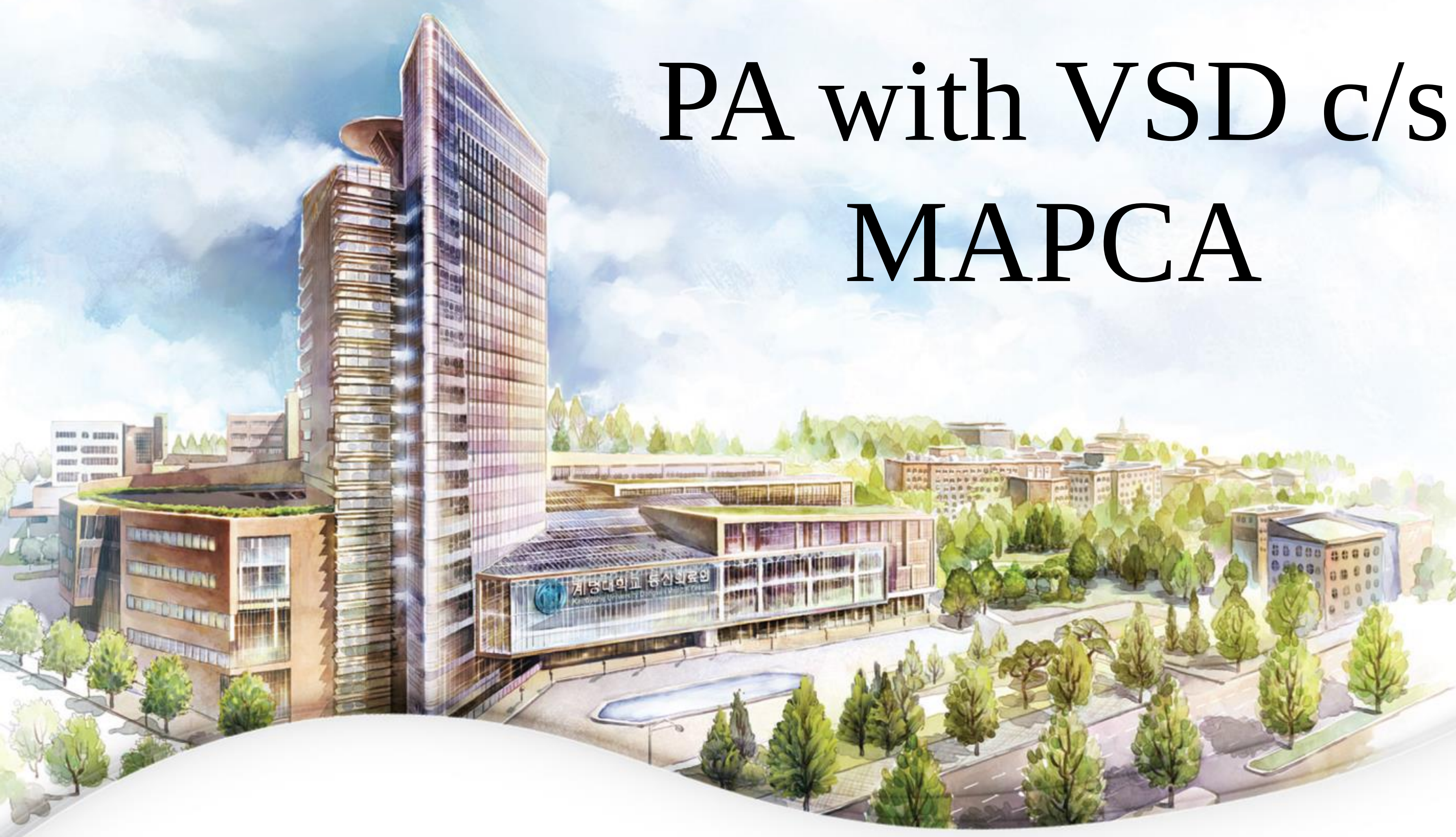
전공의 연수교육
2019.05.24

Keimyung University Dongsan Medical Center

# PA with VSD c/s MAPCA

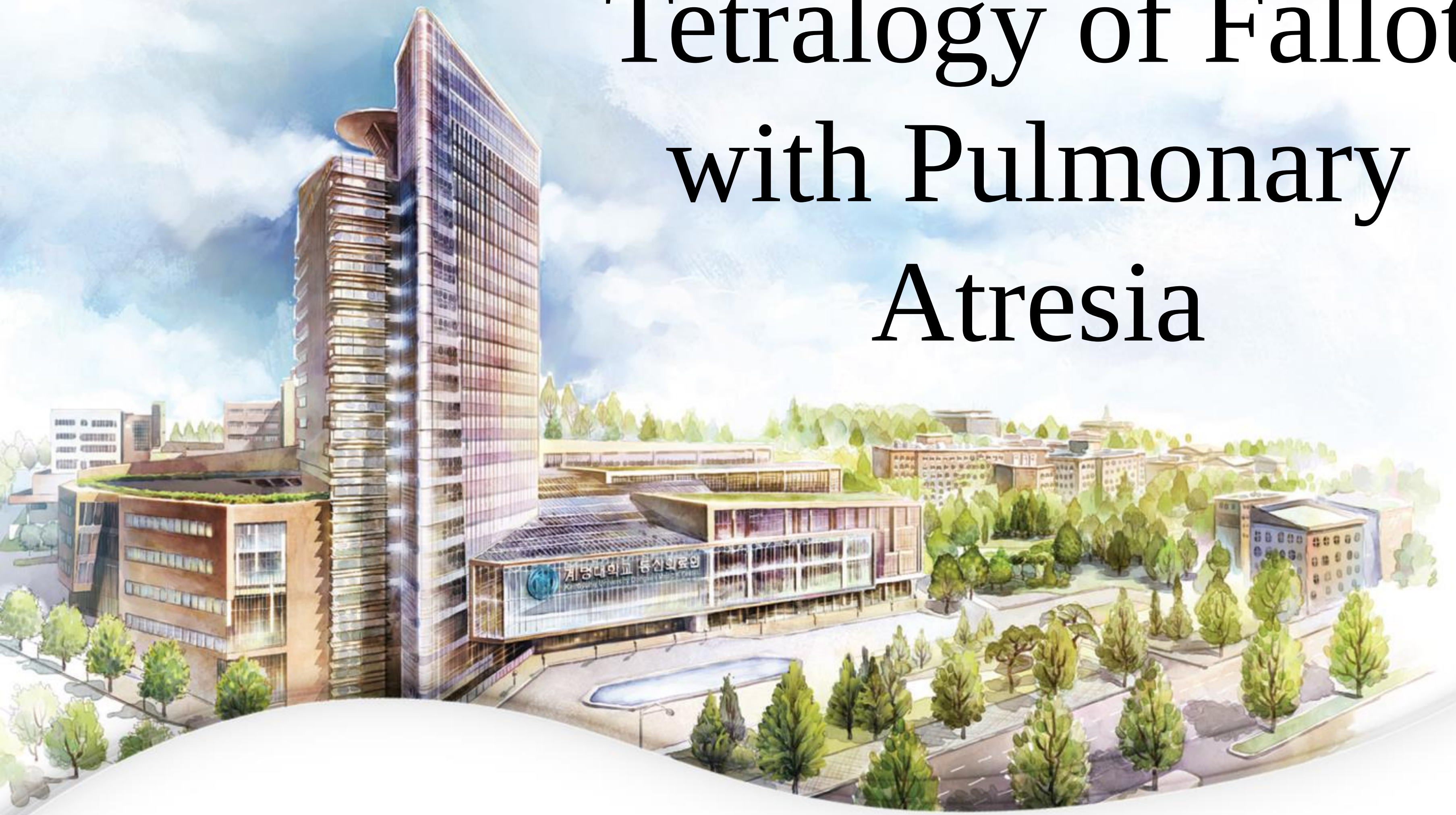



Keimyung University Dongsan Medical Center
Thoracic and Cardiovascular Surgery Department
Woo Sung Jang, MD., PhD.

전공의 연수교육
2019.05.24

계명대학교 동산의료원
Keimyung University Dongsan Medical Center

# Tetralogy of Fallot with Pulmonary Atresia



Keimyung University Dongsan Medical Center
Thoracic and Cardiovascular Surgery Department
Woo Sung Jang, MD., PhD.

# Definitions

- **PA VSD**
  - Lack of luminal continuity
  - Absence of blood flow from **either ventricle** to **pulmonary artery**
  - Discordant VA connections
    - Isomeric atrial appendages, Double inlet ventricle, or atrioventricular valvar atresia
      - Have pulmonary atresia with a hole between the ventricles
    - PA is confluent, is fed by a PDA
- **TOF with PA**
  - A specific type of PA VSD with intracardiac morphology of TOF

# TOF with PA c/s MAPCAs

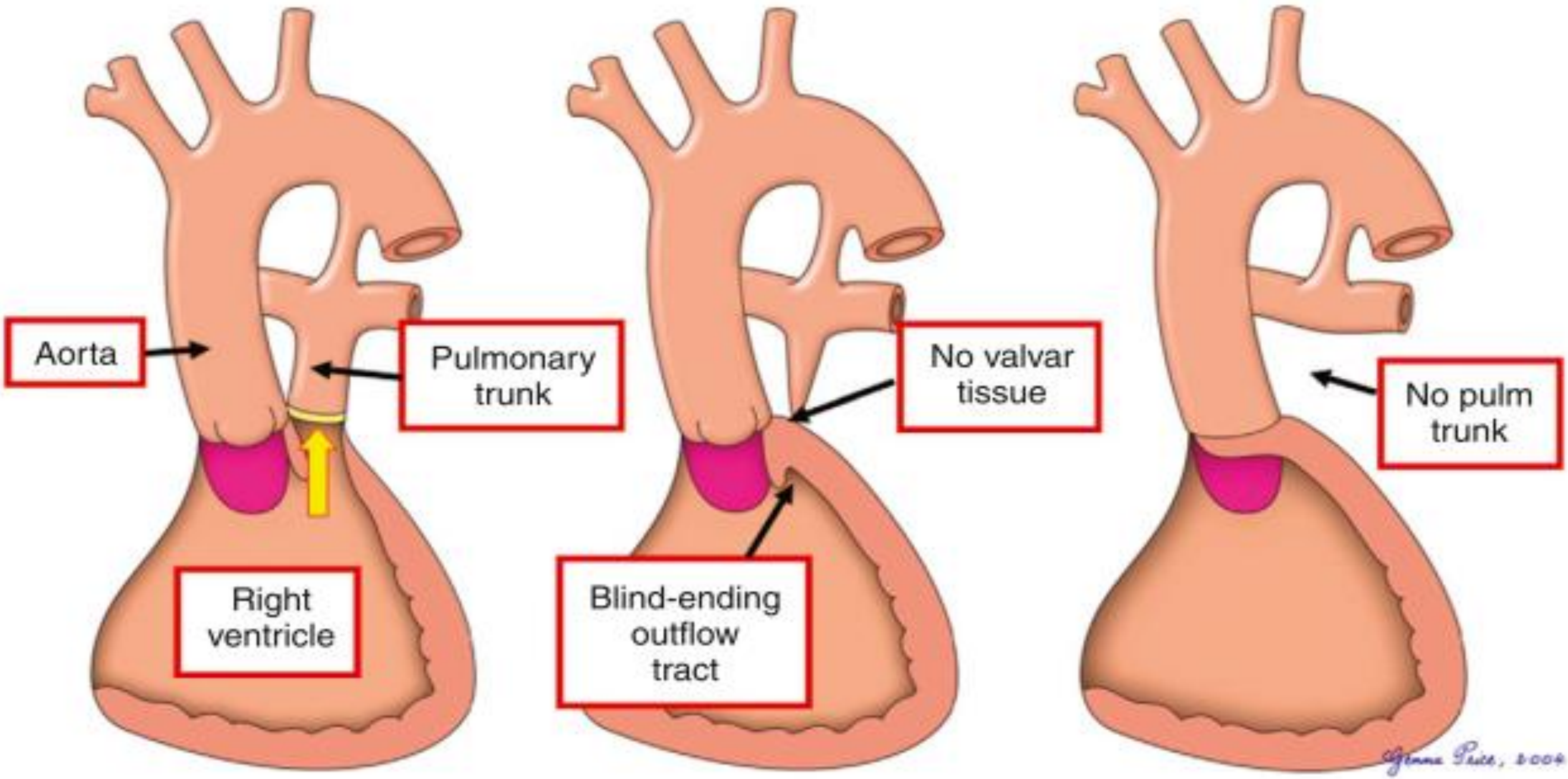
- Extreme subgroup of TOF
- Major clinical problems in the arteries that supply the pulmonary circulation
- Variable clinical presentations & different surgical strategies to that in TOF/PS

# Natural History

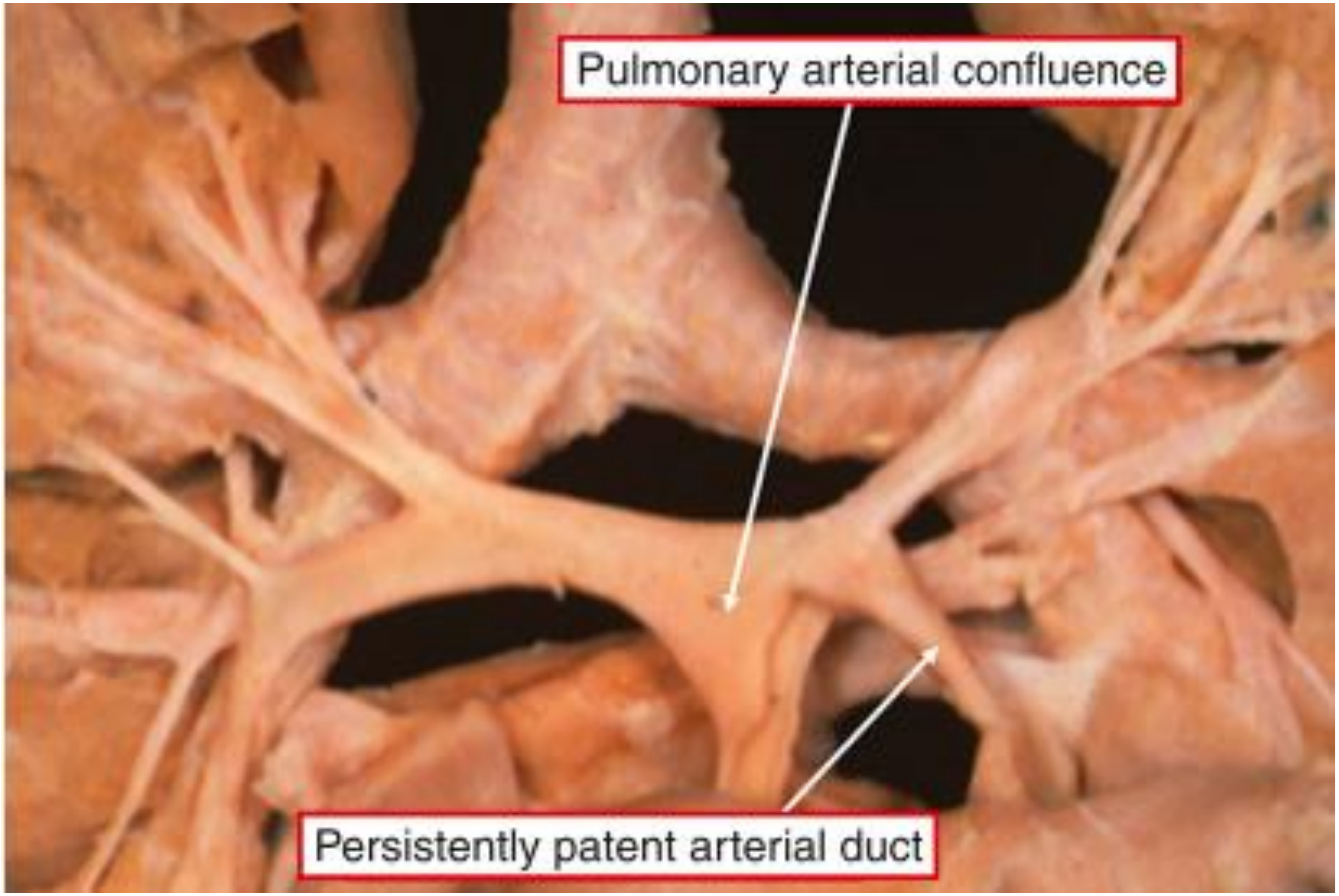
- Variable depending on the pulmonary blood flow
  - At birth, **ductus dependent** in case of true PAs
  - After ductal closure, dependent on the **collaterals**

- Excessive pulmonary blood flow : CHF, PVOD
- Moderate collateral stenosis: Balanced pulmonary blood flow
- Severe collateral stenosis : Hypoxia

# Characteristics of Pulmonary Atresia

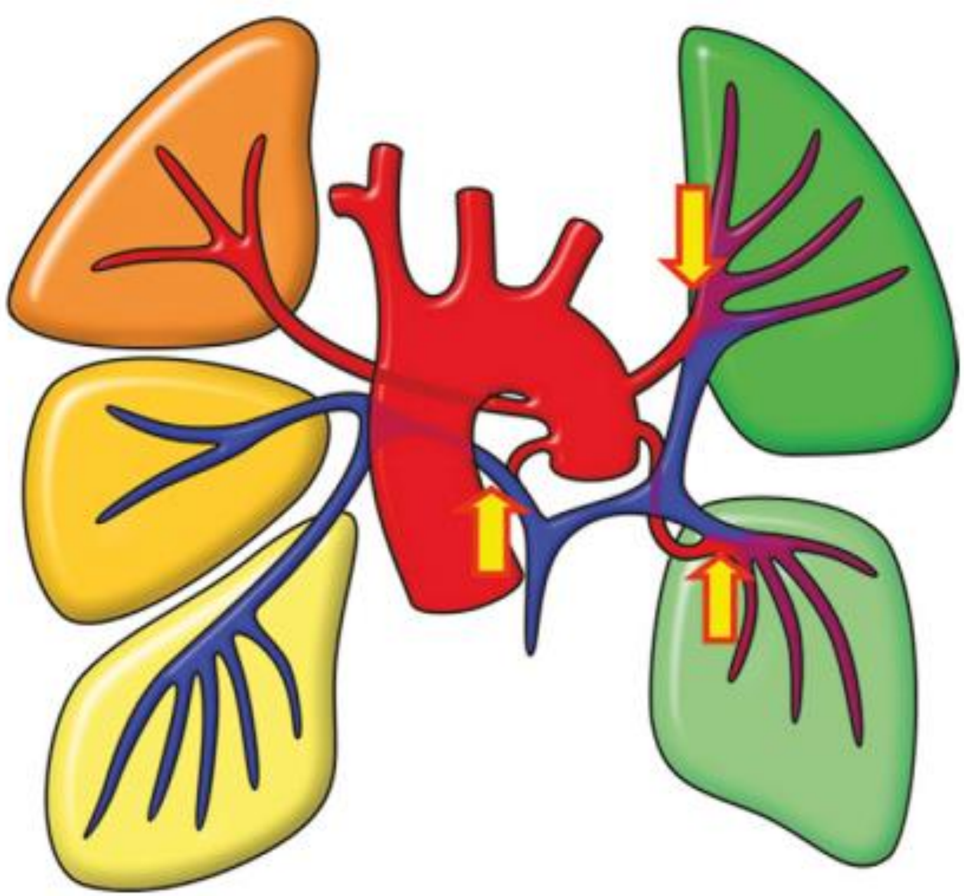


# Types of Pulmonary Circulation



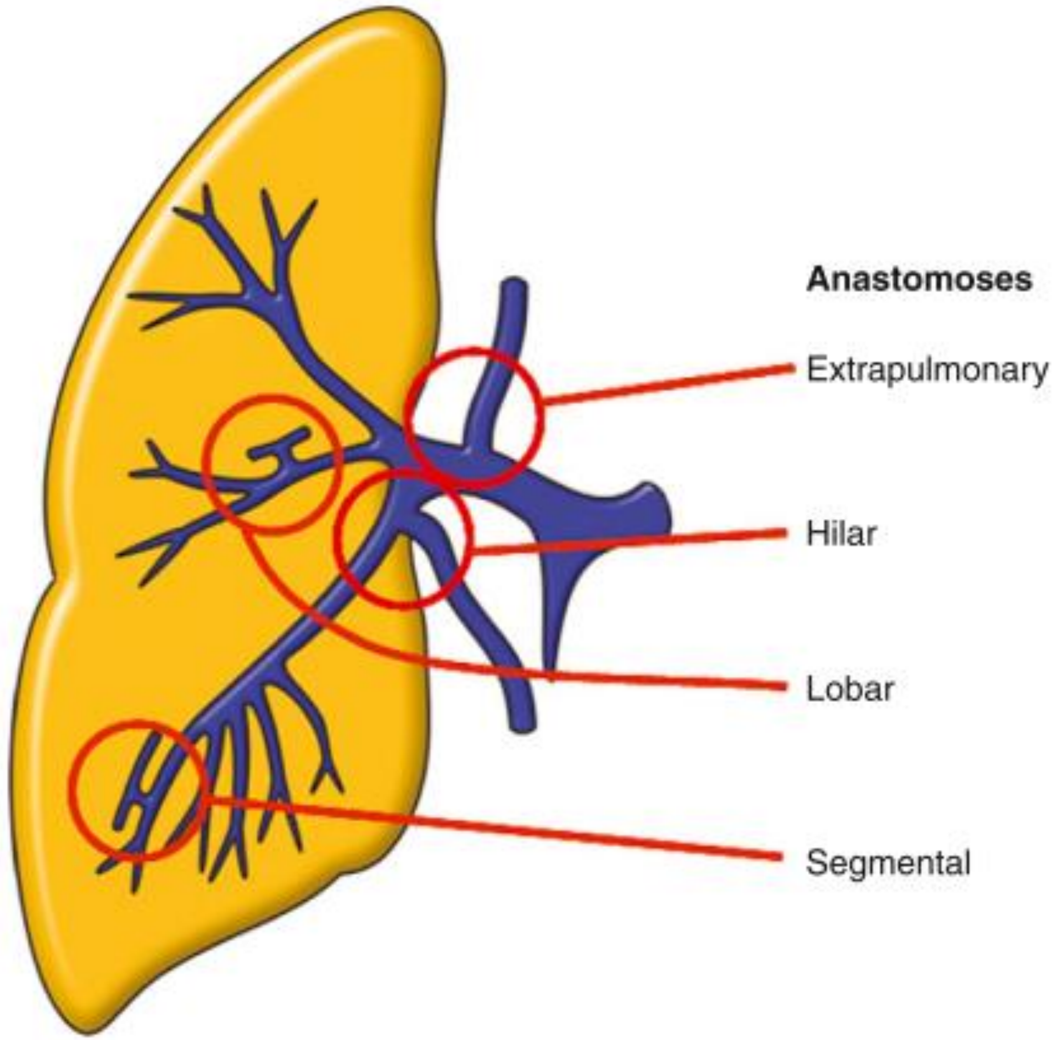

Type A 60~70%

# Types of Pulmonary Circulation

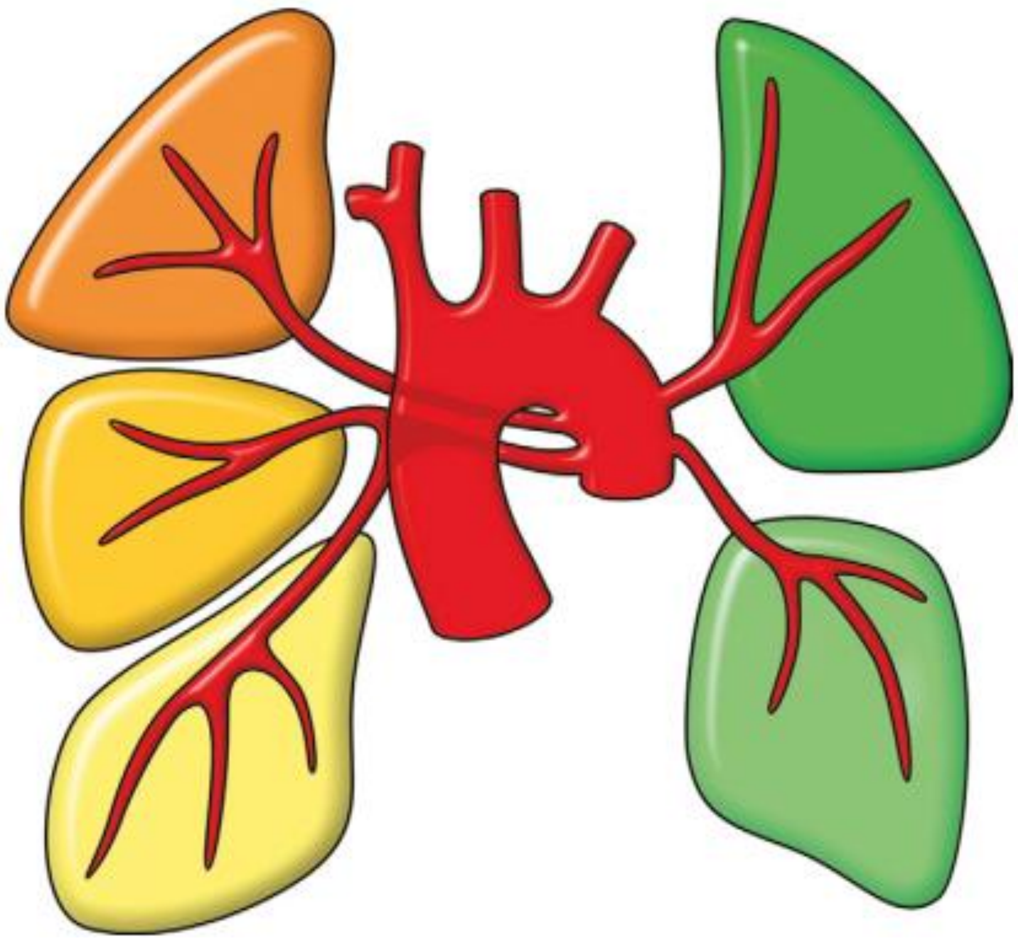


Type B 20~30%

# Communication between NPAs and MAPCAs



# Types of Pulmonary Circulation



Type C

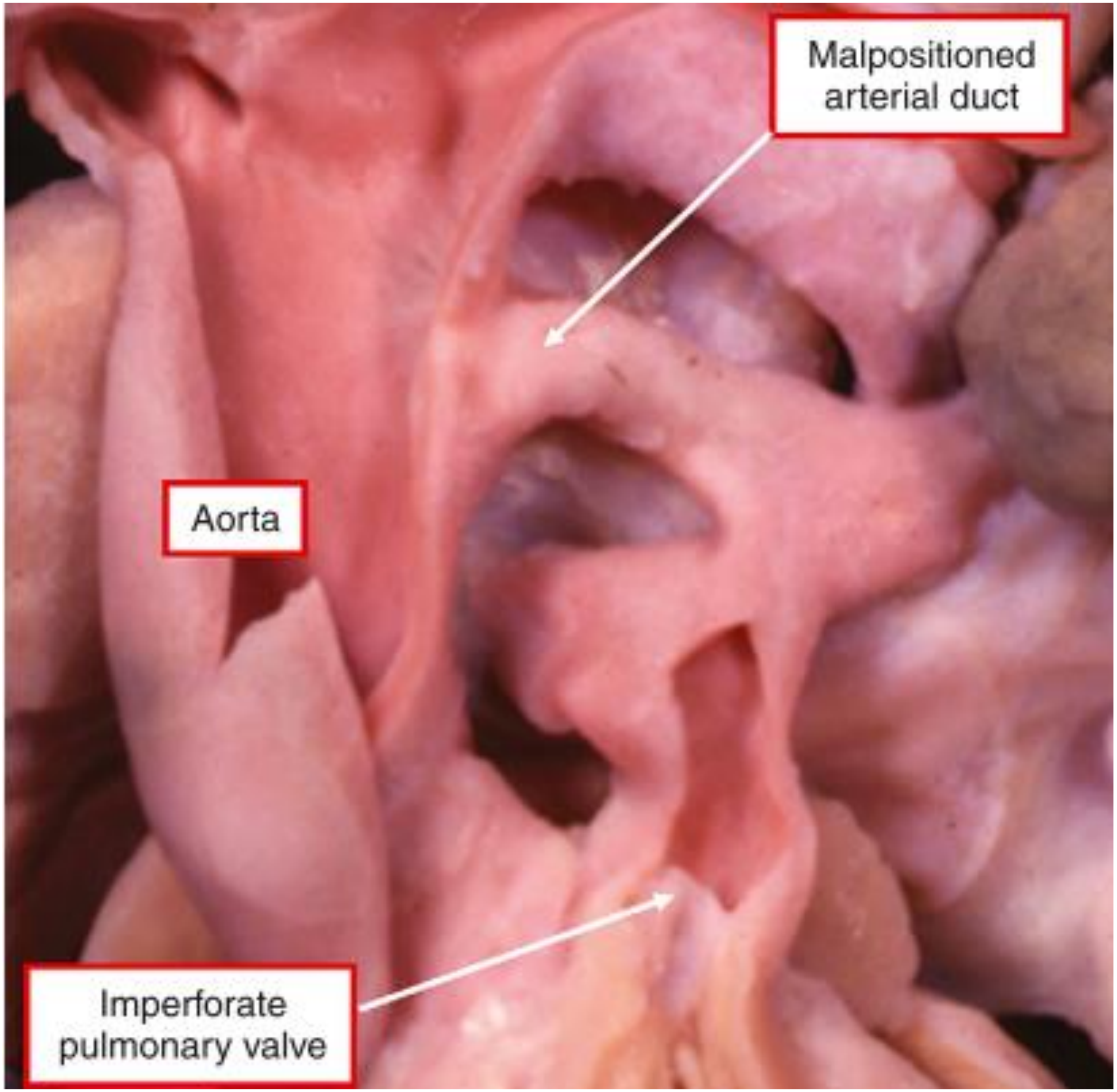
5~10%

# Sources of Pulmonary Blood Flow

- Unifocal pulmonary blood supply
  - Patent ductus arteriosus (PDA)

- Multifocal pulmonary blood supply
  - Major aortopulmonary collateral arteries (MAPCA)

# Supply from PDA



# Supply from MAPCAs

# Supply from MAPCAs

- Usually **co-exist** with intrapericardial **PAs**
- Number between **2~6**
- Usually arise from **descending thoracic aorta**
  - May originate from the aortic arch, subcalvian a, carotid a. or even the coronary arteries
- Frequently develop stenosis
- PHT and progressive PVOD
- MAPCAs connect with branches of central PAs, or constitute the only blood supply

# Characteristics of MAPCAs

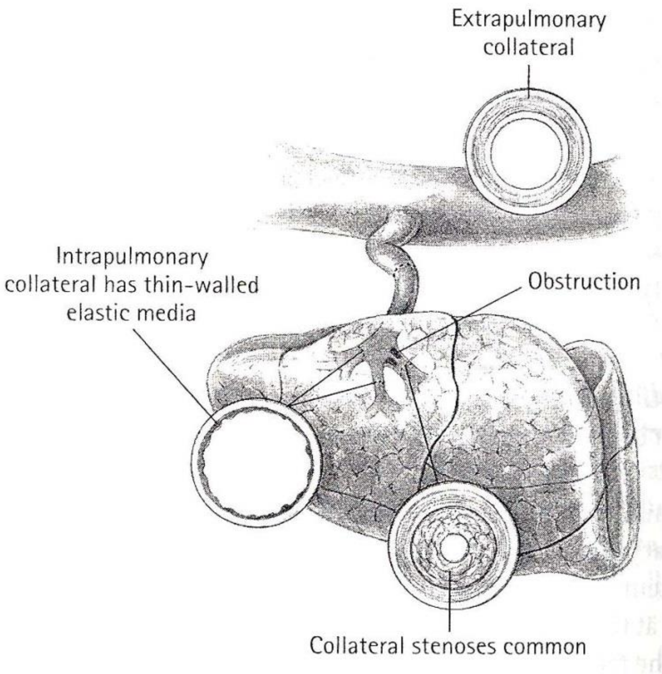
- Highly variable pulmonary arterial morphology, but some predictable pattern
  - The PDA connected to a central PA
    - **Peripheral PA distribution is normal** (no sytemic collateral arteries in that hemithorax, ductus arteriosus dose not coexist with MAPCAs in the same lung)
  - RUL and LLL segments often are supplied by single, noncommunicating MAPCAs (from subclavian artery and descending aorta)

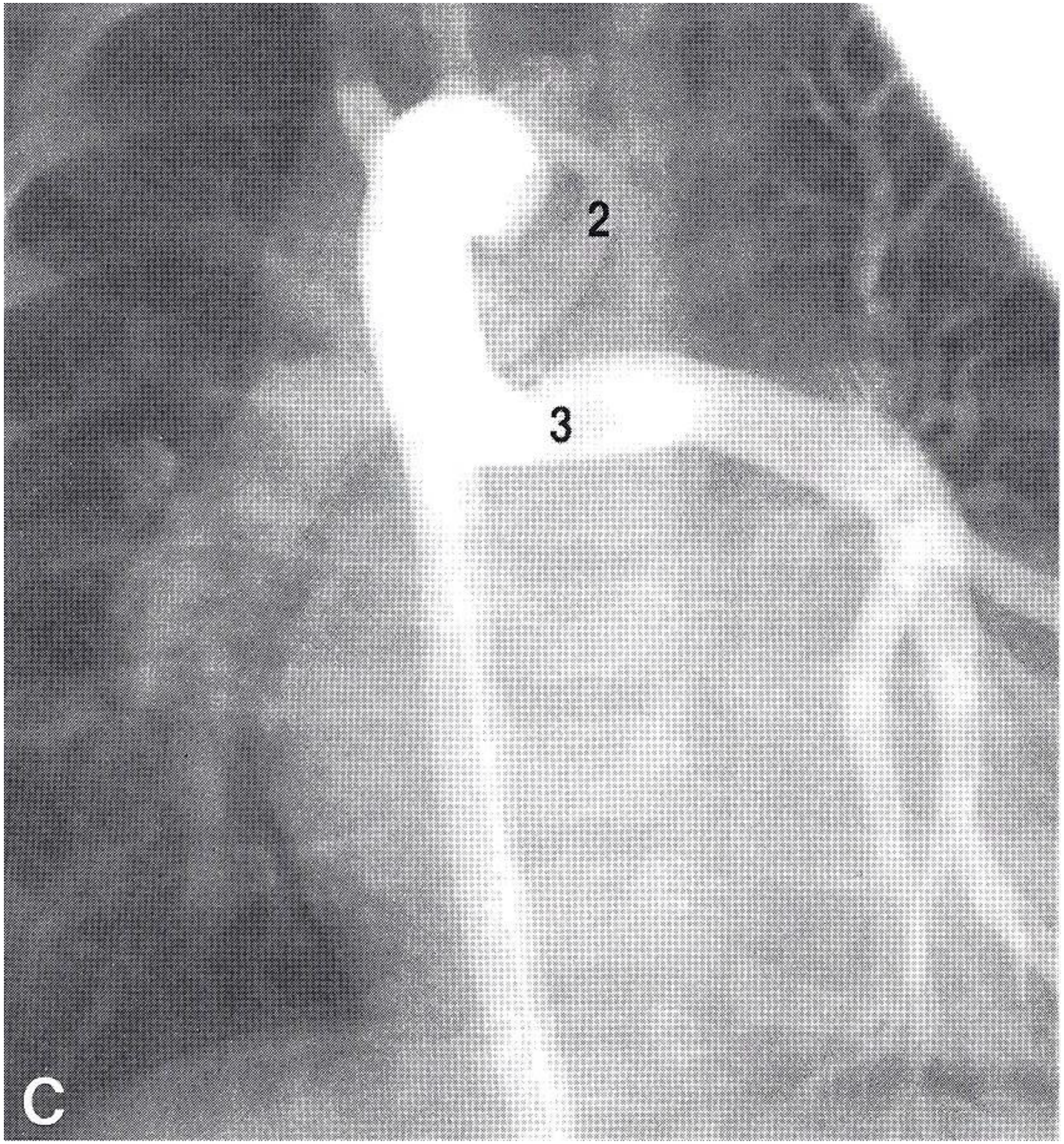
# Influence of MAPCA

- Chronic shunt & LV volume overload
  - Decreased LV function
  - Aortic annular dilatation
  - AR
- Segmental loss of lung parenchyme
  - In case of collateral stenosis
    - Hypoxia
  - In unobstructed cases
    - CHF, PVOD

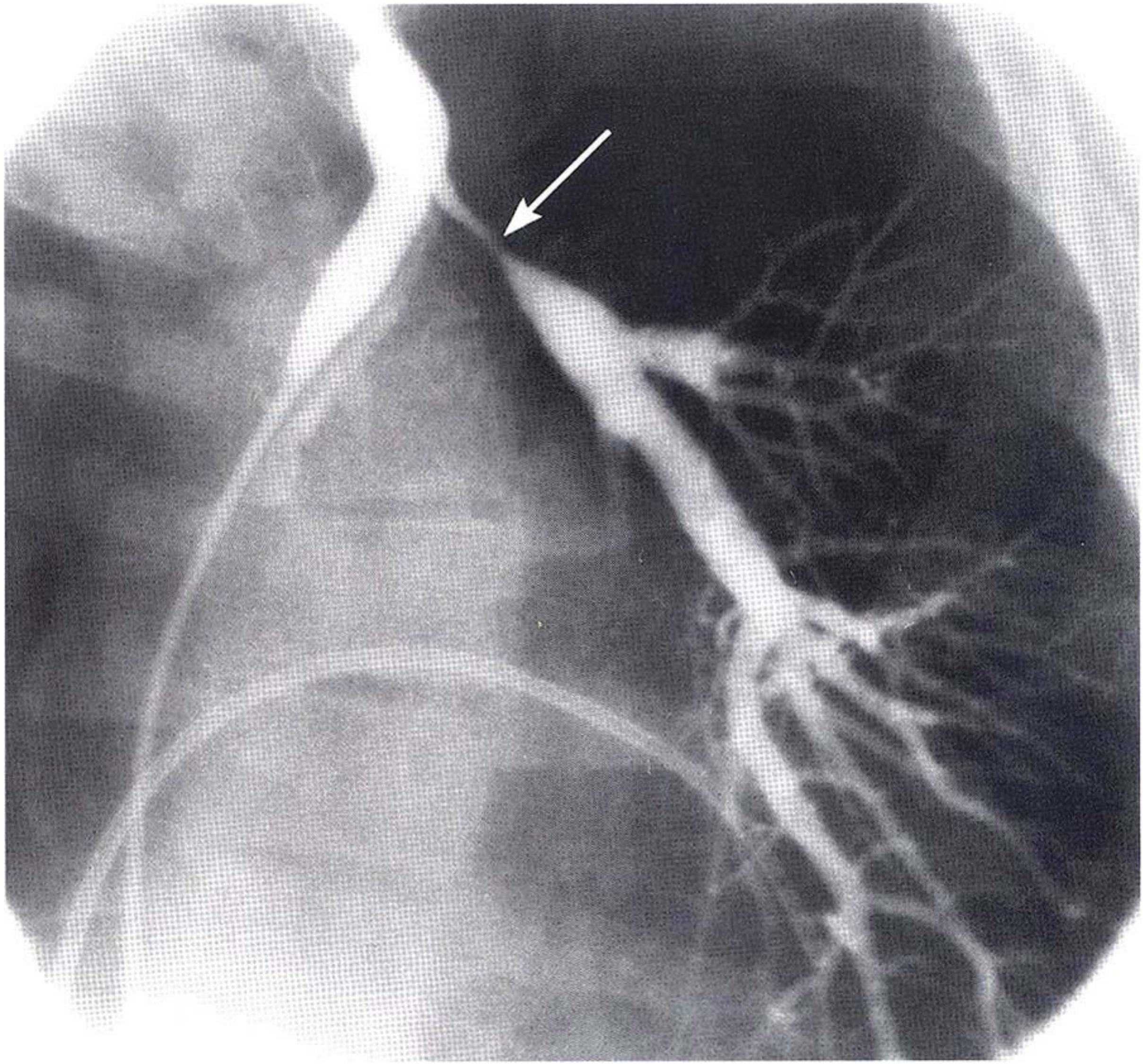
# Histologic Characteristics of MAPCAs

- **Extrapulmonary**
  - muscular artery with well developed muscular media & adventitia
- **Intrapulmonary**
  - medial muscle is gradually replaced by a thin elastic lamina resembling true PAs
- **Unobstructed MAPCAs**
  - PVOD
- Muscular segments of collaterals
  - prone to the development of severe stenosis, often progressive



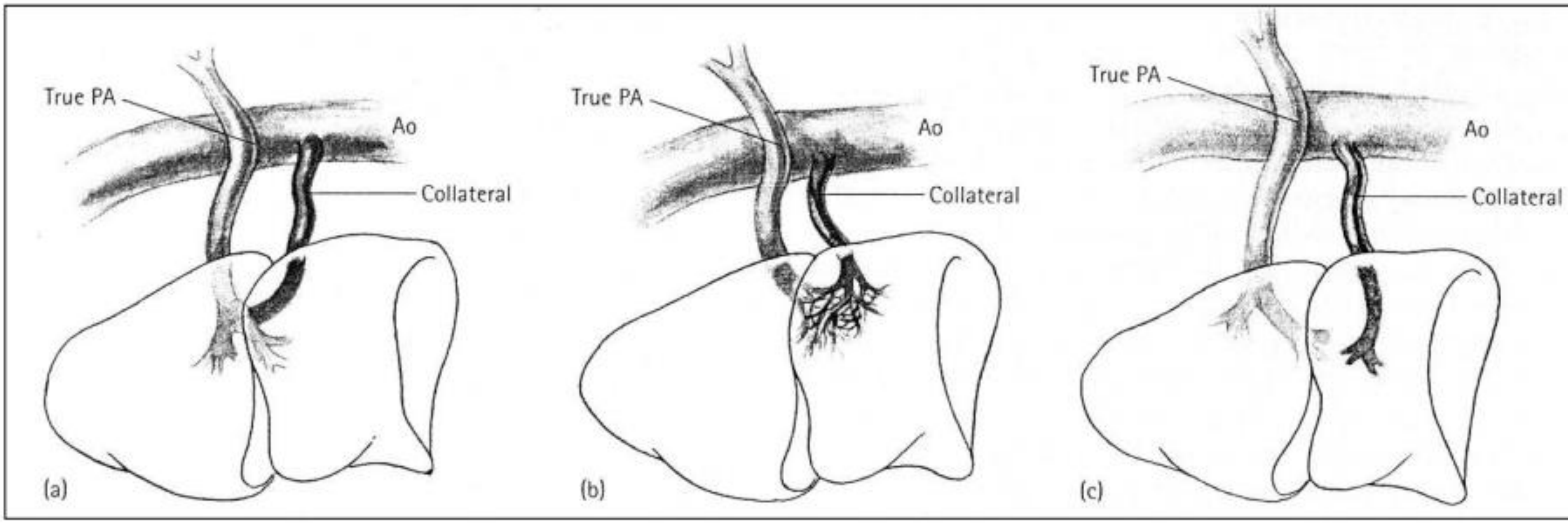



Unobstructed MAPCA



Long stenotic segment

# Connection between MAPCA and true PA



# Definitive Repair of TOF with PA c/s MAPCAs

***Ultimate goal***

: Completely separated pulmonary & systemic circulation

1. Closure of ventricular septal defect
2. Establish continuity between RV & PA
3. Occlusion of redundant collaterals & shunts / Unifocalization

# Surgery for TOF with PA s MAPCA

- **Initial palliation**
  - Shunt
  - Complete repair around 6~10 months of age
- **Primary neonatal repair**
  - Using RV-PA conduit or transannular patch
  - Foramen ovale is narrowed to 3~4mm
- Options for RVOT reconstruction
  - Conduit ± Pulmonary valve
  - Transannular patch ± pulmonary valve

# Surgery for TOF with PA s MAPCA

- **Initial palliation**
  - Shunt related complication
    - High inter-stage mortality
    - PA distortion
- **Early primary repair**
  - Early RV volume loading
  - LPA stenosis
    - Required multiple intervention

# Surgery for **TOF with** PA c MAPCAs

- **Unifocalize** the greatest number of segmental arteries together
  - Native PA and/or MPACAs
  - **Single stage vs multi-stage**
- Remove aortic sources of blood flow to segments that are dual supplied.
- **Closed VSD** (if possible) and **create RV to PA communication** achieving a RV/LV ratio of < 0.7

# Surgery for TOF with PA c MAPCAs

- Maximize the pulmonary artery
  - The size & distribution

- Maintain the adequate PBF

- Avoid the excessive PBF

# Surgical strategy in TOF with PA c MAPCA

- **Single stage repair**
  - complete unifocalization with RV to PA conduit and VSD closure at age of 4~8 months
  - High RV pressure > 80% of RV/LV
    - VSD fenestration

- **Staged repair**
  - Including growth of the central PAs with central shunt or RV-PA conduit
  - Staged thoracotomy-based unifocalization of MAPCAs
  - RV-PA conduit with VSD closure

# Rationale for early single stage unifocalization

- One can incorporate all segments of blood supply to the lung **before stenosis develop** in the MAPCAs and **before potential changes of PH** occur
- Preferred age for single stage repairs
  - **4 to 8 months of age**
    - Improved tolerance of long operations as compared to young infancy
    - Prior to the development of risks of PVOD
    - Prior to the development of MAPCA stenosis

# Advantages of single stage repair

- Eliminate the need for multiple operations
- Eliminate the use of prosthetic materials
- Establish the normal physiology early in life
  - Growth of respiratory & PA system
  - Avoid cyanosis & volume overload
  - Prevent the PVOD

# Disadvantages of single stage repair

- Increased pulmonary morbidity
  - Contusion & congestion
  - Bronchospasm
  - Phrenic nerve injury
- Magnitude of operation
- Technically more demanding
- Unknown ideal age

# Rationale for staged repair

- **Small central PAs** needs to be "rehabilitated" to normal size with shunt or RV to PA conduit
- To gain exposure to the distal MAPCA, hilar and intraparenchymal dissection is facilitated through unilateral or staged bilateral thoracotomies
- Identification and mobilization of MAPCAs is much easier through posterolateral thoracotomy than a sternotomy approach
- Single stage unifocalization is a long and tedious procedure (very stressful to a child)

# Disadvantages of staged repair

- The final repair is **achieved on an old age**
- Mediastinum & hilar regions are significantly scarred, increasing surgical risks
- **Prolonged cyanosis** & previous operation cause secondary collaterals, risks of bleeding
- The risk of drop-off before the final repair

# Unifocalization

- Definition
  - Procedures that **join the multifocal sources** of pulmonary blood flow, be they intrapericardial native pulmonary arteries or one or more collateral arteries (MAPCAs), **into a single source**

# Timing of Unifocalization

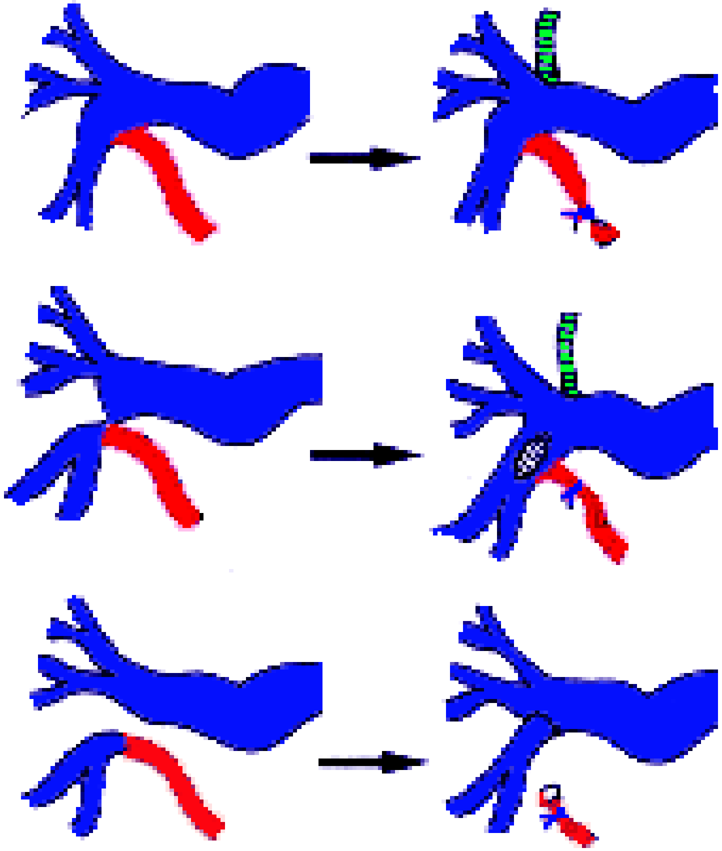
- At any age, when collaterals are large to allow technical ease without risk of thrombosis
- Variable depending on collateral size, usually older than 2~3 months
- Staged procedures may be required for the bilateral aortopulmonary collaterals

# Techniques of Unifocalization

- Procedures for collaterals
  - Ligation
  - Patch enlargement
  - Direct anastomosis
- Interposition grafts
  - Synthetic graft
  - Homograft
  - Xenograft
  - Autologous tissue including pericardium, azygos vein

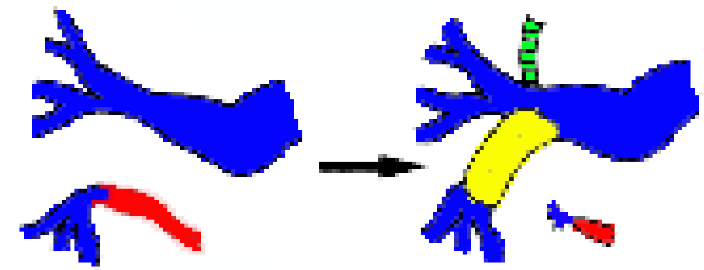
# Unifocalization procedure

- Ligation
- Angioplasty
- Anastomosis

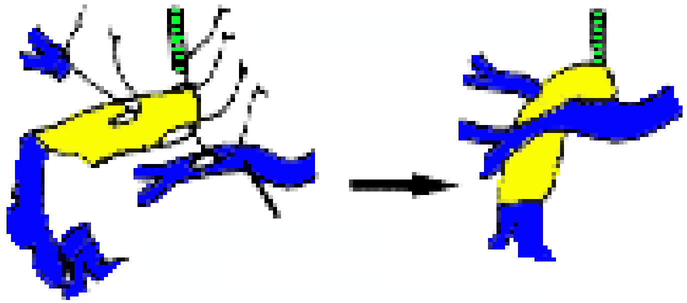


# Unifocalization procedure

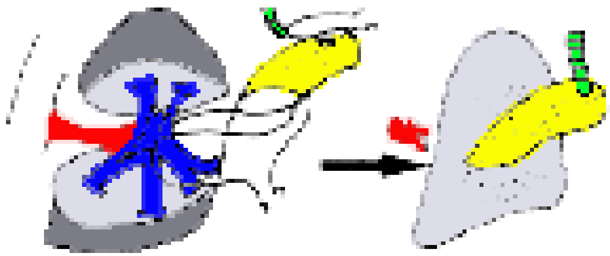
- Interposition



- Additional PA creation

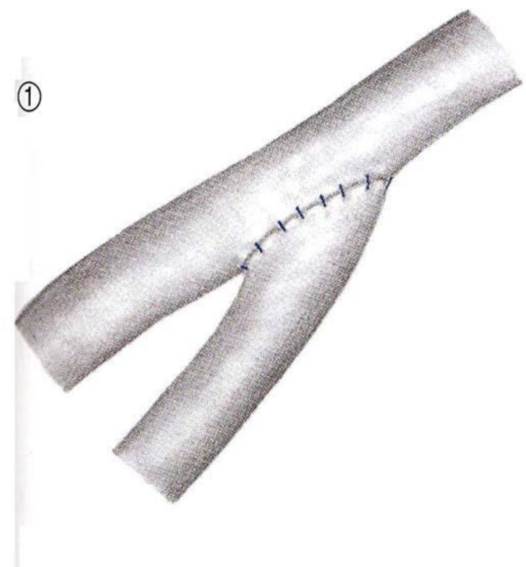


- Central PA creation

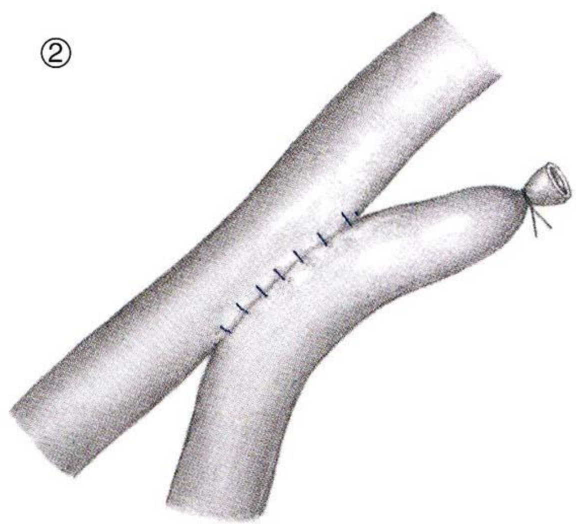


# Unifocalization procedure

-Offbypass during dissection
- Maximal use of native tissue
- Avoid circumferential use of non-viable conduits for growth potential




End-to side




Side -to side

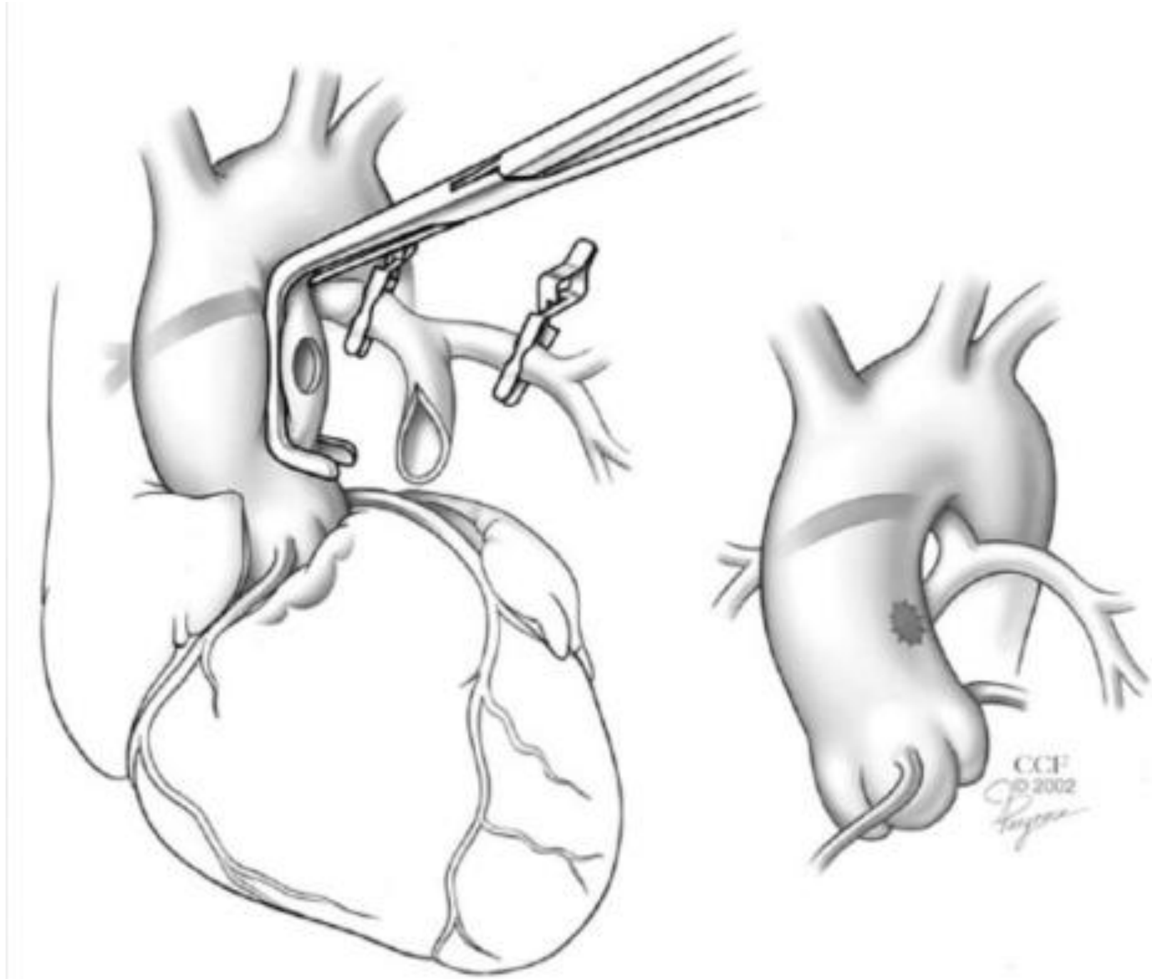
# Repair without Unifocalization

- RCH, 2009
- **Unifocalization brings no long-term benefits**
  . Unifocalization: sufficient to allow a safe repair
  but, **failed to achieve adequate growth**
  . **Dilated BAs**: limited growth potential & unstable

→ Growth of the native PA rather than recruitment of MAPCAs

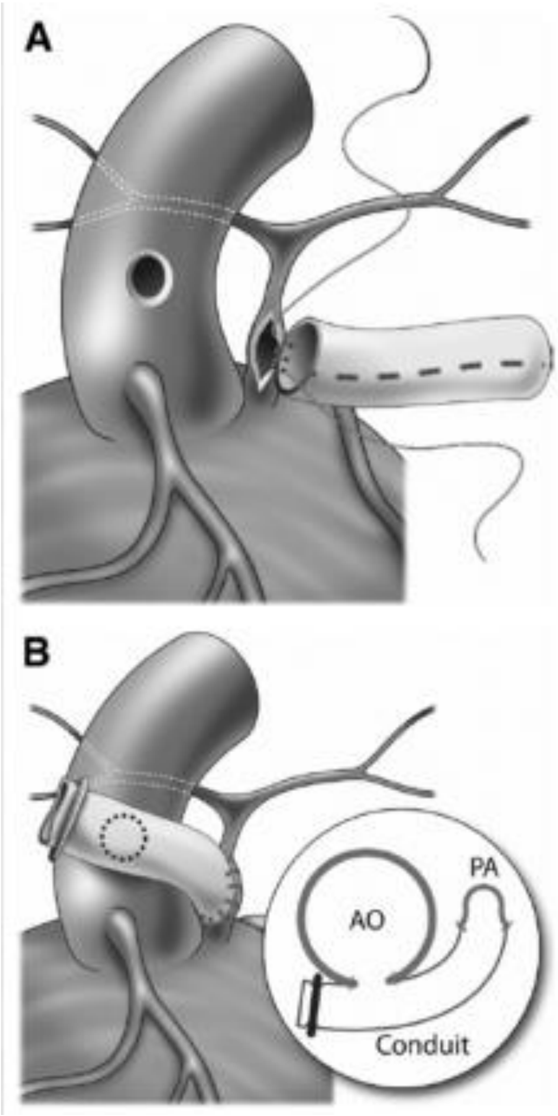
- **Multi-stage approach**
  . 4~6wks: **Modified central shunt**
  . 4~6months: **RV-PA conduit**
  . 3rd : complete repair or 2nd conduit
- 18 pts enrolled in this protocol (No Unifocalization)
  . 7 : complete repair, RVP 59% of systemic
  . 8 : awaiting repair
  . 4 MAPCAs in 17 pts: ligated

# Melbourne Shunt

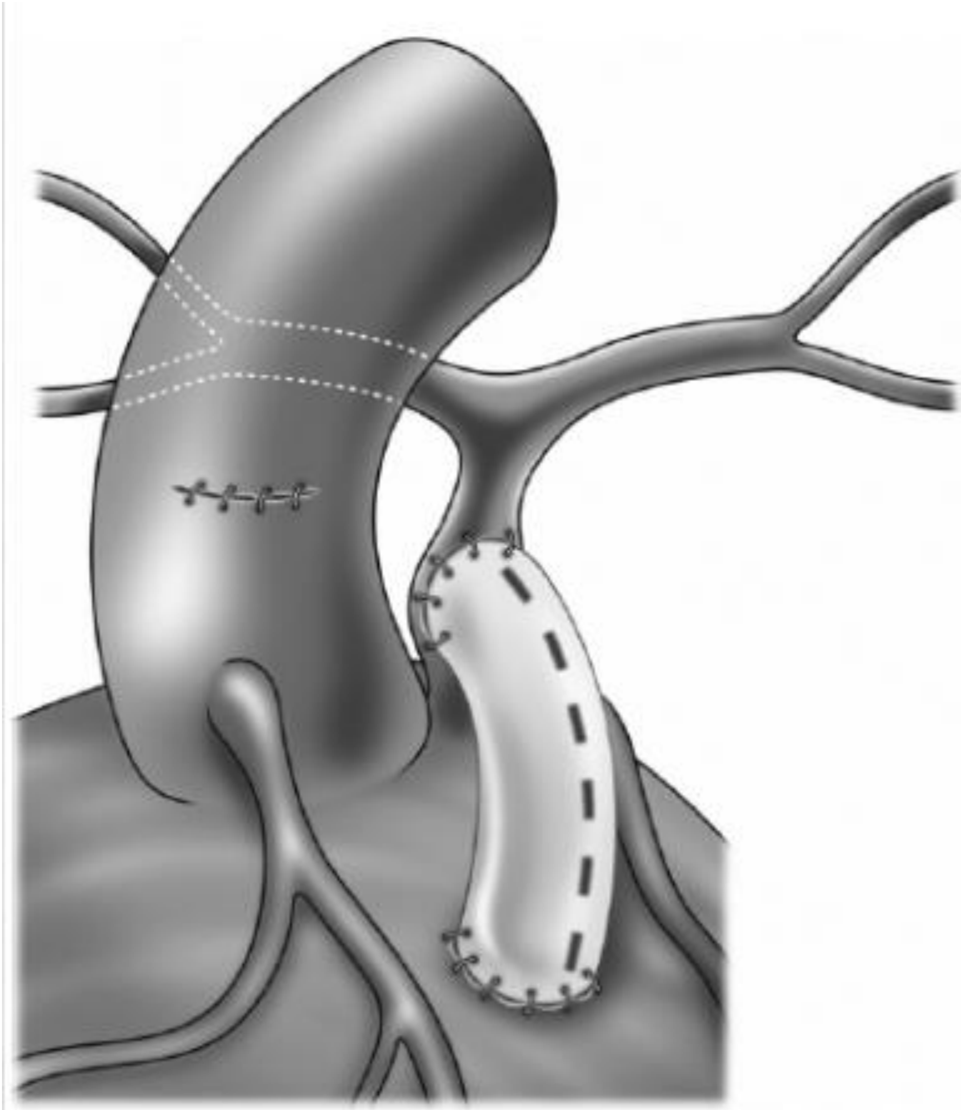



- Central end-to-side Aortopulmonary shunt
- Diminutive central pulmonary arteries

# Modified Central shunt



# RV-PA conduit



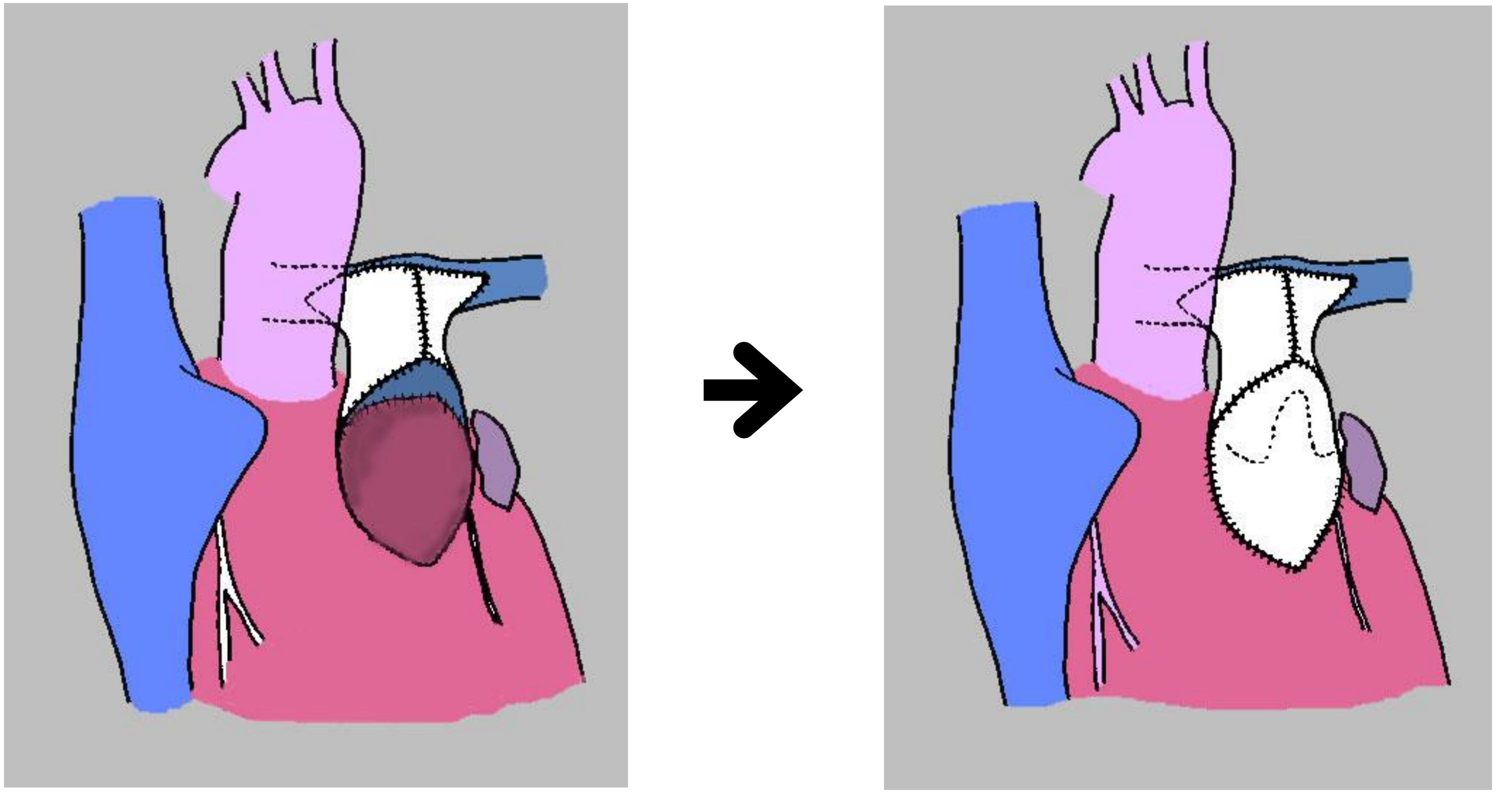
# Advantages of RV - PA Connection

- Reduction of LV volume overload
- **Pulsatile blood flow** to enhance PA growth
- Facilitating the **catheter access** for the later evaluation & intervention

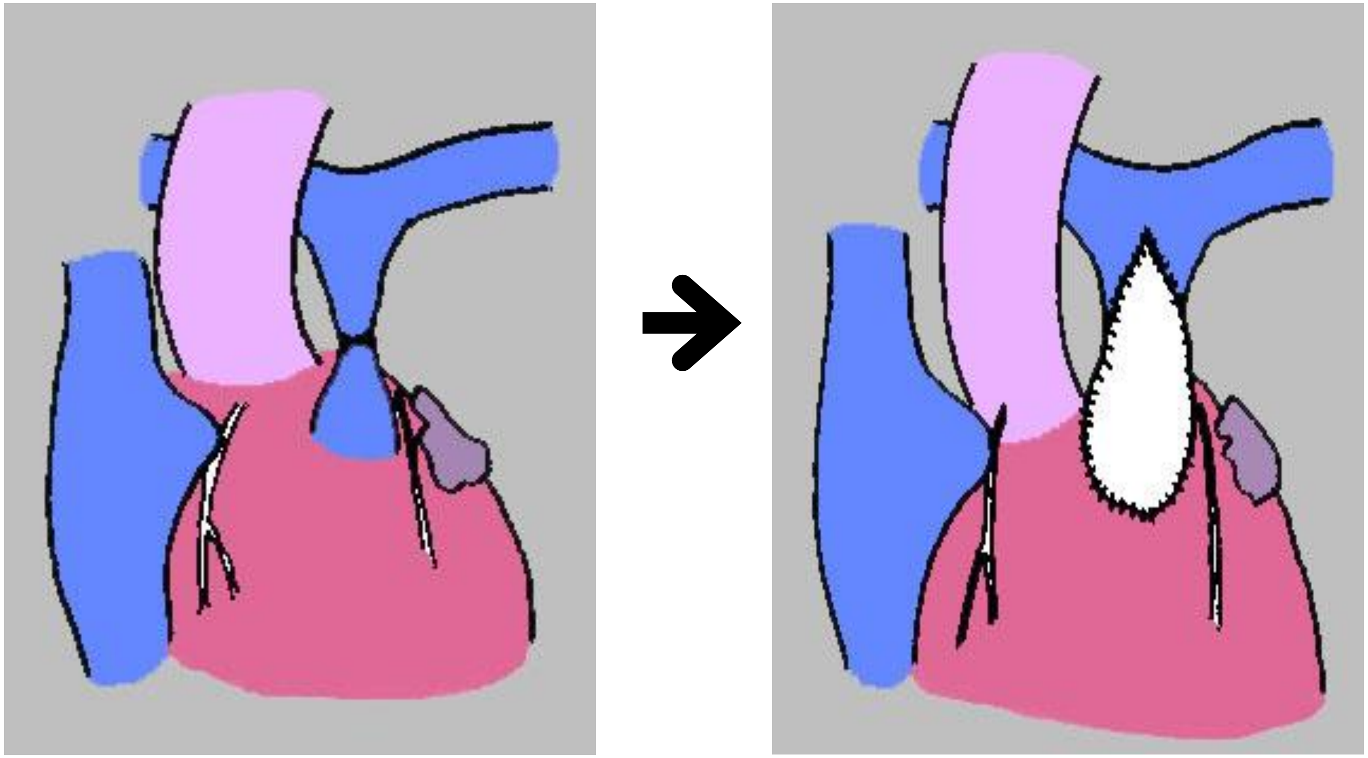
- Complications
  - aneurysm and pseudoaneurysm
  - **pulmonary flow and pressure is completely uncontrolled**

# RV-PA Reconstruction

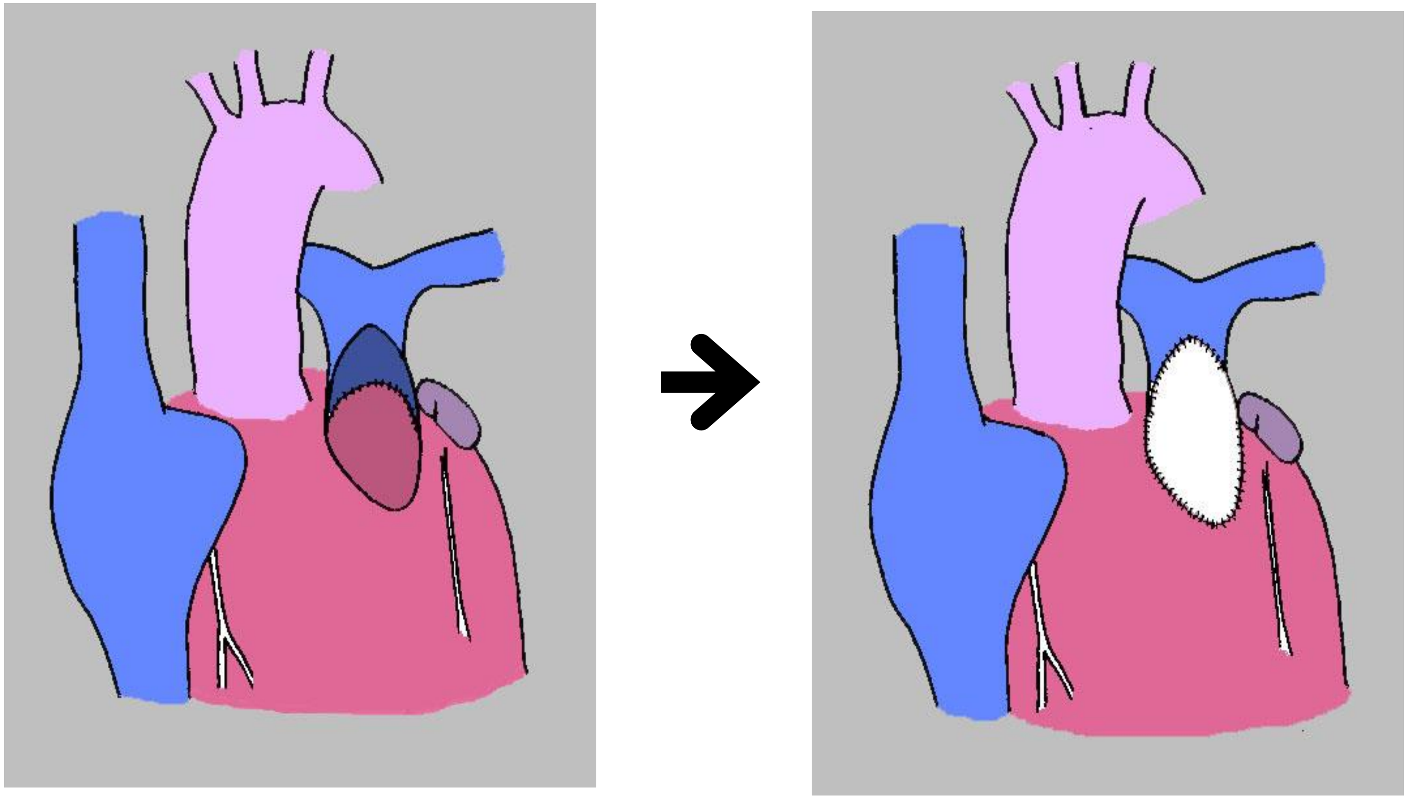
# RVOT Reconstruction with Valved Conduit



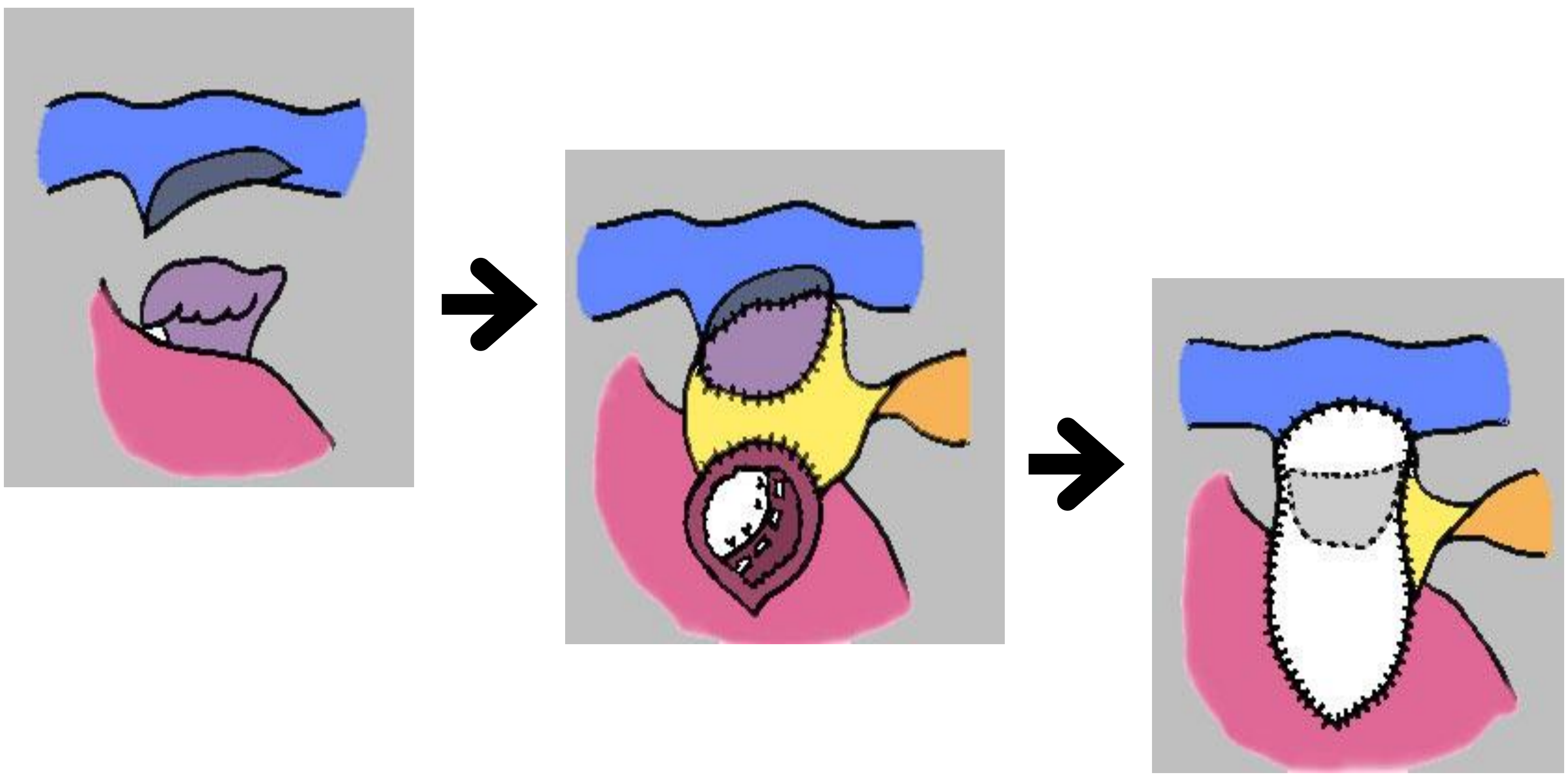
# RVOT Reconstruction with Outflow Patch



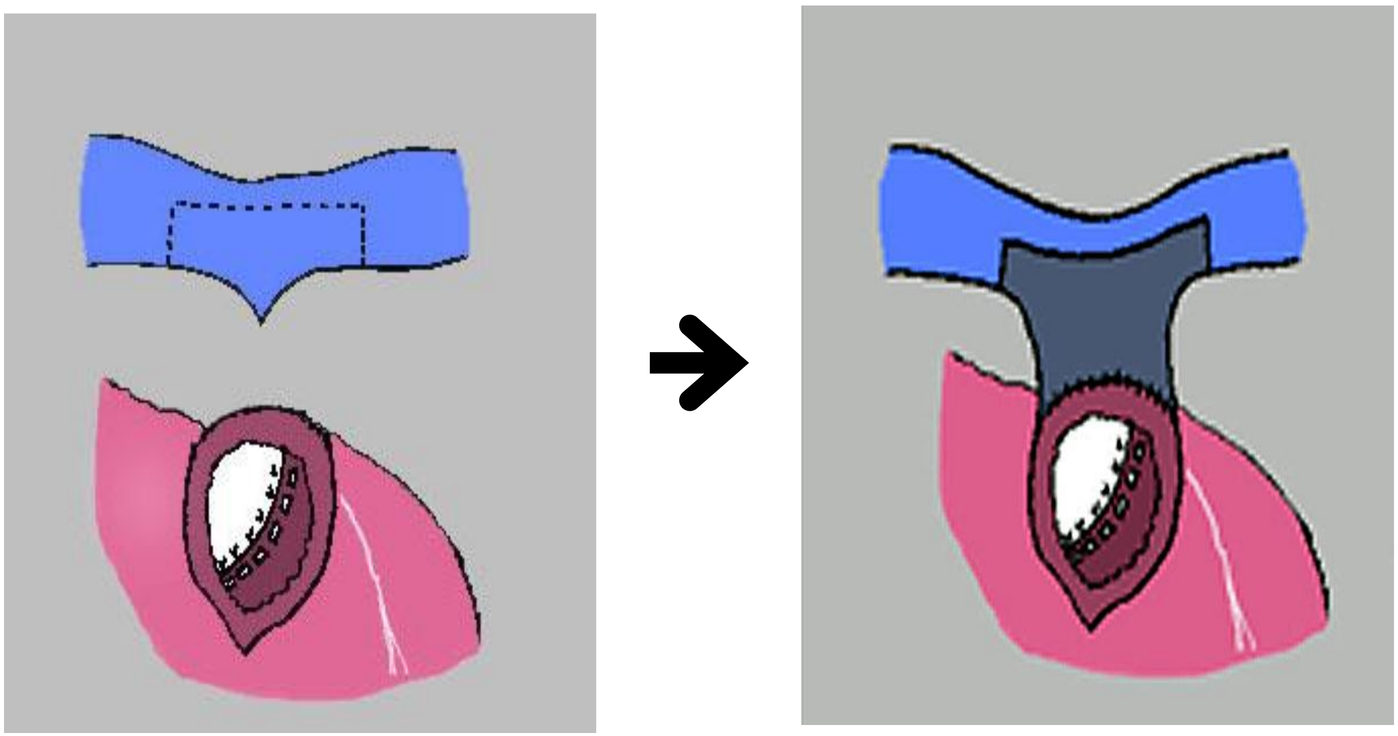
# RVOT Reconstruction with PA Reimplantation



# RVOT Reconstruction with LA Appendage



# RVOT Reconstruction with PA Flap



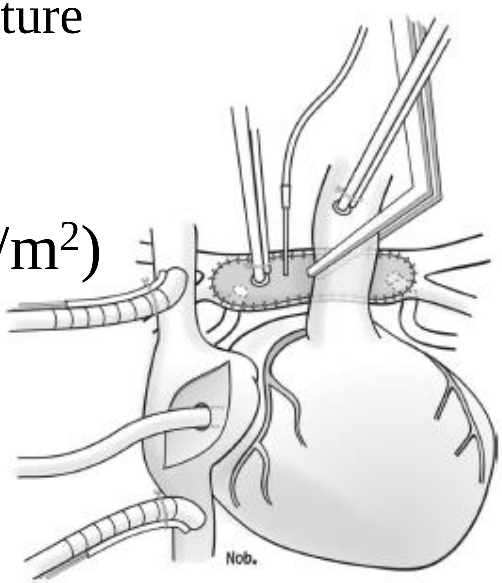
# VSD closure

# Functional Intraoperative PBF Study

* Post-repair RVSP: most reliable predictor of favorable outcome
* Data of functionality of the entire pulmonary vasculature

Hanley

- **m PAP < 25mmHg at a full flow** (2.5L/min/$m^2$) predicts RV/LV pressure ratio < 0.5



Toronto, 2009

- Close the VSD for a **mPAP of <30mmHg**
- Predict postop. physiology better than standard anatomic measures

# Anatomic Predictors of successful VSD closure

- Central PA area ≥ 50% of predicted normal
    - Puga et al JTCS 1989;98(6):1028-9
- Predicted pRV/pLV≤0.7, no MAPCA remain
  - More than 2/3 lung segments are centralized
    - Iyer and Mee, ATS 1991; 51:65-72
- Nakata index > 150 ㎟/㎡ BSA
    - Metras, EJCTS 2001;20:590-6
- TNPAI ≥200 ㎟/㎡
    - Hanley, JTCS 1997;113(5):858-66
- More than 15 of lung segments connected to native PA
    - Baker, 2002

# Functional predictors of successful VSD closure

- Net **left to right shunt**
- SpO2 is typically in the **high 80s or low 90s**
- At a cardiac index of 2.5L/min/㎡ and PA pressure of less than 30mmHg after unifocalization

# Long-Term Surgical Outcome

- Depends on
  - Number of lung segments incorporated into final repair
  - Status of pulmonary microvasculature
  - Absence of obstruction in RV-PA conduit and branch PAs.

# Surgical Results

**Table 22-1.** Summary of surgical outcomes in patients with pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries

| Authors (year) | Time period of operation | No. of patients | Strategy of unifocalization | Age at first operation | Early death | Follow-up duration | Late death | Outcomes at last follow-up |
|---|---|---|---|---|---|---|---|---|
| Reddy (2000)[71] | 1992-1998 | 85 | Single-stage | 5m (10d – 37y) | 9 (10.6%) | 22m (1 – 69m) | 7 | 84% 1YSR<br>74% 4YSR<br>93% total repair |
| Carotti (2003)[53] | 1994-2002 | 37 | Integrated approach | 39m (22d – 13y) | | 43m (1 – 85m) | | 81% 7YSR<br>85% total repair |
| Gupta (2003)[65] | 1983-2000 | 104 | Staged | 7d (3d – 22y) | 11.5% | 10.2y | 5% | |
| Duncan (2003)[49] | 1993-2001 | 46 | Staged | 7.2m (17d – 23y) | 0 | 44m (1 – 79m) | 1 (2.2%) | 61% total repair |
| d'Udekem (2005)[69] | 1975-1995 | 82 | Staged | 1.4y (7d – 34y) | 4% + 8% | 14.2y (3m – 25y) | 9 | 51% 12YSR (total repair)<br>65% total repair |
| Ishibashi (2007)[50] | 1982-2004 | 113 | Staged | 6.3y (1.1m – 34y) | | 8.8y (0.8 – 23.3y) | | 80.9% 5YSR<br>73.8% 10YSR<br>80.5% total repair |
| Davies (2009)[54] | 1989-2008 | 216 | Staged/ Single | 2y | 6% | 2.3y | 6% | 89% 3YSR<br>73% total repair |
| Honjo (2009)[67] | 2003-2008 | 20 | Single-stage | 7.7m (2-197m) | 0 | 31m (8-66m) | 5% | 94% 1YSR<br>95% total repair |
| Malhotra (2009)[38] | 1992-2007 | 462 | Single-stage | 7.7m (10d – 39y) | 5.9% | NR | NR | 86% 5YSR<br>90% total repair |

* d; days, m; months, y; years, YSR; year survival rate

# Conclusions

- MAPCAs
  - Wide spectrum of pulmonary vascular morphology and physiology, ranging
- Management
  - complex and must be individualized according to their anatomy and clinical situations

계명대학교 동산의료원
Keimyung University Dongsan Medical Center

# Thank you for your attention