

Two Airway Management Techniques in a Pediatric Patient with Temporomandibular Joint Ankylosis due to Ankyloblepharon-Ectodermal Dysplasia-Clefting Syndrome: Two-stage Fiberoptic Scope Intubation Technique and Video Laryngoscope-guided Conversion from Nasotracheal to Orotracheal Intubation – A Case Report

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ABSTRACT

Ankyloblepharon-ectodermal dysplasia-clefting (AEC) syndrome is an ectodermal dysplasia presenting with cleft lip or palate and congenital filiform eyelid fusion. This is a case report of a 1-year and 6-month-old girl with AEC syndrome presenting with temporomandibular joint ankylosis who underwent release of temporomandibular joint ankylosis, coronoidectomy, bilateral cheiloplasty, Tajima rhinoplasty, and repair of lower lip pits under general anesthesia. Fiberoptic nasotracheal intubation was done successfully using a two-stage technique originally described by Stiles. As necessitated by the surgical plan, video laryngoscope guidance was subsequently used to facilitate the conversion from nasotracheal to orotracheal intubation.

Keywords: ectodermal dysplasia, TMJ ankylosis, airway management

INTRODUCTION

Ankyloblepharon-Ectodermal Dysplasia-Clefting (AEC) or Hay-Wells Syndrome is a rare autosomal dominant genetic disease characterized by the presence of ankyloblepharon, ectodermal abnormalities, and cleft lip and/or palate.^{1,2} According to the National Organization for Rare Disorders, its exact incidence and prevalence are still unknown. Furthermore, it is considered an extremely rare disorder with fewer than 100 affected individuals described in the medical literature. Mutations in the P63 gene, which is important in limb, craniofacial, and epithelial development, are the etiology of AEC.³ A small number of identified cases, the lack of large clinical studies, and the possibility of other factors influencing the disorder prevent the physician from developing an accurate picture of associated symptoms and prognosis. Hence, observed clinical features are complex and highly variable like skin erosions, a broadened nasal bridge, hypodontia, an oval face, maxillary hypoplasia, and syndactyly.²⁻⁴ Treatment of AEC is directed toward alleviating specific symptoms.⁵

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The incidence of temporomandibular joint (TMJ) ankylosis in AEC has not been established due to the rarity of the genetic condition itself. It, however, poses a challenge to airway management that encompasses both the management of a pediatric airway and a difficult airway scenario.⁶ In this case, we performed a pediatric fiberoptic nasotracheal intubation using a guidewire and catheter (in a two-stage technique described by Stiles). A second airway management technique using a video laryngoscope was also used to shift airway access from nasotracheal to orotracheal because of the complicated surgical plan.

CASE

Our patient is a 1-year and 6-month-old Filipino girl who presented at birth with multiple congenital anomalies including a cleft lip and palate, bilateral ankyloblepharon, the inability to open her mouth, and hyperpigmented lesions on both lower extremities. She was later diagnosed to have Ankyloblepharon-Ectodermal Dysplasia-Clefting (AEC) or Hay-Wells Syndrome. Since birth, she has been fed with the use of a specialized feeding tube inserted on the left side of her mouth.

At two months, an ophthalmologist performed manual separation of the ankyloblepharon under local anesthesia with the patient fully awake. A cranial CT scan and 2D echo were requested to rule out other anomalies which showed unremarkable findings apart from the TMJ ankylosis.

The patient's parents brought her for consultation again at 13 months of age. However, the planned admission

and surgery to repair her TMJ and cleft lip were delayed due to the COVID-19 enhanced community quarantine and lockdown.

At the time of elective admission for surgery, the patient weighed 9.5 kg. Her vital signs were as follows: blood pressure of 89/59 mmHg, heart rate of 130 bpm, respiratory rate of 32 cpm, temperature of 37.3°C, and oxygen saturation of 99% at room air. Salient head and neck physical examination findings included a flat midface, hypertelorism, a flat nasal bridge, a wide nasal base, a unilateral left cleft lip, a cleft palate, a short chin, and no appreciable mouth opening (Figures 1A and 1B). Her neck had a full range of motion. She also presented with hyperpigmented lesions on the lateral plantar aspect of her foot (Figure 1C). Results of preoperative laboratory tests including complete blood count, serum electrolytes, bleeding parameters, and blood chemistry were all normal.

The surgical plan for this patient included the release of TMJ ankylosis, coronoidectomy, bilateral cheiloplasty, Tajima rhinoplasty, and repair of lower lip pits. At the OR, standard monitors were attached to the patient. Preoperative vital signs were: blood pressure of 90/60 mmHg, heart rate of 138 bpm, respiratory rate of 30 cpm, and oxygen saturation of 98% at room air. Preoxygenation via pediatric endoscopy face mask was done for 5 minutes. Atropine 0.2 mg (0.02 mg/kg) and midazolam 0.5 mg (0.05 mg/kg) were given intravenously. Lidocaine 10% spray and oxymetazoline spray were administered intranasally. Boluses of ketamine at 0.5 mg/kg were given until general anesthesia was achieved.

With the use of a pediatric endoscopy face mask for ventilation, a pediatric fiberoptic scope (Karl Storz, outer



Figure 1. (A) Patient with flat midface, hypertelorism, flat nasal bridge, wide nasal base, unilateral cleft lip, cleft palate, short chin. (B) Patient with nil mouth opening. (C) Hyperpigmented lesions on the lateral plantar aspect of the foot.

diameter 3.7 mm) was inserted in the left nostril and guided downward until its tip was at the level of the vocal cords (Figures 2A-1 and 2A-2). An epidural catheter (B. Braun Perifix, g. 20) was threaded through the suction channel of

the fiberoptic scope until its tip was visualized just beyond the lens of the scope. Ten mg of Lidocaine 2% was sprayed onto the glottic opening to blunt the laryngeal reflexes during insertion of the endotracheal tube (ETT) (Figure 2B).

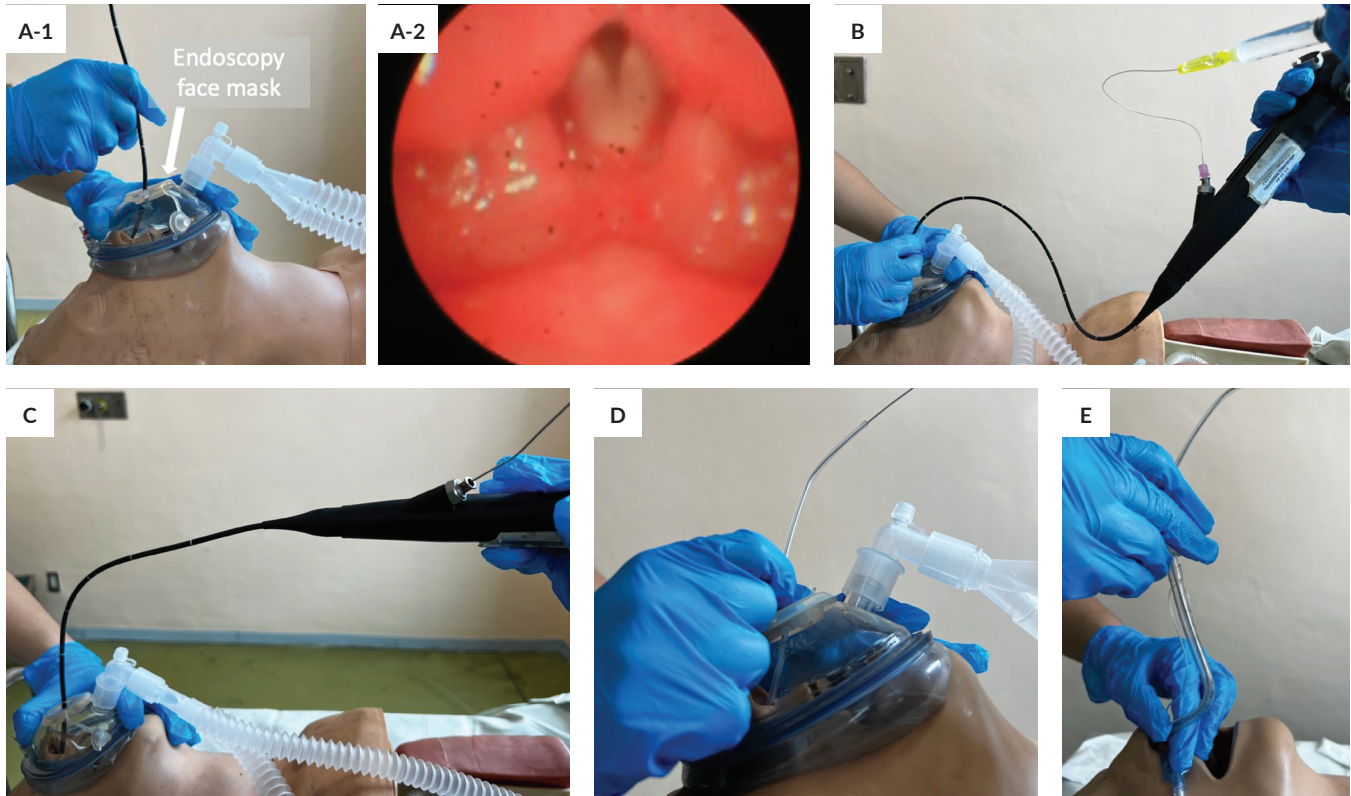


Figure 2. (A-1) Flexible fiberoptic scope inserted through the soft silicon port of the endoscopy mask into the patient's nostril. (A-2) View of the patient's glottic opening. (B) Lidocaine 2% given via epidural catheter threaded into the suction channel. (C) Guidewire inserted into the suction channel and threaded into the trachea. (D) Suction catheter threaded over the guidewire. (E) Preformed north ETT threaded over the suction catheter and guidewire into the trachea.

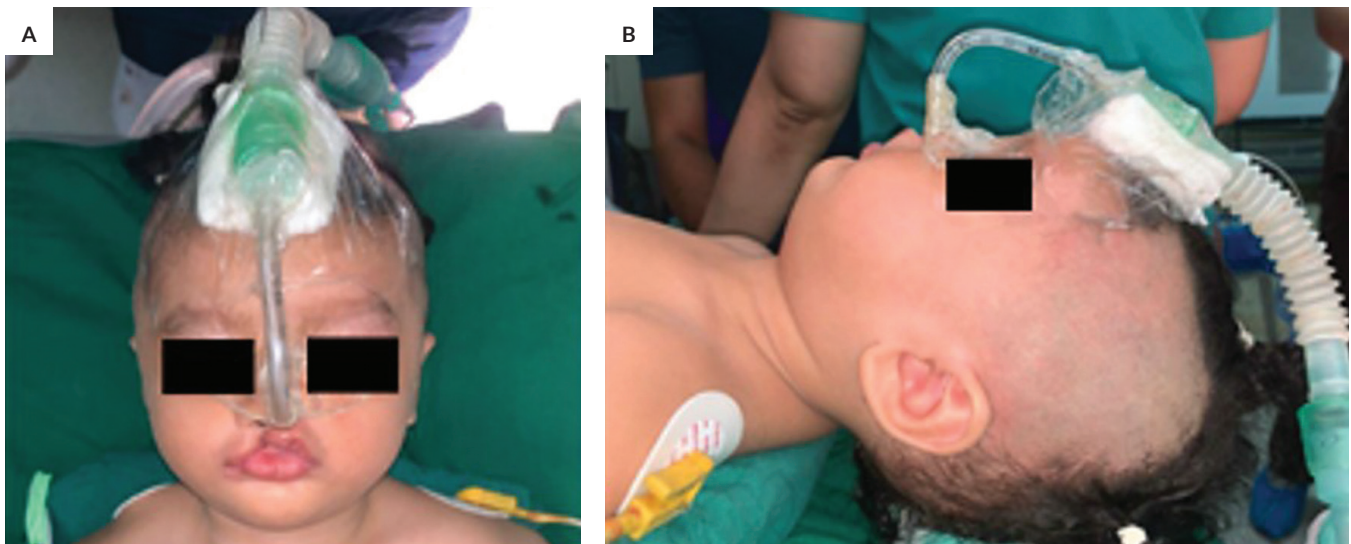


Figure 3. (A) AP view of the patient with nasotracheal tube in place. (B) Lateral view of the patient with nasotracheal tube in place.

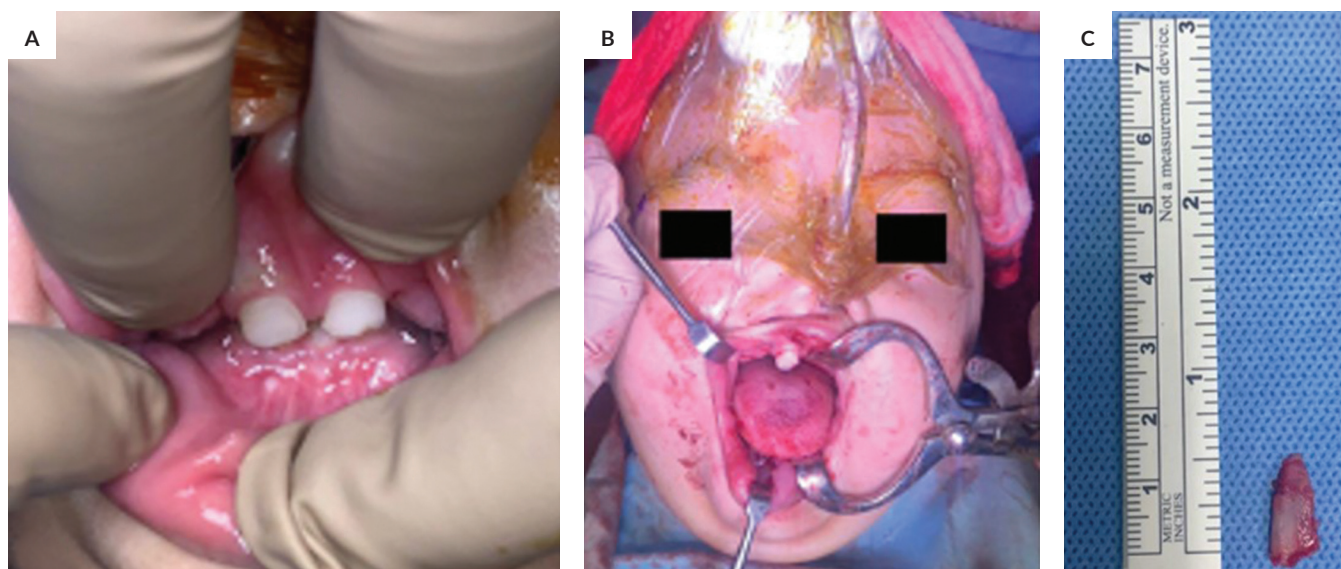


Figure 4. (A) Nil mouth opening before TMJ release. (B) 3.5 cm mouth opening after the TMJ release. (C) The portion of the coronoid process that was removed.

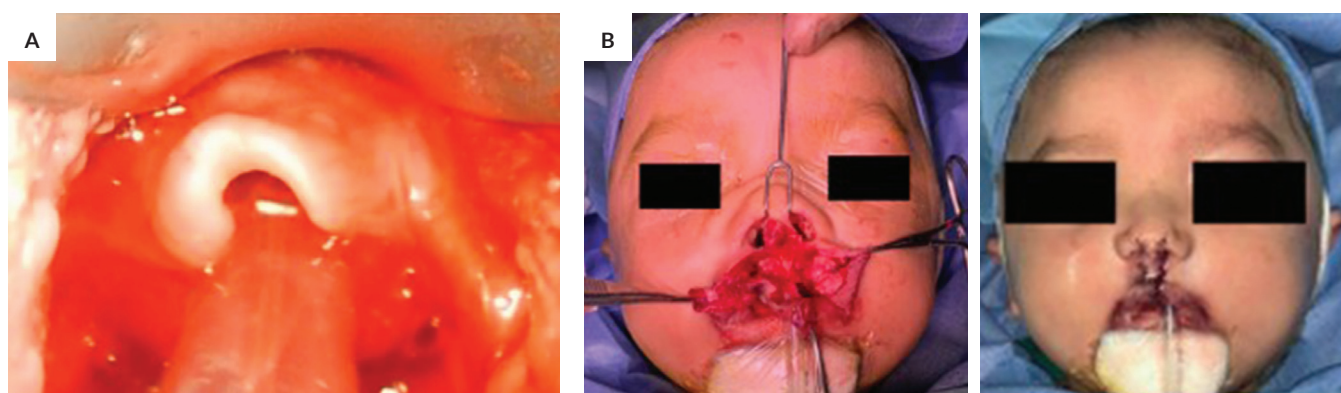


Figure 5. (A) Video laryngoscopy view of the glottic opening during conversion from nasotracheal to orotracheal intubation. (B) Patient with south ETT secured orotracheally.

The epidural catheter was then removed and a guide wire (Karl Storz 38 cm, outside diameter 0.9 mm, nitinol material) was inserted into the suction channel and advanced past the glottic opening into the trachea with about 3 cm of the guide wire within the trachea (Figure 2C). With the guide wire confirmed in place, the fiberoptic scope was removed, leaving the guide wire inside the trachea. A trimmed French 10 suction catheter was then advanced over the guide wire into the trachea (Figure 2D). A pre-formed nasal ETT (also known as north ETT) size 4.0 mm was threaded over the suction catheter and guide wire into the trachea (Figure 2E). Once at the appropriate depth, the suction catheter and guide wire were removed, leaving the ETT in place. The ETT cuff was then inflated. Its placement was confirmed by auscultation of breath sounds and the presence of capnography. The ETT was then sutured in place by the surgeon (Figures 3A and 3B).

Surgery commenced shortly after induction of general anesthesia with propofol infusion at 150–200 mcg/kg/min. A portion of the coronoid process of the mandible was removed to address the TMJ ankylosis, creating a 3.5 cm mouth opening (Figures 4A, 4B and 4C). At this point, nasotracheal intubation was converted to orotracheal intubation to facilitate bilateral cheiloplasty, Tajima rhinoplasty, and the repair of the lower lip pits. This was done with the aid of a video laryngoscope (C-MAC, Karl Storz, Germany) using a Miller blade size 1. Once the glottic opening with the ETT inside it was visualized (Figure 5A), the sutures of the north ETT were released. The north ETT was then slowly retracted cephalad and a new pre-formed oral ETT (also known as south ETT) size 4.0 mm was inserted and secured in place (Figure 5B).

The surgery lasted for 5 hours and 25 minutes. The patient was extubated fully awake. Post-operative pain

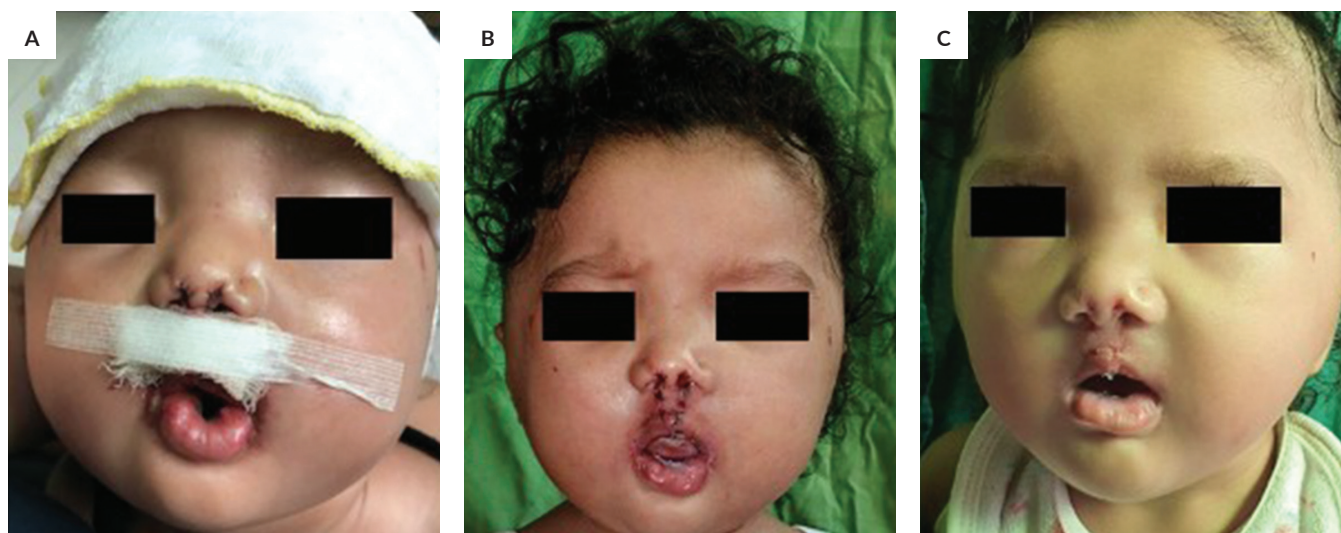


Figure 6. (A) Patient on her post-operative day 0. (B) Post-operative day 6. (C) Day of discharge, post-operative day 10.

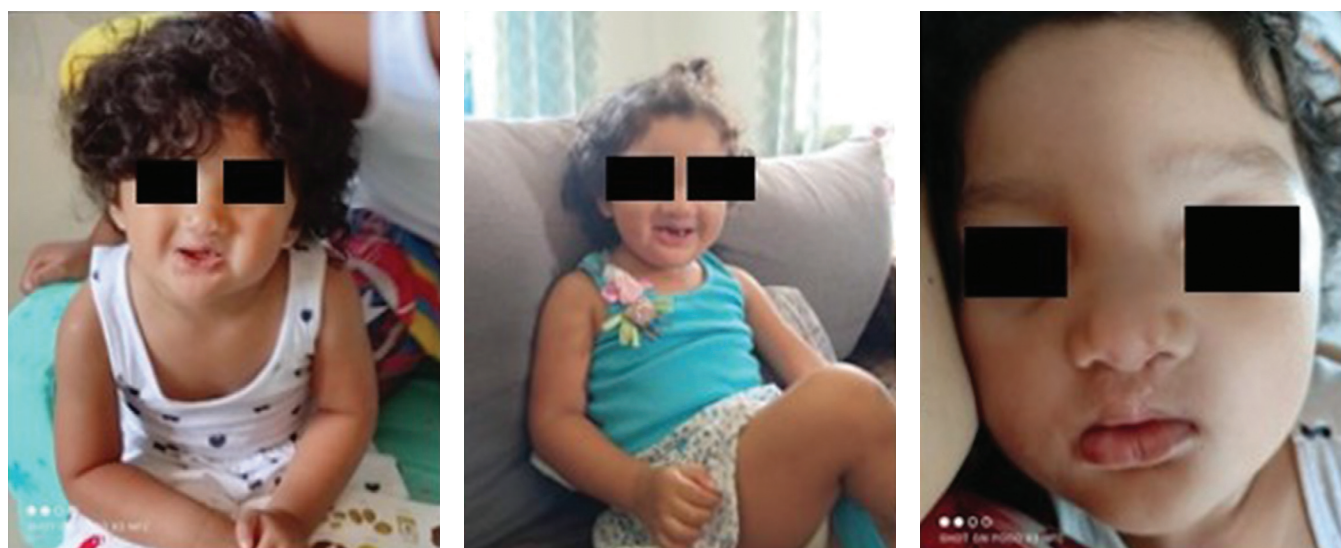


Figure 7. Six months post-operation.

medications included paracetamol 150 mg IV (15.8 mg/kg), nalbuphine 1 mg IV (0.1 mg/kg), and ibuprofen 80 mg PO (8.4 mg/kg) once feeding. She was transferred to the PACU stable and remained there without any critical events noted (Figure 6A).

On the sixth post-operative day, the sutures were removed. A dry, well-coaptated wound was appreciated (Figure 6B). The rest of the post-operative course was unremarkable, and she was discharged on the 10th post-operative day (Figure 6C). She had her follow-up via online consultation and was started on rehabilitation for strengthening of perioral muscles to improve oral feeding and stimulate language development.

Six months post-operatively (Figure 7), the mother reported that the patient's feeding had improved, and she could already babble syllables. The patient has regular follow-

up consults with the pediatric genetics and plastic surgery services in preparation for a future palatoplasty.

DISCUSSION

AEC syndrome was first reported by Hay and Wells in 1976 and is considered to be a rare type of ectodermal dysplasia.⁷ Ankyloblepharon is usually limited to small bands of vascularized connective tissue spanning the eyelids, and such is the classical feature that distinguishes AEC syndrome from other disorders that also present with ectodermal dysplasia and cleft palate and/or lip.³ Diagnosis is based on the characteristic symptoms, a detailed history, and a thorough clinical evaluation. Genetic testing may be done to detect the heterozygous missense mutation in the P63 gene

(that affects the sterile alpha motif domain in the protein). Symptoms are characterized by the abnormal development of one or more appendages of epidermal or oral ectodermal origin, and various secondary issues such as chronic sinusitis, conductive hearing loss, and otitis media.³ It is also associated with oral and auditory pathway abnormalities, including feeding difficulty, recurrent middle ear infections, and speech disarticulations.⁸ Neurologic and cognitive development are usually unaffected. Among the types of ectodermal dysplasia, TMJ ankylosis has only been identified to manifest in a child with hypohidrotic ectodermal dysplasia.⁹

Anesthetic management for TMJ ankylosis surgery presents a challenge to anesthesiologists in terms of maintaining airway patency.⁹ The inability to open the mouth makes the insertion of an oral airway, SGA device, and direct laryngoscope impossible. In a systematic review by Wahal, 210 pediatric patients with TMJ ankylosis operated on from 2010–2015 were successfully intubated with the aid of a fiberoptic endoscope. This affirms that fiberoptic intubation is the gold standard for these patients.⁸ When this fails, the only option left is a tracheostomy.

Vas and Sawant emphasized that there are no predictors of difficult intubation in the pediatric population, such as Mallampati score, Patil's sign, or Wilson's criteria. No predictive scoring exists for the airway evaluation of a child with TMJ ankylosis. Possible trismus from TMJ ankylosis further compounds the usual difficulties in securing the pediatric airway.^{10,11} Despite having protocols for airway management in syndromic children, there is currently no specific protocol for children with TMJ ankylosis.¹¹

Different methods are used to secure the airway in children with TMJ ankylosis (including fiberoptic nasal intubation, blind nasal intubation, fluoroscopic-assisted airway intubation, retrograde intubation, and the use of elastic bougie); each method poses pros and cons. Among these, the safest option is fiberoptic scope intubation while maintaining spontaneous ventilation.⁶

An awake intubation approach was not considered in this patient as she is not yet capable of cooperating. Midazolam was given to lessen agitation and crying that can propagate airway secretions. Atropine was given as an anti-sialogogue to dry up oral secretions to facilitate visualization with a fiberoptic scope. Oxymetazoline acted as a nasal decongestant to decrease nasal mucosal vascularity and reduce the risk of bleeding.⁶ Ketamine was chosen to achieve a deep level of anesthesia with the preservation of spontaneous respiration and airway reflexes. Neuromuscular blockers were not utilized during intubation as this may lead to airway collapse. In a patient with TMJ ankylosis, jaw thrust and chin lift have no value in maintaining airway patency.

A component that may be incorporated in similar scenarios is lidocaine nebulization before induction.¹⁰ This will render the tracheobronchial tree anesthetized and will prevent coughing or bucking during the threading of the guide wire and ETT.

Over the years, various techniques have been developed to secure the airway in difficult situations. Some are invasive and carry the potential for major complications. Ovassapian mentioned that if fiberoptic exposure of the larynx proves difficult, the use of an endoscopy mask provides the option of ventilating the patient during fiberoptic intubation;¹² this is a valuable option that is lacking in other intubation techniques.

The two-stage technique described by Stiles starts with positioning the fiberoptic scope at the level of the vocal cords. Then, a guide wire is advanced into the trachea. The fiberscope is removed, leaving the guide wire inside. A suction catheter is advanced over the guide wire into the trachea.¹³ Studies have shown that increasing the diameter of the anterograde guide facilitates ease of insertion of the ETT. Impingement of the tube tip at the posterior larynx tends to happen with increasing discrepancy of the diameter of guide wire and tube.¹⁴ The ETT is threaded over the guide wire and catheter into the trachea. Lastly, the catheter and guide are removed, leaving the ETT in place.¹³

Studies have reported the use of a flexible fiberoptic endoscope as an aid in passing an endotracheal tube in adult patients for whom intubation would otherwise be difficult.¹⁵ The use of a pediatric fiberoptic endoscope alone is difficult in infants since an ETT size 4.0 would not fit over the diameter of the scope. Stiles has described a method for the successful endotracheal intubation of infants using a fiberoptic endoscope and a cardiac catheter with a small, pliable guide wire. This is the basis for our procedure; no literature has been written on this intubation technique for pediatric patients with AEC syndrome presenting with TMJ ankylosis.

Like a case reported by Mohan et al., once a sufficient mouth opening has been established, nasotracheal intubation can be transitioned to oral intubation.¹⁶ This was necessary as the procedure included rhinoplasty. In this patient, we used a video laryngoscope for visualization and direct insertion of the desired orotracheal tube. In the previously cited article, direct laryngoscopy was performed to visualize the glottic opening and an elastic bougie was employed to guide ETT insertion.¹⁶

CONCLUSION

This report shows that a modified (use of endoscopy face mask for ventilation) two-stage flexible scope intubation technique is a viable option for airway management for pediatric patients with AEC Syndrome presenting with minimal mouth opening and TMJ ankylosis. Once sufficient mouth opening has been established, video laryngoscopy guidance can be used to transition from nasotracheal to orotracheal intubation if required by the surgical procedure.

Declaration of Patient Consent

Informed consent was obtained from the patient's guardian for the publication of the patient's clinical information and images.

Statement of Authorship

MRAM participated in collecting the information as well as writing both the original and final manuscript while AMHC reviewed and approved the final submitted case report.

Author Disclosure

Both authors declared no conflicts of interest.

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REFERENCES

1. Tajir M, Lyahyai J, Guaoua S, El Alloussi M, Sefiani A. Ankyloblepharon-ectodermal defects-cleft lip-palate syndrome due to a novel missense mutation in the SAM domain of the TP63 gene. *Balkan Journal of Medical Genetics*. 2020; 23(1):95-8.
2. Cabiling D, Yan A, McDonald-McGinn D, Zackai E, Kirschner R. Cleft lip and palate repair in Hay-Wells/ankyloblepharon-ectodermal dysplasia-clefting syndrome. *The Cleft Palate-Craniofacial Journal*. 2007; 44(3):335-9.
3. Guo S, Chen R, Xu Y, Mu Y, Chen L. Ankyloblepharon-ectodermal defects-cleft lip/palate syndrome. *Journal of Craniofacial Surgery*. 2017; 28(4):e349-e51.
4. Seres-Santamaria A, Arimany J, Muniz F. Two sibs with cleft palate, ankyloblepharon, alveolar synechia, and ectodermal defects: a new recessive syndrome?. *Journal of Medical Genetics*. 1993; 30(9):793-5.
5. Sutton V, Plunkett K, Dang D, Lewis R, Bree A, Bacino C. Craniofacial and anthropometric phenotype in ankyloblepharon-ectodermal defects-cleft lip/palate syndrome (Hay-Wells syndrome) in a cohort of 17 patients. *American Journal of Medical Genetics Part A*. 2009; 149A(9):1916-21.
6. Goswami D, Singh S, Bhutia O, Baidya D, Sawhney C. Management of young patients with temporomandibular joint ankylosis—a surgical and anesthetic challenge. *Indian Journal of Surgery*. 2016; 78(6):482-9.
7. Khalfi L, Hamama J, Mahroug L, Arrob A, Sabani H, El Khatib K. Syndrome de Hay-Wells : à propos d'un cas. *Archives de Pédiatrie*. 2016; 23(2):163-6.
8. Cole P, Hatef D, Kaufman Y, Magruder A, Bree A, Friedman E et al. Facial clefting and oroauditory pathway manifestations in ankyloblepharon-ectodermal defects-cleft lip/palate (AEC) syndrome. *American Journal of Medical Genetics Part A*. 2009; 149A(9):1910-5.
9. Goyal M, Pradhan G, Gupta S, Kapoor S. Hypohidrotic ectodermal dysplasia with ankylosis of temporomandibular joint and cleft palate: a rare presentation. *Contemp Clin Dent*. 2015 Jan-Mar; 6(1):110-2.
10. Wahal R. Temporo-mandibular joint ankylosis – The difficult airway. *Journal of Oral Biology and Craniofacial Research*. 2015; 5(2):57-8.
11. Vas L, Sawant P. A review of anaesthetic technique in 15 paediatric patients with temporomandibular joint ankylosis. *Pediatric Anesthesia*. 2001; 11(2):237-44.
12. Raj D, Luginbuehl I. Managing the difficult airway in the syndromic child. *Continuing Education in Anaesthesia Critical Care & Pain*. 2015; 15(1):7-13.
13. Stiles C. A Flexible Fiberoptic Bronchoscope for Endotracheal Intubation of Infants. *Anesthesia & Analgesia*. 1974; 53(6):1017-9.
14. Dhara, S.S., McGlone, D.J. and Skinner, M.W. Development of a new system for guidewire-assisted tracheal intubation: manikin and cadaver evaluation. *Anaesthesia*. 2016; 71:44-9.
15. Harwood T. Fiberoptic endoscopy and the difficult airway, 2nd edition. *Anesthesiology*. 1998; 88(2):555-6.
16. Mohan K, Mohana RL, Gopala KMS, Greeshma PG, Bhavana U. Anaesthesia for TMJ ankylosis with the use of TIVA, followed by endotracheal intubation. *J Clin Diagn Res*. 2012 Dec; 6(10):1765-7.