Lighted stylet-guided intubation via the intubating laryngeal airway in a patient with Hallermann-Streiff syndrome

L'intubation guidée avec un stylet lumineux via un masque laryngé d'intubation chez un patient souffrant du syndrome d'Hallermann-Streiff-François

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Abstract

Purpose Hallermann-Streiff syndrome is a congenital syndrome associated with oculomandibulofacial abnormalities and potentially difficult airways. This case report describes the novel use of a lighted stylet-guided tracheal tube insertion through a new supraglottic airway, the intubating laryngeal airway (ILATM), in a patient with Hallermann-Streiff syndrome who had anticipated difficult airway.

Clinical features A 26-year-old male with Hallermann-Streiff syndrome was scheduled for a vitrectomy. The patient had mandibulofacial dystocia with a bird-like appearance, a mouth opening of 4 cm, a receding chin, and a Mallampati class 3 examination. The surgeon requested muscle paralysis and no movement during surgery. After receiving midazolam, fentanyl and propofol, a size 3.5 ILATM was inserted and lung ventilation was easy to perform. A 7.5-mm internal diameter tracheal tube was mounted on a lighted stylet with its inner rigid stylet removed. After succinylcholine administration, the lighted stylet-tracheal tube assembly was inserted via the ILATM until the transillumination just vanished below the sternal notch. The lighted stylet was removed, the circuit was connected, and capnography confirmed tracheal placement of tube. The ILATM was deflated and left in situ. Upon

emergence from anesthesia, the tracheal tube, and subsequently the ILATM, were removed without complications.

Conclusions This case presents a novel use of a lighted stylet-guided tracheal tube insertion through the ILATM in a patient with Hallermann-Streiff syndrome. This intubation technique can be considered in patients with difficult airways as a primary route of intubation, or as a secondary rescue strategy.

Résumé

Objectif Le syndrome d'Hallermann-Streiff-François est un syndrome congénital associée à des anomalies oculo-mandibulo-faciales et des voies aériennes potentiellement difficiles. Cette présentation de cas décrit l'utilisation novatrice d'une sonde trachéale guidée par stylet lumineux pour l'intubation via un nouveau dispositif supraglottique de gestion des voies aériennes, le masque laryngé d'intubation ILATM, chez un patient souffrant du syndrome d'Hallermann-Streiff-François chez qui on prévoyait des voies aériennes difficiles.

Éléments cliniques Un homme de 26 ans souffrant du syndrome d'Hallermann-Streiff-François a été admis pour subir une vitrectomie. Le patient souffrait de dystocie mandibulo-faciale et présentait une apparence d'oiseau, une ouverture buccale de 4 cm, un menton effacé, et un score de Mallampati de classe 3. Le chirurgien a demandé que les muscles soient paralysés et qu'il n'y ait aucun mouvement pendant la chirurgie. Après l'administration de midazolam, de fentanyl et de propofol, un ILATM de taille 3 a été inséré et la ventilation des poumons a été facile à réaliser. Une sonde trachéale de 7,5 mm de diamètre interne a été fixée à un stylet lumineux dont le stylet intérieur rigide avait été retiré. Après l'administration de

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succinylcholine, le montage sonde trachéale – stylet lumineux a été inséré via le ILA™ jusqu'à ce que la diaphanoscopie disparaisse juste sous l'échancrure sternale. Le stylet lumineux a été enlevé, le circuit connecté, et la capnographie a confirmé le positionnement trachéal de la sonde. L'ILA™ a été dégonflé et laissé in situ. Lors du réveil de l'anesthésie, la sonde trachéale, puis l'ILA™, ont été extraits sans complications.

Conclusion Ce cas présente une utilisation innovante de l'insertion d'une sonde trachéale guidée par stylet lumineux via un ILA™ chez un patient souffrant du syndrome d'Hallermann-Streiff-François. Cette technique d'intubation peut être envisagée pour les patients présentant des voies aériennes difficiles comme voie d'intubation principale, ou comme stratégie de sauvetage secondaire.

Hallermann-Streiff syndrome is a congenital syndrome characterized by multiple maxillofacial anomalies, including microstomia, mandibular hypoplasia, dental anomalies, hypertichosis, difficult airway, and ophthalmologic abnormalities.^{1–7} In consideration of the potential airway difficulties, the Intubating Laryngeal Airway by Cookgas®, LLC (ILA™, Mercury Medical, Clearwater, FL, USA) is a new supraglottic airway device with a functionality and insertion technique similar to that of an intubating laryngeal mask airway.^{8,9} The lighted stylet has been used as an adjunct to guide the passage of a tracheal tube through an intubating laryngeal mask airway.^{10,11} We describe the successful application of a lighted stylet-guided tracheal tube insertion through an ILA™ in a patient with Hallermann-Streiff syndrome and an anticipated difficult airway. Written consent for publication of the manuscript and the patient image was granted by the patient.

Case report

A 26-year-old male (32.7 kg, 165 cm) with Hallermann-Streiff syndrome was scheduled for a vitrectomy. He had undergone multiple surgical procedures under general anesthesia since childhood, and over the previous five years, he had undergone three eye surgeries performed under general anesthesia with laryngeal mask airways. The patient had a high school education and the mental capacity to provide consent for medical procedures.

He presented with mandibulofacial dystocia involving a bird-like appearance (Fig. 1). He had a 4 cm mouth opening, a receding chin, a normal temporomandibular joint, and a stable cervical spine. The Mallampati examination was class 3. The surgeon stated the critical nature of the retinal surgery and requested assurance of complete paralysis during the

Fig. 1 Lateral profile view of a patient with typical facial features seen in Hallermann-Streiff syndrome. Note the mandibulofacial dystocia with bird-like facies, parrot-beaked nose, small mouth opening, and receding chin

procedure. The planned procedure was to induce general anesthesia using short-acting anesthetic drugs and to insert an ILA™ followed by lighted stylet-guided tracheal intubation. If this method was unsuccessful, the back-up plan was fiberoptic bronchoscope-guided tracheal intubation.

Standard monitoring included an electrocardiograph, a non-invasive blood pressure monitor, and a pulse oximeter. A 20G intravenous catheter was inserted. Anesthesia was induced with midazolam 1 mg *iv*, fentanyl 50 µg *iv*, and propofol 80 mg *iv*. A size 3.5 ILA™ (Fig. 2) was inserted in a manner similar to that for standard laryngeal mask airway insertion; then the cuff was inflated with 15 ml of air. Lung ventilation was verified by observation of chest wall movement and the presence of normal end-tidal carbon dioxide waveforms.

After successful ventilation using the ILA™, the patient received succinylcholine 60 mg *iv*. The following steps were taken to insert the tracheal tube. First, the rigid stylet was removed from the lighted stylet wand (Trachlight®, Laerdal Medical Corporation, Wappingers Falls, NY, USA; Fig. 3). Second, a conventional tracheal tube (7.5-mm internal diameter, Mallinckrodt Inc, Hazelwood, MO, USA) was mounted and clamped onto the lighted stylet in the usual manner.¹² Third, the lighted stylet-tracheal tube assembly was inserted through the ILA™. As the assembly was being advanced, a distinct dime-sized glow was observed in the anterior part of the patient's neck at the super-thyroid notch, the cricothyroid membrane, the

Fig. 2 (Top) Lateral view of the Intubating Laryngeal Airway (ILA™): (A) removable 15-mm connector; (B) transparent shaft; (C) inflatable silicone ILA™ bowl with a keyhole-shaped airway outlet and mask ridges. (Bottom) Anterior view of the ILA™ showing the inside of the ILA™ bowl: (A) keyhole-shaped airway outlet; (B) mask ridges

Fig. 3 Lateral view of a tracheal tube mounted on a lighted stylet wand with the rigid stylet removed: (A) conventional 7.5-mm tracheal tube; (B) lighted stylet wand; (C) rigid stylet

trachea, and, finally, the supra-sternal notch. The endotracheal tube was released from the lighted stylet clamp, and the lighted stylet was removed while the position of the tracheal tube was maintained. The tracheal tube cuff was inflated with 5 ml of air, and the correct tracheal tube position was confirmed by capnography. The ILA™ cuff was deflated and left in situ, and the tracheal tube was then taped and secured. The alternative was to remove the ILA™ while keeping the tracheal tube in position using a stabilizer, a technique similar to that using the intubating laryngeal mask airway. We elected to proceed with the

former approach. Anesthesia was maintained with sevoflurane, oxygen, and air and progressed uneventfully.

At the end of the surgery, while the patient remained under inhalational anesthesia, both the endotracheal tube and the ILA™ were removed. A Cormack and Lehane grade 4 view was observed under direct laryngoscopy with a # 3 Macintosh blade. The ILA™ was reinserted and sevoflurane was discontinued. The patient awakened and the ILA™ was removed without complications.

Discussion

This case describes a novel application of a lighted stylet-guided tracheal tube insertion through an ILA™ in a patient with Hallermann-Streiff syndrome.

Hallermann-Streiff syndrome, also known as Hallermann-Streiff-Francois syndrome, Francois Dyscephalic syndrome, Oculomandibulofacial syndrome, or Oculo-Mandibulo-Dyscephaly-Hypotrichosis syndrome, was initially described by Aubry in 1893.¹ The characteristic features include dyscephaly, mental retardation, bird-like facies, a hypoplastic nose, microstomia, high arched palate, mandibular hypoplasia, anterior displacement of temporomandibular joint, an anterior larynx, and dental abnormalities, including natal malformed brittle teeth.^{2,3} Multiple ophthalmological abnormalities have also been described.³ Due to the abnormal anatomy of the upper airway, difficult airway management has been described.²⁻⁶

To date, several Hallermann-Streiff syndrome studies have suggested that the anesthesiologist use, or have available, alternative airway equipment for tracheal intubation.³⁻⁶ The options for tracheal intubation include nasal intubation, oral intubation, and tracheostomy.⁴⁻⁷ However, small nares, a hypoplastic nose, and a deviated nasal septum can make nasal intubation difficult.^{3,4} In addition, a small mouth, displaced temporomandibular joint, hypoplastic mandible, and an anterior glottis may lead to difficult laryngoscopy and visualization.^{3,4} If these difficulties are encountered, awake tracheostomy has been recommended; however, a short, thick neck has often been associated with the cricoid cartilage at the level of the suprasternal notch.^{5,6}

Recent practice guidelines from the American Society of Anesthesiologists, the Difficult Airway Society, and the Canadian Airway Focus Group recommend the use of alternative airway devices, for instance, the intubating laryngeal mask airway, in the management of patients with anticipated and unanticipated difficult airways.¹³⁻¹⁵ The use of a fiberoptic bronchoscope or a lighted stylet, in conjunction with an intubating laryngeal mask airway, has also been described.^{10,11,16,17} There is a higher success rate and a decreased intubation time with lighted

stylet-guided intubation compared to blind intubation through an intubating laryngeal mask airway.^{10,11} The ILATM, a new supraglottic airway device first introduced for North American clinical use in 2004, has been recommended as an alternative device for tracheal intubation.⁹ Although the ILATM and the laryngeal mask airway share functional similarities, there are a number of notable differences. Compared to the laryngeal mask airway, the ILATM has a removable 15-mm circuit connector, no aperture bars at the ventilatory opening, and shorter shaft distances, thereby allowing insertion of larger diameter tracheal tubes.⁹ Also, the ILATM does not have a metal handle or a metal shaft; it is inserted like a standard laryngeal mask airway, and regular polyvinyl chloride tracheal tubes can be utilized.¹⁸ This report describes successful lighted stylet-guided tracheal tube insertion through an ILATM in a patient with Hallermann-Streiff syndrome.

There are several advantages to the presently described technique for tracheal intubation: (1) The ILATM is relatively inexpensive; (2) there is familiarity with the insertion technique, due to similarities with the intubating laryngeal mask airway; (3) standard tracheal tubes can be used; (4) a flexible fiberoptic bronchoscope is not required; (5) ventilation can be maintained between intubating attempts; and (6) tracheal tube advancement guided by transillumination provides confirmation of intratracheal location, and positioning the tracheal tube tip at the suprasternal notch results in an approximate mid-tracheal location.¹² However, disadvantages do exist. (1) Anatomical features of Hallermann-Streiff syndrome, such as a small mouth, high-arched palate, mandibular hypoplasia, anterior displacement of temporomandibular joint, an anterior larynx, and dental abnormalities may prevent the use of supraglottic airways.^{3,5,7} (2) Although lighted stylet-guided tracheal intubations through supraglottic devices are associated with a higher success rate, they are not universally successful. Thus, alternative techniques, including fiberoptic bronchoscopy, must be available. (3) The ILATM is only available in sizes 2.5, 3.5, and 4.5; therefore, its use in smaller children and infants is limited. Just recently, a size 1.5 ILATM became available for children weighing 10–20 kg.

In conclusion, patients with Hallermann-Streiff syndrome have multiple anatomical abnormalities and potentially difficult airways. We describe a novel application of a lighted stylet-guided tracheal tube insertion through an ILATM in a patient with Hallermann-Streiff syndrome. This intubation technique can be considered as a primary route of intubation in patients with difficult airways or as a secondary rescue strategy if a primary method fails. Further studies are needed to assess the effectiveness of ILATM-guided intubation with fiberoptic bronchoscopy.

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Conflicts of interest None declared.

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