

ANESTHETIC AIRWAY MANAGEMENT IN AN INFANT WITH CONGENITAL MACROSTOMIA ASSOCIATED WITH AN OBLIQUE FACIAL CLEFT

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ABSTRACT

This case report describes the anesthetic airway management of a 9-month-old infant with congenital macrostomia associated with an oblique facial cleft undergoing elective surgical correction. Inhalational induction was performed with sevoflurane, followed by orotracheal intubation. Initial mask ventilation presented obstruction due to posterior tongue displacement, which was successfully managed using an oropharyngeal airway. Direct laryngoscopy revealed a Cormack-Lehane IIa view, facilitated by the increased mouth opening caused by macrostomia. Extubation was performed with the patient awake to protect the airway from potential blood aspiration; however, vigorous post-extubation crying led to a hematoma and suture dehiscence, requiring surgical re-exploration under intravenous sedation. This case highlights how craniofacial malformations can differently affect various steps of airway management and emphasizes the dilemma of extubation strategy in pediatric facial reconstructive procedures.

Keywords: Macrostomia, Facial clefts, Airway Management, Anesthesia, General, Infant.

INTRODUCTION

Atypical craniofacial clefts constitute a rare and heterogeneous group of congenital malformations, representing less than 1% of all facial clefts. Their estimated incidence ranges from 1.4 to 4.9 cases per 100,000 live births, with wide anatomical and clinical diversity and variable involvement of soft tissues, bone structures, and orbital and oral components.¹ These malformations result from complex alterations in the migration, proliferation, and fusion of neural crest cells during embryogenesis and are frequently associated with other craniofacial and systemic anomalies.¹

In 1976, Paul Tessier proposed an anatomical classification of facial, craniofacial, and laterofacial clefts based on their topographic relationship with the orbit. Also termed congenital macrostomia or transverse facial cleft, it accounts for approximately 5.5% to 13.9% of craniofacial

clefts and has an estimated incidence ranging from 1:80,000 to 1:300,000 live births. In this system, the different cleft trajectories are numbered according to their anatomical location, allowing standardized description of soft-tissue and craniofacial skeletal involvement.²

Congenital macrostomia is characterized by abnormal widening of the oral opening resulting from lateral extension of the oral commissure and is classically associated with Tessier No. 7 transverse facial cleft.^{3,4} Its anatomical presentation is variable and may involve the skin, mucosa, and perioral musculature, particularly the orbicularis oris muscle.^{3,4}

Anatomical alterations of the perioral musculature may result in facial asymmetry and functional impairment. Therefore, surgical reconstruction of macrostomia aims not only to correct the aesthetic appearance of the oral commissure but also to restore the continuity and functional orientation of the musculature, oral competence, and facial symmetry (3,4).

Oblique facial clefts constitute another rare group of craniofacial malformations and may present different anatomical trajectories, involving, to varying degrees, the oral, maxillary, nasal, and orbital regions.¹

The association between different patterns of facial clefts makes the clinical presentation even more uncommon and may increase the complexity of surgical reconstruction. From an anesthetic perspective, craniofacial deformities require individualized airway assessment, since anatomical alterations may interfere with face-mask fit, ventilation, and the performance of laryngoscopy and tracheal intubation.⁵

However, the impact of a malformation on the different stages of airway management is not necessarily uniform. The presence of a craniofacial abnormality should be analyzed according to the individual anatomical characteristics and its specific impact on ventilation, laryngoscopy, and intubation.⁵

The present study aims to report the perioperative management of an infant with macrostomia associated with an oblique facial cleft who underwent surgical correction, as well as to review the main aspects described in the literature regarding the classification, clinical presentation, surgical treatment, and anesthetic implications of these malformations.

CASE REPORT

Patient I.G.R., aged 9 months, weighing 9.3 kg and measuring 72 cm in height, with a diagnosis of macrostomia associated with an oblique facial cleft, was admitted for elective surgical treatment (Figure 1).

His medical history included hydrocephalus and cow's milk protein allergy. There was no history of previous surgical procedures, and he was receiving vitamin supplementation. Regarding previous anesthesia, he had previously undergone inhalational sedation for magnetic resonance imaging, with no reported complications. The patient had been fasting since 11:00 p.m. on the previous day.

In the operating room, monitoring included noninvasive blood pressure, cardiac monitoring, and pulse oximetry, with capnography added after definitive airway control.

Considering the patient's age and the initial absence of intravenous access, inhalational induction with 8% sevoflurane was chosen. After an adequate depth of anesthesia was achieved, peripheral intravenous access was obtained with a 24G catheter in the left upper limb. Fentanyl, at a dose of 17.5 µg, and atracurium, at a dose of 5 mg, were administered.

During initial airway management, manual ventilation was possible using a No. 2 pediatric

face mask. However, upper airway obstruction attributed to posterior displacement of the tongue was observed. Placement of a No. 2 Guedel oropharyngeal airway improved airway patency and ventilation.

Direct laryngoscopy was performed with a No. 2 Macintosh blade, obtaining a Cormack-Lehane IIa glottic view. Orotracheal intubation was performed uneventfully with a No. 4 cuffed tracheal tube. Despite the presence of a craniofacial malformation potentially relevant to airway management, the macrostomia provided an increased oral opening, which, in this patient, facilitated insertion and manipulation of the laryngoscope blade and contributed to direct laryngoscopy without significant technical difficulty.

After intubation, mechanical ventilation was initiated in pressure-controlled mode, with an inspiratory pressure of 20 cmH₂O, respiratory rate of 20 breaths per minute, positive end-expiratory pressure of 2 cmH₂O, and inspired oxygen fraction of 40%. The observed tidal volume was approximately 100 mL, and ventilation was adjusted during the procedure according to clinical and capnographic parameters.

Anesthesia was maintained with sevoflurane, with an end-tidal concentration of approximately 3%, associated with continuous remifentanyl infusion at a dose of 0.1 µg/kg/min.

Adjunct medications administered included cefazolin 400 mg, clonidine 15 µg, hydrocortisone 40 mg, dipyrone 500 mg, ondansetron 1.5 mg, and dexamethasone 1.2 mg. Eye protection was provided during the procedure.

The surgical team performed local infiltration with a solution containing 2% lidocaine associated with epinephrine at a concentration of 1:100,000, totaling 100 mg of lidocaine.

During the intraoperative period, mean arterial pressure remained predominantly between 40 and 55 mmHg, heart rate between 120 and 155 beats per minute, and peripheral oxygen saturation between 95% and 99%. End-tidal carbon dioxide remained between 35 and 45 mmHg.

The surgical procedure lasted approximately three hours and thirty minutes, while the total anesthetic time was approximately four hours.

At the end of surgery, neuromuscular blockade was reversed with 0.5 mg of neostigmine associated with 0.125 mg of atropine. Considering the nature of the procedure, performed in the facial region and close to the oral cavity, and the possibility of blood and secretions from the surgical field being present in the upper airways, extubation under deep anesthesia was not performed. The tracheal tube was removed after adequate recovery of consciousness and protective airway reflexes, prioritizing reduction of the risk of pulmonary aspiration of blood.

Immediately after extubation, the patient developed vigorous crying, followed by hematoma formation at the surgical site and suture dehiscence. Given the need for a new surgical approach for hematoma drainage and resuturing, intravenous sedation with propofol 20 mg and ketamine 4 mg was performed, allowing completion of the additional procedure.

After stabilization and definitive completion of the procedure, the patient was transferred to the post-anesthesia care unit, where he remained under observation for approximately one hour. The post-anesthetic course was satisfactory, with an Aldrete-Kroulik score of 10, after which he was transferred to the ward.



Table 1: Photographic records of the perioperative period.¹⁻³ Preoperative clinical features showing the craniofacial malformation;⁴ patient after anesthetic induction;⁵ direct laryngoscopy for airway management;⁶ patient after orotracheal intubation;⁷ intraoperative appearance during surgical correction;⁸ immediate postoperative appearance, showing the result at the end of the surgical procedure.

DISCUSSION

Atypical craniofacial clefts represent a rare and heterogeneous group of congenital malformations characterized by marked anatomical and clinical variability. Although the classification proposed by Tessier remains a reference for their description, the individual presentation of each patient may differ substantially with regard to the extent of soft-tissue involvement, craniofacial skeletal involvement, and functional repercussions, requiring individualized therapeutic planning.^{1,2}

Congenital macrostomia, classically corresponding to a lateral facial cleft (Tessier No. 7), results from failure of fusion between the maxillary and mandibular processes during embryonic development. Although it may occur in isolation, its association with other atypical craniofacial clefts constitutes an even more uncommon condition, increasing the complexity of surgical reconstruction and perioperative management.^{3,4} In the present case, the coexistence of macrostomia and an oblique facial cleft posed important anatomical challenges, particularly related to airway management and the extubation strategy.

The main objectives of surgical reconstruction of macrostomia are to restore an anatomically symmetrical oral commissure, reconstruct the functional continuity of the orbicularis oris muscle, and provide adequate reconstruction of the cutaneous, muscular, and mucosal planes. Recent studies emphasize that functional success depends more on the anatomical reconstruction of the perioral musculature than exclusively on the aesthetic outcome of the skin, with repair of the orbicularis oris muscle considered a fundamental step for recovery of oral competence and facial development.^{3,4}

From an anesthetic perspective, patients with craniofacial clefts are frequently considered to have a potentially difficult airway. However, current recommendations emphasize that difficulty should not be interpreted as a single concept, but rather analyzed separately at each stage of airway

management, including mask ventilation, laryngoscopy, tracheal intubation, and extubation.^{5,6} This approach becomes particularly relevant in patients with complex anatomical abnormalities, in whom different components of the deformity may exert distinct effects on each of these stages.

In the present case, manual mask ventilation was initially hindered by dynamic upper airway obstruction resulting from posterior displacement of the tongue. This pathophysiological behavior is consistent with the loss of muscle tone induced by anesthetic agents and demonstrates that, despite the craniofacial deformity, the main ventilatory limitation was related to upper airway obstruction rather than to the seal of the face mask itself. The rapid resolution after insertion of a Guedel oropharyngeal airway demonstrates that early recognition of the mechanism of obstruction allowed a simple and effective intervention, immediately restoring adequate ventilation.

Interestingly, the difficulty observed during mask ventilation was not repeated during direct laryngoscopy. On the contrary, glottic exposure corresponding to Cormack-Lehane grade IIa was observed, and tracheal intubation was performed without significant technical difficulty. During the procedure, it was noted that the increased oral opening provided by the macrostomia itself facilitated insertion and manipulation of the laryngoscope blade, thereby facilitating glottic exposure.

This finding deserves emphasis because it illustrates that the same anatomical abnormality may exert different effects on different components of airway management. Although patients with craniofacial clefts are traditionally considered candidates for a difficult airway, the literature does not support the assertion that macrostomia systematically facilitates direct laryngoscopy. Thus, the ease observed in this case is considered an individual anatomical characteristic resulting from the greater amplitude of the patient's oral opening and should not be extrapolated to all individuals with this malformation. However, this observation reinforces the need for individualized assessment of each stage of airway management, avoiding the assumption that difficulty with ventilation necessarily implies difficulty with laryngoscopy or tracheal intubation.

Another relevant aspect concerns the extubation strategy. In reconstructive procedures involving the oral cavity and face, the decision regarding the timing of tracheal tube removal often requires balancing potentially conflicting risks. Extubation under deep anesthesia may reduce reflex responses such as coughing, respiratory effort, and agitation, theoretically decreasing tension on recently reconstructed tissues. However, reduced protective airway reflexes may increase the risk of aspiration of blood and secretions from the surgical field, especially in surgeries involving the oral mucosa and perioral region.^{5,6}

In the present case, considering the possibility of residual blood in the oropharynx after completion of the reconstruction, extubation was performed only after adequate recovery of consciousness and protective airway reflexes, prioritizing respiratory safety and reducing the risk of pulmonary aspiration. This decision was made despite recognizing that a lighter anesthetic emergence could favor the occurrence of coughing, crying, and agitation, with potential repercussions on the integrity of the surgical reconstruction.

After extubation, the patient developed vigorous crying, followed by hematoma formation at the surgical site and partial suture dehiscence, requiring a new surgical intervention for drainage and resuturing. Although a direct causal relationship between crying and hematoma development cannot be established, it is physiologically plausible that transient increases in venous pressure resulting from intense crying, expiratory effort, and agitation may contribute to bleeding from recently manipulated tissues. Thus, this episode highlights the inherent complexity of planning

anesthetic emergence in facial reconstructive surgery, in which airway protection and preservation of the surgical reconstruction do not always converge toward the same extubation strategy.

The repeat surgical intervention was performed under intravenous sedation with propofol and ketamine, allowing hematoma drainage and resuturing without the need for repeat tracheal intubation. The combination of these drugs provided adequate conditions for the short-duration procedure, preserving clinical stability and avoiding further airway manipulation, an aspect that was particularly desirable in a patient who had recently undergone facial reconstruction.

From a scientific perspective, the main aspect of this report is believed to be not only the rarity of the association between macrostomia and oblique facial cleft, but mainly the demonstration that different components of anesthetic airway management may be influenced differently by the same anatomical abnormality. While mask ventilation was impaired by dynamic obstruction resulting from posterior displacement of the tongue, the greater oral opening provided by the macrostomia facilitated direct laryngoscopy. In addition, the case illustrates the need to individualize the extubation strategy in facial reconstructive surgery, carefully balancing the risk of pulmonary aspiration and the possibility of emergence-related complications, such as coughing, crying, increased venous pressure, and bleeding at the surgical site.

CONCLUSION

Anesthetic planning for patients with craniofacial clefts should be based on detailed anatomical assessment, preparation for different airway management scenarios, and ongoing integration between the anesthesiologist and the surgical team. Rather than simply considering these patients as having a difficult airway, it is essential to understand how each anatomical characteristic may individually affect ventilation, laryngoscopy, intubation, and extubation, allowing safer and more targeted planning for each case.

REFERENCES

1. Civantos AM, Shakya P, Shaye DA. Atypical facial clefts: Tessier number 3 and 4 clefts. *Curr Opin Otolaryngol Head Neck Surg*. 2024 Aug 1;32(4):248-256.
2. Tessier P. Anatomical classification facial, cranio-facial and latero-facial clefts. *J Maxillofac Surg*. 1976 Jun;4(2):69-92.
3. Kobraei EM, Lentz AK, Eberlin KR, Hachach-Haram N, Hamdan US. Macrostomia: A Practical Guide for Plastic and Reconstructive Surgeons. *J Craniofac Surg*. 2016 Jan;27(1):118-23.
4. Khorasani H, Boljanovic S, Amma M, Knudsen K, Jakobsen LP. Erratum to "surgical management of the Tessier 7 cleft: A review and presentation of 5 cases" [JPRAS open. Volume 22, December 2019, pages 9-18]. *JPRAS Open*. 2021 Feb 20;28:141. doi: 10.1016/j.jpra.2021.02.003. Erratum for: *JPRAS Open*. 2019 Jul 23;22:9-18.
5. Lima LC, Cumino DO, Vieira AM, Silva CHRD, Neville MFL, Marques FO, Quintão VC, Carlos RV, Fujita ACG, Barros HÍM, Garcia DB, Ferreira CBT, Barros GAM, Módolo NSP. Recommendations from the Brazilian Society of Anesthesiology (SBA) for difficult airway management in pediatric care. *Braz J Anesthesiol*. 2024 Jan-Feb;74(1):744478.
6. Van Slyke AC, Burge J, Bos R, Parker G, Chong DK. The Anatomical Subunit Approach to Managing Tessier Numbers 3 and 4 Craniofacial Clefts. *Plast Reconstr Surg Glob Open*. 2022 Sep 28;10(9):e4553.

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