Morquio Syndrome: A Challenging Case
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**BACKGROUND**
- Morquio syndrome (MS) or MPS-IV
- Mucopolysaccharide storage disease
- Autosomal recessive inheritance pattern
- Most info from single or small case series
- Organ systems typically involved:
  - Neurological
  - Airway/respiratory
  - Skeletal
  - Cardiac
- Most will undergo multiple surgical procedures
- Lack of familiarity with the disease and available literature adds to the difficulty of managing these patients perioperatively

**REFERENCES**
1 Anesthetic care and perioperative complications of children with Morquio syndrome. Theroux MC
2 A retrospective audit of anesthetic techniques and complications in children with mucopolysaccharidosis. Frawley G.
3 Obstructive airway in Morquio A syndrome, the past, the present and the future. Tomatsu S.
4 Anesthetic care and perioperative complications of children with Morquio syndrome. Theroux.
5 Special airway concerns in patients with mucopolysaccharidosis. Steven Sims H.

**CASE REPORT**
A wheelchair-bound 19 year old male with Morquio Syndrome presented for an aortic valve replacement due to severe aortic insufficiency. He had undergone multiple surgical procedures in the past, including occiputo-cervico-thoracic spinal fusion. A review of prior anesthetics revealed a failed Glidescope intubation by two attending anesthesiologists with a poorly seated LMA placed as a rescue device during the most recent anesthetic. During a pre-surgical testing visit, after discussing at length with the patient the concerns for airway management, the patient adamantly refused to have an awake fiberoptic intubation after a traumatic attempt in the past. On the day of surgery, the patient was induced and spontaneous ventilation maintained. Once successful hand ventilation was proven, the patient was paralyzed to optimize intubating conditions. Initial attempts at Glidescope intubation were unsuccessful due to an inability to manipulate the pre-fashioned stylet in the patient's aberrant airway. While awaiting a fiberoptic bronchoscope for a Glidescope-assisted fiberoptic intubation, a second attending successfully intubated the patient. With incremental withdrawal of the blade, a 6.0 cuffed tube was advanced and secured in place. In addition to difficult airway management, his anatomic abnormalities, including a large head-to-body ratio, cervical fusion and pectus carinatum presented further challenges with central line placement, as well as surgical retraction. At the end of the case, patient was allowed to breath spontaneously. He was then extubated and transferred to the pediatric ICU. His postoperative course was uncomplicated.

**CONCLUSIONS**
- Morquio syndrome (MPS-IV)
- Autosomal recessive
- Deficiency of a lysosomal enzyme
- N-acetylgalactosamine 6-sulfate sulfatase
- Accumulation of byproducts in:
  - Bone and cartilage
  - Spine and joint dysplasia
- Other features include:
  - Normal IQ
  - Short stature
  - Large head-to-body ratio
  - Pectus carinatum
  - Cervical instability
  - Spinal cord compression
  - Heart valve problems.
- Majority will require multiple surgeries
- Airway problems are not uncommon
- Combination of factors
  - MPS deposits in the airway
  - Tracheal compression by brachiocephalic artery
  - Cervical instability or immobility after fusion
- Case reports have shown successful airway management videolaryngoscopy and fiberoptic intubation.
- Studies have suggested neuromonitoring during intubation if cervical instability is suspected.
- Preoperative assessment of airway, lung functions and cardiac functions are cornerstones for successful management of the Morquio syndrome patient.