PRE OP EVALUATION OF TOF

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I INTRODUCTION / DEFINITION

- **Definition:** Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart defect. It stems from a singular morphogenetic anomaly: the anterocephalic deviation of the infundibular septum.
- **The Tetrad:** This single deviation produces four anatomical hallmarks: a large, malaligned subaortic Ventricular Septal Defect (VSD), overriding of the aorta, Right Ventricular Outflow Tract (RVOT) obstruction (multilevel: infundibular, valvular, or supravalvular), and consequent Right Ventricular Hypertrophy (RVH).
- **Goal of Pre-Operative Assessment:** To systematically map the RVOT, evaluate the pulmonary arterial tree, rule out dangerous coronary anomalies, and optimize the patient's hemodynamic and hematologic status prior to surgical intervention.

II CLINICAL FEATURES & RISK STRATIFICATION

- **Clinical Presentation:** Ranges from asymptomatic murmurs in the neonatal period ("pink tet") to profound cyanosis, clubbing, and severe exertional dyspnea in older infants.
- **Auscultation:** A loud, harsh crescendo-decrescendo systolic ejection murmur at the left upper sternal border. **Crucial Exam Point:** The murmur originates from the RVOT obstruction, *not* the VSD (which is large and unrestrictive). The *shorter* and *softer* the murmur, the more severe the obstruction and right-to-left shunting.
- **Pre-Operative Risk Stratification:**
  - **Anatomical High-Risk Features:** Anomalous Left Anterior Descending (LAD) artery crossing the RVOT (5–9% prevalence), hypoplastic branch pulmonary arteries (Nakata Index < 150 mm^2/m^2), and the presence of Major Aortopulmonary Collateral Arteries (MAPCAs).
  - **Syndromic Risk (DiGeorge Syndrome):** 22q11.2 microdeletion is present in ~15% of TOF patients. It mandates pre-operative screening as it is associated with thymic aplasia (immune deficiency) and profound post-operative hypocalcemia.

III BASIC INVESTIGATIONS (ECG, CXR, LABS)

- **12-Lead Electrocardiogram (ECG):**
  - **Findings:** Right Axis Deviation (RAD) and prominent Right Ventricular Hypertrophy (RVH). Evidenced by tall R waves in V1, deep S waves in V5/V6, and a positive T wave in V1 after 3 days of age (a reliable sign of systemic RV pressure). Right Atrial Enlargement (RAE) may present as peaked P waves in lead II.
- **Chest X-Ray (CXR):**
  - **Findings:** Classic **"Coeur en Sabot" (Boot-shaped heart)**. Caused by the uplifted cardiac apex (due to RVH) and a concave main pulmonary artery segment.
  - **Pulmonary Vasculature:** Decreased pulmonary vascular markings (oligemia) due to restricted pulmonary blood flow.
  - **Aortic Arch:** Right-sided aortic arch is visible in 25% of patients (crucial for planning palliative shunt laterality).
- **Laboratory Investigations:**
  - **Complete Blood Count (CBC):** Essential to evaluate for secondary polycythemia. A normal or low hemoglobin in a cyanotic TOF patient indicates severe **relative iron deficiency anemia**, which drastically increases the risk of hyperviscosity syndrome and stroke.
  - **Metabolic Panel:** Baseline renal function and serum calcium (anticipating DiGeorge syndrome hypocalcemia).
  - **Genetic Testing:** Microarray analysis for the 22q11.2 deletion is standard pre-operative care.

IV ADVANCED DIAGNOSTIC ALGORITHM (ECHO, CT, CMR, CATH)

- **Transthoracic Echocardiography (TTE) (Class I):**
  - **Primary Modality:** Defines VSD size and exact degree of aortic override (< 50% for TOF).
  - **Annular Sizing:** Measures the Pulmonary Valve (PV) annulus diameter to calculate the **Z-score**. A Z-score of **< -3 or -4** strongly predicts the surgical necessity of a Transannular Patch (TAP).
- **Computed Tomography Angiography (CTA) (Class I/IIa):**
  - **Current Status:** The gold standard for pre-operative anatomical mapping. Exquisitely delineates anomalous coronaries crossing the RVOT (preventing fatal surgical transection), calculates the true Nakata Index for distal branch pulmonary arteries, and maps MAPCAs.
- **Diagnostic Cardiac Catheterization (Class IIa/IIb):**
  - **Current Status:** Routine use is obsolete. Strictly reserved for mapping complex MAPCAs invisible on CTA or measuring exact Pulmonary Vascular Resistance (PVR) in delayed, older presentations.

V MEDICAL MANAGEMENT & PHARMACOTHERAPY

- **Hypercyanotic ("Tet") Spell Management:**
  - **Immediate Posture:** Knee-to-chest position (increases systemic vascular resistance [SVR], forcing blood across the RVOT) and 100% supplemental Oxygen.
  - **Morphine Sulfate:** 0.1 to 0.2 mg/kg IV or SC. Depresses the respiratory center to break the hyperpnea cycle.
  - **IV Fluid Bolus:** 15 to 20 mL/kg of isotonic crystalloid to rapidly increase RV preload and physically distend the RVOT.
  - **Beta-Blockers (Class I):** Esmolol 0.5 mg/kg IV slow bolus over 1 minute (followed by 50 mcg/kg/min infusion), or Propranolol 0.1 mg/kg IV slow push. Directly relieves dynamic infundibular muscle spasm.
  - **Vasopressors (Refractory):** Phenylephrine 5 to 10 mcg/kg IV bolus, followed by 0.1 to 0.5 mcg/kg/min infusion to maximally drive up SVR.
- **Pre-Operative Optimization:** Oral iron supplementation (Ferrous Sulfate 3-6 mg/kg/day of elemental iron) if relative anemia is detected to improve red blood cell deformability.

VI INTERVENTIONAL & SURGICAL MANAGEMENT

- **Elective Complete Primary Repair (Class I):**
  - **Timing:** Optimally performed between **3 to 6 months of age**. Includes VSD patch closure and relief of RVOT obstruction (via infundibular muscle resection and, if necessary, a transannular patch).
- **Pre-Operative Palliation (Class IIa):**
  - **Indications:** Symptomatic neonates with unfavorable anatomy (severe pulmonary hypoplasia, extreme prematurity) unable to tolerate primary repair.
  - **Current Paradigm:** Percutaneous RVOT stenting is rapidly replacing the surgical modified Blalock-Taussig-Thomas (mBTT) shunt as it promotes highly symmetrical branch pulmonary artery growth.

VII MANAGEMENT OF COMPLICATIONS

- **Peri-Operative Arrhythmias:** Junctional Ectopic Tachycardia (JET) is a classic post-operative complication following VSD patch suturing near the AV node. Managed with core cooling (mild hypothermia 34-35°C), minimizing inotropes, and Amiodarone infusion (5 mg/kg over 1 hour).
- **Restrictive Right Ventricular Physiology:** Post-operative RV diastolic dysfunction causing low cardiac output. Managed by prolonging mechanical ventilation, maintaining high central venous pressures (CVP 12-15 mmHg) to drive pulmonary flow, and judicious use of Milrinone (0.5 mcg/kg/min).

VIII LIFESTYLE, FOLLOW-UP & PROGNOSIS

- **Endocarditis Prophylaxis:** Required for 6 months post-complete repair, or lifelong if residual defects remain adjacent to prosthetic patch material.
- **Long-Term Surveillance:** All patients face the inevitable consequence of **severe Pulmonary Regurgitation (PR)** if a transannular patch was used. Annual Echocardiography and serial CMR (starting in late childhood/adolescence) are mandated to track RV End-Diastolic Volume Index (RVEDVi) to appropriately time Percutaneous Pulmonary Valve Implantation (PPVI) before irreversible RV failure occurs.

IX CURRENT STATUS, EVIDENCE & PARADIGM SHIFTS (2025 UPDATES)

- **The Valve-Sparing Paradigm:** Aggressive shift away from routine transannular patches. Advanced intraoperative balloon dilation and meticulous infundibular sparing techniques now allow preservation of native pulmonary valves even with borderline pre-op Z-scores (-2.5), significantly delaying the onset of PR and the need for future re-intervention.
- **Advanced 3D Modeling:** High-resolution pre-operative CTA datasets are now routinely utilized to 3D-print patient-specific cardiac models in complex TOF variants (e.g., crossing coronaries). This enables the surgical team to physically pre-plan the specific geometry of the RVOT baffle, helping to shorten operative and bypass times.