Pulmonary atresia – Ventricular septal defect

Nguyễn Văn Đáng MD
Huỳnh Thanh Kiều MD
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 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 severe in patients with MAPCAs than in those with PDA.
- The size of the PA: amount of collateral arteries, position where they connect 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 $\rightarrow$ ↑ 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.
Echocardiography

- Mcgoon index
  \[ \frac{LPA + RPA}{\text{descending Ao}} \]
  LPA–RPA: prebranching----Desceding Ao: just above diaphragm
  Normal value: 2-2.5

- Nakata index
  \[ \text{cross-sectional area of } \frac{(LPA + RPA)}{\text{BSA}} \]
  Normal value: 330 ±30 mm²/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.
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 \(\rightarrow\) 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%)  
  - Hypoplastic PAs

  ![Diagram]

  - Central shunt  
    - RV-PA connection + Unifocalization + VSD closure, later
  - RV-PA connection  
    - Unifocalization  
    - VSD closure

- **Nonconfluent PAs**  
  - RV-PA conduit  
    - Unifocalization  
    - VSD closure
  + MAPCAs

  ![Diagram]

  (6-8 mm homograft)
Reference

- Moss and Adam’s Heart Disease in infants, Children, and Adolescent: Including the fetus and Young Adult, 7th Ed
- Pediatric Cardiology for Practitioner, 5th Ed
Thanks for your attention