Clinical Reports

Perioperative management of paediatric microstomia

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Paediatric microstomia may occur congenitally in the whistling face syndrome but is more often acquired after accidental thermal injuries such as biting an electrical extension cord or ingesting household lye. The surgical correction of microstomia includes separation and cosmetic reconstruction of the fused lips and postoperative oral splinting. Microstomia from lye ingestion may be associated not only with limited mouth opening but also with such severe intraoral scarring that common landmarks guiding either rigid or flexible fiberoptic laryngoscopy are obscured, rendering oral and nasotracheal intubation difficult or impossible. We report a case of paediatric microstomia after lye ingestion in which conventional direct laryngoscopy, flexible fiberoptic laryngoscopy, and multiple blind nasal approaches to tracheal intubation were unsuccessful. However, tracheostomy was avoided and successful tracheal intubation was accomplished using a new rigid tubular pharyngolaryngoscope.

La microstomie pédiatrique peut être congénitale comme dans le syndrome du « whistling face » mais elle est plus souvent acquise après des lésions thermiques accidentelles survenant après avoir mordu une extension électrique ou ingéré un caustique d'entretien ménager. La correction chirurgicale de la microstomie inclut la séparation et la reconstruction cosmétique des lèvres fusionnées et la fixation orale postopératoire. La microstomie survenant après ingestion de caustique peut être associée non seulement avec une limitation de l'ouverture buccale mais aussi avec de telles scarifications sévères intra-orales que les points de repère de la laryngoscopie fibroptique flexible ou de la laryngoscopie rigide sont obscurcis rendant l'intubation orale

et nasotrachéale difficiles ou impossibles. On rapporte le cas d'une microstomie pédiatrique après ingestion de caustique chez qui la laryngoscopie conventionnelle directe, la laryngoscopie par fibroscope flexible et des multiples approches par voie nasale aveugle pour l'intubation trachéale furent sans succès. Cependant, la trachéostomie fut évitée et l'intubation trachéale fut accomplie avec un nouveau pharyngolaryngoscope tubulaire rigide.

Paediatric microstomia may occur congenitally in rare instances but is more often acquired after thermal injury or chemical ingestion. Microstomia limits the oral intake of solid foods and often restricts oral intake to liquids and semisolids that can be aspirated by a straw. The surgical management of microstomia includes separation of fused lips, cosmetic and functional lip reconstruction, and postoperative oral splint therapy to prevent recurrence due to scar contractures.

Flexible fiberoptic laryngoscopy has been recommended for nasotracheal intubation in children with airways compromised by decreased mouth opening.¹ In severe cases of microstomia, as in any difficult airway, successful airway management may require multiple capabilities for difficult airway management in addition to flexible fiberoptic endoscopy during initial and subsequent reconstructive procedures. We report a case of acquired paediatric microstomia in which caustic oral burns made conventional, fiberoptic, and blind techniques of tracheal intubation impossible. However, tracheostomy was averted and tracheal intubation for anaesthesia was accomplished with a new tubular pharyngolaryngoscope with an intraluminal light source.

Case report

A 21-month-old, 15 kg female infant was referred to our institution for evaluation and management of acquired microstomia from accidental lye ingestion two months earlier. On admission, the child appeared well nourished but could only open her mouth 10 mm in the midline by forcefully pursing her lips and could eat and drink only

Key words

AIRWAY: obstruction;

ANAESTHESIA: paediatric.

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Accepted for publication 4th November, 1990.

FIGURE 1 Maximum mouth opening in a 21-month-old female infant who had accidentally ingested lye two months earlier.

with a straw (Figure 1). The lips were fused to the gingiva circumferentially and to each other laterally. Tongue protrusion was severely limited by sublingual burn scar contractures. The lower buccal sulcus was obliterated by gingival fusion. There was no obvious upper respiratory tract airflow obstruction when the child was awake or asleep.

A series of staged plastic procedures was planned to expand the oral opening and included: (1) bilateral oral commissurotomies with release of lip scar contractures; (2) labial mucosal advancement flaps to free the lips circumorally; and (3) mandibular vestibuloplasty with dermal skin grafting to enlarge the oral cavity and allow unrestricted jaw motion through the temporomandibular joints.

Since there was no upper airway obstruction, the patient was given premedication with meperidine, 15 mg, hydroxyzine, 15 mg, and atropine, 0.2 mg iv, and she entered the operating room resting comfortably and was easily arousable. An iv infusion was started in the left hand and preoxygenation with 100% oxygen (O₂) begun. Intraoperative monitoring commenced with ECG, precordial stethoscope auscultation, automated BP determination, pulse oximetry, and continuous measurement of inspired and end-tidal gases at the tracheal tube connector by mass spectrometry.

Ketamine, 10 mg, was administered for iv anaesthesia with spontaneous ventilation during endoscopy. The nostrils were sprayed with 0.5 ml 4% cocaine, and a 4.5 ml endotracheal tube was inserted through the right nostril into the nasopharynx. A bronchoscopic swivel adaptor was attached to the nasopharyngeal tube for O₂ insufflation throughout the fiberoptic endoscopy. As a 5.0 mm

outside diameter flexible fiberoptic laryngoscope* would not fit through the 4.5 mm endotracheal tube in the right nostril, the fiberoptic laryngoscope was advanced instead through the left nostril to provide direct visual guidance of the endotracheal tube from the nasopharynx into the trachea.

Due to intraoral scarring, common landmarks such as the tongue, epiglottis, aryepiglottic folds, and arytenoid cartilages could not be identified. In addition, the nasopharyngeal tube could not be advanced blindly into the trachea with multiple passes during spontaneous ventilation. Retrograde catheter-guided tracheal intubation was not attempted because of the infant's size and healed oesophageal burns. A conventional paediatric laryngoscope could not be inserted into the patient's mouth for direct suspension laryngoscopy without extensive surgery and the risk of bleeding into an unprotected upper airway (Figure 1). Bilateral commissurotomies to enlarge the mouth opening just enough to permit the introduction of a modified tubular laryngoscope† (Figure 2) were necessary for translaryngeal airway management.

Nasopharyngeal insufflation of 0.5–2.0% inspired halothane in oxygen was begun during spontaneous ventilation. At an adequate anaesthetic depth, the fused portions of the lips were infiltrated with 5 ml, 1% lidocaine with 1:200,000 epinephrine. Bilateral commissurotomies of the lips were performed to enlarge the oral opening sufficiently to receive the tubular pharyngolaryngoscope. The tubular laryngoscope slid easily and atraumatically through the lip opening, past the contracted tongue, and toward the epiglottis. With appropriate cervical extension, the tip of the tubular laryngoscope came to rest at the glottis, permitting clear visualization of moving true vocal cords. A sterile 4.5 mm oral endotracheal tube was inserted through the tubular laryngoscope and into the glottis under direct vision with bright, intraluminal fiberoptic lighting. The tubular laryngoscope was then split into its two halves and each half was removed separately, leaving the endotracheal tube in position to be secured. The nasopharyngeal tube was removed and the oral tracheal tube was secured to some deciduous teeth. Anaesthesia was maintained with halothane, 0.5–1.5%, and nitrous oxide 60% in oxygen.

Following the establishment of endotracheal anaesthesia, mouth reconstruction proceeded for the next six hours

*Olympus LF-1 Tracheal Intubation Fiberscope. Olympus Corporation, 4 Nevada Drive, Lake Success, N.Y. 11042-1179.

†Pediatric Pharyngo-Laryngoscope, Model #52-2220 Modified, Pilling Instruments, 420 Delaware Drive, P.O. Box 7014, Fort Washington, PA 19034.

FIGURE 2 (A) The two handle-blades and the knurled screw to hold the handle-blades of the paediatric laryngoscope together during orotracheal intubation. (B) Demonstration of the intraluminal insertion of an endotracheal tube through the assembled tubular laryngoscope during illumination provided by fibreoptic light sources in each handle-blade.

and included full lingual commissurotomies, labial mucosal advancement flaps to resurface the lips, and multiple scar releases and buccal mucosal rotation flaps to recreate the oral cavity and free the lips and cheeks from scar contractures. Maintenance and replacement fluids were provided throughout the surgical procedures. No blood transfusion was required.

At the end of the surgical procedure, the prosthodontist made an impression of the new oral cavity with a tracheal tube groove at its roof. Using the mouth impression, the prosthodontist created a mouthpiece to maintain maximum mouth opening, permit unobstructed oral air flow, and allow for an orotracheal airway when indicated.

The patient was allowed to awaken from the anaesthetic with the oral endotracheal tube held securely in place by the prosthodontic retention splint. The patient was then permitted to breathe spontaneously through the 4.5 mm oral endotracheal tube attached to a 40% inspired

FIGURE 3 Appearance eight days after surgery. Patient returned to the operating room for removal of the retention sutures shown and for replacement of the temporary oral splint shown. Note the upper midline groove in the oral splint for an endotracheal tube or a straw.

oxygen source and 5 cmH₂O continuous positive airway pressure. The trachea was extubated without complications on the second postoperative day.

On the eighth postoperative day the patient returned to the operating room for removal of the retention sutures which held the labial advancement flaps in place and for the application of an additional external oral commissural splint (Figure 3). These procedures were carried out under *iv* ketamine with 100% inspired nasal oxygen. The patient was discharged on a liquid diet the following day. Prosthodontic splint therapy continued for several weeks.

One year later, the patient returned for final plastic reconstruction by bimandibular vestibuloplasties with dermal grafts harvested from the buttocks. The anaesthetic and airway management were uncomplicated. At this time, conventional oral direct laryngoscopy was performed easily for orotracheal intubation. The patient tolerated the final cosmetic procedure well and was discharged on the third postoperative day tolerating a soft diet. The patient continues to grow and develop normally, with markedly improved mouth opening, increased range of jaw motion through the temporomandibular joints, and normal dental development.

Discussion

The thermal injury most commonly associated with acquired microstomia in children is an electrical burn to the commissures of the lips caused by biting or sucking on a live extension cord or electrical outlet.² As in the case presented, the chemical ingestion most commonly associated with acquired microstomia in children is caustic lye ingestion of household cleaning products containing

hydroxides of sodium, potassium, or ammonia.^{3,4} The rare congenital causes of microstomia in children include the whistling face syndrome⁵ and complete mandibular agenesis.⁶

In paediatric microstomia, airflow in the upper respiratory tract is not compromised. Nasopharyngeal airflow patterns will even permit unobstructed nasal ventilation by mask but often prohibit blind nasotracheal intubation due to anatomical distortion, as in the case reported. The most important feature of microstomia complicating airway management for surgical anaesthesia is limited mouth opening making oral access to the trachea difficult. In addition, oral scarring may obliterate most common landmarks leading to the glottis such as the tongue, epiglottis, and aryepiglottic folds.

The anaesthetic management of paediatric microstomia may be divided into two phases, (1) anaesthesia for initial tracheal airway establishment, and (2) anaesthesia for extensive intraoral reconstructive surgery. During initial airway establishment, children with microstomia should be allowed to breathe spontaneously as the sights (air bubbles at the glottis), sounds (airflow noises in the pharynx), and other indicators of upper airway airflow (expired CO₂) may be the best indicators of a passage to the trachea. If upper respiratory tract airflow is compromised, respiratory depressants, such as barbiturates and narcotics, should be avoided as premedicants.

As in the case reported, flexible fibreoptic and blind techniques may be unsuccessful due to anatomical distortion and burn scar contractures. Flexible fibreoptic endoscopy may also be limited by the age and size of the child and the calibre of the endotracheal tube selected.¹ Commonly available flexible fibreoptic laryngoscopes will not pass through endotracheal tubes with an internal diameter of ≤ 5 mm.²

Orotracheal intubation by direct laryngoscopy can be performed in children with microstomia only after the mouth opening is enlarged by bilateral commissurotomies. Commissurotomies may be performed under local anaesthesia, ketamine anaesthesia, or inhalational anaesthesia during spontaneous ventilation.

We chose ketamine for fibreoptic examination of the pharynx and attempted tracheal intubation to provide a combination of anaesthesia and spontaneous ventilation. We chose nasopharyngeal insufflation of inhaled anaesthetics for bilateral commissurotomies, tubular pharyngolaryngoscopy, and oro-tracheal intubation for a number of reasons.⁷ First, there was a nasopharyngeal tube already in place, inserted under flexible fibreoptic guidance. Second, nasal insufflation would permit complete and unhurried access to the mouth. Lastly, an inhalational anaesthetic would provide a deeper plane of anaesthesia

for oro-tracheal intubation during spontaneous ventilation. In the event of laryngospasm, positive pressure ventilation could be easily provided by face mask.

A tubular laryngoscope blade may improve glottic visualization after flexible endoscopic failures by creating viewing space out of oedematous mucosa or scar tissue and illuminating the laryngopharynx with a bright intraluminal light source that cannot be obstructed by unsuspected anatomical changes, bloody secretions, or oedematous mucosa.⁸ Standard nontubular laryngoscope blades with open sides and flexible fibreoptic laryngoscopes do not create viewing space within the laryngopharynx in the presence of circumferential oedema or scarring.⁸ Conventional and flexible fibreoptic light sources are also obstructed and obliterated by the enveloping effects of oedematous mucosa, scar tissues, blood, or thick oral secretions.⁸ Tubular laryngoscopes may offer considerable advantages in establishing an oro-tracheal airway in patients with congenital or acquired microstomia and may offer advantages in other airway obstructive lesions with circumoral mass effect, such as congenital cystic hygroma, giant oral haemangioma, or oral arteriovenous malformations, by offering gentle compression of surrounding tissues and unobstructed intraluminal fibreoptic lighting.

Awake tracheal extubation is recommended postoperatively for several reasons. First, cosmetic lip reconstruction contraindicates tight face mask application for positive pressure ventilation. Second, the oral splint may obstruct the upper airway in any patient whose upper airway protective reflexes are obtunded by anaesthetics or sedatives. Finally, reintubation of the trachea immediately after microstomia reconstruction could disrupt floating skin grafts and delicate suture lines and compromise the success of the entire oral reconstructive procedure.

In summary, endotracheal intubation by flexible fibreoptic laryngoscopy may be impossible in infants with severe microstomia from accidental lye ingestion. The use of a paediatric tubular pharyngolaryngoscope may permit oro-tracheal intubation and avert a tracheostomy for intraoperative airway management. Successful oro-tracheal intubation may also require limited separation of the lips by bilateral commissurotomies before tubular laryngoscopy.

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