Mucopolysaccharidoses – airway management step by step

A. S. Fernandes¹, J. Borges², M. Martins¹

¹Anesthesiology Service, Centro Hospitalar Lisboa Norte E.P.E.

²Anesthesiology Service, Hospital do Divino Espírito Santo de Ponta Delgada E.P.E.

Euroanaesthesia 2018
THE EUROPEAN ANAESTHESIOLOGY CONGRESS
COPENHAGEN 02-04 JUNE

Background

Inherited genetic diseases with progressive course, glycosaminoglycans accumulate

Multisystemic, variable presentation with severe morphological changes

Complex anesthetic management, particularly the airway

Clinical cases

- C., ♂, 23 years old, 40 kg, 1.16 m, ASA III
- PH: MPS type VI, previous difficult intubation
- Listed for corneal transplant
- Proposed for fiberoptic bronchoscopy airway mapping

- S., ♀, 28 years old, 37 kg, 1.34 m, ASA III
- PH: MPS type VII, previous difficult intubation
- Listed for aortic prosthesis revision
- Proposed for fiberoptic bronchoscopy airway mapping

Preparation

- Standard ASA monitorization + BIS
- Premedication with midazolam bolus
- Seated positioning
- Topical sprayed lidocaine 10% - naso- and oropharynx
- Lidocaine 2% nebulization
- Pre-oxygenation with 100% O₂ via endoscopy mask

Procedure

- Remifentanil TCI target 1,5-2,5 ng/mL
- Fiberscope nasally introduced
- Lidocaine – *spray-as-you-go*

Patients remained awake, calm and cooperative with maintained spontaneous ventilation.
No airway obstruction or desaturation episodes occurred. They remained hemodynamically stable.

Discussion and conclusion

It has been reported that **25%-50%** of MPS patients have problematic airways and **82%** of MPS patients receiving anesthesia require urgent airway interventions.

We present the anesthetic management of patients with MPS with a high risk of difficult intubation and ventilation in whom awake fiberoptic airway mapping was performed under light sedation with midazolam and remifentanil, with associated appropriate topical anesthesia.

We encountered no major problems and emphasize that careful planning and experienced support for difficult airway management are important when anesthetizing such patients.