PBLD – March 14, 2015 6:50a-8:00a – Table 18

Rhythm and Blues: A case of corrected congenital heart disease presenting for post-tonsillectomy hemorrhage

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Learning Objectives:

1. Review the relevant peri-operative issues when caring for a patient with Trisomy 21.
2. Develop a plan for patients presenting to the operating arena with pacemakers for emergency surgery.
3. Develop an anesthetic plan for a patient with corrected congenital heart disease.
4. Review the relevant concerns with obstructive sleep apnea in the peri-operative period.

Case:

Three-year-old male child with history of Trisomy 21 who presents nine days after tonsillectomy through your hospital's emergency department with hematemesis. The patient's mother reports an uneventful surgical course and anesthetic nine days prior at a hospital much farther from home than your current hospital. The patient has a history of congenital heart disease, repaired at 3 months of age. Additionally, she reports that her son had a pacemaker placed in a subsequent surgery, which was interrogated prior to his tonsillectomy and was working well per her discussion with the cardiologist. The patient has a history of sleep apnea, which is why the tonsils were removed and the patient stayed in the hospital overnight following surgery.

Per your discussion with the emergency room physician, you discern the patient produced approximately 20mL of bloody emesis. The patient is not vomiting blood actively at this time. The patient is posted for examination under anesthesia and possible cauterization of post-tonsillectomy hemorrhage. He is NPO for solids for 9 hours and liquids for 7 hours.

Questions:

1. What are the risk factors for post-tonsillectomy hemorrhage?
2. When does post-tonsillectomy hemorrhage usually occur?

3. What cardiac defects are common with a history of Trisomy 21?
4. What cardiac history is relevant before proceeding with this case?
5. Does this patient require any cardiac work-up that should delay this case?
6. What information about the pacemaker is needed before the operating room?
7. How can information about the pacemaker be obtained without an interrogation if it is not available?
8. What is suggested by his history of overnight stay following tonsillectomy for obstructive sleep apnea?
9. What is the natural course of obstructive sleep apnea following tonsillectomy?

Case Continued:

Vital Signs: BP 98/40, HR 92, RR 20, Room Air O2 Sat 99%.

Labs: WBC 6.1, Hgb 10.2, HCT 31, Plt 120
Na 141, K 3.6, Cl 102, HCO3 22, BUN 22, Cr 0.38

EKG: P waves are seen, followed by a pacemaker spike and QRS complex.

CXR: Clear lung fields, normal appearance of the airway. There are sternal sutures appreciated over the midline. There is a pacemaker generator in the right upper quadrant of the abdomen with pacemaker wires seen coursing to the right atrium and apex of the heart.

Physical Exam: Appropriate-size three-year-old boy, uncomfortable, but in no acute distress. He is anxiously clinging to his mother. Lungs: CTAB. CV: RRR, faint systolic murmur. Skin: No rash, capillary refill <2 seconds, normal turgor.

The patient had an IV placed in the emergency department for fluids given his recent hematemesis. He presents with his mother to the preop holding area. The patient's mother has signed blood consent, anesthesia consent, and surgical consent. The patient has 2 units of blood available in the operating suite. Since it is Saturday morning at 4am, no cardiologist or electrophysiology nurse is available to interrogate the pacemaker before going to the OR.

Questions Continued:

10. What are the options for alleviating patient anxiety in this case?
11. The patient's mother insists on being present for induction. She was present for induction for the tonsillectomy. Is parental presence appropriate?
12. What is an appropriate induction technique for this patient?
13. What airway or surgical equipment is needed in the room prior to induction?
14. Are there any relevant airway concerns given the history of Trisomy 21?
15. Should a magnet be placed on the pacemaker for the duration of surgery?

Case Continued:

The patient undergoes preoxygenation and rapid sequence induction with placement of a 4.5 cuffed oral RAE tube using direct laryngoscopy. There was some blood in the hypopharynx, but none appeared to be in the airway. The case proceeds and the surgeon is able to cauterize the suspected site of bleeding without immediate complication. There is little additional blood loss during the case.

Questions Continued:

16. Is this patient a candidate for extubation, either deep or awake?
17. How long should this patient be monitored with pulse oximetry post-op?
18. Does the patient need post-operative cardiac monitoring or interrogation of the pacemaker?

Discussion:

Post-tonsillectomy Hemorrhage

Tonsillectomy is one of the most common surgical procedures in children. Hemorrhage is the most significant complication of a tonsillectomy. Coagulation screening prior to tonsillectomy is not advised. Children with histories suggesting a mild bleeding disorder or with abnormal PT or PTT, used to screen for abnormal coagulation, were not more likely to develop post-tonsillectomy hemorrhage [1]. In another prospective study of children that underwent extensive preoperative coagulation testing, this testing failed to predict risk of major bleeding during tonsillectomy [2].

Reported rates of post-tonsillectomy hemorrhage are incredibly varied. A Norwegian study of all patients from 1999-2005 revealed that 0.5% of patients will be admitted postoperatively for post-tonsillectomy hemorrhage and only 0.1% will require reoperation [3]. As that study shows, post-tonsillectomy hemorrhage is much more likely to resolve spontaneously or with emergency department cauterization than it is to require re-operation. Generally, rates of post-tonsillectomy hemorrhage average less than 5% [4] [5] [6], but some report up to 15% [7]. If a patient has a small post-tonsillectomy hemorrhage, then they are more likely to develop a larger subsequent hemorrhage, which is described as the sentinel bleed [7].

Obstructive sleep apnea is associated with upregulation of prothrombotic factors. Patients with this diagnosis may be less likely to bleed following tonsillectomy versus patients with chronic tonsillitis [6]. Patients with increased levels of pain after the procedure or with increasing severity of pain over the first few post-op days are also at risk for hemorrhage [8]. Surgeon experience, drugs used perioeperatively, and the age of the child all affect rates of hemorrhage.

Primary hemorrhage is the development of post-operative hemorrhage in the first 24 hours following surgery, generally felt to be the result of surgical technique. Secondary hemorrhage occurs after 24 hours, but often occurs 5-10 days following tonsillectomy [9]. Secondary hemorrhage is likely related to healing factors and occurs when the fibrin clot sloughs off the surgical site. Less than 1% of patients require blood transusion, but these patients should have a type and screen if they present with post-tonsillectomy bleeding [4].

Trisomy 21 (Down's Syndrome)

This genetic syndrome occurs in approximately 1 in 800 live births. These patients have a long life expectancy that has improved over the years (median age of death 49 years). Common diagnoses at death in these patients are: congenital heart defects (29.1%), dementia (21.2%), hypothyroidism (20.3%), and leukemia (1.3%) [10]. Common anesthetic complications with Trisomy 21 include: severe bradycardia (3.66%), airway obstruction (1.83%), post-extubation croup (1.83%), and difficult intubation (0.54%) [11].

Atlantoaxial Instability (AAI)
Up to 20% of patients with Trisomy 21 will have radiographic evidence of AAI [12]. However, AAI is frequently asymptomatic. While the official position of the American Academy of Pediatrics (AAP) had been to limit the activity of children with Trisomy 21 and AAI prior to 1995 [13], this activity limitation was later reversed in favor of treating patients symptomatically [14]. Symptomatic AAI occurs in about 2% of patients with Trisomy 21 [15].

Anesthesiologists were surveyed regarding AAI in Trisomy 21 and their usual practices. Given the lack of guidelines on the topic, this provided some insight into how this condition is usually managed. Screening for symptoms related to AAI is a common practice in these patients. A minority of survey respondents obtain neck radiographs or subspecialist consultation in asymptomatic patients. A majority of survey respondents would obtain neck radiographs and subspecialist consultation in symptomatic patients. Approximately half of the respondents would hold the head in neutral position in asymptomatic or symptomatic patients for intubation [16].

If radiographic screening is to be undertaken, then lateral neck x-rays in flexion and extension are the recommended screening tool [17]. CT scans are taken with the patient supine and their head in the neutral position; this may lead to false-negative studies [18]. In a review of 488 patients with Trisomy 21 undergoing noncardiac procedures with general anesthesia, there were no reported neurologic compliations [11]. Signs of AAI include neck pain, difficulty walking, abnormal gait, ankle clonus, muscle weakness, positive Babinski sign, or hyperreflexia at the Achilles tendon [19].

Tonsillectomy and other otolaryngology procedures may require unfavorable neck positioning for patients with AAI. Grisel's syndrome, which is atlantoaxial subluxation, is reported rarely in the literature following ENT procedures [20]. However, the recommendation is that the surgeon identifies children with symptoms in the preoperative visit and considers consultation or radiography at that time [21]. Generally speaking, hyperextension of the neck should be avoided in children with AAI presenting for T&A [22].

Subglottic Stenosis
Subglottic stenosis is more common in children with Trisomy 21 [23]. This condition is probably related to the higher rate of post-extubation croup and difficult intubation in Trisomy 21 [11]. The tracheal diameter is also smaller in patients with Trisomy 21. An endotracheal tube that is smaller than the patient's age might predict may be required. Checking a leak after intubation is advisable so that intubation injury does not worsen subglottic stenosis [24] [25].

Difficult Airway
Difficulty intubating the trachea is the result of a number of features of Trisomy 21. These patients are small for their age and their craniofacial and airway proportions

are reduced [11]. As discussed previously, their tracheal diameter is small and subglottic stenosis is common. Macroglossia is present with midface and mid-mandibular hypoplasia [26]. They have adenotonsillar hypertrophy which may also contribute to an obstructed view [11]. The epiglottis is short and tends to collapse against the posterior pharynx [27]. While Trisomy 21 is not frequently thought of when considering syndromes that are difficult intubations, its frequency in the population and higher rate of difficult intubation than the general population should be considered.

Congenital Heart Disease

There are several cardiac anomalies commonly associated with Trisomy 21. Congential heart disease occurs with a frequency of about 50% in this population [28]. Which cardiac anomalies are most common varies depending on the race of the patient. In Caucasian patients, the most commonly identified lesions are: atrioventricular septal defect (AVSD) in 45%, ventricular septal defect (VSD) in 35%, secundum atrial septal defect (ASD) in 8%, patent ductus arteriosus (PDA) in 7%, and Tetralogy of Fallot (ToF) in 4% [28]. In Asian patients, the most common lesions are: VSD in 39%, PDA in 34%, ASD in 23%, and AVSD in 16%. In the same Asian study group, AVSD occurred concomitantly with 25% of ToF cases [29].

Congenital heart disease is common with Trisomy 21. Echocardiography screening for structural defects is advocated after birth [30]. Congential heart disease repair is generally well tolerated in this population. The defects, as discussed above, are not associated with a high surgical mortality rate. Interestingly, a recent study showed children with Trisomy 21 undergoing repair of congenital heart defects are more likely to survive to discharge than children without Trisomy 21 when Congenital Heart Surgery risk category, premature birth, major noncardiac structural anomaly, and age are controlled for [31]. Mortality rates of children with unrepaired congenital heart disease is increased following noncardiovascular surgery versus those patients without congenital heart disease [32].

Patients with partial AV canal (primum ASD, no VSD, separate AV valves with cleft mitral valve) rarely have symptoms as infants and undergo repair at ages 2-4. Patients with complete AV canal (large primum ASD, VSD, and common AV valve), more common in Trisomy 21, have large left to right shunts and develop heart failure in infancy. However, due to increased PA pressures in Trisomy 21, these patients are somewhat protected against heart failure in infancy [33]. Also, patients with Trisomy 21 and complete AV canal develop pulmonary hypertension more quickly than children without Trisomy 21, so they may require earlier repair [34]. There is increased risk of complete heart block in complete AV canal due to displacement of the AV node.

The mortality rate for correction of AVSD is certainly less than 10% and may actually be <1% [35] [36] [37]. AVSD associated with hypoplasia of the right or left ventricle may require single ventricle palliation rather than biventricular repair [38]. While quality of life issues and short life expectancies once dominated the

debate regarding congenital heart disease repair in Trisomy 21, low surgical mortality rates and longer life expectancy in Trisomy 21 now favors repair [39] [40]. Without correction, complete AVSD mortality is 96% by 5 years of age [41]. Risk of reoperation for permanent pacemaker implantation within 10 years is 2% and reoperation for all reasons is 14% [42].

Preoperative Cardiac Evaluation

As with many cardiac conditions, evaluation of functional status and symptoms are key to deciding whether a patient requires further diagnostic testing or subspecialist evaluation prior to anesthesia. Specifically asking about functional status versus peers and changes in functional status over time are useful screening questions. Auscultation of a complete repair may reveal a quiet precordium. However, left ventricular outflow tract obstruction, AV valve stenosis or regurgitation, or residual VSD may be associated with murmur or abnormal heart sounds.

Contrary to popular belief, pacemaker response to a magnet was not programmed for the convenience of anesthesiologists and surgeons. Instead, it is present for electrophysiologic diagnostic use. A pacemaker for complete heart block, which would have to sense dual chambers to be maximally effective, is likely programmed to DDD and a magnet would change the mode to DOO with a preset rate. Reasons a pacemaker may not respond normally to a magnet include: low battery life, pacemaker is programmed to ignore the magnet, or the device is too deep from the skin to sense the magnet. The best course of action when interrogation is not possible is to confirm the type of device (pacemaker or AICD) using a chest X-ray, EKG to confirm the underlying rhythm or pacemaker dependence, and subsequent EKG following magnet application (to confirm new rhythm with the magnet) [43] [44].

Interference is likely to occur if monopolar electrocautery is used in head and neck procedures. Placement of the return pad on the shoulder will ensure that current does not pass through the leads or device since the device is implanted below the diaphragm. This will ensure minimal interference and minimize damage to the device and leads. Bipolar electrocautery further minimizes risk. EKG, CXR, and possibly an information card about the device carried by the parent provide all the information obtainable about the device in an urgent situation. The manufacturer of the device can be called for perioperative management information if the information card on the device is available.

Obstructive Sleep Apnea

Almost 60% of children with Trisomy 21 have obstructive sleep apnea. Parental concerns about sleep habits in their children correlate poorly with presence of sleep apnea. Therefore, polysomnography has been recommended in all Trisomy 21 patients between 3 and 4 years of age [45]. With routine screening recommended and incidence of sleep apnea being high, Trisomy 21 patients present frequently for adenotonsillectomy.

In one study, overnight polysomnography studies were abnormal in 100% of patients with Trisomy 21 [46]. Sleep apnea does not explain all of the sleep disturbances seen in Trisomy 21, polysomnography has shown increased sleep fragmentation as well [47]. The associated diagnoses frequently seen with obstructive sleep apnea are also common in Trisomy 21 patients including: pulmonary hypertension, behavior problems, and failure to thrive [46].

Adenotonsillectomy is almost 100% successful in treatment of obstructive sleep apnea in non-syndromic children. However, it is only about 50% successful in children with Trisomy 21 [48]. As MRI sleep studies have shown, there are multiple levels of obstruction in these patients [49]. Nasopharyngeal obstruction, collapse of the soft palate on the posterior pharyngeal wall, macroglossia, lingual tonsil hypertrophy, and epiglottic collapse all contribute to the sleep apnea [22]. Tracheotomy is reserved for the most severe cases of sleep apnea.

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