PALLIATIVE SURGERIES IN TETRALOGY OF FALLOT (TOF): CURRENT STATUS

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I THE CONTEMPORARY PARADIGM SHIFT

- **Primary Complete Repair as the Gold Standard:** In modern congenital cardiac surgery, **elective single-stage complete intracardiac repair between 3 and 6 months of age** is the standard of care for most infants with Tetralogy of Fallot (TOF).
- **Restricted Role of Initial Palliation:** Palliative procedures are no longer performed routinely; they are reserved as a **staged bridge to complete repair** for symptomatic neonates and young infants with high-risk anatomical or physiological features in whom neonatal cardiopulmonary bypass carries excessive morbidity and mortality:
- **Prematurity or Very Low Birth Weight (less than 2.0 to 2.5 kg)**.
- **Severely Hypoplastic Confluent Branch Pulmonary Arteries** (Nakata Index less than 150 mm2/m2; palliation promotes flow-mediated pulmonary arterial growth).
- **Anomalous Coronary Artery Crossing the RVOT:** Specifically an **anomalous LAD arising from the RCA (or a large single coronary ostium) coursing across the subpulmonary infundibulum** (present in 5 to 9 percent of TOF patients), which precludes a standard transannular patch in a small neonate and requires later RV-to-PA conduit repair or tailored double-barrel/transatrial repair.
- **Severe Active Extracardiac Comorbidities:** Neonatal sepsis, necrotizing enterocolitis, intracranial hemorrhage, or tracheoesophageal anomalies.

II SURGICAL PALLIATIVE PROCEDURES

- **Modified Blalock-Thomas-Taussig (mBTT) Shunt (Standard Surgical Palliation):**
  - **Technique:** Interposition of a **polytetrafluoroethylene (Gore-Tex) tube graft (typically 3.0 to 4.0 mm in neonates)** connecting the **subclavian or innominate artery** to the **ipsilateral branch pulmonary artery** (typically performed on the side opposite the aortic arch).
  - **Advantages over the Classical Blalock-Taussig Shunt:** Preserves blood supply to the ipsilateral arm (avoiding subclavian steal and limb ischemia), regulates shunt flow via the fixed graft diameter, and is easier to take down at complete repair.
  - **Limitations:** Continuous **systolic-diastolic runoff from the aorta** lowers systemic diastolic blood pressure, creating a risk of **coronary and splanchnic (necrotizing enterocolitis) diastolic steal**, and can cause **tethering, stenosis, or distortion of the branch pulmonary artery** at the anastomotic site.
  - **Central Aortopulmonary Shunt (Mee / Melbourne Shunt):**
  - **Technique:** A short Gore-Tex interposition graft or direct anastomosis connecting the **ascending aorta directly to the main pulmonary artery**.
  - **Indication:** Reserved for neonates with **extremely diminutive, hypoplastic, yet confluent central pulmonary arteries** (where a peripheral mBTT shunt would thrombose or distort the tiny branch PA), delivering symmetric high-volume flow into both right and left branch PAs simultaneously.
  - **Right Ventricular Outflow Tract (RVOT) Non-Valved Patch / Sano-Type Shunt:** Surgical decompression of the infundibulum without VSD closure in selected centers to grow hypoplastic native pulmonary arteries via antegrade pulsatile flow.
  - **Obsolete Historical Shunts (Abandoned):** The **Waterston Shunt** (side-to-side ascending aorta to right PA) and **Potts Shunt** (side-to-side descending aorta to left PA) have been completely abandoned in TOF due to uncontrollable pulmonary overcirculation, unilateral Pulmonary Vascular Obstructive Disease (PVOD), and severe kinking/distortion of the pulmonary arteries that was extremely difficult to take down at complete repair.

III THE TRANSCATHETER PALLIATION REVOLUTION

- **Right Ventricular Outflow Tract (RVOT) Stenting:**
  - **Technique:** Percutaneous femoral or internal jugular venous deployment of a bare-metal premounted coronary or peripheral stent across the obstructed muscular infundibulum and pulmonary valve into the main PA.
  - **Hemodynamic Superiority Over mBTT Shunt:** Unlike an aortopulmonary shunt, RVOT stenting directs **purely systolic, pulsatile, antegrade venous blood** from the right ventricle into the pulmonary trunk—**completely preserving aortic diastolic pressure (zero coronary or mesenteric diastolic steal)**, avoiding left heart volume overload, and promoting **superior, symmetrical growth of both branch pulmonary arteries (higher Nakata Index gain)**.
  - **Trade-Off:** When the stent crosses the pulmonary valve annulus, it cages the native valve leaflets, committing the infant to a transannular patch or valved conduit at the time of definitive surgical repair (when the stent is longitudinally incised and partially or fully explanted).
  - **Patent Ductus Arteriosus (PDA) Stenting:**
  - **Technique and Indication:** Transcatheter stenting of the ductus arteriosus via carotid, axillary, or femoral access; preferred in **duct-dependent TOF with Pulmonary Atresia** or critical PS with a tortuous/favorable vertical ductus, achieving lower procedural mortality, less ICU stay, and more symmetrical PA growth than surgical mBTT shunting (**Glatz et al., *Circulation* 2018**), though associated with a higher rate of planned catheter re-interventions.

Palliative Modality	Flow Pattern & Hemodynamics	Impact on Aortic Diastolic Pressure	Impact on Pulmonary Artery Anatomy	Primary Limitation / Trade-Off
Modified BTT Shunt (Surgical)	Continuous systolic-diastolic flow from subclavian artery	**Decreased** (coronary & mesenteric **diastolic steal**)	Asymmetric PA growth; risk of **ipsilateral PA stenosis/distortion**	Acute shunt thrombosis; phrenic or recurrent laryngeal nerve injury; seroma
RVOT Stenting (Transcatheter)	**Pulsatile systolic antegrade flow** from RV	**Preserved** (zero diastolic runoff from aorta)	**Symmetric, rapid bilateral branch PA growth**	Commits pulmonary annulus to transannular patch at final repair; frame fracture
PDA Stenting (Transcatheter)	Continuous systolic-diastolic flow via ductus	Decreased (diastolic runoff, similar to mBTT)	**More symmetric PA growth** than mBTT; avoids surgical scar	Higher rate of re-intervention for in-stent neointimal proliferation; ductal spasm

IV COMPLICATIONS OF PALLIATIVE PROCEDURES

- **Acute Shunt or Stent Thrombosis (Life-Threatening Emergency):** Sudden occlusion of an mBTT shunt or PDA stent presents with **precipitous, profound cyanosis, refractory hypoxemia, and disappearance of the continuous shunt murmur**. Managed immediately with **intravenous unfractionated heparin bolus (50 to 100 U/kg)**, increasing systemic blood pressure (phenylephrine/epinephrine) to drive flow across the shunt, emergency **catheter thrombectomy/angioplasty/stenting**, or emergency surgical revision. **Low-dose Aspirin (3 to 5 mg/kg/day)** is mandatory after all shunts and stents to prevent thrombosis.
- **Pulmonary Overcirculation and Congestive Heart Failure:** Occurs if the mBTT graft (e.g., >4.0 mm in a small neonate) or PDA stent is oversized, causing high pulmonary blood flow, pulmonary edema, left ventricular volume overload, and **diastolic steal-induced necrotizing enterocolitis or myocardial ischemia**.
- **Surgical Local Complications (mBTT Shunt): Perigraft Seroma** (ultrafiltration of plasma across the porous PTFE graft causing a mass/pleural effusion), **Horner Syndrome**, **Phrenic Nerve Palsy** (hemidiaphragmatic paralysis), **Recurrent Laryngeal Nerve Injury**, and **Chylothorax**.

V PRACTICAL CONSIDERATIONS IN THE INDIAN CLINICAL CONTEXT

- **RVOT Stenting in Malnourished, Late-Presenting Infants:** In Indian public and tertiary hospitals, infants with TOF frequently present with **intractable hypercyanotic spells, severe protein-energy malnutrition (weight under 3 to 4 kg), or active lower respiratory tract infection** where immediate open-heart repair carries high mortality; **percutaneous RVOT stenting** rapidly stabilizes arterial saturation (to 85 to 92 percent), avoids an ICU thoracotomy, and allows 4 to 6 months of nutritional rehabilitation and branch PA growth prior to elective complete repair.

KEY REFERENCES

- **Glatz AC, Petit CJ, Goldstein BH, et al.:** Comparison Between Patent Ductus Arteriosus Stent and Modified Blalock-Taussig Shunt as Palliation for Infants With Ductal-Dependent Pulmonary Blood Flow, *Circulation*, 2018.
- **Sandoval JP, Chaturvedi RR, Benson L, et al.:** Right Ventricular Outflow Tract Stenting in Tetralogy of Fallot Infants With Risk Factors for Early Primary Repair, *Circulation: Cardiovascular Interventions*, 2016.
- **Libby P, Bonow RO, Mann DL, Tomaselli GF, Bhatt DL, Solomon SD:** *Braunwald's Heart Disease: A Textbook of Cardiovascular Medicine*, 12th Ed., Elsevier, 2022.