

Evolving challenges of airway management in Morquio

A syndrome: a case report

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In Brief

Patient: 17 YO with Morquio A Syndrome (MPS IV)

Procedure: Cricotracheal resection (CTR)

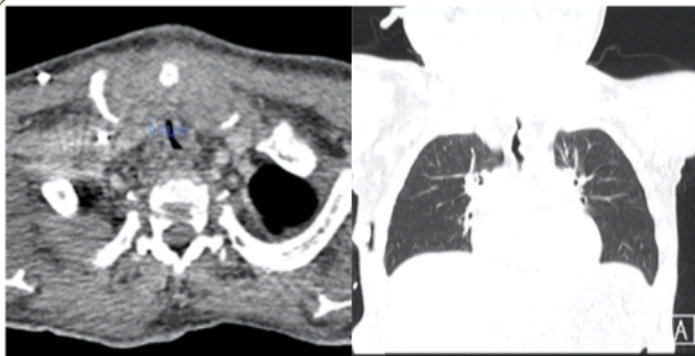
Indication: Severe tracheal tortuosity and obstruction worsening with age

Significance: First reported CTR in this population without use of CPB

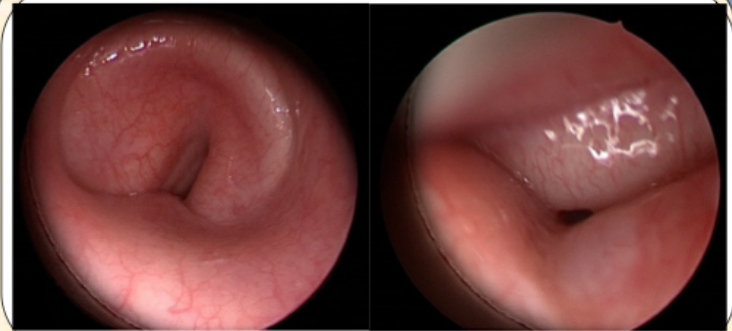
Learning Points:

1. Intubation success rates are improving in MPS with advances in treatment however overall airway complications remain high
2. Children with Morquio die from progressive tracheal obstruction. Death following failed extubation has been described
3. Maintenance of spontaneous ventilation is critical. Use of flexible bronchoscopy should be a mainstay of airway management. Delayed extubation to NIV should only occur after spontaneous ventilation has been well established

Case Description



- Spontaneous ventilation was maintained using TIVA for microlaryngobronchoscopy (MLB)
- MLB confirmed multi-level tracheal obstruction
- Bag mask ventilation was challenging during a brief apnoea requiring 2 hand technique and oropharyngeal airway
- Laryngoscopy was C+L I
- A flexible bronchoscope was utilised to position the tip of the ETT distal to the tracheal folds and tortuosities
- Mechanical ventilation commenced post ETT placement



- The most significant deformity was resected (5 tracheal rings)
- The ETT tip was surgically positioned under direct vision at 22cm, distal to the remaining obstruction
- During closure the ETT slipped to 20cm and ventilation became impossible: ETCO₂ trace was lost
- Flexible bronchoscopy revealed complete obstruction distal to ETT tip but failed to traverse it as did a rigid bronchoscope
- Preparations were made for repeat tracheostomy
- Anaesthesia was lightened and with the return of spontaneous effort ventilation improved and tracheal obstruction visibly reduced
- A final flexible bronchoscopy attempt successfully navigated the tortuosity

Discussion

- Morquio patients die from severe tracheal tortuosity and obstruction which worsens with age¹
- This is the first report of CTR without use of CPB
- Enzyme replacement therapy has improved intubation success rates but airway complications remain high²
- Death secondary to failed extubation has been described³
- The flexible bronchoscope is advocated for intubation of MPS pts⁴. We used it to position ETT tip distal to tracheal tortuosities at initial intubation and as airway rescue. Its subtle manoeuvrability prevailed where a rigid scope failed
- Spontaneous ventilation was equally essential for airway rescue
- Dynamic intrathoracic airway lesions produce most obstruction to flow with high intrathoracic pressure
- Paralysis of the diaphragm reduces the transpleural pressure gradient which distends airways distal to an obstruction
- The child's PedsQL score has significantly improved
- He now runs 1km from home to his village with his running frame

Acknowledgements

We would like to thank the patient and his family for consent to present this case.

References

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