonary artery (LPA) acute-angle correction angioplasty. (A–B) The main pulmonary artery (MPA)

Jang et al. Ann Thorac Surg 2017;103:862-8

Weaning from bypass

- **Residual RVOTO**
  - Less than 70~80% of systemic pressure
  - Dynamic obstruction of RV
    - Relative hypovolemia
    - Inotrope induced hypercontractility

Weaning from bypass

- **Residual VSD**
  - ↑↑LA pressure, systemic hypotension
  - ABGA at RA and PA
  - Undetected muscular VSD

Weaning from bypass

- **Residual VSD**
  - Poorly tolerated
    - **Peripheral PA**
      - Thin walled and distensible
      - Not elevated PVR
      - Very large L → R shunt effect with LV and RV volume overload and dilation
    - **Transannular patch and TR**
      - Exacerbate RV volume overload
      - Poorly tolerated in the setting of diastolic dysfunction
    - **Ventricle**
      - Adapted to a state of relative pressure and not volume overload prior to repair

Weaning from bypass

- Coronary obstruction
  - RVOT patch suture line close to coronary artery
  - Tension within the epicardium
    - Partial obstruction of the coronary artery
  - Use interrupted pledgetted sutures with the pledgets lying on the endocardial surface of the free wall

Thank you for your attention