Tetralogy of Fallot

Surgical Specialty:	Cardiac
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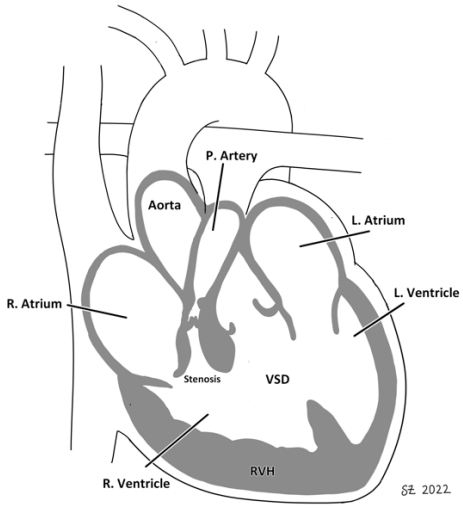
Background:

- General Considerations
  - Tetralogy of Fallot (TOF) is the most common cyanotic heart disease resulting from infundibular underdevelopment, resulting in the 4-characteristic cardiac anomalies[1]:

Tetralogy of Fallot (TOF) anomalies See figure. (Source credit: authors)

1. Unrestricted ventricular septal defect (VSD)
2. Right ventricular outflow tract obstruction (RVOTO): fixed (valvular/ supravalvular) and dynamic (Infundibular spasm)
3. Overriding of the aorta
4. Right ventricular hypertrophy (RVH)



  - Anatomic variations can range from TOF with mild RVOTO to complete RVOTO, or mild pulmonary stenosis to pulmonary atresia.
  - Presentation and surgical approach to TOF with pulmonary stenosis is different from TOF with pulmonary atresia due to the presence of major aortopulmonary collaterals in the latter.[3] Another variant, TOF with absent pulmonary valve also has different implications. Thus, this case guide will focus on *TOF with pulmonary stenosis*.
  - The goal of surgical repair of TOF with pulmonary stenosis is to establish pulmonary blood flow by relieving or bypassing the RVOTO and closing the VSD. Surgical approaches include trans-atrial, ventricular, and trans-pulmonary. A right ventricle (RV) to pulmonary artery (PA) conduit may be needed (e.g., anomalous coronary arteries, occurring in 5-12%).[3]
  - Timing of elective repair varies according to patient characteristics and institutional preference, ranging from neonates to infants 4 to 6 months of age. Delaying the surgery allows time for somatic growth, which improves the technical conditions for complete repair; this benefit is achieved at the expense of longer hypoxic exposure.

- Patient Considerations
    - Clinical presentation varies with the anatomic variation of TOF and the extent of fixed and dynamic RVOTO.[2]
        - Neonates with TOF and severe RVOTO can be critically ill at presentation depending on ductal flow requiring prostaglandin-E1 (PGE1) infusion. If the patient is a poor candidate for full repair, staged repair with initial palliation via modified Blalock-Taussig-Thomas Shunt (mBTTS), central shunt, or transcatheter-based PDA stenting or RVOT balloon dilation/stenting is initially performed with full repair performed later.
        - "Pink Tets": Mild pulmonary stenosis. Physiology resembles a patient with a VSD resulting in pulmonary over circulation and congestive heart failure (CHF). Poorly compensated CHF causes frequent respiratory illnesses and failure to thrive. All “pink tets” eventually become “blue tets”.
        - "Blue Tets": risk of cyanotic spells due to sudden R to L shunting. Triggers: agitation, pain, crying (sympathetic surge causing dynamic obstruction from infundibular spams and increased pulmonary vascular resistance (PVR) to a lesser degree), and dehydration, fever or other factors decreasing systemic vascular resistance (SVR) and preload. Medications: propranolol to decrease infundibular contractility and lower heart rate (HR) to increase diastolic filling time. Instinctive squatting behavior is observed in older kids to increase SVR and reverse R to L shunt.
        - Failure to thrive to very robust infants resulting from feeding often to appease crying and agitation that could lead to hypoxemia.
        - Polycythemia and clubbing are measures of longstanding cyanosis. Risks of hyperviscosity with thrombotic risk and impaired coagulation.

- Disease Specific Considerations
    - Associated Comorbidities/Syndromes[1,2]
        - 22q11 deletion syndrome, VACTERL/CHARGE association, and Alagille syndrome
        - Associated cardiac anomalies - Bicuspid pulmonary valve, main and branch pulmonary artery stenosis, partial or total anomalous pulmonary venous return, aberrant subclavian artery origin, atrioventricular septal defect, and left superior vena cava (SVC).
        - 25% of TOF patients present with a right-sided aortic arch, and mirror image branching of the head vessels. Coronary abnormalities occur in 5-12% of patients with TOF, including a left anterior descending artery crossing the RVOT inferiorly. Failure to detect these preoperatively can have serious consequences for a successful outcome.

Anesthetic Planning:

- Pre-Anesthetic Evaluation[2,3]
    - Evaluation of the anatomy of TOF, including coronary arteries and other associated cardiac and extra-cardiac anomalies (Echocardiography, EKG, and CXR, +/-cardiac CT scan)

- Review vital signs (SpO_2 at rest and when most cyanotic), current medications, growth history, prior hospitalizations (respiratory illnesses, Tet spells). Inquire about squatting behavior in older children.
- Physical examination: airway, evaluate for signs of CHF, clubbing, and for vascular access sites.
- Pre-operative discussion with surgery/cardiology/intensive care unit (ICU) teams regarding anticipated surgical approach, plans for early extubation and other relevant patient-specific perioperative considerations (e.g., syndrome related).
- Discuss the risks/benefits/alternatives with the family. Verify code status and ECMO option, and consent for blood product transfusion.
- Type and crossmatch (consider irradiated blood or blood products for children younger than six months or diagnosed with 22q11 deletion), complete blood cell coun, comprehensive metabolic profile, and coagulation profile.
- Avoid prolonged fasting.

- Specific or Unique Room Set-Up Requirements[2,3]
  - Airway - Oral vs. Nasal endotracheal tube (ETT)
  - Premedication to allay anxiety and prevent tet spells for at-risk patients
  - Inhalational vs. intravenous (IV) vs. intramuscular (IM) induction.
    - Pre-induction IV allows for controlled induction and prompt access for fluid/vasoactive medication administration. Con: Painful/difficult IV placement could trigger R to L shunt.
    - Inhalational induction facilitates IV placement in challenging patients. Con: Excessive vasodilation can promote R to L shunt. Therefore, titrate to effect and hemodynamics.
    - IM induction is a reasonable alternative to facilitate induction in patients with difficult IV access and challenging controlled inhalational induction. Con: Pain on injection and slower onset of action than the IV route.
  - Specific medications:
    - Phenylephrine (2-10 mcg/kg) for treatment of tet spells in at-risk patients
    - Vasoactive Infusions - To treat pre-bypass hypotension, vasopressin is preferred over epinephrine due to the concern of worsening RVOTO with epinephrine. After adequate surgical relief of RVOTO, use epinephrine as needed for inotropic support.
    - Opioid boluses and infusion, Ketamine, IV acetaminophen +/- regional block
    - Esmolol for negative inotropy and decrease in RVOTO to help mitigate cyanotic tet spells
    - Esmolol or vasodilator infusion should be available for potential postoperative hypertension.
    - Pain control and sedation- Opioid bolus and infusion, dexmedetomidine
    - Antifibrinolytics (tranexamic acid or aminocaproic acid)
    - Heparin
    - Protamine
    - Calcium
  - Monitors/ vascular access
    - Standard American Society of Anesthesiologists (ASA) monitors with 5 lead ECG, near infra-red spectroscopy.
    - Transesophageal echocardiography (TEE).
    - Arterial line and central venous line (spare femoral sites for future percutaneous valve placement), peripheral IV (PIV).

- Point-of-care (POC) viscoelastic testing
- Local anesthetic for incision sites and/or regional block for postoperative pain control
- Blood products (irradiated) – Red blood cells (RBC), fresh frozen plasma (FFP), cryoprecipitate, and platelets
- ICU bed for postoperative care.

Intraoperative Considerations:

General

- Risk of cyanotic spells during pre-CPB period.[2,3]
  - Mitigate triggers such as agitation/hypoxemia/hypercarbia/hypovolemia, which would cause sympathetic surge leading to infundibular spasm. Avoid decrease in SVR, either of which would lead to increased R to L shunt, causing severe cyanosis and hypoxic cardiac arrest.
  - Use 100% O_2 and ventilate with low airway pressures
  - If agitation and light anesthesia suspected, increase the anesthetic depth and titrate while monitoring for decreased SVR (decreases sympathetic surge relieving infundibular spasms)
  - Place patient in knee-chest position or perform abdominal compression to increase SVR and preload.
  - Phenylephrine (2-10 mcg/kg) to increase SVR and decrease the R to L shunt
  - Esmolol at 0.5 mg/kg to decrease inotropy and mitigate RVOTO
  - Fluid bolus (Eliminate air in IV line to avoid paradoxical air embolism)
  - When all these measures fail, and the patient continues to deteriorate, the surgeon should open the chest quickly, do aortic compression to temporize, and prepare to initiate CPB urgently.

- Induction (see Anesthetic Planning section for more details)
  - Inhalation induction with sevoflurane to secure PIV. Administer neuromuscular blocker and intubate.
  - IV induction with ketamine (2mg/kg), fentanyl (1-2mcg/kg), and rocuronium (0.6 mg/kg).
  - IM induction with ketamine 4mg/kg and midazolam 0.1mg/kg)to facilitate PIV placement in the uncooperative child. Supplement with IV or inhalational agents, neuromuscular blocker, and intubate.
  - IV fluids for euvolemia.
  - Post-induction arterial and central venous line.
  - TEE probe insertion.
- Maintenance
  - Inhalational agent with IV opioids and adjuvant sedation (dexmedetomidine infusion 0.2 to 0.7 mcg/kg/hr)
  - Maintain SVR to decrease R to L shunt
- Hemodynamic/Physiologic goals
  - To decrease R to L shunting and reduce hypoxemia[2]
    - Adequate preload and normal heart rate to minimize dynamic RVOTO.
    - Maintain afterload (SVR) to reduce R to L shunt and promote pulmonary blood flow.
    - Avoid hypoxemia, hypercarbia, and acidosis.
- Surgical Considerations[1]
  - Aberrant coronaries crossing RVOT might require an RV to PA conduit.

  - Pre-repair intraoperative TEE to confirm cardiac lesions. Post-repair TEE to assess residual VSD, RVOT gradients, pulmonary valve function, and ventricular function. The RV: LV pressure ratio can aid in determining the adequacy of repair (0.5-0.75 or less is considered acceptable)
  - Difficulty in weaning from CPB most often results from residual anatomical defect[1]
  - Heart block may occur due to the proximity of the conduction system to the VSD patch—use a temporary pacemaker with appropriate settings. Right-sided bundle branch block is common after ventriculotomy.[1]
  - Predisposition to junctional ectopic tachycardia (JET) in the postoperative period. Treatment measures include amiodarone and cooling to 35 degrees C.
- Emergence/Disposition
  - Post CPB bleeding- Treat with cryoprecipitate, platelet, and FFP transfusions as guided by POC viscoelastic testing.
  - RV dysfunction: consider milrinone, dopamine and/or epinephrine infusions, and right atrial/left atrial line to assist in volume treatment.
  - Esmolol infusion if residual obstruction or post-repair hypertension. Add vasodilator infusion if needed for post-repair hypertension.
  - Thick RV necessitates adequate preload to support cardiac output
  - Early extubation if hemodynamically stable without the need for escalating support and with minimal bleeding.[2]
- Post-op Care
  - Post-op transfer to ICU
  - Analgesia (opioids+/- regional block) and sedation (dexmedetomidine infusion)

Case Specific Complications/Pitfalls

- Arrhythmias – JET, complete heart block[2]
- Residual lesions: VSD, RVOTO, expected pulmonary regurgitation with transannular patch repair.[1]
- Restrictive RV physiology.[2]

References

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