There is no terror in the bang, only in the anticipation of it: Anesthetic challenges of a patient with Heterotaxy Syndrome
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Case
HPI: LP is an 8 year old girl with a history of heterotaxy syndrome who presents for outpatient dental procedure and gastrostomy tube replacement.

PMH:
1. Heterotaxy syndrome
2. Dextrocardia, LV dominant
3. Double outlet RV with D-malposition of the great vessels
4. Total anomalous pulmonary venous return
5. Asplenia
6. Bilateral SVC
7. Supraventricular tachyarrhythmia
8. Right bundle branch block
9. Moderate/Severe AV regurgitation
10. Chronic pulmonary infections
11. Reactive airway disease
12. Anxiety with sleep disorder and food aversion requiring G-tube placement

PSH:
1. S/p left modified Blalock-Taussig
2. S/p AV valvuloplasty, placement of left bidirectional Glenn
3. S/p Fontan procedure
4. Ladd's band procedure
5. Fundoplication with gastrostomy tube placement

Allergies:
milk protein, synagis (Palivizumab)

Medications:
Lactobacillus, Amoxicillin, Aspirin, Clotrimazole, Enalapril, Furosemide, Atrovent, Lansoprazole, Albuterol, Bactrim

VS: BP 102/73 HR 101 Temp 36.3 RR 24 O2 90% on RA
Ht: 38 inches Wt: 27 kg
Gen: crying and scared
Airway: MP II, good distances, FROM neck, poor dentition
Pulmonary: bilateral rhonchi (L>R) CV: holosystolic murmur

Diagnosis:
Vehicle:
%O2: 21
EtCO2: 35
PetCO2: 34
PetO2: 100
FiO2: 0.21

Anesthetic Management

Preoperative:
- Patient was cardiovascularly optimized, adequately NPO, and stable in regards to her respiratory symptoms, and it was decided to proceed with surgery
- Premedication with PO ketamine and midazolam
- Child Life counselor and parental was present in OR for induction

Intraoperative:
- Standard ASA monitors placed inc. 5-lead ECG; defibrillator pads placed prior to induction
- Inhalation induction with sevoflurane on room air oxygen
- 22G PIV placed; IV succinylcholine and fentanyl given, spontaneous ventilation resumed
- DL with Miller 2; grade I view of VC. oral 5.5 cm cuffed ETT placed atraumatically
- IV fentanyl for analgesia and LR infusion to maintain preload
- Patient was extubated deep with ventilatory support in OR until breathing spontaneously and baseline etCO2

Postoperative:
- Patient was observed in PACU for 1 hour and once awake, tolerating liquids, she was discharged home

Anesthetic Goals

1. Maintain pulmonary and systemic blood flow; avoid positive pressure ventilation
2. Maintain PVR with fraction of inspired oxygen to maintain O2 sats at baseline
3. Avoid ‘steal phenomenon’ of pulmonary blood flow from the systemic circulation
4. Avoid stress response to surgery with increases in SVR, HR or PVR which could lead to worsening of AV regurgitation, hypoxemia, or arrhythmia
5. Maintenance of adequate intravascular volume
6. Anticipate and be ready to treat arrhythmia