Tetralogy of Fallot

Congenital cyanotic cardiac lesion with right-to-left intracardiac shunt resulting in decreased pulmonary blood flow and arterial hypoxemia: the classic tetralogy includes 1) VSD, 2) over-riding aorta, 3) RVOT obstruction (pulmonic stenosis) and 4) RV hypertrophy

ANESTHETIC CONSIDERATIONS:

- Pediatric / neonatal / premature patient
- Complex congenital heart defect anatomy and physiology:
  - **Risk of worsening R-to-L shunt** (dependent on PVR:SVR ratio; avoid ↑ PVR / ↓ SVR)
  - RVOT obstruction + RV dysfunction
  - Unrepaired vs. palliative vs. corrected anatomy
  - implications for procedures (i.e. CVL, PA catheter, etc.)
- Compensation / complications of TOF
  - **Hypercyanotic attacks** ("Tet spells")
  - Chronic hypoxemia, **polycythemia**, **coagulopathy**
  - Risk for paradoxical embolism → **strict avoidance of IV air**
  - Infection: **IE prophylaxis**
- Other congenital defects / syndromes :
  - Cardiac – ASD (pentalogy of F.), R Ao arch, PDA, multiple VSD, right sided LAD, P/T APVR
  - Systemic – Trisomy 21, DiGeorge, Velocardiofacial
- Altered drug pharmacokinetics
  - Enhanced onset of action of IV (R→ L shunt) and IH (L→ R shunt) agents
- Considerations of repaired TOF
  - Potential conduction abnormalities (a. fib / flutter, **RBBB**), pacemaker, AICD
  - Potential RV overload / dysfunction secondary to pulmonic regurgitation
  - Infective endocarditis prophylaxis
  - Difficult IV/arterial access due to previous procedures

ANESTHETIC GOALS:

- Minimize R-to-L shunting by observing HD goals:
  - **Preload**: full
  - **PVR**: avoid precipitants of ↑ PVR (+ RVOT obstruction = infundibular spasm)
  - **SVR**: strictly maintain
  - **Contractility**: avoid ↑ (as would ↑ infundibular spasm; continue perioperative beta-blockers)
  - **Rate**: normal (avoid tachycardia)
  - **Rhythm**: strict NSR
  - **MVO_2**: monitor for ischemia
- Remember precipitants of ↑ PVR
  - Hypoxia, hypercarbia, hypothermia, acidosis, atelectasis, elevated airway pressures (especially decreased expiratory time), pulmonary vasoconstrictors, light anesthesia/pain, sympathetic stimulation, and polycythemia

HISTORY

- Pink "Tet" vs. cyanotic "Tet"
  - Degree of cyanosis and frequency of "Tet" spells → intermittent or continuous
  - Precipitants (crying, feeding = young kids, exercise = older kids)
  - Frequency of squatting (↑ SVR), tachypnea, LOC and seizures
- Exercise tolerance (older children will generally have decreased exercise capacity rather than overt cyanosis)
- Developmental history (weight gain, keeping up with other kids physically etc.)
- Prior surgery: palliative shunts (e.g. BT shunt) vs. completed repair
- Medications: beta blockers etc.
- Other complications of TOF:
  - Cerebrovascular accidents: due to either venous thrombosis (polycythemia), severe arterial hypoxemia, or paradoxical emboli
  - Cerebral abscess common in severe TOF and presents with headache, fever, lethargy, NV and seizure, focal deficit
  - Infective Endocarditis → TOF is high risk as per AHA Guidelines
- Chromosomal anomalies in 15% (Trisomy 21, DiGeorge or Velocardiofacial syndromes)
- CHF rarely develops (more common in palliated patients who now have too high pulmonary BF)

PHYSICAL

- **VITALS** - HR, BP, R/A SpO_2
- **GENERAL** - Cyanosis (peripheral / central) in nail bed and under tongue / lips during hypercyanotic spells (under tongue most sensitive for central cyanosis), clubbing
- **CVS** - Harsh SEM along LLSB (d/t RVOT obstruction, not VSD since unrestrictive) which may disappear during hypercyanotic spells (minimal PBF), loud single S2, RV heave & palpable thrill

INVESTIGATIONS

- **Labs**
  - CBC & INR / PTT for Hb / polycythemia, PLT, coagulation profile (coagulopathy secondary to polycythemia, hepatic congestion and likely chronic microvascular DIC/hypofibrinogenemia triggered by hyperviscosity)
  - +/- ABG (assess room air PaO_2, often < 50 mmHg; preoperative pH / $PaCO_2$, often normal)
- **Imaging**
  - ECG (conduction defects, RVH, RAD)
  - ECHO (clarify anatomy, assess severity of RVOT obstruction, R-L shunting, other anomalies)

- +/- CATH (coronary anatomy e.g. anomalous LAD off of R side, chamber pressures etc.)
- +/- CXR (if suspect CHF, normal TET findings include "boot" shaped heart, small PAs)

OPTIMIZATION

- Judicious pre-medication with goal of cooperative child but avoid hypercapnia → avoid agitation if possible (i.e. crying with IM injections) d/t risk of "Tet" spell
- Continue preoperative beta blockers (e.g. propranolol) to minimize infundibular spasm
- Ensure patient well hydrated, particularly if polycythemic:
  - Avoid excessive NPO periods → can precipitate sludging due to polycythemia and hypovolemia exacerbates RVOT obstruction
- Consider euvolemic phlebotomy, autologous blood donation, ANH in polycythemic patients
- IE prophylaxis

ANESTHETIC OPTIONS

- Any technique acceptable as long as hemodynamic goals are met
- GA generally preferred
- Cautions with regional:
  - Dramatic ↓ SVR (particularly with SAB), thus general anesthetic preferred
  - Ensure coagulation status is normal d/t high risk of coagulopathy

ANESTHETIC SETUP

- **Drugs**
  - Phenylephrine (1 mcg/kg)
    - Avoid ephedrine as β_1-agonism may exacerbate infundibular spasm
  - Esmolol (0.5 mg/kg)
  - Propranolol (0.1 mg/kg)
  - $NaHCO_3$ (1 mEq/kg)
- **Equipment**
  - Standard CAS monitors + temperature monitoring + pediatric / neonatal setup
    - Two pulse oximeters (in case 1 fails)
      - Arterial and SpO2 must be on side opposite BT shunts (due to potential mixing of SVC to right subclavian artery)
    - $EtCO_2$ may be unreliable due to significant dead space ventilation (↓ pulmonary perfusion)
      - $ETCO_2$ significantly *under*estimates true $PaCO_2$ with R → L shunting
  - Arterial line monitoring
  - +/- CVP, ECHO
  - Forced air warmers, fluid warmers

MANAGEMENT OF ANESTHESIA

- **Induction**
  - Meticulous airway management to avoid hypoxemia and hypercarbia
  - Generally recommend IV induction with goals to maintain SVR and avoid precipitants of ↑ PVR (including stimulation with laryngoscopy)
  - **Ketamine** (1-2mg/kg IV) widely accepted induction agent due to maintenance of SVR
  - Inhalational induction can be performed (theoretically prolonged with R-L shunt) but decrease in SVR must be anticipated and aggressively treated (Halothane preferred as decreases contractility and maintains SVR)
- **Maintenance**
  - Ventilatory strategy to minimize intrathoracic pressure (decreased venous return / increased PVR) yet avoidance of hypoxemia or hypercapnia (increased PVR)
    - Generally small Vt, high rate (short I times), avoid PEEP
  - **Ketamine + 50% N_2O favored for hemodynamic stability** +/- small dose benzodiazepine
    - If N_2O is used, do not use > 50% concentration:
      - Although ↑ PVR in adults, this effect not seen in infants at concentrations < 50%
      - Risk of ↑ VAE
  - Volatiles are OK and may relax RV infundibulum but watch for ↓ SVR
  - NMB – avoid histamine releasing drugs for their effect on SVR
  - **Opioids are very stable** in cyanotic congenital heart disease
- **Emergence**
  - Ensure awake, warm, comfortable, reversed NMB prior to considering extubation
  - With major procedure, plans should be made for postoperative PICU

DISPOSITION & MONITORING

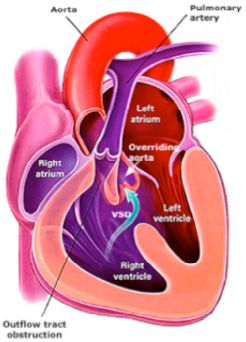
- High acuity unit
- Need for apnea and bradycardia monitoring for PCA < 52 weeks
- Cardiac monitoring (ventricular dysrhythmias common after TOF repair)
- Pain control depending on procedure – commonly opioid infusions post cardiac surgery

COMPLICATIONS

- **Hypercyanotic spell ("Tet" spell)**
  - Infants and children, not seen in adolescents or adults
  - Thought to be associated with infundibular cardiac muscle spasm (which increases RVOT obstruction) or sudden drops in SVR
  - Sudden hypoxemia, increasing cyanosis and tachypnea, sometimes progresses to loss of consciousness, seizures, CVA or sudden death
  - Often associated with crying, agitation, exercise
  - Treatment
    - Decrease PVR

        - Hyperventilation with 100% FiO_2
    - Increase SVR
        - Knees to chest
        - ***IV fluid bolus*** (improves RV filling and pulmonary flow)
        - Phenylephrine 1 mcg/kg IV
    - Morphine 0.1-0.2 mg/kg (questionable value in anesthetized patients)
    - Treat infundibular spasm
        - IV β-blocker: propranolol (0.1-0.2 mg/kg); esmolol (0.5 mg/kg)
    - Consider bicarbonate (1 mEq/kg) if anticipate acidosis as potential trigger
    - If all above fail, emergency aorticopulmonary shunt
- **Polycythemia**
    - Secondary to hypoxemia
    - Associated with increased risk of thromboembolism, esp. with HCT > 70%
    - Associated with other coagulation abnormalities – deficiency in Vitamin K dependent clotting factors and impaired platelet aggregation
- **CVA**
    - Secondary to arterial thrombosis and hypoxemia
    - Dehydration exacerbates polycythemia and increases thrombosis risk
- **Cerebral Abscess**
    - Presents with headache, fever, lethargy, progresses to persistent emesis, seizure activity, sepsis
    - ? Arterial seeding of infection in previously infarcted areas
- **Infective endocarditis**
    - High risk lesion with high mortality
    - All dental and surgical procedures require antibiotic prophylaxis
- **Right heart failure**
- **Arrhythmias**
- **Paradoxical air embolus**

Tetralogy of fallot



PATHOPHYSIOLOGY

- Cyanotic from birth or within first year of life
- Mortality > 50% by age 3 in unrepaired TOF
- Tetralogy of Fallot:
    - Infundibular pulmonic stenosis
    - Right ventricular hypertrophy
    - Ventricular septal defect
    - Overriding aorta over the VSD
- Spectrum of presentation with degree of R-L shunt (and thus cyanosis) dependent on:
    - Degree of RVOT obstruction
    - Size of VSD
- RVOT obstruction had both **fixed** and **dynamic** (infundibular spasm) components
- Thus, shunt has both **obligate** and **dependent** (i.e. dependent on ratio of PVR:SVR) components
- Generally, **PAP and PVR are low** due to decreased pulmonary blood flow
- Associated abnormalities
    - TOF + ASD = Pentalogy of Fallot
    - Right sided Ao arch
    - Associated coronary anatomical abnormalities (e.g. LAD arising from RCA)
    - 15% associated with specific chromosomal abnormalities (DiGeorge, Trisomy 21, Velocardiofacial)
- Surgical Options
    - **Repair** – patch repair VSD, enlarge RVOT (may need to ligate previous BT shunt, may need RV to PA conduit)
    - **Palliative** – BT shunt, central shunt of aorta to PA
        - Reserved for kids not yet able to tolerate full repair e.g. small PAs or pulmonary atresia

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