AORTO PULMONARY COLLATERALS IN CHD

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I INTRODUCTION AND DEFINITION

- **Definition:** Aortopulmonary Collaterals (APCs) are systemic-to-pulmonary arterial vessels arising from the aorta or its major branches that conduct blood to the pulmonary arterial bed.
- **Embryology and Etiology:** Major Aortopulmonary Collateral Arteries (MAPCAs) represent persistence of embryonic splanchnic (intersegmental) arteries that normally regress once the sixth aortic arch connects the truncus arteriosus to the intrapulmonary vascular plexus. In cyanotic Congenital Heart Disease (CHD) with severely restricted or absent antegrade pulmonary blood flow, these vessels serve as the primary or sole source of pulmonary perfusion.
- **Core Pathology:** Classic feature of **Pulmonary Atresia with Ventricular Septal Defect (PA-VSD)** (also termed **Tetralogy of Fallot with Pulmonary Atresia [TOF-PA]**), where 30 to 65 percent of patients have MAPCAs. Acquired APCs also develop in **Tricuspid Atresia** and complex **Single Ventricle** physiology (particularly after Bidirectional Glenn or Fontan palliation).

II CLASSIFICATION OF COLLATERALS

A Major Aortopulmonary Collateral Arteries (MAPCAs) vs. Acquired APCs

- **Major Aortopulmonary Collateral Arteries (MAPCAs):** Congenital, large (greater than 2 to 3 mm) muscular systemic arteries, typically numbering 2 to 6. Most commonly originate from the anterior/lateral wall of the **descending thoracic aorta** (greater than 70 percent, near the carina), followed by the **subclavian arteries**, aortic arch, internal mammary arteries, bronchial arteries, or rarely the abdominal aorta/celiac trunk and coronary arteries. They frequently supply lung segments that lack connection to the central pulmonary arteries.
- **Acquired Collaterals:** Small, tortuous, and numerous vessels arising from bronchial, intercostal, or internal mammary arteries postnatally in response to chronic hypoxemia and non-pulsatile low pulmonary flow (commonly seen in older unoperated cyanotic patients and post-Glenn/Fontan circuits).

B Barbero-Marcial Classification of Pulmonary Supply in PA-VSD

- **Type A:** All bronchopulmonary segments are connected to confluent central pulmonary arteries supplied by a Patent Ductus Arteriosus (PDA); no MAPCAs are present.
- **Type B:** Both central pulmonary arteries (typically hypoplastic) and MAPCAs are present, sharing pulmonary blood supply (most common subtype).
- **Type C:** Complete absence of native central pulmonary arteries; pulmonary parenchyma is supplied exclusively by MAPCAs.

C Functional Supply: Essential (Sole Supply) vs. Dual Supply

- **Essential (Non-Communicating / Sole-Supply) Collaterals:** The bronchopulmonary segment is perfused **only** by the MAPCA with no connection to the native central pulmonary arterial tree. Ligating or coiling an essential collateral without surgical unifocalization causes **segmental lung infarction**.
- **Dual-Supply (Communicating / Non-Essential) Collaterals:** The lung segment is supplied by **both** the MAPCA and an arborizing native central pulmonary artery. These can be safely coiled or ligated without causing infarction.

III CLINICAL ROLE AND SIGNIFICANCE

- **Life-Sustaining Perfusion:** Provide essential pulmonary blood flow for gas exchange in the absence of a patent right ventricular outflow tract.
- **The Hemodynamic "Two-Edged Sword":**
  - **Volume Overload:** Large, unrestrictive collaterals act as a massive left-to-right shunt, causing pulmonary overcirculation and **Left Atrial and Left Ventricular dilation and heart failure**.
  - **Segmental Pulmonary Hypertension:** Unprotected MAPCAs arising from the high-pressure aorta expose individual pulmonary capillary beds to systemic pressures, causing rapid intimal proliferation and irreversible **Pulmonary Vascular Obstructive Disease (PVOD)** in those specific lung segments (while stenotic MAPCAs protect distal segments from PVOD).
  - **Systemic and Coronary Vascular Steal:** Significant diastolic runoff from the aorta into the low-resistance pulmonary bed lowers aortic diastolic pressure (producing bounding pulses and wide pulse pressure), potentially compromising coronary, cerebral, or mesenteric perfusion and increasing preload on a single ventricle.

IV BASIC INVESTIGATIONS (ECG, CXR, LABS)

- **12-Lead ECG:** Demonstrates **Right Ventricular Hypertrophy (RVH)** and **Right Axis Deviation**. In patients with massive collateral flow and left heart volume overload, **Biventricular Hypertrophy (BVH)** and Left Atrial Enlargement are seen.
- **Chest X-Ray (CXR):** Shows a **boot-shaped heart (coeur en sabot)** with a concave main pulmonary artery segment and a right aortic arch in 25 to 50 percent of cases. The hallmark parenchymal finding is **asymmetric pulmonary vascularity**, where some lung zones appear oligemic (dark) while adjacent zones appear plethoric.
- **Laboratory Evaluation:** Secondary erythrocytosis (elevated hemoglobin and hematocrit) in cyanotic patients, mandatory **iron studies** (as iron-deficient microcytosis increases hyperviscosity and stroke risk), and **22q11.2 deletion (DiGeorge Syndrome) genetic testing** (present in 40 to 50 percent of PA-VSD with MAPCAs).

V ADVANCED DIAGNOSTIC ALGORITHM

- **Transthoracic Echocardiography (TTE):** Defines intracardiac anatomy (large malalignment VSD, overriding aorta, absent RV-to-PA continuity, arch sidedness), but cannot adequately map the number and distal segmental arborization of MAPCAs.
- **Computed Tomography Angiography (CTA):** Crucial non-invasive 3D roadmap that maps the exact origin, retroesophageal or mediastinal course, stenoses, and segmental connections of every collateral, and measures native PA confluence size using the **Nakata Index** and **McGoon Ratio**.

Morphometric Index	Calculation Formula	Normal Value & Surgical Cut-off
Nakata Index (PAI)	(Cross-sectional area of RPA + LPA in mm2) divided by BSA in m2	Normal approx. 330 mm2/m2; greater than 150 to 200 mm2/m2 required for VSD closure
McGoon Ratio	(Diameter of RPA + Diameter of LPA) divided by Aortic diameter at diaphragm	Normal approx. 2.1; greater than 1.2 to 1.5 acceptable for repair
Total Neopulmonary Artery Index (TNPAI)	(CSA of Native PAs + CSA of Unifocalizable MAPCAs in mm2) divided by BSA in m2	Greater than 200 mm2/m2 predicts low post-repair RV/LV pressure ratio

- **Cardiac Catheterization and Selective Angiography:** Complementary to CTA because CTA cannot measure pressure. Catheterization measures **distal pressure** inside each MAPCA (identifying segments at risk of PVOD) and uses selective angiography and balloon occlusion trials to prove **dual supply vs. sole supply**.

VI MANAGEMENT STRATEGIES

- **Interventional (Transcatheter Coiling / Vascular Plugs):** Indicated for **dual-supply (non-essential) MAPCAs** or acquired APCs to reduce left heart volume overload, eliminate competitive flow, or control hemoptysis.
- **Surgical Unifocalization (Stanford / Hanley Paradigm):**
  - **Procedure:** Through a midline sternotomy, all sole-supply MAPCAs are detached from the aorta, mobilized, and anastomosed ("unifocalized") to the native central pulmonary arteries to construct a single, low-pressure **neopulmonary arterial tree**.
  - **Single-Stage Complete Repair:** If the reconstructed neopulmonary bed is adequate (confirmed by an intraoperative flow study at 2.5 to 3.0 L/min/m2 maintaining mean PA pressure at 25 mmHg or less), the VSD is closed and a valved **RV-to-PA conduit** is placed in the same operation. If PA pressure remains above 25 mmHg, the VSD is left open or fenestrated.

VII MANAGEMENT OF COMPLICATIONS

- **Hemoptysis:** Rupture of fragile, high-pressure tortuous MAPCAs or bronchial collaterals is managed via airway protection and urgent **transcatheter coil or vascular plug embolization**.
- **Post-Repair RV Hypertension and Low Cardiac Output:** Occurs if the cross-sectional area of the unifocalized pulmonary bed is insufficient or affected by segmental PVOD. Managed with **inhaled Nitric Oxide (iNO)**, **Milrinone**, avoidance of hypoxia/acidosis, and VSD fenestration if suprasystemic RV pressure persists.
- **Reperfusion Pulmonary Edema:** Acute unilateral or segmental pulmonary edema occurring in previously stenotic, hypoperfused lung segments suddenly exposed to full pulsatile flow after unifocalization; managed with PEEP and diuretics.

VIII CURRENT STATUS, EVIDENCE AND RECENT ADVANCES

- **Early Single-Stage Unifocalization (3 to 6 Months):** Large surgical series demonstrate greater than 85 percent complete repair and survival rates when unifocalization is performed early at **3 to 6 months of age**, minimizing the duration lung segments are exposed to systemic aortic pressures.
- **3D Virtual and Printed Surgical Planning:** Patient-specific 3D CT reconstructions are used to plan complex hilar and retroesophageal MAPCA anastomoses preoperatively, reducing cardiopulmonary bypass and cross-clamp times.
- **RV-to-PA Conduit Advances:** Decellularized pulmonary/aortic homografts and bovine jugular vein conduits (**Contegra**) are used for surgical RVOT reconstruction, while transcatheter pulmonary valves (**Melody** or **SAPIEN** in dysfunctional RV-PA conduits; **Harmony** in the native or patched RVOT) enable subsequent non-surgical treatment of RVOT dysfunction.

KEY REFERENCES

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