MEDICAL AND SURGICAL TREATMENT OF TETRALOGY OF FALLOT (TOF)

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I INTRODUCTION AND PATHOPHYSIOLOGIC TARGETS

- **The Core Principle:** Management of Tetralogy of Fallot (TOF) focuses on balancing the systemic and pulmonary circulations by addressing its four anatomical components (resulting from anterocephalad malalignment of the infundibular septum): a large non-restrictive subaortic **Ventricular Septal Defect (VSD)**, **Right Ventricular Outflow Tract (RVOT) obstruction**, **overriding aorta** (less than 50 percent override), and **concentric Right Ventricular Hypertrophy (RVH)**.
- **The Clinical Goal:** To maintain adequate pulmonary blood flow, prevent or abort acute hypercyanotic **"Tet Spells"**, and perform timely complete intracardiac repair (ideally preserving native pulmonary valve competence) to prevent chronic right ventricular failure and fatal arrhythmias.

II MEDICAL MANAGEMENT AND THE HYPERCYANOTIC SPELL (TET SPELL)

A Acute Emergency Management of Hypercyanotic Spells

- **Knee-Chest Position / Squatting:** Immediately place the infant in the knee-chest position on the caregiver's shoulder to calm the child and kink the femoral arteries, which **acutely increases Systemic Vascular Resistance (SVR)**, reduces right-to-left shunting across the VSD, and forces blood into the RVOT.
- **Humidified 100% Oxygen:** Administered to reduce hypoxic pulmonary vasoconstriction and improve systemic arterial oxygen saturation.
- **Morphine Sulfate (0.1 to 0.2 mg/kg IV, IM, or SC) or Ketamine (1 to 2 mg/kg IV / 3 to 5 mg/kg IM): Morphine** suppresses the medullary respiratory center to abolish paroxysmal hyperpnea and blunts sympathetic tone (Ketamine is an effective alternative that simultaneously sedates the child and elevates SVR).
- **Intravenous Fluid Bolus (10 to 20 mL/kg Isotonic Crystalloid):** Normal saline bolus increases right ventricular preload, distends the dynamic subpulmonary infundibulum, and lowers blood viscosity. (Add **Sodium Bicarbonate 1 to 2 mEq/kg IV** if severe metabolic acidosis persists, breaking acidemia-driven hyperpnea.)
- **Intravenous Beta-Blockers: Propranolol (0.05 to 0.15 mg/kg slow IV push)** or **Esmolol (500 mcg/kg IV bolus over 1 minute followed by 50 to 200 mcg/kg/min infusion)** relieves catecholamine-driven dynamic **infundibular muscle spasm** and slows heart rate to augment diastolic RV filling.
- **Alpha-Adrenergic Vasopressor (Phenylephrine): Phenylephrine (5 to 20 mcg/kg IV bolus followed by 0.1 to 0.5 mcg/kg/min infusion)** potently elevates SVR, raising left ventricular pressure to drive blood from the RV into the pulmonary artery.
- **Refractory Spell Rescue:** Intubation, general anesthesia, and neuromuscular paralysis, followed by emergency **transcatheter RVOT stenting**, **Modified Blalock-Thomas-Taussig (mBTT) shunt**, or urgent primary surgical repair.

B Chronic Medical Optimization (Bridge to Surgery)

- **Prostaglandin E1 (Alprostadil, 0.01 to 0.05 mcg/kg/min IV):** Mandatory in severely cyanotic neonates with critical RVOT obstruction or pulmonary atresia to maintain patency of the ductus arteriosus until neonatal intervention.
- **Oral Propranolol (1 to 4 mg/kg/day in 3 divided doses):** Used as a temporary medical bridge to prevent recurrent hypercyanotic spells in infants awaiting surgical repair.
- **Iron Supplementation and Avoidance of Dehydration:** Iron deficiency (even with an apparently "normal" hemoglobin of 11 to 13 g/dL, which represents relative anemia in cyanotic CHD) produces rigid, microcytic erythrocytes that precipitate hypercyanotic spells and **cerebral thromboembolism/stroke** in children under 2 years of age; oral iron supplementation is mandatory.

III SURGICAL TREATMENT: PALLIATIVE VS. DEFINITIVE REPAIR

- **Optimal Timing of Elective Primary Repair:** Contemporary standard of care is **elective single-stage complete intracardiac repair between 3 and 6 months of age** in stable infants, avoiding prolonged cyanosis and RV fibrosis.

A Palliative Procedures (Bridge to Complete Repair in High-Risk Neonates)

- **Indications:** Prematurity or low birth weight (less than 2.5 kg), severely hypoplastic branch pulmonary arteries (Nakata Index less than 150 mm2/m2), anomalous **LAD arising from the RCA crossing the RVOT**, or severe extracardiac comorbidities.
- **Modified Blalock-Thomas-Taussig (mBTT) Shunt:** Interposition of a **3.0 to 4.0 mm Gore-Tex (PTFE) tube graft** between the subclavian/innominate artery and the ipsilateral branch pulmonary artery (preserving the native subclavian artery, unlike the classical Blalock-Taussig shunt).
- **Transcatheter RVOT Stenting or PDA Stenting:** Increasingly preferred over surgical shunts in high-risk neonates, providing pulsatile antegrade pulmonary blood flow (RVOT stent) and symmetrical branch PA growth without a thoracotomy or sternotomy.

B Definitive Complete Intracardiac Repair

- **Transatrial-Transpulmonary Approach (Modern Standard):** Performed via the right atrium (through the tricuspid valve) and main pulmonary artery, minimizing or avoiding a large right ventriculotomy incision to prevent late scar-related ventricular arrhythmias.
- **Patch Closure of the VSD:** Dacron or bovine pericardial patch closure of the malalignment VSD, baffling the overriding aorta exclusively to the left ventricle while avoiding the **Bundle of His** along the posteroinferior margin of the defect.
- **Relief of RVOT Obstruction:**
  - **Valve-Sparing Repair (Preferred Strategy):** Resection of obstructing infundibular muscle bundles combined with pulmonary valvotomy/commissurotomy (and intraoperative balloon dilation if needed) when the pulmonary valve annulus **Z-score is greater than -2 to -3**, preserving pulmonary valve competence.
  - **Transannular Patch (TAP):** Required when the pulmonary annulus is severely hypoplastic (**Z-score less than -3**); an incision is extended across the pulmonary valve ring onto the infundibulum and augmented with a pericardial patch (often with a monocusp valve), which relieves obstruction at the price of **chronic free Pulmonary Regurgitation (PR)**.
  - **RV-to-PA Valved Conduit:** Required in **TOF with Pulmonary Atresia** or when an **anomalous LAD crosses the RVOT** (precluding a transannular incision).

IV LATE POST-OPERATIVE COMPLICATIONS AND SURVEILLANCE (ESC 2020 ACHD GUIDELINES)

- **Chronic Severe Pulmonary Regurgitation (PR):** The hallmark late hemodynamic lesion after a transannular patch, leading to progressive **Right Ventricular (RV) dilation, RV systolic dysfunction, functional Tricuspid Regurgitation, right heart failure, and exercise intolerance**.
- **Arrhythmias and Sudden Cardiac Death (SCD): Right Bundle Branch Block (RBBB)** is present in over 90 percent of repaired patients. Progressive RV stretch and surgical scars (between the VSD patch/ventriculotomy and tricuspid/pulmonary annuli) create slow-conduction anatomical isthmuses that cause **Intra-Atrial Reentrant Tachycardia (IART)** and **Sustained Monomorphic Ventricular Tachycardia**, with a **QRS duration of 180 ms or greater (Gatzoulis sign)** and **CMR Late Gadolinium Enhancement (LGE)** predicting SCD.
- **Aortic Root Dilation and Aortic Regurgitation:** Progressive dilation of the overriding aortic root occurs in 15 to 30 percent of adults with repaired TOF (due to intrinsic medial cystic necrosis and early volume load), occasionally causing aortic regurgitation.

V TRANSCATHETER AND SURGICAL PULMONARY VALVE REPLACEMENT (TPVR / PVR — ESC 2020 ACHD)

- **Devices:** Balloon-expandable valves (**Melody** and **Edwards SAPIEN 3**) for prior surgical conduits/bioprostheses, and self-expanding platforms (**Harmony** and **Alterra Adaptive Prestent**) for large native/transannular-patched RVOTs.

Patient Category (Repaired TOF with Severe PR)	ESC 2020 Class of Recommendation	Objective Clinical / Cardiac MRI (CMR) Criteria for PVR / TPVR
Symptomatic Patients	**Class I (Level C)**	Severe PR (CMR regurgitant fraction >30–40%) and/or moderate-to-severe RVOTO with **cardiovascular symptoms or exercise intolerance**
Asymptomatic Patients (CMR Volumetric / Functional Triggers)	**Class IIa (Level C)**	Severe PR/RVOTO with **any one** of: (1) **RVEDVi >=160 mL/m2**; (2) **RVESVi >=80 mL/m2**; (3) Progressive RV systolic dysfunction; (4) Progressive moderate-to-severe **Tricuspid Regurgitation**; (5) **RVSP >80 mmHg**; or (6) Sustained tachyarrhythmias

VI PRACTICAL CONSIDERATIONS IN THE INDIAN CLINICAL CONTEXT

- **Late-Presenting Adolescent and Adult Unoperated TOF:** In India, unoperated TOF frequently presents in adolescence or adulthood (including acyanotic **"Pink TOF"** with mild RVOT obstruction). Older unoperated patients exhibit severe concentric RV hypertrophy, myocardial fibrosis, LV underfilling (risking post-operative transient LV pulmonary edema after repair), and prominent **aortopulmonary collaterals**; preoperative **MDCTA** and **selective coil embolization of large dual-supply collaterals** are essential to prevent intraoperative steal and postoperative pulmonary hemorrhage.
- **Prioritizing Valve-Sparing Repair:** Because long-term serial Cardiac MRI follow-up and costly repeat surgical or transcatheter pulmonary valve replacements (**TPVR**) pose major financial barriers in India, Indian congenital surgeons prioritize **valve-sparing transatrial-transpulmonary repair** (accepting a mild residual RVOT peak gradient of 20 to 30 mmHg) over routine transannular patching whenever the annulus allows.

KEY REFERENCES

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