Emergent non-cardiac surgery in a patient with congenital heart disease and WPW: perioperative management of acute hemoptysis and SVT

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Objectives: Participants in this PBLD will:
1. Review anatomy and physiology pertinent to the care of patients with congenital heart disease, specifically single ventricle physiology following stage II palliation
2. Develop a differential diagnosis for hemoptysis in the setting of Glenn cardiac anatomy and a tracheostomy
3. Discuss induction and airway management strategies for a compromised airway
4. Discuss Wolf-Parkinson-White syndrome including recognition and management of unstable supraventricular tachycardia
5. Review the pediatric anesthesiologist role in the perioperative surgical home for patients with congenital heart disease presenting for non-cardiac surgery

Case history:
A 3-year-old 14kg female with severe subglottic stenosis status post tracheostomy presents emergently after hours in apparent distress due to acute onset copious bleeding from her tracheostomy site. The bleeding began after the patient finished dinner. As the only in-house anesthesiologist you are called by the emergency department for airway management. The limited past medical history you are able to collect includes tricuspid atresia status post Glenn palliation and Wolff-Parkinson-White (WPW). The patient’s current medications are sildenafil and half a baby aspirin every day.

Questions:
What is tricuspid atresia and Glenn palliation? What physiologic considerations are there for anesthesia in someone with tricuspid atresia and Glenn palliation? How do the physiologic considerations change for those with Fontan palliation? Specifically, how is the venous drainage affected in both of these palliative procedures and how does this relate to pulmonary blood flow?
When you arrive in the emergency room the patient appears to be moderately distressed with relatively frequent hemoptysis via her 4.5 uncuffed Bivona tracheostomy tube. You see bloodstains over her clothing and face as well as the stretcher and you note perioral cyanosis. You note moderate tachycardia and sporadic pulse oximetry with readings in the 70’s. Your ENT colleague arrives at approximately the same time.

Questions:
What is the expected peripheral oxygen saturation for this patient? What does your initial evaluation/examination consist of and what treatment can you provide this patient in the emergency room? Your ENT colleague wants to proceed emergently to the OR. How do you respond? What is your differential for the source and etiology of the patient’s bleeding? Are there additional colleagues or resources available that you would call for at this time?

After completing all of the temporizing measures you have described, the condition of the patient appears unchanged to slightly improved and you take the patient emergently to the OR.

Questions:
How would this patient scenario alter your OR setup? What are the possible airway management and induction and maintenance strategies for this child with hemoptysis with a tracheostomy? How would your approach change for the same patient without a tracheostomy? How does the cardiac anatomy/physiology and WPW affect your chosen strategy and drug choice?

After inducing anesthesia and securing the airway in the operating room the hemoptysis appears to subside by visual inspection, suctioning, auscultation, and pulse oximetry. After induction the five lead EKG is noted to be sinus tachycardia with pre-excitation delta waves and you now notice questionable ST segment changes in all leads.

Questions:
What are your goals for peripheral oxygen saturations in this patient? How will you manage the FiO2 for your stated goals and what are the implications of 100% FiO2 in Glenn physiology? As the bleeding appears to be stable at this time will you place additional vascular access or invasive monitoring? Does Glenn cardiac anatomy affect your placement/use of venous access? Will you do additional preparations or testing before proceeding with rigid bronchoscopy? How will you evaluate the ST segment changes you are questioning?

Attention is then given to obtaining adequate vascular access, twelve lead EKG and intraoperative transthoracic echocardiogram (TTE). Arterial access is easily placed in the radial artery, however adequate peripheral venous access proves challenging necessitating central venous access. A twelve lead EKG is reassuring showing sinus rhythm with pre-excitation but no evidence of ischemia. TTE is also
reassuring with good concentric function of the single left ventricle without significant wall motion abnormality or mitral valve regurgitation. The operating table is then turned 90 degrees and the ENT surgeon commences rigid bronchoscopy. The patient initially tolerates the procedure but later requires intermittent boluses of IV anesthetics and phenylephrine to maintain both an adequate plane of anesthesia and mean arterial pressure. Right bronchial venous engorgement is identified as the likely source of bleeding. During rigid bronchoscope inspection right bronchial venous bleeding ensues.

**Questions:**

At this point would you modify your choice for maintaining anesthesia? How are you oxygenating and ventilating the patient during rigid bronchoscopy and is volatile anesthesia sufficient? Would you start any vasoactive infusions? Are you concerned about this patient’s cerebral perfusion pressure? When would you be concerned and how can you estimate this? What options do you and the surgeon have to temporize the bleeding and how will you resuscitate the patient?

Further inspection of the right bronchus appears hemostatic and the procedure is concluded without the use of electrocautery, the airway is managed via a 4.5 cuffed Bivona tracheostomy tube. Just as the bleeding is controlled and you breathe a sigh of relief, you note an almost instantaneous increase in the heart rate to 190 bpm. You look at your ECG monitor and see supraventricular tachycardia (SVT), you also note a low blood pressure on the arterial waveform with a thready pulse by palpation.

**Questions:**

What different types of SVT might present on the rhythm strip and what are the factors that could lead to SVT in this patient? How will you proceed to manage this change in hemodynamic status?

Through the methods you employed successful cardioversion leads to the return of a stable sinus rhythm.

**Questions:**

What is your disposition planning for this patient? Will you keep the patient sedated requiring a ventilator or will you plan to allow the patient to return to a conscious state? Will you consult with anyone when making this decision? What further safety concerns do you have for this patient?

**Discussion:**

Tricuspid atresia (TA) lacks a tricuspid valve in addition to a hypoplastic right ventricle, which places it into the single ventricle category and pathway. Although there are many different types of TA, necessitating differing neonatal surgical approaches, the expected course includes stage II palliation of superior vena cava (SVC) to pulmonary artery (PA) connection or Glenn anastomosis followed years later by Fontan completion. While in the Glenn single ventricle stage the pulmonary blood is derived via
passive drainage from the SVC to the PA. During anesthesia slight hypercarbia will allow cerebral arterial vasodilation resulting in increased cerebral blood flow leading to increased pulmonary blood flow and improved systemic oxygenation and delivery.

Maintaining a low pulmonary vascular resistance is important to single ventricle physiology and performance and subsequent staged palliation. When pulmonary vascular resistance is increased pulmonary vasodilator medications may be used in effort to normalize the pulmonary artery pressure. An elevation of the pulmonary artery pressure in a patient with Glenn anastomosis will be translated upstream to the SVC venous drainage and its contributing venous distribution. This is a contributor to venovenous collaterals and lower systemic oxygen saturations. The bronchial veins drain to the azygous and hemiazygous system which then drain to the SVC. Elevated pulmonary pressures in Glenn physiology can subsequently lead to tracheal bronchial venous engorgement and potential life threatening bleeding varices.

Management of the pediatric airway can be challenging especially in the emergent bleeding airway situation. The anesthetic induction approach will need to be tailored to each unique situation and experience of the anesthesiologist. No single method of airway management has been shown to succeed in all patients or situations. In our scenario having a pediatric ENT surgeon present, a working suction, and a rigid bronchoscope for airway management and bleeding is key. The cause and differential of hemoptysis is long and in our patient could be as simple as bleeding at the tracheal stoma due to irritation or trauma but turned out to be bronchial venous bleeding. Other causes of hemoptysis could include: infection, vascular malformation or aneurysm or vasculitis, pulmonary venoocclusive disease, pulmonary embolism, cancer, trauma, toxins being aspirated, coagulopathy, or foreign body aspiration, among others.

In patients with Wolff-Parkinson-White (WPW) syndrome there is an accessory conduction pathway in addition to the normal atrioventricular conduction pathway that leads to the characteristic but not always present pre-excitation delta wave and short PR interval on ECG. Abnormal myocardial activation can be seen as abnormal ST and T waves on ECG. Some AP which conduct in a retrograde manner may have a normal ECG during sinus rhythm being “concealed.” Most WPW arrhythmias are atrioventricular re-entrant tachycardias (AVRT) and ~20% are atrial fibrillation or flutter. During atrial flutter there is a circular loop in the atrium only, but the rapid rate can be conducted to the ventricle via the AP. The ventricular rate will be determined based on the refractory period of the AP, shorter refractory periods can lead to a rapid and dangerous ventricular rate and a life-threatening rhythm.

Perioperative management of WPW syndrome can be challenging and incorrect treatment can lead to clinical deterioration and even cardiac arrest. Unfortunately many anesthesia practices and factors, not always avoidable, can accentuate accessory pathway travel and arrhythmia propagation, such as: laryngoscopy, perioperative nausea and gagging, hypothermia, sympathetic blockade or sympathetic activation, hyperventilation, cholinergic medications and succinylcholine. Patient volume status and good preload may decrease the use of sympathomimetics and phenylephrine is the vasopressor of choice as it limits tachycardia and has been used as an anti-arrhythmic in WPW. Epinephrine containing solutions such as for local anesthesia should be used cautiously due to inducing tachycardia. When an
unstable arrhythmia occurs during anesthesia, rapid recognition and prompt PALS algorithms need to be employed.

For patients with congenital heart disease presenting for anesthesia or noncardiac surgery, the pediatric anesthesiologist is one of several experts who should work in a multidisciplinary team approach to patient care. Additional expert consultants can and should be sought during challenging management decisions preoperatively, intraoperatively, and postoperatively. Due to the nature of the anesthesia specialty we are positioned to direct and coordinate decision making in this regard.

Prior Presentation: No

References:


