Anesthetic Management of a 5-year old boy with untreated Hunter Syndrome (Mucopolysaccharidoses II)
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INTRODUCTION
Mucopolysaccharidoses (MPS) are a group of chronic progressive lysosomal storage diseases caused by deficiencies of enzymes required for the catabolism of glycosaminoglycans (GAGs) [1].

<table>
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<th>Type</th>
<th>GAG Storage</th>
<th>Enzyme Deficiency</th>
<th>Key Features</th>
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</table>
| MPS I | Hurler | α-L-iduronidase | Severe MPS I before age 2. Enzyme replacement therapy (ERT) before age 2.
| MPS II | Hunter | α-L-iduronidase | Enzyme replacement therapy (ERT) since 1980s reserved for type II enrolled in the Hunter Outcome Survey. |
| MPS III | Sanfilippo | α-L-iduronidase | Sanfilippo disease (type A, B, and C). |
| MPS IV | Morquio | α-D Glucuronidase | MPS IV (Morquio A and B) and MPS VI (Morquio C) are also known as Maroteaux-Lamy disease. |

INTRODUCTION


Anesthetic Concerns

- 20 years higher incidence of difficult intubation
- Thickening of soft tissues
- Enlarged tongue, TBA
- Short immovable neck
- Limited C spine and TMJ mobility, C spine instability
- Restricted lung dz + OSA 26.7% up to 80%
- Cardiac Disease in 60-100% (esp. MPS II, VI) [5]
- Behavioral problems, aggression

DISCUSSION

Anesthetic Assessment

- History: Snoring or obstruction? C-spine instability? Murmurs?
- Careful airway exam, cardiac exam
- Despite high incidence of cardiac involvement, not well studied and no guidelines. Recommendation in cardiology literatures:
  - echo and 12 lead ECG diagnosis
  - regular monitoring Q1-2 yrs for MPS I, VI 1-3 yrs for MPS II
- Prepare airway equipment
- Consider available person/setting up for surgical airway

Airway Considerations

- Incidence of difficult intubation is 25%. LMAs preferred in 52 cases.
- No cases of problems using LMA in 52 cases
- Difficult intubation in 25%
- Difficult mask ventilation in 7%
- No cases of problems using LMA in 25 cases
- Failed intubation in 2 cases (1.6%)

- Difficult mask ventilation in 14.2%
- Difficult intubation in 25%
- 1 case of dislodged LMA of 60 cases
- Failed intubation in 2 cases (10%)

- Interestingly, in the Frawley study, the highest incidence of both difficult mask ventilation (26.7%) and difficult intubation (35%) occurred in MPS II, as our patient had.

- 2) LMAs can be used as a primary airway device or used as a conduit for fiberoptic intubation
- Equivocal association between age and incidence of difficult airway.

4) ERT has not been shown to reduce the incidence of difficult airway in recent studies

- HSCT before age 2 reduces the incidence of difficult mask ventilation and difficult intubation

- Failure of studies to show any airway issues post ERT may be due to fairly recent available and thus late age at treatment initiation.

Perioperative Management

1) Airway Obstruction is common in the OR as well as in PACU so vigilance is key

- More airway problems in 12% of cases in the Frawley study
- Obstruction in about 10% of patients in the Megens study
- 2) Admit vs same day discharge? No consensus guidelines.

- 43% of patients in the Frawley series discharged same day on ICU admission required for small number of patients, especially in those with extensive airway manipulation, intraoperative cardiopulmonary event, more likely in MPS patients with pre-existing cardiac disease.

CONCLUSION

Given the 25% incidence of difficult intubation in children with MPS reported in the literature, adequate preparation and airway equipment is essential. Recent retrospective studies have shown LMAs to be particularly effective in the MPS population. Our case demonstrates the successful use of LMA as a conduit for fiberoptic intubation in a patient with HS.

LITERATURE CITED