INDICATIONS OF CARDIAC CATHETERIZATION IN CCHD

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I THE PARADIGM SHIFT (THE MULTIMODALITY IMAGING ERA)

- **Evolution from Diagnostic to Therapeutic:** With the high diagnostic accuracy of **Transthoracic/Transesophageal Echocardiography (TTE/TEE)**, **Multidetector CT Angiography (MDCTA)**, and **Cardiac MRI (CMR)** for defining intracardiac and extracardiac anatomy, routine purely anatomical diagnostic catheterization in Cyanotic Congenital Heart Disease (CCHD) has largely been replaced.
- **Current Role of Catheterization:** Invasive cardiac catheterization in CCHD is now performed for two specific mandates: (1) **Invasive Hemodynamic Assessment** (measuring pressures, transpulmonary gradients, and Pulmonary Vascular Resistance [PVR] that non-invasive imaging cannot directly measure, or performing selective angiography of complex collateral beds), and (2) **Transcatheter Palliative or Definitive Interventions**.

II DIAGNOSTIC INDICATIONS (HEMODYNAMICS AND SELECTIVE ANATOMY)

- **Assessment of Pulmonary Vascular Resistance (PVR) and Operability (Class I):** The single most critical diagnostic indication in CCHD patients with unrestricted pulmonary blood flow (e.g., Truncus Arteriosus, d-TGA with VSD, unoperated Single Ventricle, or DORV without PS) presenting beyond infancy. Direct Fick or thermodilution measurement of **PVR** and **Qp:Qs** determines whether the patient is operable (**PVR less than 3 Wood Units [Class I]** or **3 to less than 5 Wood Units [Class IIa]** in ESC 2020 ACHD guidelines; indexed PVR less than 4 to 6 Wood Units·m2 in pediatric guidelines) versus inoperable **Eisenmenger physiology**.
- **Acute Vasoreactivity Testing:** In pediatric patients with borderline elevated PVR (4 to 8 Wood Units·m2), acute vasodilator challenge using **inhaled Nitric Oxide (iNO at 20 to 40 ppm) and/or 100 percent Oxygen** assesses pulmonary vascular reactivity (a drop in PVR index by greater than 20 percent to a final PVRi below 6 WU·m2 with a fall in the PVR/SVR ratio below 0.3 favors operability, often with a fenestrated patch). (Note: When high oxygen concentration is inhaled, dissolved plasma oxygen must be included in the **Fick equation** to avoid erroneous PVR calculation.)
- **Selective Mapping and Pressure Measurement in MAPCAs (TOF with Pulmonary Atresia):** While CT Angiography maps the 3D origins of **Major Aortopulmonary Collateral Arteries (MAPCAs)**, cardiac catheterization remains essential to (1) measure the **distal segmental pressure** within each individual MAPCA (identifying lung segments protected by proximal stenosis vs. those with segmental pulmonary hypertension), and (2) perform **selective MAPCA and pulmonary vein wedge angiography** to differentiate **dual-supply (communicating)** from **sole-supply (essential)** lung segments prior to unifocalization.
- **Pre-Glenn and Pre-Fontan Single-Ventricle Hemodynamic Staging:** Mandatory prior to Bidirectional Glenn or Fontan completion to confirm **mean PA pressure less than 15 mmHg**, **PVR less than 2 to 3 Wood Units**, **transpulmonary gradient less than 5 to 6 mmHg**, **systemic ventricular end-diastolic pressure (VEDP) less than 12 mmHg**, confluent pulmonary arteries (**Nakata Index and McGoon Ratio**), and to identify **aortopulmonary or venovenous collaterals**.
- **Coronary Artery Assessment in PA-IVS:** In neonates with **Pulmonary Atresia with Intact Ventricular Septum (PA-IVS)** and a diminutive, hypertensive right ventricle, **right ventricular angiography and selective coronary angiography** are mandatory to identify **Right Ventricle-Dependent Coronary Circulation (RVDCC)** (ventriculocoronary fistulae/sinusoids with proximal coronary artery stenoses/interruptions); if RVDCC is present, decompressing the RV causes fatal myocardial infarction, mandating a single-ventricle pathway or transplant.

CCHD Clinical Scenario	Primary Catheterization Objective	Key Hemodynamic / Angiographic Target
Late-Presenting CCHD with High Pulmonary Flow (d-TGA+VSD, Truncus, Single Ventricle)	Assess **PVR, Qp:Qs, and Operability** (+/- iNO + O2 vasoreactivity)	**PVR <3 WU (Class I)** or **3 to <5 WU (Class IIa)** in adults; pediatric PVRi <4–6 WU·m2 and PVR/SVR <0.3
TOF with Pulmonary Atresia & MAPCAs	Selective MAPCA angiography & distal pressures; PV wedge injections	Differentiate **sole-supply (essential)** vs. **dual-supply** segments; coil dual-supply collaterals
Pre-Glenn / Pre-Fontan Staging	Measure passive cavopulmonary hemodynamics and collateral burden	**mPAP <15 mmHg, PVR <2–3 WU, VEDP <12 mmHg**, Nakata Index >200 mm2/m2
Pulmonary Atresia with Intact Septum (PA-IVS)	RV angiogram + aortogram/coronary angiogram	Rule out **RV-Dependent Coronary Circulation (RVDCC)** before attempting RV decompression

III THERAPEUTIC AND INTERVENTIONAL INDICATIONS

- **Balloon Atrial Septostomy (Rashkind Procedure — Class I):** Emergency life-saving bedside or cath-lab procedure for neonates with **d-Transposition of the Great Arteries (d-TGA)** and a restrictive interatrial communication presenting with profound hypoxemia and metabolic acidosis (also indicated in Tricuspid Atresia, Pulmonary Atresia, or HLHS with a restrictive ASD). Enlarges the foramen ovale to allow bidirectional interatrial mixing and decompress the left atrium.
- **Transcatheter Patent Ductus Arteriosus (PDA) Stenting:** Frontline neonatal alternative to a surgical modified Blalock-Thomas-Taussig (mBTT) shunt in neonates with **duct-dependent pulmonary blood flow** (e.g., Pulmonary Atresia, Tricuspid Atresia, severe TOF), promoting symmetrical branch PA growth with lower procedural morbidity and shorter ICU stay.
- **Right Ventricular Outflow Tract (RVOT) Stenting:** Palliative transcatheter bridge to complete surgical repair in high-risk, low-birth-weight, or severely cyanotic infants with **Tetralogy of Fallot** and refractory hypercyanotic spells or hypoplastic branch pulmonary arteries.
- **Balloon Pulmonary Valvuloplasty (BPV) and Radiofrequency Valve Perforation:** Definitive emergency therapy for neonates with **Critical Valvular Pulmonary Stenosis** (duct-dependent right-to-left atrial shunt), or **radiofrequency wire perforation followed by BPV** in membranous **PA-IVS** with an adequate tripartite RV and no RVDCC.
- **Transcatheter Embolization of Vascular Collaterals:** Coil or vascular plug occlusion of **dual-supply MAPCAs** (to eliminate left heart volume overload before bypass), **acquired aortopulmonary collaterals** in Glenn/Fontan circuits, or **venovenous collaterals** and **pulmonary arteriovenous malformations (PAVMs)** causing progressive cyanosis.
- **Conduit, Branch PA, and Fontan Fenestration Interventions:** Balloon angioplasty and stenting of stenotic **RV-to-PA conduits** or branch pulmonary arteries, **Transcatheter Pulmonary Valve Replacement (TPVR)**, and percutaneous creation, stenting, or device closure of a **Fontan fenestration**.

IV PRACTICAL CONSIDERATIONS IN THE INDIAN CLINICAL CONTEXT

- **High Volume of Late-Presenting Operability Studies:** Because many Indian children with CCHD and increased pulmonary blood flow or large MAPCAs present in late childhood or adolescence, invasive **PVR operability assessment** and selective collateral catheterization remain everyday procedures in Indian tertiary centers.
- **Transcatheter Palliation in Malnourished Infants (RVOT and PDA Stenting):** Indian infants with severe TOF or duct-dependent CCHD frequently present with **severe protein-energy malnutrition, low weight (under 3 to 4 kg), or active respiratory infection** where neonatal cardiopulmonary bypass carries high mortality; **RVOT stenting or PDA stenting** serves as a cost-effective, life-saving bridge allowing somatic growth and pulmonary artery rehabilitation over 4 to 6 months before definitive surgical repair.
- **Stabilization for Transport (Rashkind Septostomy):** In neonates with d-TGA born outside major cardiac surgical hubs, urgent **Echo-guided or fluoroscopic Rashkind Balloon Atrial Septostomy** stabilizes arterial oxygenation, allowing weaning from costly **Prostaglandin E1 (Alprostadil)** infusions and safe transport to a surgical center.

KEY REFERENCES

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