PRE OP ASSESSMENT OF TOF; CURRENT STATUS; RECOMMENDATIONS

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I INTRODUCTION AND EMBRYOLOGICAL DEFINITION

- **Morphogenetic Definition (Van Praagh Monology):** Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart defect beyond the neonatal period, resulting from a single primary developmental abnormality: **anterocephalad (anterior and superior) malalignment/deviation of the outlet (infundibular/conal) septum** relative to the trabecular muscular septum. This single embryological shift produces all four components of the tetrad: (1) a large non-restrictive **subaortic malalignment Ventricular Septal Defect (VSD)**, (2) **Right Ventricular Outflow Tract (RVOT) obstruction** (subvalvular infundibular, valvular, and/or supravalvular), (3) **overriding of the aorta** (less than 50 percent override over the RV; >50 percent RV origin is classified as **Double Outlet Right Ventricle [DORV]**), and (4) **concentric Right Ventricular Hypertrophy (RVH)** secondary to systemic RV pressure.
- **Objectives of Pre-Operative Assessment:** To define every level of RVOT and branch pulmonary artery stenosis, measure the pulmonary valve annulus Z-score, **exclude anomalous coronary arteries crossing the RVOT**, identify additional muscular VSDs or aortopulmonary collaterals, screen for genetic syndromes (22q11.2 deletion), and select the optimal timing for **valve-sparing primary repair vs. staged palliation**.

II PRE-OPERATIVE RISK STRATIFICATION

A High-Risk Anatomical Features (Dictating Surgical Modification)

- **Anomalous Coronary Artery Crossing the RVOT (Present in 5 to 9 Percent of TOF):** Most commonly an **anomalous Left Anterior Descending (LAD) artery arising from the Right Coronary Artery (RCA) or right sinus of Valsalva**, a single coronary ostium, or a large dual LAD/conal branch coursing directly across the anterior subpulmonary infundibulum. **Surgical Impact:** Precludes a standard transannular ventriculotomy incision (as inadvertent transection causes fatal intraoperative anterior myocardial infarction); mandates either a **transatrial-transpulmonary repair**, a **tailored oblique ventriculotomy parallel to the coronary**, a **double-barrel technique**, or an **extracardiac RV-to-PA conduit**.
- **Hypoplastic Branch Pulmonary Arteries and Annular Z-Score:**
  - **Pulmonary Valve Annulus Z-Score:** A **Z-score greater than -2 to -3** generally permits **valve-sparing repair**, whereas a severely hypoplastic annulus (**Z-score less than -3 to -4**) predicts the requirement for a **Transannular Patch (TAP)**.
  - **Nakata Index (PAI) and McGoon Ratio:** A **Nakata Index less than 150 mm2/m2** or **McGoon Ratio less than 1.2** indicates severe branch pulmonary artery hypoplasia, predicting suprasystemic post-repair RV pressure and favoring **initial palliation (RVOT stent or mBTT shunt)** to grow the pulmonary arterial tree before VSD closure.
  - **Major Aortopulmonary Collateral Arteries (MAPCAs) and Discontinuous PAs:** Present in **TOF with Pulmonary Atresia (PA-VSD)**; mandates complete preoperative CTA + catheter mapping to plan **surgical unifocalization**.
  - **Additional ("Swiss-Cheese") Apical Muscular VSDs or Complete AVSD ("Tet-Canal"):** Unrecognized second muscular VSDs cause acute postoperative left-to-right volume overload and heart failure after RVOT relief, while **TOF with complete Atrioventricular Septal Defect** (strongly associated with **Down Syndrome / Trisomy 21**) requires complex two-patch or modified single-patch repair.

B Clinical, Syndromic, and STAT Mortality Risk Factors

- **Patient Factors: Prematurity, low birth weight (less than 2.5 kg)**, and **prostaglandin-dependent neonatal cyanosis or refractory ventilated hypercyanotic spells** carry higher morbidity with neonatal bypass and often favor initial **transcatheter RVOT/PDA stenting**.
- **Genetic Syndromes (Present in 20 to 30 Percent of TOF):**
  - **22q11.2 Microdeletion (DiGeorge / Velocardiofacial Syndrome — Present in ~15% of classical TOF and up to 40–50% of TOF with Pulmonary Atresia or Right Aortic Arch):** Mandates preoperative screening (FISH / chromosomal microarray) because **thymic hypoplasia** requires **irradiated, leukoreduced blood products** (to prevent fatal transfusion-associated graft-versus-host disease), **parathyroid hypoplasia** causes severe **perioperative hypocalcemia**, and velopharyngeal/airway anomalies complicate extubation.
  - **Other Associations: Trisomy 21** (associated with TOF + AVSD), **Alagille Syndrome (*JAG1*)** (peripheral branch PA stenosis + cholestasis), and **CHARGE / VACTERL** associations.
  - **STAT Mortality Categories (Society of Thoracic Surgeons — EACTS):** Standard primary TOF repair is **STAT Category 1 to 2** (operative mortality **~1 percent**), whereas **TOF with Pulmonary Atresia and MAPCAs** or neonatal repair with severe comorbidities rises to **STAT Category 4**.

III MULTIMODALITY PRE-OPERATIVE DIAGNOSTIC ALGORITHM

Imaging Modality	Guideline Status	Specific Pre-Operative Parameters Evaluated
Transthoracic Echocardiography (TTE)	**Class I (Mandatory First-Line in All Patients)**	Malalignment VSD size & **<50% aortic override** (excludes DORV); multilevel RVOT obstruction; **PV annulus diameter & Z-score**; proximal coronary origins; **Right Aortic Arch (present in 25%)**; additional muscular VSDs, ASD/PFO, PDA, and **Left SVC to Coronary Sinus (10%)**
Cardiac CT Angiography (MDCTA)	**Class I / IIa (Non-Invasive Reference for Extracardiac & Coronary Anatomy)**	Definitive exclusion of **anomalous LAD crossing RVOT**; 3D branch PA measurements (**Nakata Index & McGoon Ratio**); aortic arch branching (aberrant subclavian artery); and complete mapping of **MAPCAs**
Cardiac Catheterization & Angiography	**Class III for Routine Uncomplicated TOF; Class I for Specific Hemodynamic/Interventional Indications**	Reserved for: (1) **TOF with PA & MAPCAs** (measuring distal segmental MAPCA pressures and proving dual vs. sole supply); (2) **Coiling large dual-supply APCs**; (3) **RVOT or PDA stenting**; or (4) assessing **PVR** in rare late-presenting pink TOF

IV PRE-OPERATIVE MEDICAL STABILIZATION AND TET SPELL PROTOCOL

- **Hypercyanotic Spell Emergency Protocol:**
  - **Physical Maneuver & Oxygen: Knee-chest position** (increases SVR) and **100% humidified oxygen**.
  - **Sedation and Hyperpnea Suppression: 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**).
  - **Volume Expansion and Acidosis Correction: Isotonic crystalloid bolus 10 to 20 mL/kg IV** (increases RV preload) and **Sodium Bicarbonate 1 to 2 mEq/kg IV** if metabolic acidosis persists.
  - **Relief of Dynamic Infundibular Spasm (Class I): Esmolol 500 mcg/kg (0.5 mg/kg) IV bolus over 1 minute followed by 50 to 200 mcg/kg/min infusion**, or **Propranolol 0.05 to 0.15 mg/kg slow IV push**.
  - **SVR Augmentation (Refractory Spells): Phenylephrine 5 to 20 mcg/kg IV bolus followed by 0.1 to 0.5 mcg/kg/min infusion**.
  - **Outpatient Pre-Surgical Optimization: Oral Propranolol (1 to 4 mg/kg/day)** for spell prophylaxis, **oral iron supplementation** (to maintain MCV >80 fL and prevent microcytic hyperviscosity stroke), dental/infection clearance, and **avoidance of dehydration**.

V CURRENT SURGICAL TIMING AND VALVE-SPARING RECOMMENDATIONS (AATS / STS CONSENSUS)

- **Elective Primary Repair at 3 to 6 Months of Age (Class I):** Asymptomatic or mildly cyanotic infants with standard anatomy undergo elective single-stage complete repair between **3 and 6 months of age** (performing repair before 3 months increases transannular patch rates and ICU stay, while delaying beyond 12 months increases chronic hypoxia, RV hypertrophy/fibrosis, and spell morbidity).
- **Symptomatic Neonates (<30 Days) or Weight <2.5 kg:** High-volume neonatal centers may perform primary neonatal repair in selected cases, but **staged palliation—preferentially via transcatheter RVOT Stenting (or PDA stenting / surgical mBTT shunt)**—followed by complete repair at 4 to 8 months achieves lower early morbidity and allows the pulmonary annulus to grow for a subsequent valve-sparing repair.
- **The Valve-Sparing Transatrial-Transpulmonary Paradigm:** To prevent late RV failure from chronic free pulmonary regurgitation, contemporary surgery prioritizes a **transatrial-transpulmonary approach** combining infundibular muscle resection through the tricuspid valve, pulmonary commissurotomy, and **intraoperative balloon dilation of the pulmonary annulus** to preserve the native valve whenever the pulmonary annulus **Z-score is -2.5 to -3.0 or greater**.

KEY REFERENCES

- **Expert Consensus Panel (Villafañe J, Feinstein JA, Jenkins KJ, et al.):** Hot Topics in Tetralogy of Fallot, *Journal of the American College of Cardiology*, 2013.
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- **Baumgartner H, De Backer J, Babu-Narayan SV, et al.:** 2020 ESC Guidelines for the management of adult congenital heart disease, *European Heart Journal*, 2020.