Pulmonary atresia – Ventricular septal defect

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Pulmonary atresia – Ventricular septal defect (PAVSD)

- PA-VSD is the most severe form of tetralogy of Fallot (TOF), accounted for 20.3% of all forms of TOF and about 2% of congenital heart disease
- The intra-cardiac anatomy : TOF + membraneous or complete atresia of the pulmonary valve
- It is one of the common causes of cyanosis and hypoxemia in the neonate

Embryology

- Day 27, the arterial branches of the paired 6th aortic arches form an the pulmonary vascular plexus→The lungs have a dual blood supply
- The larger vessels will form the pulmonary arteries . The smaller vessels form bronchial arteries.
- The disconnection RV- PA → the lungs: PDA, major aorto–pulmonary collateral arteries(MAPCAs)

Pathology

- PA anomalies: hypoplasia, nonconfluence, and abnormal distribution, are more serve in patient with MAPCAs than in those with PDA
- The size of the PA: amount of collateral arteries, position where they connected to pulmonary arteries.
- Blood of the lungs : PDA,MAPCAs, occasionally coronary artery, and plexuses of bronchial or pleural arteries.

Pathology

- PDA:
  - No branches, less tortuous than collaterals.
  - Normal narrowing occurs →hypoplasia of the pulmonary arteries becomes more severe
  - Unstable source of pulmonary blood
- MAPCAs:
  - From descending thoracic aorta, subclavian arteries,abdominal aorta, coronary arteries
  - Stenoses are present in nearly 60% of collateral arteries
  - Stable source of pulmonary blood flow

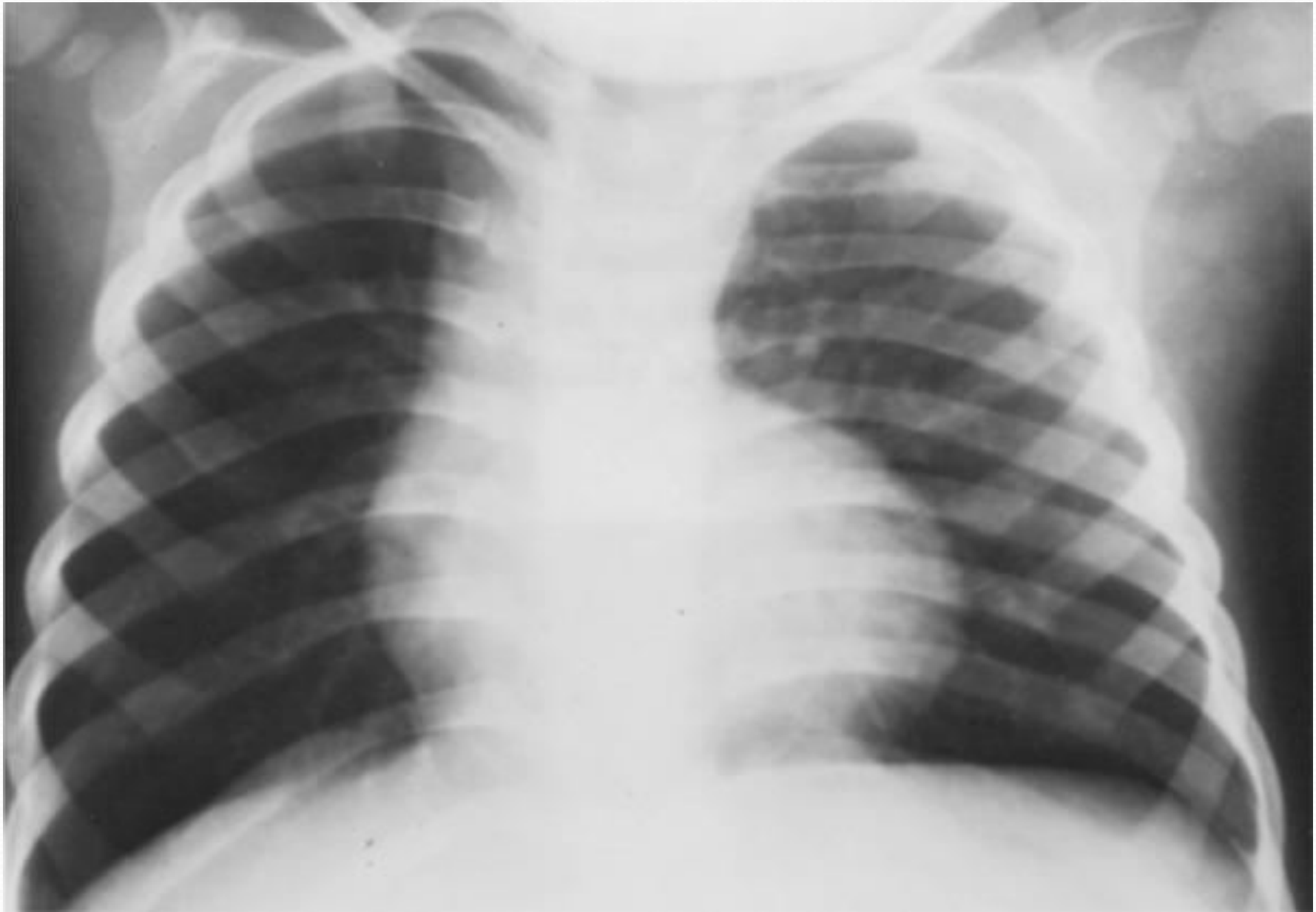
Pathology

- The VSD: membranous or infundibular, larger than isolated VSD
  - The aorta : predominantly from the right ventricle
  - 50% ASD
  - Right ventricular hypertrophy : moderate to severe
  - In most cases, the origin and distribution of the coronary arteries are normal

CLINICAL MANIFESTATIONS

- Cyanosis at birth or heart failure (rare). The degree of cyanosis depends on whether the ductus is patent and how extensive MAPCAs are.
- PDA closure → ↑ cyanosis.
- Continuous murmur from the PDA (during the first 4 to 6 weeks of life) or collaterals. The S2 is loud and single.
- The ECG shows RAD and RVH

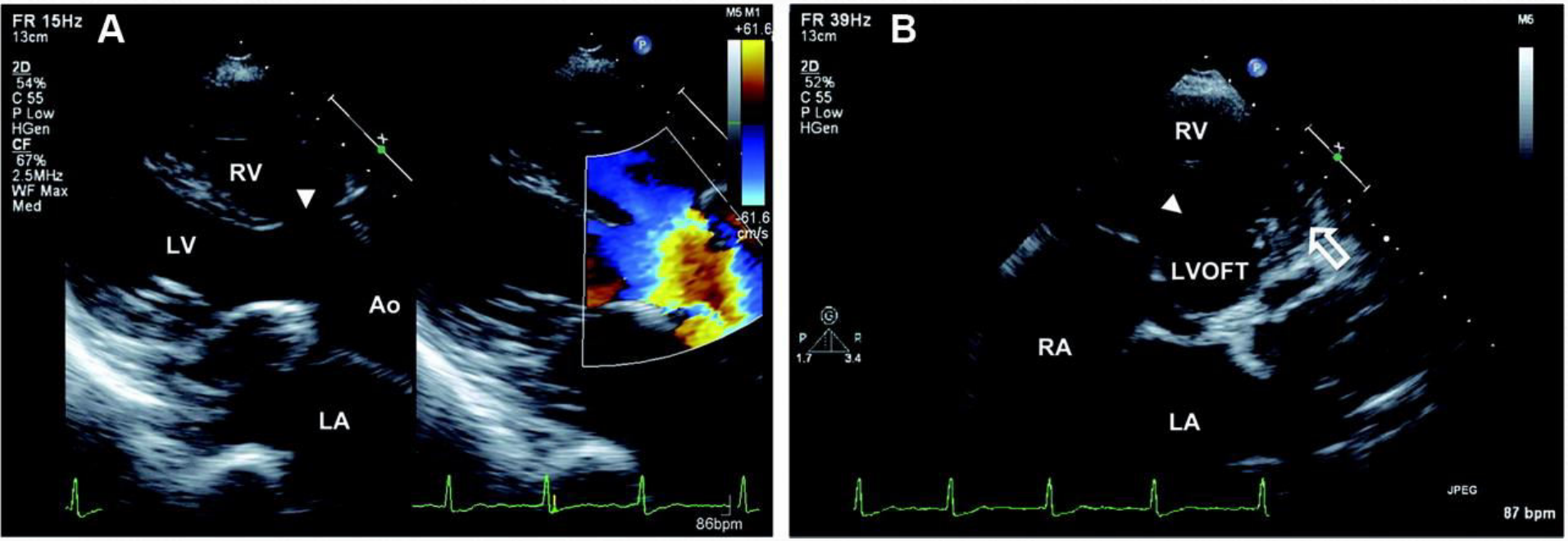
Chest x-ray films show a normal heart size. The heart often appears as a boot-shaped silhouette



Echocardiography

- VSD : number, position, the extent, relation to the valve
- PA : define pulmonary valve atresia , size, confluence, distribution →Mcgoon index, Zscore, Nakata index.
- The presence of PDA and MAPCAs
- Coronary arteries, additional defects : ASD....

Transthoracic echocardiographic examination.



Fukui D et al. Circulation 2011;124:2155-2157

American Heart Association
Learn and Live

Echocardiography

- Mcgoon index

  =(LPA + RPA)/ descending Ao

  LPA–RPA: prebranching-----Desceding Ao: just above diaphragm

  Normal value: 2-2.5

- Nakata index

  cross-sectional area of (LPA + RPA) / BSA

  Normal value: 330 ±30 mm^2/BSA

Clasification

Barbero-Marcial & Jatene, 1990

Group A

- All bronchopulmonary segments are supplied by central pulmonary arteries. Source of pulmonary blood flow usually from PDA or MAPCA.
- *Group A1:* Left and right NPA are either normal in size or hypoplastic, but are confluent and nonstenotic.
- *Group A2:* Central NPA are either stenotic or nonconfluent

Group B

- Some bronchopulmonary segments are supplied by central NPA and others by MAPCA.

Group C

- All bronchopulmonary segments supplied exclusively by MAPCA.

Tchervenkov & Roy, 2000

Type A

- NPA are present. There are no MAPCA.

Type B

- Both NPA and MAPCA present.

Type C

- No NPA. MAPCA only.

Cardiac Catheterization and Angiocardiography

- Pulmonary arteries: confluence, size, distribution, the true pressure and resistance→ Mcgoon, Nakata index, Z score
- PDA,MAPCAs:
  - the number, location and degree
  - the extent of the pulmonary arterial tree supplied by each collateral vessel and to determine which type of pulmonary artery connection is present
  - selective balloon occlusion techniques
- The existence of multiple VSDs and anatomy of the coronary arteries

Management

- Medical: PGE1 infusion→ keep PDA open
- Surgical: A connection the RV and true PA as early in life as possible.

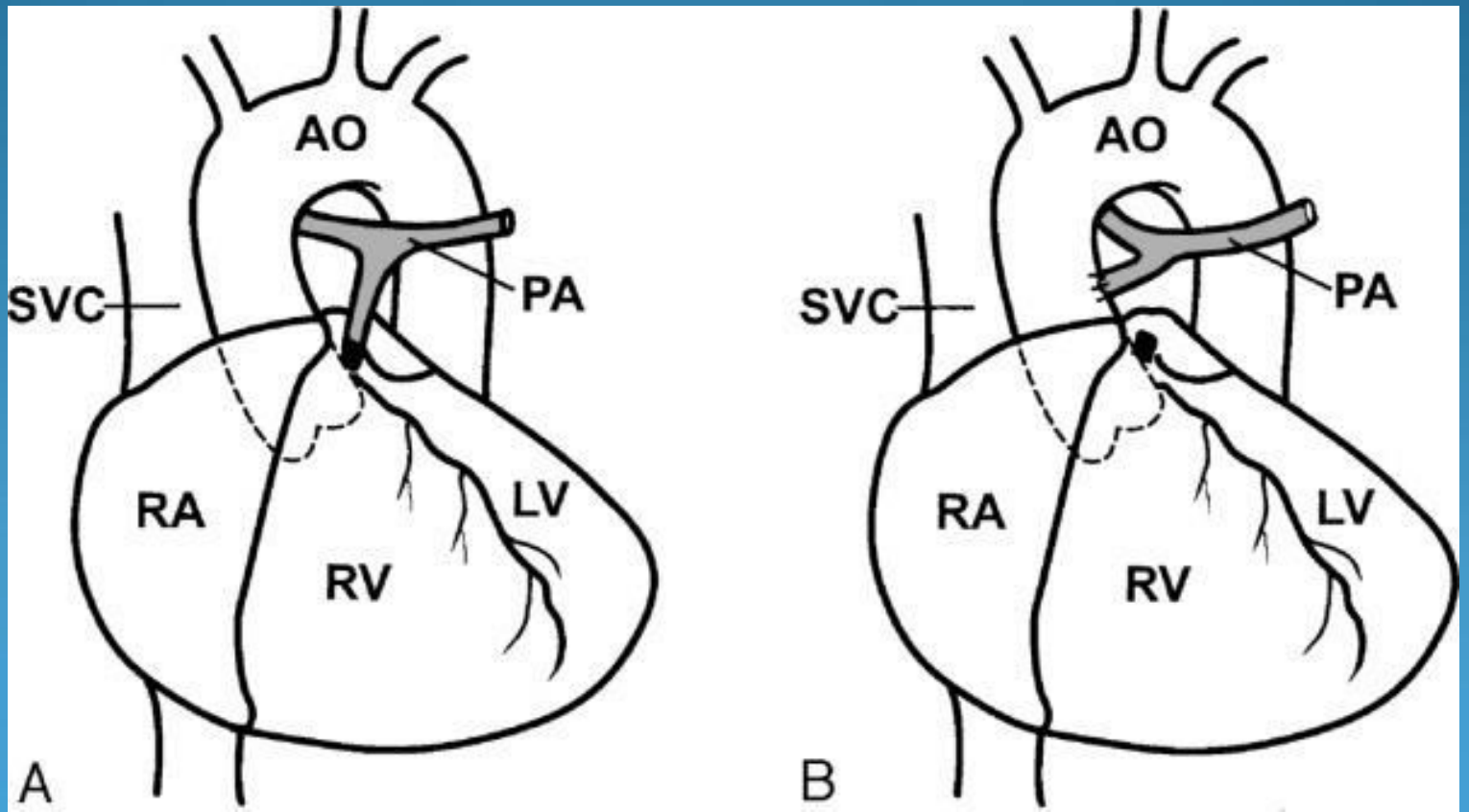
Management

Palliative operation:

systemic-pulmonary artery shunts, unifocalization

Cyanosis, small PA (Nakata index < 200), PDA stenosis

Central shunt > BT shunt



Management

Complete repair: RV- PA connection + closure VSD

1. Single-stage repair:
   - PA confluence, true PA provide most or all PBF + SpO2 >75%
   - the central PA connects without obstruction to sufficient regions of the lung
   - Nakata index ≥ 200
   - The mortality rate 5% and 20%

   →Closing the VSD, establishing continuity between the RV and the unifocalized PA

Management

2. Multiple-stage repair: three steps

- Step 1: RV–to–hypoplastic PA conduit, small homograft conduit (6 to 8 mm internal diameter)

  Catheterization : 3 to 6 months later

  occlude MAPCAs

  define the PA distribution

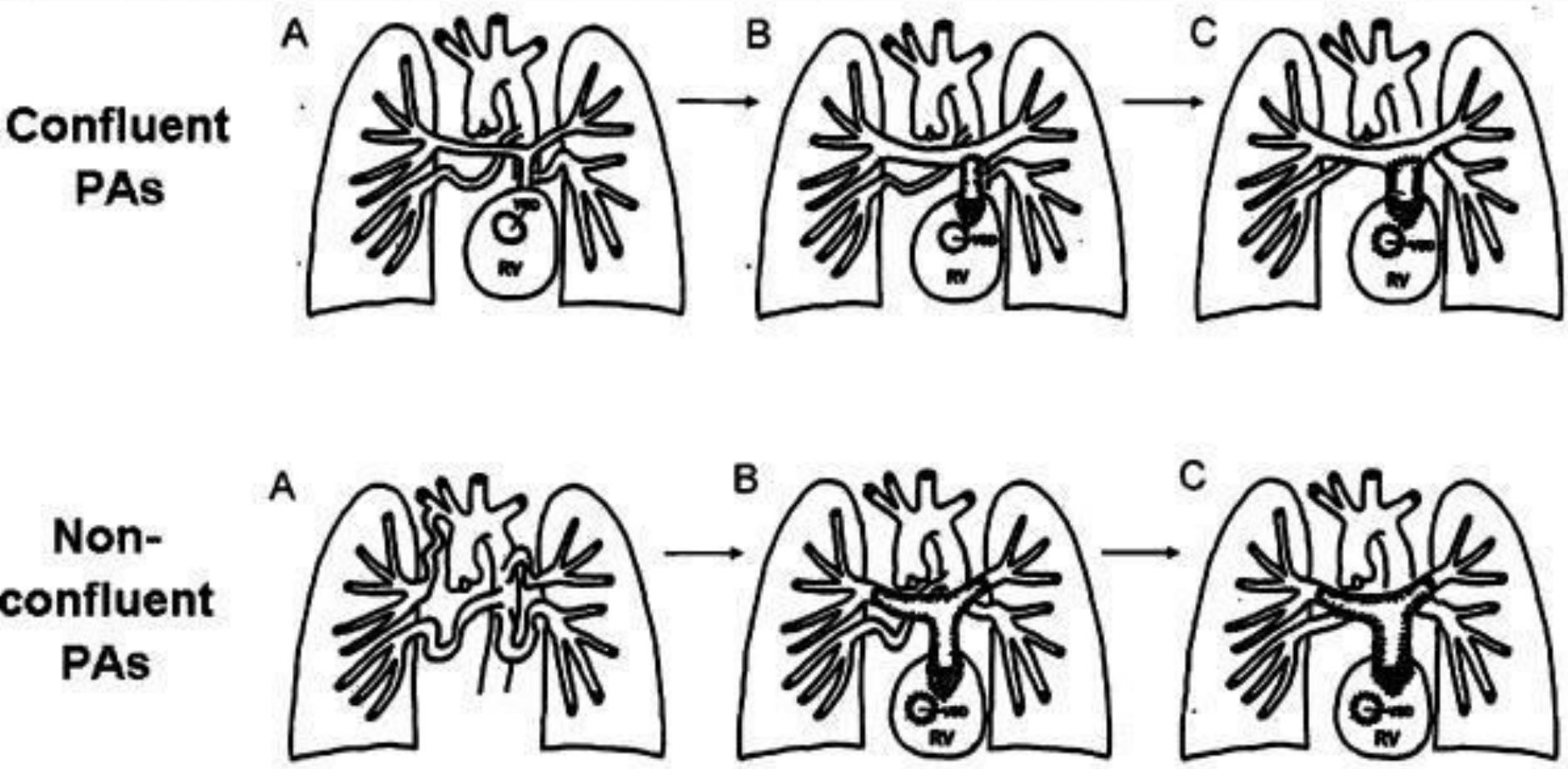
Management

- Step 2:
  - A unifocalization procedure
  - Catheterization 3 to 6 months later
    1. identify multiple peripheral stenosis in both the true and unifocalized collaterals
    2. balloon dilatation with or without stenting → further unifocalization procedures

Management

- Step 3:
  - Closure of VSD at 1 to 3 years of age.
  - The homograft conduit PA - RV.
    - RV pressure : 50% - 10% to 20% systemic pressure by ballooning or stenting
    - Central fenestration of 3 to 4 mm

Management



Tetralogy of Fallot with Pulmonary Atresia (or Pulmonary Atresia and VSD)

- **Confluent PAs with:**
  - **Favorable PA anatomy** (True PAs providing most PBF with O_2 sat >75%) → Single stage repair (VSD closure + RV-to-unifocalized PA connection)
  - **Hypoplastic PAs**
    - Central shunt → RV-PA connection + Unifocalization + VSD closure, later
    - RV-PA connection → Unifocalization → VSD closure
- **Nonconfluent PAs + MAPCAs** → RV-PA conduit → Unifocalization → VSD closure (6-8 mm homograft)

Reference

- Moss and Adam's Heart Disease in infants, Children, and Aldolescent : Including the fetus and Young Adult, 7th Ed
- Pediatric Cardiology for Practioner, 5th Ed

Thanks for your attention