*Revista da Sociedade Portuguesa de Anestesiologia*
http://revistas.rcaap.pt/anestesiologia
https://dx.doi.org/10.25751/rspa.21883

CASO CLÍNICO

# The role of Anaesthesiologists at Different Levels of Difficult Airway Management Care: A Case Report

## *O Papel do Anestesiologista em Diferentes Níveis da Abordagem da Via Aérea Difícil: Um Caso Clínico*

Marta Pacheco[1*], Rita Fernandes[2], José Aguiar[3], Jorge Órfão[1]

**Afiliação**
[1] Departamento de Anestesiologia, Cuidados Intensivos e Emergência, Centro Hospitalar e Universitário do Porto, Porto, Portugal.
[2] Serviço de Anestesiologia, Centro Hospitalar São João, Porto, Portugal.
[3] Hospital Lusíadas Porto, Porto, Portugal.

**Keywords**
Airway Obstruction; Anesthesia; Anesthesiologists; Mucopolissacaridoses; Tracheostomy
***Palavras-chave***
*Anestesia; Anestesiologistas; Mucopolissacaridoses; Obstrução das Vias Respiratórias; Traqueostomia*

## ABSTRACT

Mucopolysaccharidoses are a group of lysosomal storage diseases. The deficiency of specific lysosomal enzymes leads to accumulation of glycosaminoglycans (GAGs) in the tissues and organs. Patients had difficult airway (DA) criteria: long epiglottis, anterior larynx, small trachea, short neck, temporo-mandibular joint rigidity, cervical instability and complications with difficult ventilation and oxygenation. We describe a case that illustrates the role of the anaesthesiologist in the approach and maintenance of a DA during the disease progression. DA criteria made the maintenance of spontaneous ventilation a primary goal and fiberoptic should be the first approach. A precarious definitive airway and previous respiratory arrest linked to infraglottic obstruction, made manipulation of the tracheostomy cannula stressing, dangerous and not definitive. Trained anaesthesiologists in DA, fiberoptic and FROVA allowed safe diagnosis and exchange the cannula safely.

## RESUMO

As mucopolissacaridoses pertencem ao grupo das doenças lisossomais de sobrecarga, nas quais a deficiência de enzimas lisossomais leva à acumulação de glicosaminoglicanos nos tecidos e órgãos. Os doentes apresentam habitualmente critérios de via aérea difícil: epiglote longa, laringe anterior, traqueia e pescoço curtos, rigidez da articulação temporo-mandibular, instabilidade cervical e ventilação e oxigenação difíceis. Descrevemos um caso que ilustra o papel do anestesiologista na abordagem e manutenção da via aérea difícil durante a progressão da doença. Neste caso, os critérios de via aérea difícil tornaram a manutenção da ventilação espontânea um objetivo primordial, sendo a abordagem por fibroscopia a técnica de eleição. A via aérea definitiva precária e a paragem cardiorrespiratória prévia associada à obstrução infraglótica tornaram a manipulação da cânula de traqueostomia um ponto crítico e perigoso. Anestesiologistas treinados em via aérea difícil e fibroscopia foram o elemento-chave para o diagnóstico e a manipulação segura da cânula de traqueostomia.

Autor Correspondente/Corresponding Author*:
Marta Pacheco
Morada: Largo Prof. Abel Salazar, 4099-001 Porto, Portugal.
E-mail: martapacheco29@gmail.com

## INTRODUCTION

Mucopolysaccharidoses (MPS) are a heterogeneous group of lysosomal storage diseases which the lack or deficiency of specific lysosomal enzymes leads to an accumulation of GAGs in the tissues[1-3] with progressively organ dysfunction.[2] The classification includes seven major types (Table 1).[1] Musculoskeletal, central nervous, heart and respiratory systems are the most affected (Table 2).[1-3]

Maroteaux-LamySyndrome, is characterized by enzyme N-acetylgalactosamine-4-sulfatase deficiency, which leads to dermatan sulfate accumulation. It is autosomal recessive inherited, with an incidence of 1: 30 000.[3]

Patients typically exhibit: difficult ventilation criteria with facial mask: macroglossia, obstructive sleep apnoea (OAS), tracheomalacia; and difficult intubation criteria: long epiglottis, anterior larynx, reduced caliber of the trachea, short neck, temporo-mandibular joint rigidity and cervical instability that makes cervical hyperextension impossible.

The perioperative morbidity and mortality of these patients is high and the main causes of death in this period are related to the difficulty of maintaining the airway permeable, even with tracheostomy.[1] Post-intubation problems included respiratory failure, abundant secretions, stridor, bronchospasm, lower

**Table 1. Classification of mucopolysaccharidoses[1]**

| Type | Syndrome denomination | Enzyme deficiency | Accumulated substract (GAGS) |
|---|---|---|---|
| **MPS I** | Hurler<br>Hurler-Scheie<br>Sheie | α-L-Iduronidase | Dermata sulfate/Heparin sulphate |
| **MPS II** | Hunter | Iduronate-2-sulfatase | Dermatan sulfate/Heparin sulphate |
| **MPS III** | Sanfilippo A<br>Sanfilippo B<br>Sanfilippo C<br>Sanfilippo D | Heparan-N-sulfatase<br>α-N-acetyl-glucosaminidase<br>AcetylCoA:a-glucosaminide acetyltransferase<br>N-acetilglucosamine-6-solfatase | Heparin sulphate |
| **MPS IV** | Morquio tipo A | N-acetilglucosamine-6-solfatase | Keratin sulphate |
| | Morquio tipo B | B-galactosidase | |
| **MPS VI** | Maroteaux-Lamy | N-acetylgalactosamine-4-sulfatase | Dermatan sulfate |
| **MPS VII** | Sly | β-glucuronidase | Chondroitin-6-sulphate / Dermatan sulfate/Heparin sulphate |

**Table 2. Main associated clinical conditions[1,2]**

| Neurological | Orthopedic | Cardiac | Respiratory | Airway | Ocular |
|---|---|---|---|---|---|
| - Intellectual disability<br>- Behaviour disorders<br>- Communicating hydrocephalous<br>- Seizures<br>- Sensor neural deafness<br>- Spinal cord compression<br>- Peripheral neuropathy | - Skeletal deformities: kyphoscoliosis, valgus and varus limbs<br>- Joint stiffness<br>- atlanto-axial and odontoid instability | - Systemic arterial blood hypertension<br>- Heart failure<br>- Valvulopathy<br>- Ischemic cardiomyopathy<br>- Cardiac conduction abnormalities | - Restrictive lung disease<br>- Recurrent pneumonia | - Obstructive sleep apnea<br>- Tracheomalacia<br>- Macroglossia<br>- Rhinitis, tonsillitis, laryngitis and otitis media | - Glaucoma<br>- Corneal opacity<br>- Retina degeneration |

airway collapse and infection.[2] Other complications are laryngeal or subglottic oedema with incapability to maintain patency after extubation,[1] which could make extubation difficult or even impossible.

We report perioperative care plan of a patient with case of MPS type VI (Maroteaux-Lamy syndrome) and demonstrate the role of the anaesthesiologist in the approach, maintenance of the airway management during hospitalar stay and outline a plan for discharge.

## CASE REPORT

Female patient, 19 years old, medical history of Maroteaux-Lamy Syndrome, with multisystem involvement, under enzymatic substitution treatment with weekly Galsulfase.

Pre-operative evaluation:

- Cardiac: Electrocardiogram - Sinus rhythm, 80 bpm. Echocardiogram - with mild mitral and aortic stenosis, normal bi-ventricular function;
- Respiratory: Respiratory insufficiency with OAS – under nocturne non-invasive ventilation (NIV). Lung function tests (LFTs) with moderate restrictive lung disease;
- Musculoskeletal and neurological: Neck computed tomography (CT) showed atlanto-axial instability, pronounced intracanal ligament thickening causing stenosis of the spinal canal, without spinal cord compression. Severe attainment of skeletal muscle with articular limitation;
- The airway physical exam showed inadequate mouth opening, macroglossia, Mallampati class 4, short neck, inadequate range of neck and inaccessible cricothyroid membrane. In view of these findings, the patient had a difficulty airway. No previous surgical or anaesthetic history.

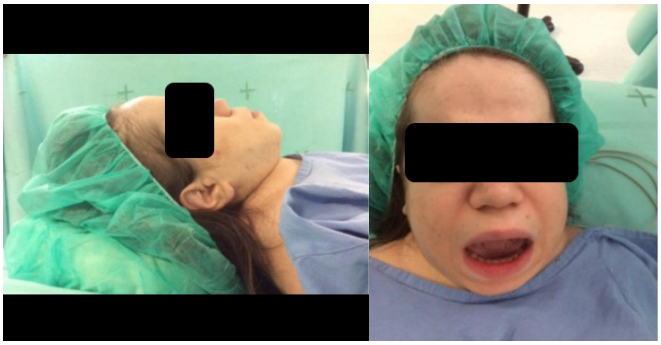

**Figure 1. Patient difficulty airway criteria**

Analytics values within normal ranges.

Functional capacity < 4 METs. ASA Physical Status Classification System: IV.

Patient was proposal for microlaryngoscopy in suspension to exeresis of a vallecular cyst. Airway approach was done by intubation through nasal fiberscopy after unsuccessful videolaryngoscopy, both attempts in spontaneous ventilation

(SV) with remifentanil perfusion and topical application of local anaesthetic to the airway. Due to significant oedema at the end of the intervention, was decided to maintain intubation and surveillance in the intensive care unit (ICU) in the immediate postoperative.

After 6 days in the ICU, due to persistent oedema of the airway, extubation was not safe and a tracheostomy was performed without complications. She proceeded to the ventilatory weaning and NIV was resumed during night. The patient developed an episode of airway haemorrhage and further investigation showed granulomas below the tracheostomy cannula, with decrease of the tracheal diameter (~25% usable area). A rigid bronchofiberscopy was then performed by Pneumology, with application of topic mitomycin in the granulomas. During an attempt to remove the tracheostomy cannula for inspection of the tracheostomy, airway collapsed with complete obstruction of the tracheal lumen. Rapid progression to respiratory arrest was observed, with recovery after emergent replacement of tracheostomy cannula to ensure airway.

Cervical computed tomography showed thickening of the pharyngolaryngeal mucosa, with collapse of the airway in the plane of the base of the tongue and uvula, apparently due to oedema.

Given the difficulties experienced in the previous intervention, the patient was proposed for tracheal cannula exchange in the operating room (OR). At that time, patient was conscious, cooperative and oriented, eupnoeic in spontaneous ventilation for de tracheostomy cannula without supplemental $O_2$, under nocturnal NIV, and hemodynamically stable.

Before the start of the procedure, it was outlined alternative plans for the case of obstruction and hypoventilation / apnoea, as well as the preparation of emergency material, namely for upper airway ventilation and tracheostomy stoma ventilation:

- swabs for cover the stoma, facemask, oral and nasal airway adjuncts, supraglottic airway device to ventilate through the upper airway;
- videolaryngoscopes with appropriate tubes;
- paediatric facemask to ventilate through the stoma;
- smaller tracheostomy tube and 6.0 cuffed tracheal tube for stoma intubation;
- bougie/Frova;
- arrest trolley and inflating bag.

In OR, under standard ASA monitoring, on SV with supplemental $O_2$, an infusion of with remifentanil 20 ug/mL was started in a dose of 0.1 ug/kg/min. Nasal fiberscopy was performed by an Anaesthesiologist trained in DA. Oedema/ granulomas and pharyngeal stenosis were observed, especially at the nasopharynx, extending at the level of the posterior wall, making it impossible to pass the fiberscope through the vocal cords. Then, fiberscopy was performed through the tracheostomy cannula.


**Figure 2. Fiberscopy images showing tracheal cannula and horizontal procidency (arrow)**

Below the cannula, normal mucosa folds with horizontal procidency to the tracheal were found occluding partially its lumen (Fig. 2). A FROVA stylet was placed through the tracheal cannula with the objective of oxygenation simultaneously with a guiding function for the exchange of the tracheal cannula.

Patient tolerated the entire procedure, without intercurrences. At the end, patient was awakened in SV and was transported to the ICU.

Posteriorly, the procidency found was diagnosed for computed tomography neck as an anatomic variant of right subclavian artery, with a high risk of bleeding to the trachea. The risk benefit analysis determines no surgical indication because of high bleeding risk.

Verified these facts, it was chosen a longer silicone cannula that allowed to overcome the protuberant lesion and the following cannula exchange procedures were performed in an OR environment by an anaesthesiologist trained in DA. Approach was performed in a similar way to the first, without intercurrences.

These procedures allowed the discharge of the patient in security to the domicile.

The plan passes through the periodic exchange of the tracheostomy cannula programmed in the OR.

## DISCUSSION

Given the obvious DA criteria in this patient, the maintenance of spontaneous ventilation was the primary goal, and fiberoptics for intubation should be the first approach. Despite apparently having a definitive airway, the previous history of respiratory arrest and the tracheomalacia observed in the intraoperative period, precludes the manipulation of the tracheal cannula, making this airway insecure, dangerous and not definitive. It is advisable that tracheal cannula exchanges should be done by the same team and we decided to do the manipulation on OR with trained anaesthesiologist in DA.

As discussed in the previous literature,[4] the algorithm for manipulating tracheostomy in situations of obstruction or displacement implies the use, among others, of devices such as fibroscope and introductors. FROVA Stylet use allowed

the guidewire function and the possibility of oxygenation if necessary.

This case shows the role that of anaesthesiologists may play at different levels of airway management care. Trained anaesthesiologists in DA, fiberoptics and FROVA Stylet allowed safe approach with good oxygenation, problems identification and exchange of the cannula.

**Ethical Disclosures**
**Conflicts of Interest:** The authors have no conflicts of interest to declare.
**Financing Support:** This work has not received any contribution, grant or scholarship.
**Confidentiality of Data:** The authors declare that they have followed the protocols of their work center on the publication of data from patients.
**Patient Consent:** Consent for publication was obtained.
**Provenance and Peer Review:** Not commissioned; externally peer reviewed.

**Responsabilidades Éticas**
**Conflitos de Interesse:** Os autores declaram a inexistência de conflitos de interesse na realização do presente trabalho.
**Fontes de Financiamento:** Não existiram fontes externas de financiamento para a realização deste artigo.
**Confidencialidade dos Dados:** Os autores declaram ter seguido os protocolos da sua instituição acerca da publicação dos dados de doentes.
**Consentimento:** Consentimento do doente para publicação obtido.
**Proveniência e Revisão por Pares:** Não comissionado; revisão externa por pares.

**ORCID**
Marta Pacheco https://orcid.org/0000-0001-9243-2545
Rita Fernandes https://orcid.org/0000-0003-2697-6987
José Aguiar https://orcid.org/0000-0003-2667-9755
Jorge Órfão https://orcid.org/0000-0003-2922-9725

Received: 22nd of December, 2020 | Submissão: 22 de dezembro, 2020
Accepted: 25th of May, 2021 | Aceitação: 25 de maio, 2021
Published: 20th of September, 2021 | Publicado: 20 de Setembro, 2021



## REFERENCES

1. Spinello C, Novello L, Pitino S, Raiti C, Murabito P, Stimoli F, et al. Anesthetic Management in Mucopolysaccharidoses. ISRN Anesthesiology. 2013;2013.
2. Walker R, Belani KG, Braunlin EA, Bruce IA, Hack H, Harmatz PR, et al. Anaesthesia and airway management in mucopolysaccharidosis. J Inherit Metab Dis. 2013;36:211-9.
3. Margarida M, Luísa M, Maria P. Abordagem Anestésica das Mucopolissacaridoses. Rev Soc Port Anestesiol. 2012; 21:
4. B. A. McGrath, L. Bates, D. Atkinson and J. A. Moore. Multidisciplinary guidelines for the management of tracheostomy and laryngectomy airway emergencies. Anaesthesia. 2012; 67:1025–41.