Lesson 292: PreAnesthetic Assessment of the Patient With Mucopolysaccharidosis

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REVIEW DATE: May, 2011

Read this article, reflect on the information presented, then go online and complete the lesson post-test and course evaluation before the termination date below. (CME credit is not valid past this date.) You must achieve a score of 80% or better to earn CME credit.

TIME TO COMPLETE ACTIVITY: 2 hours
RELEASE DATE: June 1, 2011
TERMINATION DATE: June 30, 2012

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Needs statement

Mucopolysaccharidosis (MPS) describes a group of genetic disorders that can complicate the anesthetic care of patients—in particular, management of the airway. Patients with MPS should be managed by experienced anesthesiologists at centers that are familiar with these disorders. Rarely encountered disease states have been identified as important topics in the continuing education of clinical anesthesiologists.
Learning Objectives

At the end of this activity, the participant should be able to:

1. Define MPS.
2. Describe the pathophysiology of MPS.
3. Explain how tissue dysfunction develops in patients with such rare lysosomal storage disorders.
4. Cite the incidence of MPS.
5. List the clinical manifestations of MPS.
6. List specific problems associated with the administration of anesthesia to patients with MPS.
7. Present treatment options for patients with MPS.
8. Define appropriate preoperative evaluation of these patients.
10. Anticipate potential problems in airway management, postoperatively.

Case History

A 2-year-old boy with MPS type I (Hurler syndrome) presented for bilateral inguinal hernia repair. The child often made grunting sounds while asleep, according to his parents. He had been given several antibiotics in the past for frequent respiratory infections, and was found to have a mild heart defect. The patient had no allergies and had never received an anesthetic. He weighed 14 kg. A physical examination was remarkable for coarse facies, macroglossia, short neck, and hepatosplenomegaly. A cardiac examination was positive for a 2 of 6 murmur. Mild aortic regurgitation with normal left ventricular systolic function and no wall motion abnormalities were observed with 2-D echocardiography. Cervical spine and chest x-rays were within normal limits.

Mucopolysaccharidosis (MPS) describes a group of rare lysosomal storage disorders characterized by a deficiency or complete lack of lysosomal enzymes necessary for the stepwise breakdown of glycosaminoglycans (GAGs), also known as mucopolysaccharides.1‐3 Consequently, fragments of GAGs accumulate intracellularly in the lysosomes, which results in cellular enlargement that causes disruption and dysfunction of tissues. This process leads to numerous clinical abnormalities. The incidence of all types of MPS is reported to be between 1 in 10,000 to 1 in 30,000 live births.1

Pathophysiology

GAGs are long-chain complex carbohydrates comprising repeating disaccharide units of sulfated acidic and amino sugars. GAGs usually are linked to proteins to form proteoglycans, which are the major constituents of the ground substance of connective tissue, the lubricant in joint fluid, and the surface coating that initially binds growth factors to cells. The major GAGs are chondroitin-4-sulfate, chondroitin-6-sulfate, heparan sulfate, dermatan sulfate, keratan sulfate, and hyaluronic acid. Within organisms, these substances are degraded by the sequential action of lysosomal enzymes, leading to a stepwise shortening of the terminal sulfate, acidic, and amino sugar residues. Deficient or dysfunctional activity of the degrading enzymes results in MPS disorders, of which there are 11 types based on level of severity (Table). Clinical phenotypes of the disorder depend on the distribution and turnover of the substrate affected by the deficiency, instead of the distribution of the enzyme.1‐6
### TABLE: MPS DISORDERS

<table>
<thead>
<tr>
<th>Type</th>
<th>Eponym</th>
<th>Enzyme Deficiency</th>
<th>GAG Stored</th>
<th>Craniofacial Abnormalities</th>
<th>Joint and Skeletal Deformities</th>
<th>Cardiac Involvement</th>
<th>Visceral, Visual, Neurologic Manifestations</th>
</tr>
</thead>
<tbody>
<tr>
<td>MPS I (severe)</td>
<td>Hurler syndrome</td>
<td>α-L-iduronidase</td>
<td>Dermatan sulfate, heparan sulfate</td>
<td>Macrocephaly, coarse facies, macroglossia, hydrocephalus</td>
<td>Joint stiffness, thoracolumbar kyphosis, possible odontoid deformity, hypoplasia, short neck, short stature</td>
<td>Coronary intimal and valvular thickening, mitral regurgitation, cardiomegaly</td>
<td>HSM, umbilical and inguinal hernias, corneal clouding, severe mental retardation</td>
</tr>
<tr>
<td>MPS I (attenuated)</td>
<td>Scheie syndrome</td>
<td>α-L-iduronidase</td>
<td>Dermatan sulfate, heparan sulfate</td>
<td>Coarse facies, macroglossia, prognathism</td>
<td>Short neck, normal stature</td>
<td>Aortic regurgitation</td>
<td>HSM, umbilical and inguinal hernias, corneal clouding</td>
</tr>
<tr>
<td>MPS I (attenuated with different features)</td>
<td>Hurler-Scheie syndrome</td>
<td>α-L-iduronidase</td>
<td>Dermatan sulfate, heparan sulfate</td>
<td>Macrocephaly, coarse facies, macroglossia, micrognathia</td>
<td>Diffuse joint limitation, short neck, short stature</td>
<td>Mitral and aortic valve thickening and regurgitation</td>
<td>HSM, umbilical and inguinal hernias, corneal clouding</td>
</tr>
<tr>
<td>MPS II (severe)</td>
<td>Hunter syndrome (severe)</td>
<td>Iduronate sulfatase</td>
<td>Dermatan sulfate, heparan sulfate</td>
<td>Macrocephaly, coarse facies, hydrocephalus</td>
<td>Diffuse joint limitation, short neck, short stature</td>
<td>Coronary intimal thickening, ischemic cardiomyopathy</td>
<td>HSM, corneal clouding</td>
</tr>
<tr>
<td>MPS II (attenuated)</td>
<td>Hunter syndrome (mild)</td>
<td>Iduronate sulfatase</td>
<td>Dermatan sulfate, heparan sulfate</td>
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<tr>
<td>MPS IIIA (symptoms appear after the first year of life)</td>
<td>Sanfilippo A syndrome</td>
<td>Heparan-N-sulfatase</td>
<td>Heparan sulfate</td>
<td>Coarse facies, heavy eyebrows that meet in center of face above the nose</td>
<td>Mild stiffness of joints, short stature, dysphagia</td>
<td>Minimal to none</td>
<td>Severe retardation, behavioral problems, diarrhea</td>
</tr>
<tr>
<td>MPS IIIB</td>
<td>Sanfilippo B syndrome</td>
<td>α-N-acetyl glucosaminidase</td>
<td>Heparan sulfate</td>
<td>Coarse facies</td>
<td>Mild stiffness of joints, short stature, walking problems, lumbar-vertebral dysfunction, dysphagia</td>
<td>Minimal to none</td>
<td>Developmental delay, behavioral problems</td>
</tr>
<tr>
<td>MPS IIIC</td>
<td>Sanfilippo C syndrome</td>
<td>Acetyl CoA: α-glucosaminide N-acetyltransferase</td>
<td>Heparan sulfate</td>
<td>Coarse facies</td>
<td>Mild stiffness of joints, short stature, lumbar-vertebral dysfunction, dysphagia</td>
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<tr>
<td>MPS IID</td>
<td>Sanfilippo D syndrome</td>
<td><em>N</em>-acetylg glucosamine-6-sulfatase</td>
<td>Heparan sulfate</td>
<td>Coarse facies</td>
<td>Mild stiffness of joints, short stature, lumbar-vertebral dysfunction, dysphagia</td>
<td>Minimal to none</td>
<td>Developmental delay, behavioral problems</td>
</tr>
<tr>
<td>MPS IVA</td>
<td>Morquio syndrome type A</td>
<td>Galactose-6-sulfatase</td>
<td>Keratan sulfate, chondroitin-6-sulfate</td>
<td>Coarse facies</td>
<td>Joint laxity, severe kyphoscoliosis, odontoid hypoplasia, short neck, C1-C2/C2-C3 subluxation, short stature</td>
<td>Aortic regurgitation</td>
<td>Mild corneal opacities, HSM</td>
</tr>
<tr>
<td>MPS IVB</td>
<td>Morquio syndrome type B</td>
<td>β-galactosidase</td>
<td>Keratan sulfate, chondroitin-6-sulfate</td>
<td>Coarse facies</td>
<td>Joint laxity, severe kyphoscoliosis, odontoid hypoplasia, short neck, C1-C2/C2-C3 subluxation, short stature</td>
<td>Aortic regurgitation</td>
<td>Mild corneal opacities, HSM</td>
</tr>
<tr>
<td>MPS VI</td>
<td>Maroteaux-Lamy syndrome</td>
<td><em>N</em>-acytylgalactosamine-4-sulfatase</td>
<td>Dermatan sulfate, heparan sulfate</td>
<td>Macrocephaly, coarse facies, macroglossia, hydrocephalus</td>
<td>Mild stiffness of joints, kyphoscoliosis, odontoid hypoplasia, short stature</td>
<td>Mitral and aortic valve thickening and regurgitation</td>
<td></td>
</tr>
<tr>
<td>MPS VII</td>
<td>Sly syndrome</td>
<td>β-glucuronidase</td>
<td>Dermatan sulfate, heparan sulfate, chondroitin-4,6-sulfate</td>
<td>Macrocephaly, coarse facies</td>
<td>Joint flexion, contractures, thoracolumbar deformity, hip dysplasia, odontoid hypoplasia, short stature</td>
<td>Mitral and aortic valve thickening and regurgitation</td>
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<tr>
<td>MPS IX</td>
<td></td>
<td>Hyaluronidase</td>
<td>Hyaluronan</td>
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**GAG**, glycosaminoglycans; **HSM**, hepatosplenomegaly; **MPS**, mucopolysaccharidosis

Modified and adapted from reference 11.

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**Classification**

Of the 11 MPS disorders, there are 7 major types classified I through IX. (MPS V, formerly Scheie syndrome, and MPS VIII are no longer recognized types.) The types of MPS disorders are differentiated by clinical features and age at presentation, and biochemically by the associated enzyme deficiency. As a general rule, impaired degradation of heparan sulfate is more closely associated with mental deficiency, and impaired degradation of dermatan, chondroitin, and keratan sulfates results in mesenchymal abnormalities.5,6 Overall, MPS disorders can be grouped into 4 broad categories
according to the dominant clinical features expressed:

- soft tissue storage and skeletal disease, with or without brain disease (MPS I, II, and VII);
- soft tissue and skeletal disease (MPS VI);
- primarily skeletal disease (MPS IVa and IVb); and
- primarily disease of the central nervous system (CNS; MPS IIIa-IIIId).

**Clinical Manifestations**

MPS disorders are characterized by progressive craniofacial, joint, and skeletal deformities, progressive cardiac involvement, and early death (often during childhood) from pulmonary infection or cardiac failure.\(^1\)\(^-\)\(^18\) Hurler syndrome is the prototypical MPS disorder, occurring in 1 in 100,000 live births (Figure). Deposits of GAGs lead to thickened heart valves, causing valvular insufficiency more often than stenosis. Myocardial hypertrophy, ventricular dysfunction, and cardiomyopathy result from accumulation of GAGs in the myocardium, frequently ending in congestive heart failure and death.

Intimal deposition of GAGs causes coronary luminal narrowing and occlusion that can be progressive.\(^1\)\(^-\)\(^13\) GAGs also are deposited in abdominal viscera, leading to hepatosplenomegaly (HSM) in most, if not all, patients. Umbilical and inguinal hernias can result from abdominal protuberance from HSM. Ineffective support of connective tissue of the anterior abdominal wall often develops.

In addition to heart disease and HSM, most infants have chronic pulmonary disease caused by restriction of the thoracic cage due to kyphoscoliosis, airway obstruction secondary to deposition of GAGs in the upper airways, recurrent pulmonary infections, pulmonary hypertension, and cardiomyopathy. Tongue protrusion and excessive tracheobronchial secretions are common.\(^1\)\(^-\)\(^13\)

**Diagnosis**

A physician should suspect MPS when a child presents with coarse facies, HSM, bone disease, and heart disease with or without CNS abnormalities. However, the initial presentation may be subtle and signs may be variable, depending on the type and severity of MPS. Measuring urinary concentration of GAGs can assist in identifying MPS; a definitive diagnosis is made by assaying enzyme activity in peripheral blood leukocytes.

Skeletal radiography may reveal the characteristic pattern of abnormalities known as dysostosis multiplex. An eye examination should be performed to assess corneal clouding and glaucoma, which is most common in MPS I, II, VI, and VII. A complete cardiac evaluation is necessary to adequately assess valvular and myocardial disease. A comprehensive neurologic examination is required to assess the potential for spinal cord compression and hydrocephalus.\(^1\)\(^-\)\(^6\),\(^13\)
Complications

Cardiopulmonary complications are the most common cause of death in patients with MPS.

Respiratory: Respiratory abnormalities are the result of airway obstruction, neurologic compromise, recurrent infections, skeletal restrictions, and organomegaly—all of which can lead to pulmonary insufficiency, severe sleep apnea, and sudden death from central apnea. Patients with MPS IV are especially prone to high cord compression secondary to atlantoaxial instability and odontoid dysplasia, which can lead to depressed respiration or sudden respiratory arrest.

Upper airway obstruction can result from redundant airway tissue caused by MPS deposition in the soft tissues of the nasopharynx. Enlarged tonsils and adenoidal tissue, macroglossia, and thickened gums also may be present. Secretions are excessive due to chronic or recurrent ear and sinus infections. Treatment focuses on maintenance of a stable airway. Airway obstruction can be reduced temporarily by removal of the tonsils and adenoids, along with use of positive airway pressure.11,16

Cardiac: Cardiac abnormalities are well documented. Valvular disease is caused by progressive thickening of the mitral and aortic valves; it is common in MPS types I, II, and VI. The defect typically results in heart failure; in severe cases, valve replacement may be required. The narrowing of coronary vessels secondary to the intimal deposition of MPS impairs blood flow; cardiac ischemia results. Pulmonary hypertension may exacerbate right-sided heart failure.13,16

Skeletal and Connective Tissue: Complications develop as GAGs accumulate in the bones, joints, and ligaments. Patients with MPS types I, II, VI, and VII are known to be affected by dysostosis multiplex; those with types I, IV, and VII are affected by odontoid hypoplasia. Hypoplasia can lead to atlantoaxial instability, C1 to C2 subluxation, and high spinal cord compression. Prophylactic cervical fusion should be undertaken to prevent progressive cord compression.

Vertebral subluxation and kyphoscoliosis develop throughout the spinal column, which can compromise the spinal cord; patients may need spinal fusion for stabilization. Unfortunately, patients typically heal poorly from these surgeries and often develop complications requiring repeat surgery.

Short stature is a common finding throughout the spectrum of MPS types. Patients often present with joint stiffness secondary to accumulation of MPS in the synovial fluid and other connective tissues of the joints.

Gastrointestinal: Complications include recurring inguinal and umbilical hernias, and HSM with increased intra-abdominal pressure. More than one surgical repair is often necessary.11,13,16

Neurologic: Neurologic complications are well documented. Developmental delay and progressive neurologic decline characterize the severe forms of MPS I, II, III, and VII. Communicating hydrocephalus frequently develops in types I, II, III, VI, and VII because of the engorgement of arachnoid granulations by storage material; this impedes resorption of cerebrospinal fluid (CSF) and increases intracranial pressure.

In MPS III, hydrocephalus is secondary to ventricular enlargement because of cerebral atrophy in later stages of the illness. Pachymeningitis cervicalis, a progressive thickening and scarring of the meninges
around the cervical spinal cord caused by accumulation of MPS, is another neurologic complication. Such thickening may form a sleeve around the spinal cord that impedes the flow of CSF, and progressively compresses the cervical cord. Cord compression from pachymeningitis cervicalis and odontoid dysplasia can result in progressive ascending paresis and paralysis.11,15

**Ophthalmic and Auditory:** Vision and hearing complications are common. Ophthalmologic manifestations include corneal clouding; blindness can develop. Eye examinations should be conducted at the time of diagnosis and annually thereafter.

Auditory manifestations include conductive and neurosensory deafness. Hearing loss may be attributable to frequent ear infections, defective ossification in the middle ear, scarring of the tympanic membrane, or nerve damage. Annual audiologic examinations are warranted, and are particularly important for patients with MPS type I. Hearing aids are beneficial.

**Therapy**

The treatment of patients with MPS disorders is usually symptomatic and not specific. Bone marrow transplantation has been used successfully to treat some of the disorders in the MPS spectrum. In most patients with successful engraftment, transplantation reduces HSM, increases joint mobility, decreases airway obstruction, improves cardiac function, decreases CSF pressure, and especially in younger patients may stabilize mental regression. Unfortunately, BMT does not correct skeletal disorders or prevent decline of the CNS in severe cases.

Immunosuppressant treatment is required. The therapy routinely is offered only to patients with Hurler syndrome younger than approximately 2 years. Immunosuppressant therapy is less commonly used in patients with mild MPS types II, VI, and VII. Cord blood is another potential source for transplantation.19

Emerging treatments for MPS include enzyme replacement therapy, substrate reduction therapy, chaperone-mediated therapy, and gene therapy. Although clinical efficacy has not been shown completely for any of these therapies, many clinicians are optimistic that future developments will lead to a disease-modifying treatment.20,21 Enzyme replacement therapy with recombinant iduronate-2-sulfatase (idursulfase) is under clinical investigation, and weekly IV infusions have been found to improve many of the symptoms and signs of MPS.20

**Anesthesia Considerations**

**Preoperative Evaluation:** Assessments of neurologic function, the probability of a difficult airway and ventilatory management, cardiac complications, skeletal disease, and visceral manifestations should be considered preoperatively.1,2,11,13 Chest x-ray, arterial blood gas analysis, and pulmonary function tests may be indicated in patients with chronic pulmonary infections or kyphoscoliosis.

Vital capacity, functional residual capacity, and total lung capacity often are reduced by skeletal restrictions. Preoperatively, the goal should be to optimize lung capacity; this may include physiotherapy, pulmonary toilet, and antibiotics if infection is present.1,2,11,13

To assess clinically relevant spinal disease, radiography should be used to identify atlantoaxial
subluxation—especially in patients with Morquio or Hurler syndrome. Flexion–extension cervical films may confirm the potential for subluxation and demonstrate tracheal collapse on flexion. Atlantoaxial subluxation contraindicates cervical extension during endotracheal intubation.

Spinal cord compression due to subluxation frequently occurs within the spectrum of MPS. Patients presenting with clinical manifestations such as abnormal gait, sensory changes, or weakness in the lower extremities should be evaluated by a neurologist. Somatosensory-evoked potentials can be used to detect early cord compression and guide the timing of surgical intervention.

Patients with MPS I who undergo spinal surgery are at increased risk for major complications, including spinal cord infarction and spinal instability. Communicating hydrocephalus may be present, and if suspected, a measurement of CSF pressure should be considered. With increased intracranial pressure, a ventriculostomy may successfully reduce CSF pressure, but typically does not significantly reverse clinical disease.

An enlarged heart and pulmonary congestion should prompt an evaluation by 2-D echocardiography, which can detect right ventricular hypertrophy with strain, conduction blocks, left atrial enlargement, tachydysrhythmias, and ischemic changes. Systolic murmurs are common and this too should prompt an echocardiographic evaluation. If the patient experiences chest pain, or clinical symptoms that suggest ischemia, more invasive diagnostic testing (eg, angiography) is indicated.

Case report findings have suggested that the contribution of cardiac involvement—particularly mitral insufficiency and cardiomegaly—was minimal in stress tolerance related to anesthetic management; however, severe and extensive coronary obstruction was cited as a cause of 2 deaths, intraoperatively.

Patients with moderate to severe skeletal disease should be monitored continually by an orthopedic surgeon. Spine deformities may require fusion; acetabular hip dysplasia can be managed with osteotomy; and genu valgum with epiphyseal stapling. Carpal tunnel release can provide relief and return some function to the hands.

Visceral manifestations of disease are common. Normally, inguinal hernias have been repaired before disease diagnosis. Umbilical hernias often recur, probably due to HSM. Because the most common clinical manifestations include chronic upper respiratory infections, it is important to identify any potentially infectious processes. Tonsillectomy and adenoidectomy should be considered for all patients in whom the airway is compromised.

Patients with MPS typically undergo a routine annual examination of ears, nose, and throat. A careful preanesthetic assessment can be invaluable, however, if underlying pathophysiologic processes are subclinical and have not been identified.

**Drugs:** Premedication sedatives should be administered cautiously, if at all, because of risks for upper airway obstruction, respiratory depression, hypercarbia, and cardiorespiratory arrest. Opioids should be avoided in these patients if airway problems are anticipated because of respiratory depression. Oropharyngeal secretions can be controlled with anticholinergics, such as scopolamine or glycopyrrolate.
Hurler syndrome, being the prototypical and most severe form of the MPS disorders, results in difficult tracheal intubation in as much as 50% of these patients. Some studies have suggested IV induction for younger patients with a lesser degree of craniofacial involvement, and inhaled induction for older patients in whom airway difficulties are established or anticipated.\(^{3,4,8}\) Other studies maintain that inhalation induction is preferable; however, IV induction may be necessary in patients with severe mental retardation or who are uncooperative. Many studies have suggested that induction with intramuscular ketamine is preferred to inhalation induction.\(^{3,4,8}\)

Patients with MPS seem not to be at increased risk for malignant hyperthermia. Maintenance anesthesia is usually achieved with an inhalational agent.\(^{3,4,8}\) The muscle relaxant of choice is often short-acting and nondepolarizing.

**Airway Management:** In patients with MPS, ventilation may be difficult as a result of abnormal facies. An air-cushioned pediatric face mask may be applied upside down, with the broad chin edge of the mask over the patient’s brow and nose, and the narrow nasal bridge of the mask over the open mouth and protruding tongue.\(^{11}\) Advanced instruments for managing the airway should be available, including an assortment of face masks, endotracheal tubes, laryngoscope blades and handles, fiber-optic equipment, a video laryngoscope, and the difficult airway cart, in addition to a surgeon standing by in case emergency tracheostomy is necessary.

Direct laryngoscopy in awake orotracheal intubation will be difficult. Airway manipulation is much easier in a deeply sedated, spontaneously ventilating patient.\(^{11}\) As mentioned, atlantoaxial subluxation secondary to odontoid hypoplasia/dysplasia with spinal cord and brainstem compression may occur during cervical hyperextension.\(^{11}\) Cervical traction can be used to prevent manipulation of the neck.

Deposits make it extremely difficult to feel the trachea; thus, retrograde catheter-guided tracheal intubation is not recommended. Blind nasotracheal intubation and tracheostomy carry significant risks, and are recommended only in emergency situations. Some clinicians believe that a fiber-optic bronchoscope should be available as part of the anesthetic management of all known cases of difficult intubation.\(^{11}\)

**Postoperative Management:** The pediatric patient emerging from anesthesia may experience difficulty in breathing against the high airway resistance of an endotracheal tube. Pulmonary hypertension can be exacerbated, and negative pressure pulmonary edema may ensue and require immediate management, including mechanical ventilatory support.

Multiple attempts at intubation should be avoided because it can lead to symptomatic glottic and subglottic edema. Such iatrogenic conditions are very difficult to treat because of a progressive narrowing of the tracheal lumen by MPS deposits.\(^{8,11}\) Conducting a fiber-optic intubation, and postoperatively leaving the endotracheal tube immediately in place, minimizes airway complications—particularly in cases in which not all of the extubation criteria have been met. After tracheal extubation, humidified oxygen, chest physiotherapy, and postural drainage should be instituted, and continued until the patient is ambulatory and able to expectorate excessive secretions.\(^{8,11}\)
Management of the Case Presented

Two anesthesiologists accompanied the child (who was not sedated) to the operating suite. Both a laryngeal mask airway and fiber‐optic tower were available. Sizes 4.0, 4.5, and 5.0 cuffed endotracheal tubes were prepared. American Society of Anesthesiologists standard monitors were applied, and a mask induction with sevoflurane was facilitated by placement of an oral airway while the second anesthesiologist secured peripheral IV access with a 22‐gauge angiocatheter. Propofol, 30 mg IV, was administered. Laryngoscopy, performed with a Macintosh 2 blade, revealed a grade III view.

The first attempt at laryngoscopy with a 4.5 cuffed endotracheal tube was unsuccessful. A second attempt with manipulation of the larynx improved the view to grade II. Correct placement of the tube was confirmed by capnography and bilateral breath sounds. The patient was placed in the lateral position and a caudal block performed in sterile fashion with a 22‐gauge angiocatheter and 15 mL of ropivacaine 0.2%.

Anesthesia was maintained with sevoflurane in oxygen. After an injection of ropivacaine 0.2% to the surgical site and closure of the wound, the patient’s trachea was extubated while the patient was fully awake. The post‐operative course was uneventful and the patient was discharged home the next day.

Conclusion

The perioperative management of patients with MPS often is difficult, and although new treatments are providing hope, many challenges remain. Understanding the pathophysiology of this group of diseases increases awareness of the potential risks associated with anesthesia and surgery. Ideally, pediatric patients with MPS should be managed by anesthesiologists familiar with the disease process to minimize complications and reduce morbidity and mortality.

Dr. Elizabeth A.M. Frost, who is the editor of this continuing medical education series, is clinical professor of anesthesiology at The Mount Sinai School of Medicine in New York City. She is the author of Clinical Anesthesia in Neurosurgery (Butterworth‐Heinemann, Boston) and numerous articles. Dr. Frost is past president of the Anesthesia History Association and former editor of the journal of the New York State Society of Anesthesiologists, Sphere. She is also editor of the book series based on this CME program, Preanesthetic Assessment, Volumes 1 through 3 (Birkhäuser, Boston) and 4 through 6 (McMahon Publishing, New York City).
REFERENCES


Post-test

1. Mucopolysaccharidosis (MPS) is a glycogen storage disorder characterized by:
   a. a deficiency in enzymes that break down glycosaminoglycans
   b. a defined genetic abnormality
   c. excess serotonin
   d. hyperbilirubinemia

2. The incidence of MPS is:
   a. estimated to be 1 in 10,000 to 1 in 30,000
   b. so rare as to be unknown
   c. variable depending on the country
   d. estimated to be 1 in 50,000 to 1 in 100,000

3. In patients with MPS, respiratory abnormalities can result from:
   a. neurologic compromise
   b. skeletal restrictions
   c. organomegaly
   d. All of the above are correct.

4. Which of the following is descriptive of severe Hunter syndrome?
   a. Elevated high-density lipoprotein
   b. Corneal clouding
   c. High level of serum magnesium
   d. Low level of serum potassium

5. A well-documented skeletal complication within the spectrum of MPS disorders is:
   a. dysostosis multiplex
   b. spina bifida
   c. osteogenesis imperfecta
   d. Marfan syndrome
6. Which of the following is not a concern for the anesthesiologist managing a surgical patient with MPS?

a. Securing the airway
b. Oropharyngeal secretions
c. Skeletal abnormalities
d. Higher risk for malignant hyperthermia

7. Patients with MPS I (Hurler syndrome) are not usually noted to have:

a. delayed development
b. severe mental retardation
c. shortened life expectancy
d. unusually tall stature

8. Within the spectrum of MPS disorders, the rate of difficult tracheal intubation can be as high as:

a. 5%
b. 100%
c. 40%
d. 50%

9. Visceral manifestations are common within the spectrum of MPS disorders; the most common reason for recurrence of umbilical hernias in these patients is:

a. obesity
b. hepatosplenomegaly
c. ascites
d. heavy lifting

10. Which of the following therapies has not been used successfully for patients with MPS?

a. Bone marrow transplantation
b. Enzyme replacement
c. Substrate reduction
d. Serotonin reuptake inhibitors and dopamine antagonists