



## Case report

# Anesthesia for an adult with mucopolysaccharidosis I

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**Abstract** We describe the anesthetic management difficulties of a man with mucopolysaccharidosis I. We also briefly review the anesthesia literature related to this disease.  
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## 1. Introduction

Hurler syndrome (mucopolysaccharidosis I [MPS I]) is a rare metabolic disorder that is associated with a high degree of anesthetic risk. We describe the anesthetic management in a 21-year-old patient with Hurler syndrome who required a transoral odontoidectomy, posterior spinal fusion, and tracheostomy secondary to odontoid hypoplasia. Anesthesiologists can expect to see more of these patients in the future as new therapies such as bone marrow transplant and enzyme replacement therapy extend life expectancy [1,2]. This will create a new population of patients living longer and requiring surgical intervention for a variety of indications, including cervical decompression and stabilization [3].

## 2. Case report

A 21-year-old white man with a history of MPS I was scheduled for the aforementioned procedure. His medical history was significant for sleep apnea, mitral valve prolapse, mild asthma, and chronic otitis media. He presented with

1.5-year history of progressive spastic quadriplegia, extreme pain with walking, and lower back and leg numbness. His past surgical history included bilateral carpal tunnel release, capsulectomy of metacarpophalangeal joints, repair of umbilical hernia, tonsillectomy, and adenoidectomy. His last anesthetic was administered 4 years before admission, but no anesthetic records were available. His medications included enzyme replacement therapy with  $\alpha$ -L-iduronidase once weekly and albuterol inhaler as needed.

On physical examination, the patient was 152.4 cm and weighed 50 kg. The rest of the physical examination was notable for a short neck with limited mobility, an inability to open his mouth fully and a Mallampati IV airway, and flexed wrists and elbows (Fig. 1). His vital signs were heart rate, 91; blood pressure, 135/90; and respiratory rate, 20.

Laboratories and chest x-ray were unremarkable. An echocardiogram revealed a normal ejection fraction and no valvular abnormalities (other than mild mitral valve prolapse). Pulmonary function tests showed a restrictive pattern with the forced vital capacity of 44% of the reference value. A sleep study revealed episodes of frank apneas that lasted up to 45 seconds and were associated with desaturations down to 82%. Magnetic resonance imaging of the cervical region demonstrated a soft tissue mass compressing the entire upper cervical cord down to the C2-3 disk space. The

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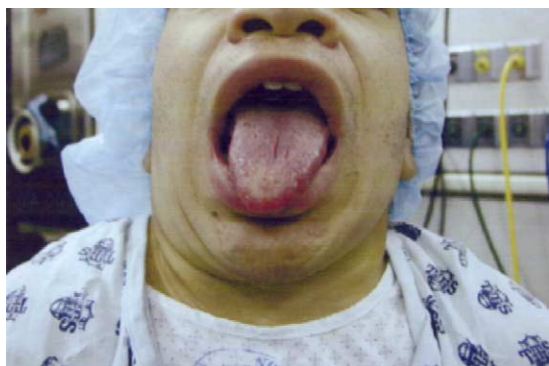
remainder of the spinal canal was narrowed from posterior compression (Fig. 2).

From the preoperative assessment, a difficult intubation was anticipated and an awake fiber-optic intubation was planned. The patient was pretreated with 0.4 mg of glycopyrrolate. On arrival to the operating room, routine monitors were placed, an 18-gauge intravenous line was established, and oxygen was administered via nasal canula. Scopolamine 0.3 mg intravenous and droperidol 2.5 mg intravenous were administered for sedation. The patient was acceptably cooperative. Airway topicalization was accomplished with 5 mL of lidocaine 4% and nebulization, and lidocaine 2% gel was applied to the pyriformis fossa to block the superior laryngeal nerve. We could not perform a transtracheal or direct superior laryngeal block because of distorted anatomy. Lidocaine 2% was sprayed on the vocal cords under direct vision with the fiberoptic scope. Despite all this preparation, intubation took 4 attempts before it was successfully completed. Copious oral secretions hampered visualization and proved the greatest obstacle to successful intubation. Visualization was improved by changing the patient's position from supine to sitting.

General anesthesia was induced with propofol, fentanyl, and cisatracurium after correct position of the endotracheal tube was confirmed and the patient moved all extremities. A second peripheral intravenous line (16 gauge) as well as 20-gauge arterial catheter was placed after induction of general anesthesia.

He was positioned supine for tracheostomy, which was followed by odontoidectomy. This lasted 10 hours. The patient was then turned prone for the occipitocervical fusion (down to C5 level).

Anesthesia was maintained with oxygen, N<sub>2</sub>O, isoflurane, fentanyl, and cisatracurium boluses. The surgery took nearly 24 hours, but it was completed uneventfully. The patient lost 2000 mL of blood. Eight liters of Ringer's lactate, 4 U of packed red blood cells, and 2 U of fresh frozen plasma were administered intraoperatively. The patient remained hemodynamically stable throughout the procedure and did not require vasoactive drugs. He was able to breathe without ventilator support by postoperative day 2.



**Fig. 1** Adult with MPS I.



**Fig. 2** T2-weighted sagittal cervical spine magnetic resonance image showing severe irreducible cervicomedullary compression secondary to MPS I.

The remainder of his hospital stay was uneventful, and he was discharged to long-term rehabilitation.

### 3. Discussion

Mucopolysaccharidoses (lysosomal storage diseases) are a group of inherited metabolic disorders caused by a deficiency of a specific lysosomal enzyme required for a normal degradation of glycosaminoglycans. In Hurler syndrome, the most common MPSs, there is a mutation of the  $\alpha$ -L-iduronidase enzyme, which leads to excessive accumulation of partially degraded glycosaminoglycans within cells. As a consequence organ, skeletal and connective tissue involvement is diffuse and worsens with time [4]. Dwarfism, hypertelorism, progressive mental retardation, hepatosplenomegaly, kyphosis, cardiac valvular disease, cardiomyopathy, and coronary artery disease characterize Hurler syndrome [4,5]. Expected life span is 5 to 10 years, with death most commonly due to respiratory or cardiac failure [6-8] This is changing as bone marrow transplant and enzyme replacement therapy become more widely available [1,2].

Patients with MPS I show widespread progressive involvement of many organs and tissues. Because of this, frequent surgical intervention is common and is associated with high complication rates. One report estimated overall perioperative mortality at about 20% [4].

The difficulty in taking care of these patients cannot be understated. Death from failed intubation [9], failed tracheostomy [4], and postoperative respiratory arrest [5] have all been reported.

For the anesthesiologist, the establishment and maintenance of an adequate airway represent the most commonly encountered problem in MPS I [5,9,10]. Several anatomical abnormalities predispose these patients to upper airway obstruction, sleep apnea, and difficult intubation; these include a short neck with limited mobility, relatively high epiglottis, a deep cranial fossa that narrows the nasopharynx, hypoplastic mandible, ankylosis of the temporomandibular joints, infiltration of the pharyngeal tissues and tracheal cartilages, intraluminal narrowing of the conductive airways, and excessive secretions. Also, macroglossia, friable oral mucosa, and tonsillar hypertrophy can add to the difficulty [4-6] In this particular patient, the unstable atlantoaxial joint also had to be considered when managing the airway.

The anatomical abnormalities of this disease mandate a careful airway plan with ample backup support. Even with advanced planning and skillful execution, airway management can prove difficult. Most authors agree that pretreatment with an anticholinergic is necessary [6]. Despite pretreatment with glycopyrrolate, our patient still presented with copious secretions. We found a seated position to be helpful in managing secretions. Because of reported deaths after sedation [6], we avoided narcotic and benzodiazapine premedications. We used scopolamine and droperidol hoping this would provide some sedation without respiratory depression. This combination did not seem very effective at sedating our patient, but neither did it cause respiratory depression.

The literature reports different approaches to airway management, but most would agree on maintaining spontaneous ventilation until the airway is secure [6]. Because our patient was able to cooperate, we chose an awake fiberoptic technique for securing the airway. The oral route is usually easier than nasal because of anatomical abnormalities and dried secretions [4]. Smaller-than-anticipated tracheal tubes are often needed. The fiberoptic scope and the laryngeal mask airway have both been used successfully with these patients [11,12].

Other perioperative problems in patients with MPS I include positioning due to progressive joint immobilization and ventilation secondary to restrictive lung disease and frequent respiratory tract infections. Cardiac involvement is

typical and responsible for intraoperative deaths [13], so careful evaluation and monitoring are mandatory. Arterial catheter placement was also difficult because of wrist flexion. A dorsalis pedis arterial line is a useful alternative in these cases.

The decision to extubate these patients can be as perilous as intubation. Fortunately, our patient received a tracheostomy intraoperatively, which made further airway management much safer.

In summary, we report the successful anesthetic management of a 21-year-old man with Hurler syndrome undergoing a cervical decompression and stabilization procedure. Anesthesiologists will see more of these patients as new therapies extend life expectancy.

## References

- [1] Kakkis ED, Muenzer J. Enzyme-replacement therapy in mucopolysaccharidosis. *N Engl J Med* 2001;344(3):182-8.
- [2] Guffon N, Souillet G, Maire I, Straczek J, Guiband P. Follow-up of nine patients with Hurler's syndrome after bone marrow transplantation. *J Pediatr* 1998;133:119-25.
- [3] Kachur E, Del Maestro R. Mucopolysaccharidoses and spinal cord compression: case report and review of the literature with implications of bone marrow transplantation. *Neurosurgery* 2000;47(1):223-9.
- [4] King DH, Jones RM, Barnett MB. Anaesthetic considerations in the mucopolysaccharidoses. *Anaesthesia* 1984;39:126-31.
- [5] Baines D, Keneally J. Anaesthetic implications of the mucopolysaccharidoses: a fifteen-year experience in a children's hospital. *Anaesth Intensive Care* 1983;11:198-202.
- [6] Sjogren P, Pedersen T, Steinmetz H. Mucopolysaccharidoses and anesthetic risk. *Acta Anaesthesiol Scand* 1987;31:214-8.
- [7] Walker RWM, Darowski P, Morris P, Wraith JE. Anaesthesia and mucopolysaccharidoses. A review of airway problems in children. *Anaesthesia* 1994;49:1078-84.
- [8] Semenza GL, Pyeritz RE. Respiratory complications of mucopolysaccharide storage disorders. *Medicine* 1988;67(4):209-19.
- [9] Moores C, Rogers JG, McKenzie IM, Brown TCK. Anaesthesia for children with mucopolysaccharidoses. *Anaesth Intensive Care* 1996; 24:459-63.
- [10] Kempthorne PM, Brown TCK. Anaesthesia and the mucopolysaccharidoses: a survey of techniques and problems. *Anaesth Intensive Care* 1983;11:203-7.
- [11] Khan AK, Khan HK. Use of laryngeal mask airway in mucopolysaccharidoses. *Pediatr Anaesth* 2002;12(5):468.
- [12] Wilder RT, Belani KG. Fiberoptic intubation complicated by pulmonary edema in a 12-year-old child with Hurler's syndrome. *Anesthesiology* 1990;72(1):205-7.
- [13] Belani KG, Krivit W, Carpenter B, Braulin E, et al. Children with mucopolysaccharidoses: perioperative care, morbidity, mortality, and new findings. *J Pediatr Surg* 1993;28(3):403-10.