ANESTHESIOLOGY™2014
OCTOBER 11-15 | NEW ORLEANS, LA

Session: L024
Session: L096

Tonsillectomy and Adenoidectomy in a Child With Down Syndrome More to It Than You Think!

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Disclosures: These presenters have no financial relationships with commercial interests

Stem Case and Key Questions Content

An eight-year-old girl with Down syndrome and congenital heart disease (CHD) is scheduled for a tonsillectomy and adenoidectomy for OSA.
Upon entering the examination room, you note an active, talkative girl playing in the corner. Once she sees you, however, she cries out and quickly runs to her parents. Her parents apologetically tell you that “she’s been in the hospital so much that she really doesn’t like doctors.”

1) What are signs of preoperative anxiety in children?

2) Are there specific things that make them afraid?

3) What other factors may be contributing to this patient’s anxiety?

Her parents report that their daughter was born at term and stayed in the hospital for an abdominal surgery for an “intestinal blockage” and cardiac evaluation. Since then, she has had several surgeries, including a patch closure of an atrial septal defect (ASD) 15 months ago.
4) What is the intestinal blockage to which her parents are referring?

5) What are common gastrointestinal issues in patients with Down syndrome?

6) How common are cardiac anomalies in patients with Down syndrome?

7) Are some cardiac anomalies more common than others?

8) Does this history affect your decision regarding SBE prophylaxis?

9) What are the current American Heart Association (AHA) guidelines for SBE prophylaxis?

She is having surgery today for OSA. Her parents state that she had a sleep study several weeks ago that showed moderate obstructive sleep apnea (Apnea -Hypopnea Index, AHI, of 10) with an oxygen nadir of 82% during sleep.6.7 They admit that they did not want her to have another surgery, but that when they were counseled to begin using continuous positive airway

pressure (CPAP), they realized that “there was no way she was going to wear that mask to sleep.”

10) What is AHI? What is the significance of an AHI of 10 in the pediatric patient?
Her parents deny any other pertinent past medical history. They state that she had “normal” cervical spine radiographs when she was three years old, but no follow-up radiographs since.

11) How sensitive is a cervical spine radiograph in determining cervical spine instability?

12) What are the current recommendations from the American Academy of Pediatrics (AAP) regarding routine cervical spine radiographs in patients with Down syndrome?

They deny any neurologic signs, including gait instability, bowel or bladder incontinence, or radiculopathy. As you continue talking to the patient’s parents, you notice her eyeing you warily. As you approach her to perform a physical exam, she starts to cry.

13) How will you transport this patient to the operating room?

14) Would you consider giving premedication to ease separation from her parents? If so, what medication would you give and why?

15) What are your concerns regarding premedication in patients with OSA?

16) Would you consider other techniques (e.g., parental presence, for example)?

On physical exam, you note an alert child with low set ears and a protruding tongue. She has a short neck and well-healed scar on her sternum. Chest auscultation reveals regular rate and rhythm, no murmur, and transmitted upper airway sounds.
Her father accompanies you to the operating room for induction of anesthesia.

17) Will you perform an intravenous or inhalational induction? Why?

The patient pulls off the pulse oximeter probe you place on her toe. After your third attempt, you reluctantly decide to proceed with inhalation induction without the monitor.

18) What are your concerns during an inhalation induction in this patient?

Her father leaves for the waiting area. You replace the pulse oximeter probe, which reads 89%, 35 beats per minute.

19) How will you treat this bradycardia? Is this heart rate expected or unexpected?

You effectively treat the bradycardia and place a peripheral IV.

20) How will you intubate this patient? What are your concerns during intubation?

21) What is your analgesia plan?

The rest of the procedure is uneventful. During emergence, the ENT resident asks, “Can this patient go home from the recovery room?”

22) Where will you send the patient - home, an inpatient unit, or an intensive care unit?

23) What are possible airway risks to consider in this patient post-tonsillectomy?

24) After what time would you consider the patient “safe” to be discharged?

25) What are the ASA guidelines for postoperative management of patients with OSA?

Key questions
1) What are signs of preoperative anxiety in children?

2) How common are cardiac anomalies in patients with Down syndrome?

3) What are the AHA guidelines for SBE prophylaxis?

4) How sensitive are cervical spine radiographs in determining cervical spine instability?

5) What are the current recommendations from the AAP regarding routine cervical spine radiographs in patients with Down syndrome?

6) What issues do you need to address when premedicating patients with OSA?

7) What are your concerns during an inhalation induction in this patient?

8) What are your considerations while intubating this patient?

9) What are possible airway risks to consider in this patient post-tonsillectomy?

10) What are the ASA guidelines for postoperative management of patients with OSA?

Model Discussion Content

Introduction

Down syndrome, or trisomy 21, is the most common genetic anomaly, occurring once in every 600-800 live births.1 The presence of additional chromosomal material leads to classic phenotypic characteristics and anatomic anomalies.

Preoperative assessment

One of the first challenges the anesthesiologist faces is establishing rapport with the pediatric patient. Preoperative anxiety occurs in up to 60% of all pediatric patients. Children can be anxious for a number of reasons, including separating from their parents, loss of control, and uncertainty regarding a surgery or anesthetic. While some patients can communicate their concerns regarding an upcoming procedure, others express their anxiety with behavioral

changes, such as trembling, avoiding eye contact, crying, ceasing play or talk, and running away. When assessing for preoperative anxiety, several factors should be considered, including age and developmental maturity, previous experience with surgical procedures or illness, individual anxiety level, and situational factors.2

Down syndrome patients may be more likely to have preoperative anxiety because of their multiple past interactions with the healthcare system and varying degrees of mental retardation, which range from mild (IQ 50-70) to severe (IQ 20-35). Assessment for preoperative anxiety in these patients is essential.

With this information, the anesthesiologist must determine what will be the safest, least traumatic way to proceed to the operating room. Options to consider include premedication with benzodiazepines, alpha-2 agonists, distraction techniques, parental presence, or some combination of the above. If the patient receives sedative medication, however, she must remain in a monitored setting, as airway obstruction is a potential complication in a Down syndrome patient.

Coexisting conditions
The airway of the Down syndrome patient can be challenging due to her large tongue, short neck, and laryngomalacia. In addition to airway obstruction caused by preoperative anxiolytic medication, obstruction during inhalation induction should be expected.1

The diagnosis of OSA is common in patients with Down syndrome, occurring in 50-79% of patients. The AAP recommends screening for OSA in Down syndrome patients as early as one month of age.5 In most cases, the pediatrician screens the patient for the presence of snoring and other sleep apnea symptoms. If present, the patient is then referred for polysomnography and/or referral to a specialist. While the STOP questionnaire is an effective screening tool for OSA in adult patients, its use has not been validated in the pediatric population.6,9

In addition to obstruction, atlanto-axial instability can occur in 12-32% of Down syndrome patients.5 Manipulation of the neck for intubation and surgery (including turning the head for myringotomy, placement of ear tubes, and tonsillectomy) should be performed very cautiously. Historically, routine cervical spine radiographs were used to evaluate for neck instability. However, this modality has several limitations. First, the physician must wait until the child reaches at least 3 years of age for cervical spine radiographs to be valid, as adequate bone mineralization does not occur until this time.5 Furthermore, normal cervical spine radiographs at a single point in time do not rule out future cervical abnormalities. For these reasons, the AAP does not recommend cervical spine radiographs in asymptomatic Down syndrome patients. Instead, the clinician should evaluate for upper motor neuron signs or symptoms, including gait instability, radiculopathy, and bowel or bladder incontinence. Additionally, all involved clinicians (i.e., physicians, nurses, surgical technicians) should be counseled to avoid excessive neck extension in the operative and perioperative period.5

In contrast, symptomatic patients should undergo cervical spine imaging. If these studies are abnormal, patients should be immediately referred to a pediatric neurosurgeon or orthopedic surgeon who specializes in atlanto-axial evaluation and management.5

Up to 40% of patients with Down syndrome have CHD. The most common anomaly is complete atrioventricular canal (CAVC), occurring in 40% of those with CHD, followed by ventricular septal defect (VSD, 25%) and ASD (10-15%).1 Although these anomalies may be corrected prior to surgery, concerns regarding residual defects, alterations in pulmonary vasculature, and the need for SBE prophylaxis should be addressed.1

The current AHA guidelines for SBE prophylaxis were published in 2007 and have greatly decreased the indications for SBE prophylaxis.8 Per the guidelines, antibiotic prophylaxis is now indicated only in patients with the following conditions:

a) prosthetic cardiac valve or prosthetic material used for cardiac valve repair
b) a history of infective endocarditis (IE)
c) unrepaired cyanotic CHD, including palliative shunts and conduits, or
completely repaired congenital heart defect with prosthetic material during the first six months after the procedure, or repaired CHD with residual defects
d) cardiac transplantation recipients with valvulopathy.

The type of surgery also determines the need for SBE prophylaxis. Dental procedures that manipulate gingival tissue, periapical region of teeth, or perforation of oral mucosa carry the highest risk of IE. Other procedures that require antibiotic prophylaxis include those on the respiratory tract, infected skin structures, or musculoskeletal tissue.

Patients with Down syndrome have an increased incidence of bradycardia during inhalation induction with sevoflurane. In a 2010 study performed in Philadelphia, the incidence of bradycardia during induction was 57% in patients with Down syndrome, compared to 12% in those without Down syndrome.3 This increased incidence of bradycardia was independent of a CHD diagnosis. The mechanism behind this phenomenon has not been determined, but is thought to be due to genetic alterations in cardiac cell size, number, and quality. Administration of intravenous antimuscarinic agents did not prevent bradycardia in patients to whom they were given.

Other concomitant processes in patients with Down syndrome may include hypothyroidism, gastrointestinal narrowing such as duodenal or anorectal atresia, gastroesophageal reflux disease, and Hirschsprung disease.1 Older Down syndrome patients are at increased risk for leukemia and Alzheimer disease.1

OSA and opioid requirements in the pediatric patient
When planning an analgesic regimen for the pediatric patient with OSA, the anesthesiologist should pay attention to the oxygen nadir recorded during polysomnography. In a study

performed by Brown, postoperative morphine requirements were significantly correlated to preoperative oxygen saturation and patient age; the lower the oxygen nadir and patient age, the smaller the amount of opioid required for analgesia postoperatively.10 This decreased opioid requirement is thought to be due to hypoxia-induced increased number of μ opioid receptors. The use of adjunct analgesic medications, including dexamethasone, acetaminophen, dexmedetomidine and ibuprofen, may also have an opioid-sparing effect.

Postoperative care
Decisions pertaining to the postoperative care of a patient with OSA depends on the patient, underlying disease processes, type of surgery, and analgesia. ASA guidelines on the perioperative management of patients with OSA can help the anesthesiologist identify patients at risk for perioperative complications and the subsequent decision making process.
When considering inpatient versus outpatient status, the ASA recommends taking the following nine factors into consideration: sleep apnea status, anatomic and physiologic abnormalities, status of coexisting diseases, type of surgery, type of anesthesia, need for postoperative opioids, patient age, adequacy of post-discharge observation, and the capabilities of the outpatient facility.

The ASA also recommends that patients with increased risk from OSA should not be discharged to an unmonitored setting until they are no longer at risk for respiratory depression. Patients no longer at risk for respiratory compromise should be able to sleep in a non-stimulating environment and maintain a normal oxygen saturation while breathing room air. The guidelines note that for any given procedure, recovery for a patient with OSA may be longer than recovery for a patient without OSA.

When applying the ASA guidelines to this case, one notes that a history of Down syndrome, moderate OSA, airway surgery, and parenteral opioid requirement all place the patient at significantly increased risk for complications.4 Additionally, the possibility of bleeding and the presence of postoperative edema in a congenitally narrowed airway with laryngomalacia places the patient at further risk for airway obstruction. Admitting this patient for observation is the best choice. Where to admit the patient (i.e., an inpatient or intensive care unit) depends on the capabilities of the institution, with the decision being made in conjunction with the surgeon and other pediatric specialists.

Though tonsillectomy and adenoidectomy remains the first-line treatment for pediatric patients with OSA, post-operative assessment must be performed to assure resolution of symptoms.6 For the pediatric patient with refractory OSA, the use of CPAP and weight loss are recommended.6

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