Twenty Years of Anesthetic and Perioperative Management of Patients with Tetralogy of Fallot with Absent Pulmonary Valve

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Background:
- Tetralogy of Fallot with Absent Pulmonary Valve (ToF APV) is a rare variant of ToF
- Aneurysmal dilation of the pulmonary arteries (PAs) can cause airway compression resulting in tracheo- and bronchomalacia
- Smaller intraparenchymal airways are also obstructed by abnormal branching of segmental pulmonary arteries
- This can result in varying degrees of airway symptoms including respiratory failure in the neonatal period
- Few data in the anesthesia literature regarding airway management and risk of complete obstruction
- AIM: To review our institutional experience with this group of patients

Methods:
- Retrospective chart review
- January 1995 – August 2014

Patients N = 44
- Median age: 53 d, range 2d - 3.75 years
- Median weight: 3.8 kg, range 1.5-15 kg
- Male gender: 25 (57%)
- Genetic disorder: 21 (48%)
- Cardiorespiratory symptoms: 27 (61%)
- Preoperative location: Cardiac ICU 16 (36%), Cardiology inpatient 10 (23%), Same-day admission 18 (41%)
- Intubated/mechanically ventilated preoperatively: 16 (36%)
- Supine: 11 (69%)
- Prone: 4 (25%)
- Lateral: 1 (6%)

Anesthetic Management and Intraoperative Events
- Induction: Intravenous 27 (61%), Combination 6 (14%), Inhaled 5 (11%), Could not be determined 6 (14%)
- Airway obstruction at induction: 1 (2%)
- Airway obstruction after intubation: 0 (0%)
- Non-supine intubation: 0 (0%)
- Intraoperative cyanosis: 0 (0%)
- ECMO initiation: 0 (0%)

VSD closure 44 (100%)
- Pulmonary artery: 37 (84%)
- Plication alone: 25 (58%)
- Plication & excision: 11 (40%)
- Central PA: 1 (3%)
- Lecompte maneuver: 3 (7%)
- Management of RVOTO: Transannular patch 23 (52%), Homograft 19 (43%), Gortex conduit 1 (2%), Melody valve 1 (2%)

Postoperative Procedures
- Rigid bronchoscopy 8 (18%)
- Tracheostomy 5 (11%)
- Lobectomy 1 (2%)
- Tracheopexy 0 (0%)

Conclusions:
- Low incidence of perioperative airway obstruction (2%)
- No episodes of cardiopulmonary arrest or ECMO initiation
- High respiratory-related morbidity, associated with genetic syndromes and overall mortality low which likely reflects improved surgical and critical care management