

Anesthetic Issues in the Child with Morquio Syndrome: no such thing as a minor anesthetic.

Moderators: Robert P. Brislin, DO, Mary C. Theroux, MD

Institution: Nemours A.I. DuPont Hospital for Children

Objectives:

1. Discuss the anatomical and physiological airway difficulties in children with Morquio A Syndrome, or mucopolysaccharidosis (MPS) type IV A.
2. Value the importance of a thorough airway evaluation along with pulmonary and cardiology consultation prior to all procedures, even minor ones.
3. Recognize the essential steps in induction and airway management.
4. Discuss the importance of patient positioning during procedures and the indications for neuromonitoring.
5. Discuss the specific extubation criteria in children with Morquio's and anticipate the anatomical and physiological contributors to failed extubation.

Case History:

A ten year old girl with Morquio A Syndrome presents for cervical decompression and occiput to C4 fusion with placement of halo. Medical imaging reveals occipitocervical instability with compression and kyphosis at C3.

The family is originally from Iraq, moving to the USA five years ago. An older sibling also with Morquio's had a cervical fusion at another hospital. An emergency tracheotomy was done due to inability to ventilate or intubate on induction. Subsequently in the PICU the airway was lost during a tracheostomy change and the patient expired.

Questions:

What are the major manifestations of Morquio's Syndrome? What are the commonalities and differences between Morquio's and the other mucopolysaccharidoses? What type of cervical instability is typically seen in Morquio's? How might cervical myelopathy manifest?

Case History cont:

The child was born at full term via SVD. Birth weight was 3 kg. She is able to ambulate short distances, limited by knee pain due to lower extremity abnormalities. While living in Iraq the father relates she rarely ventured outside of the house. She exhibits no muscle weakness or bowel or bladder issues, but does exhibit brisk reflexes and several beats of clonus in the lower extremities. She has no allergies. She has mild aortic stenosis with insufficiency along with hypertension for which she is prescribed atenolol 12.5 mg BID. There were no prior surgeries.

She is an alert interactive girl. Her appearance is one of short stature and musculoskeletal manifestations of Morquio. Her current weight is 16 kg. Height 88. BMI is 21. Her HR is 86, RR 20, BP 101/58. She has a small barrel chest with pectus carinatum. Lungs are clear to auscultation. There is a 2/6 SEM. Examination of the airway reveals a short neck with limited flexion/extension, fair mouth opening with large tongue, Mallampati 3, and poor dentition with multiple chips and carries. Hematology and chemistry labs are normal.

A total spine MRI was done two weeks previously. The MRI revealed occipitocervical instability in flexion-extension views, spinal cord compression and kyphosis at C3, with lessening of compression on extension. There is also thoracolumbar kyphosis with wedging at L2 and L3. No sedation was used for the MRI due to the father's anxiety over the use of sedatives or anesthesia and the potential loss of the child's airway.

Questions:

What additional consults and diagnostic evaluations do you want preoperatively? What are you looking for specifically regarding 1) upper and lower airway anatomy and dynamics, 2) pulmonary function, 3) cardiovascular anatomy and function?

Case History cont:

Pulmonary evaluation reveals restrictive lung disease with poor thoracic cage compliance. Airway fluoroscopy demonstrates moderate tracheomalacia. Baseline ETCO₂ 44, SaO₂ on room air 98%. CXR reveals bibasilar atelectasis. Cardiology consultation reveals moderate aortic stenosis with a peak gradient of 33 mm Hg (mean 18 mm Hg), low moderate aortic regurgitation with moderate left heart enlargement but normal systolic function. There is a significantly dilated ascending aorta.

Questions:

Any additional study or studies that would assist in evaluating the airway? How would you optimize this child's pulmonary status?

How would you design the anesthetic induction and management of the airway with the patient's upper and lower airway issues in mind? What are the potential problems you may encounter during an inhalation or intravenous induction? How does the aortic stenosis and regurgitation affect your plan? What is essential in head and neck positioning during induction? What issues might you encounter with a fiberoptic intubation? Or videolaryngoscope? When is neuromonitoring indicated during intubation?

Intraoperative:

Following induction and successful intubation a halo was placed and the child is positioned prone.

Questions:

What intravenous access and monitoring would you use? What blood products would you have available? What anesthetic maintenance would you use during the cervical fusion? What are the positioning issues in the prone position? How does the patient's lower airway pathology affect your mechanical ventilation strategy?

What are your interventions if there is a change in motor-evoked potentials?

Intraoperative conclusion:

At the conclusion of the surgery, the child is returned supine.

Questions:

The father is extremely anxious over the timing of extubation and the monitoring of his child once extubated. What are your extubation criteria in this child? What is your post-extubation plan? What are the issues that make a failed extubation more likely in this child? What are the issues postoperatively from a cervical fusion that make a failed extubation more dangerous? What respiratory modalities may be useful following extubation? How will you communicate with the PICU consultants about a plan in case of respiratory difficulties during the first postoperative night?

Post-operative:

The child is extubated and taken to the PICU for cardiorespiratory and neurologic monitoring. Pain Service is consulted for analgesia.

What are your recommendations for analgesia?

Epilogue

This child returns at a later date for femoral osteotomies. How are the airway issues different following a cervical fusion? What are the positioning issues intra-operatively? Is neuromonitoring indicated during intubation? Is neuromonitoring indicated during the procedure? Would you place an epidural?

This child returns for a bilateral myringotomy with tube placement. What information do you require preoperatively? What is your anesthetic plan?

What airway issues become more significant as the child reaches later adolescence and into adulthood? What about other systemic issues?

Discussion

Morquio A Syndrome (MS), also known as mucopolysaccharidosis (MPS) type IVA, is a progressive lysosomal storage disorder with autosomal recessive inheritance. Accumulation of glycosaminoglycans (GAG) deposits, mainly keratan sulfate, cause anatomic and physiologic abnormalities throughout the body. Children with Morquio's appear normal at birth, but accumulation of GAG deposits cause the manifestation of skeletal dysplasia with prominent forehead, kyphoscoliosis, pectus carinatum, and genu valgum. Cervical spine abnormalities are a significant issue in children with Morquio's due to odontoid hypoplasia, ligamentous laxity, incomplete ossification, and extradural GAG deposits. Cervical myelopathy may progress to quadriplegia or death if untreated. Cervical myelopathy may manifest as progressive exercise intolerance.

Children with Morquio are especially prone to have a difficult airway. A large tongue, short neck, submucosal and cartilaginous GAG deposits, barrel chest, and either an unstable cervical spine or an immobile cervical spine following cervical fusion can make mask ventilation and endotracheal intubation difficult. In addition, emergency tracheotomy may be hazardous due to the short neck, abnormal tracheal rings and mucosa, fused cervical spine, and the possibility of the trachea being located beneath the sternum and inaccessible to traditional surgical approach. Positioning of the head and neck during airway management is particularly important. The airways of MS patients can easily obstruct when flexed due to anterior buckling of the abnormal tracheal walls. MS patients often align themselves with their head looking up as this position optimizes airway patency and unobstructed breathing. This position should be maintained during anesthesia. Neuromonitoring may be required during induction and intubation in a child with an unstable cervical spine.

Pulmonary and otolaryngology consultations are essential to document upper and lower airway abnormalities due to the accumulation of GAG causing thickened, stiff upper airway structures, and weakened, deformed tracheo-bronchial walls. PFTs may demonstrate significant restrictive disease with elevated airway resistance. A chest CT can delineate deviation or obstruction of the trachea. Morquio's patients are also prone to obstructive sleep apnea. Cardiology evaluation is necessary to document valvular and aortic integrity and ventricular function. As Morquio's patients reach late adolescence they can develop intimal sclerosis of the coronary arteries. Accumulation of GAG deposits in the myocardium may lead to significant ventricular dysfunction.

Children with Morquio's require diligent preoperative evaluation even for minor procedures as upper and lower airway, pulmonary, cardiovascular, and neurologic sequelae may have significantly progressed since the last anesthetic. Recommended actions include: strict attention to head and neck positioning, proper preparation for airway management with the necessary airway equipment and assistance, special airway device such as a videolaryngoscope accompanied by tongue retraction, and a formulated plan for extubation criteria and measures to optimize upper airway and lower airway function after extubation. Especially in longer cases, these children may be at risk for spinal cord ischemia while supine depending on the extent of extradural compression from GAG deposits and the thoracolumbar kyphosis. Careful padding and support of the spine is necessary and sometime intraoperative neuromonitoring is required.

References

Theroux MC, Nerker T, Ditro C, Mackenzie WG. Anesthetic care and perioperative complications of children with Morquio Syndrome. *Pediatr Anaesth* 2012; **22**: 901-907.

Tomatsu S, Mackenzie WG, Theroux MC, Mason, RW, et al. Current and emerging treatments and surgical interventions for Morquio A syndrome: a review. *Research and Reports in Endocrine Disorders* 2012; **2**: 1-13.

Solanki GA, Martin KW, Theroux MC, Lampe C. Spinal involvement in mucopolysaccharidosis IV A (Morquio-Brailsford or Morquio A syndrome): presentation, diagnosis, and management. *J Inher Metab Dis* 2013; **February 6 Online**: Springerlink.com.

Tong CKW, Chen JCH, Cochrane DD. Spinal cord infarction remote from maximal compression in a patient with Morquio syndrome. *J Neurosurg Pediatrics* 2012; **9**: 608-612.

Rodriguez M, Miller TL, Mackenzie WG, Ditro C, Chidekel AS, Shaffer TH. Characteristics of impulse oscillometry and thoracoabdominal motion in children with thoracic cage disorders. *Pediatr Pulmonol.* 2010; **45(7)**: 679-686.

Tobias JD. Anesthetic care for the child with Morquio syndrome: general versus regional anesthesia. *J Clin Anesth* 1999; **11**: 242-246.