CASE REPORT

Successful intubation with air-Q in Pierre Robin syndrome

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SUMMARY

Airway access is particularly difficult in infants and children with some anatomical deformities, usually associated with congenital syndromes. Craniofacial abnormalities are commonly seen in the Pierre Robin Syndrome (PRS), Treacher Collins and Goldenhar syndromes. The Pierre Robin sequence consists of micrognathia and relative macroglossia with or without cleft palate. In the severe case, airway obstruction and feeding difficulties are present. Endotracheal intubation may be difficult, or in some case even impossible. We present a case report of intubation of a child, suffering from PRS, by using air-Q, a new intubating LMA, and use of tongue stitches to maintain airway during recovery.

Key words: Pierre Robin Syndrome; Pierre Robin sequence; Craniofacial abnormalities; Micrognathia; Macroglossia; Glossoptosis; Air-Q intubating laryngeal mask airway; Tongue stitches; Intubation

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INTRODUCTION

Pierre Robin Syndrome (PRS) is a congenital syndrome, consisting of multiple anomalies, including, micrognathia, receding and short mandible, cleft lip and/or palate. These children are either diagnosed before birth with the help of USG or immediately after birth. They have to undergo multiple surgeries, which usually have to be performed under general anesthesia. The major problem in PRS, which is faced by anesthesia team, is maintenance of airway due to glossoptosis resulting in obstructive sleep apnea (OSA). The anatomic obstruction along the entire airway can lead to cardio-pulmonary arrest and even death.[1–3]

We present a case report of successful management of airway in a one and half year old child, suffering from PRS and scheduled to undergo closure of her cleft palate. We intubated him with the help of air-Q intubating laryngeal mask airway (ILMA) or masked laryngeal airway, and had to use tongue sutures to keep the airway patent.

CASE REPORT

A two years old baby girl, weighing 8 kg, reported for repair of her cleft palate under general anesthesia. Since birth she had had feeding problems due to her congenital anomalies. The only way of feeding for the initial fourteen months had been through her nasogastric tube, which had to be replaced after every few days. She had recurrent chest infections due to repeated aspirations. For the previous ten months she could tolerate small liquid feeds, but no solids. She was a known case of PRS, had short receding jaw and mouth opening limited to 1 cm, tongue was large, plastered to the mouth floor and retracted posteriorly. There was no other associated congenital abnormality. The breathing was labored and noisy due to obstruction of the airways by bulky tongue and her resting SpO_2 was 90% on room air. An attempt to visualize the oral structures failed due to lack of cooperation by the crying child. Her blood count was normal and Hb was 11.2 gm/dl, WBC count 14.9, RBC count 5.03 and hematocrit 36. Her echocardiography revealed no abnormality of her heart. She was anticipated to be a particularly difficult case as regard to airway management.

In the operating room, inj. atropine 0.2 mg was given IV and infusion was started. She was planned to be

anesthetized by inhalational technique, without loosing spontaneous respiration. Sevoflurane in 100% oxygen was administered through face mask, starting with 3% and gradually increasing to 8%. Standard monitoring was attached. When adequate depth of anesthesia was achieved, laryngoscope McIntosh blade No. 1 was tried to be inserted in the oral cavity but failed to advance beyond a few centimeters. An attempt with Miller blade also failed. There was just not sufficient space within her oral cavity to accommodate the laryngoscope blade. The child was continued to be ventilated by face mask, although full effort was required to keep her mouth open by pushing her mandible downward and forward. An i-gel No. 1.5 was then inserted and the anesthesia circuit attached to it. The child was now easily ventilated even by manual compression of the reservoir bag. But the surgery could not be accomplished with i-gel within the pharynx. Then air-Q No. 1.5 was brought in, prepared and inserted. The cuff was inflated. The circuit was attached and successful manual ventilation was confirmed by capnography and by chest auscultation. After ventilation for five minutes with sevoflurane in 100% oxygen, the circuit and the connector of the air-Q were removed, and a well-lubricated endotracheal tube (ETT) No. 3.5 was inserted through the proximal end of the air-Q. It was felt to pass through the glottis, its cuff inflated to 25 cmH_2O pressure and the circuit was attached. Again successful manual ventilation was confirmed by capnography and by chest auscultation. The cuff of air-Q was then deflated, the connector of the ETT was removed and the removal stillet inserted in the proximal end of the ETT. Keeping the stillet as well as the ETT steady, the air-Q was pulled out. Manual ventilation was resumed through the ETT and equal air entry confirmed bilaterally. The child was paralysed with inj. atracurium 5 mg IV. Inj. nalbuphine 2 mg and inj. midazolam 1 mg were given IV. Mechanical ventilation was started in PCV mode, respiratory rate of 25-30/min, I:E ratio 1:1.5, FiO_2 of 50% and sevoflurane 2-3%.

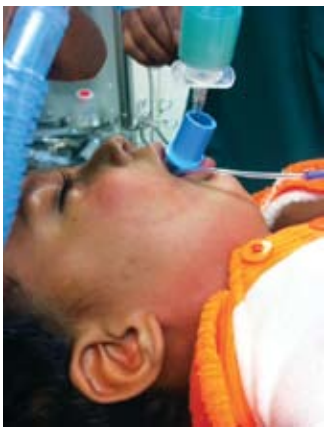

Figure 1: ETT in place through the air-Q

The surgery lasted for about thirty minutes, after which the anesthetic agent was stopped and the child was ventilated with 100% oxygen. Respiratory rate and the inspiratory pressures were gradually decreased so as to let CO_2 build up to 40-45%. On resumption of spontaneous breathing effort, neostigmine and glycopyrrolate were given. The child maintained adequate tidal volume on spontaneous breathing and the recovery went smooth until extubation was done, when she started crying. The surgeon demanded that the child be kept calm and stress free, to avoid bleeding from the surgical site. Hence, she was sedated with inj. midazolam 0.5 mg IV, after which upper airway obstruction by the tongue was noted and SpO_2 dropped to 80%. Airway was only partly patent even by forceful jaw holding. To raise SpO_2 from 90% to $\geq$95%, air-Q was reinserted, anesthesia circuit attached and 100% O_2 given. This cycle was repeated three times. Every time air-Q was removed and the child allowed to resume full recovery, she cried; and every time she was sedated, her SpO_2 fell due to airway obstruction by the tongue. The surgeon was requested to intervene and stitch the tongue outside. He stitched it at two places through the alveolar margins pulling it forward, after which the stridor was terminated and the child maintained SpO_2 despite being sedated. She was then shifted to the pediatric ICU for postoperative care. The stitches in the tongue were removed the next day and she was discharged after few days.

DISCUSSION

Airway management and endotracheal intubation in children usually present no difficulties for the experienced anesthesiologists. In infants, access may be slightly more problematic because of certain anatomical variations. Airway access is particularly difficult in infants and children with some anatomical deformities, usually associated with congenital syndromes. Craniofacial abnormalities are commonly seen in the PRS, Treacher Collins and Goldenhar syndromes. The Pierre Robin sequence consists of micrognathia and relative macroglossia with or without cleft palate. In the severe case, airway obstruction develops in the first four weeks of life.[4] When the baby is supine the tongue will fill the nasopharynx (via the cleft palate if present) and cause varying degrees of airway obstruction. In our case the child had all of the classical signs of PRS, so a difficult airway was anticipated at the time of pre-anesthesia assessment.[5] We took all the appropriate precautions to deal with adverse situation.

While pre-operative sedation of the pediatric patients remains individual choice, it's common for anesthesiologists to give oral premedication to reduce perioperative anxiety, ease induction, and increase parental satisfaction.[6,7] We chose not to sedate her preoperatively, due to uncertain timing of the surgery and inadequate protocols for transferring pediatric patients from the ward to the operating rooms.

The repair of the cleft palate necessitates the choice of general anesthesia with intubation. The airway is shared by the anesthesiologist and the surgeon, so we cannot

opt for the laryngeal mask airway (LMA) or i-gel, as both these devices fill the small buccal cavity leaving no space for the surgical maneuvers. The combination of severe micrognathia and relative macroglossia and the cephalad placement of the larynx can make the larynx almost invisible with conventional equipment. Some authors have advocated awake LMA insertion followed by inhaled induction of anesthesia in neonates with the PRS and severe upper airway obstruction presenting for glossopexy or mandibular distraction.[8,9] This option cannot be exercised in pediatric cases due to lack of cooperation.

In similar situations in an adult patient an awake fibreoptic technique would often be advocated, but this cannot be done in children since cooperation is required to gain good bronchoscopic views. Pediatric flexible fibreoptic bronchoscope was available with us, but its use in an emergency requires considerable expertise and practice.

Atropine premedication was administered to dry up secretions and to counteract bradycardia due to vagal stimulation. We used oxygen with sevoflurane by a spontaneous breathing method. Muscle relaxants were withheld until the airway was secure. Intubation was performed under deep inhalational anesthesia for fear of loss of airway. Use of a muscle relaxant during induction of anesthesia may result in a situation where one can neither manually inflate the patient's lungs nor intubate, and must therefore gain a surgical airway rapidly. Maintenance of spontaneous breathing allowed us a way out, should the airway had proved impossible to secure.[10]

The use of a supraglottic airway device for the management of a difficult airway and as a conduit for tracheal intubation is recommended by many guidelines, e.g. that of the American Society of Anesthesiologists.[11] In the meantime a multitude of such devices has been developed and made commercially available.

It is usually impossible to insert even the smallest Macintosh or Miller blades in the pharynx for intubation in these children. The alternatives available are blind intubation, the use of conventional LMA or i-gel or intubating LMA. Blind intubation may be associated with a high incidence of failure and the risk of hypoxia. Intubation through LMA Classic or i-gel, although mentioned in the literature, but is not favoured due to low rate of success. The original intubating laryngeal mask airway (Fastrack) was specifically designed to facilitate intubation without using a laryngoscope in difficult cases, however, it is not available in our country. Recently another similar device has been launched with the name of air-Q (Cookgas, St. Louis, Missouri, USA). Its design includes a curved shaft, the lack of a grill in the ventilating orifice and an easily removable airway adapter. However, reports and studies concerning this device are scarce in the literature.[12,13] The air-Q ILA offers several advantages in children over traditional laryngeal masks, the most significant being the ability to pass an ordinary cuffed tracheal tube through its airway tube, although it failed to be reliable facilitators for blind intubation. In contrast, the Fastrach ILMA had a 95% success rate and proved to be the best approach among those tested for securing the airway and facilitating blind intubation. The additional use of a fibrescope led to a near-100% success rate.[14] But it requires special, purpose built ETT to be used with it. The air-Q has shown a performance similar to the Cobra-PLUS regarding quality of ventilation, but was slightly better for blind intubation. However, 43% failed attempts seems rather high considering that this device is used in an emergency setting.[15] Traumatisation by supralaryngeal airways and blind intubation have been described in the literature.[16-18] A new generation air-Q has been described to be more efficient in intubation.[19] We successfully used air-Q for intubation in our patient in first attempt and atraumatically. An ordinary cuffed ETT was used and the air-Q was removed over the tube easily.

During recovery, sedation is advisable to keep the child cool and calm and avoid bleeding and aspiration. The sedation, on the other hand, causes tongue to lose tone and results in respiratory obstruction even in lateral or prone position in PRS patients. Some maneuvers described to overcome this obstruction include insertion of small diameter tubes (or nasal airways if available in suitable size) through both of the nostrils and/or tying tongue to the chin.[20] In a survey based study conducted via e-mail to 2080 members of the American Cleft Palate-Craniofacial Association, with a total of 396 responders, tongue suture in postoperative airway management of the cleft palate patient was used by 41.1% of the respondents, equal to those who never used it.[21]

In our case the surgeon tied the tongue at two points with silk passing through the alveolar ridges (gums). Both these methods are useful and the child can be kept sedated. In extreme cases, these children are best nursed prone, but surgical intervention such as tracheostomy, bilateral mandibular distraction or glossopaxy may prove necessary.[4,22]

CONCLUSION

Airway maintenance during anesthesia is the prime duty of the anesthesiologists, and air-Q ILMA as well as tongue stitches can provide great help and come to their rescue in difficult cases like Pierre Robin syndrome.

REFERENCES

1. Tomaski SM, Zalzal GH, Saal HM. Airway obstruction in the Pierre Robin sequence. Laryngoscope.1995;105:111–4. [PubMed]
2. Guilleminault C. Obstructive sleep apnea syndrome and its treatment in children: Areas of agreement and controversy. Pediatr Pulmonol. 1987;3:429–36. [PubMed]
3. Cohen SR, Simms C, Burstein FD, Thomsen J. Alternatives to tracheostomy in infants and children with obstructive sleep apnea. J Pediatr Surg. 1999;34:182–6. discussion 187. [PubMed]
4. Gözü A, Genç B, Palabiyik M, Unal M, Yildirim G, Kavuncuoğlu S, Ozsoy Z. Airway management in neonates with Pierre Robin sequence. Turk J Pediatr. 2010 Mar-Apr;52(2):167-72. [Medline]
5. Smith MC, Senders CW. Prognosis of airway obstruction and feeding difficulty in the Robin sequence. Int J Pediatr Otorhinolaryngol. 2006 Feb;70(2):319-24. [Medline] Epub 2005 Aug 19.
6. Barone CP, Pablo CS, Barone GW. Postanesthetic care in the critical care unit. Crit Care Nurse. 2004;24(1):38-45.[Context Link]
7. Jordi M, Elena C, Anna H. Evidence for the use of a numerical rating scale to assess the intensity of pediatric pain. EurJ Pain. 2009;13(10):1089-1095. [Context Link]
8. Markakis D, Sayson S, Schreiner M. Insertion of the laryngeal mask airway in awake infants with the Robin sequence. Anesth Analg 1992; 75: 822–824. [Medline] [Free Full Article]
9. Stricker P, Budac S, Fiadjoe J et al. Awake laryngeal mask insertion followed by induction of anesthesia in infants with the Pierre Robin sequence. Acta Anaesthesiol Scand 2008; 52: 1307–1308. [Medline] doi: 10.1111/j.1399-6576.2008.01751.x.
10. Walker RWM. Management of the difficult airway in children. J R Soc Med. 2001 July;94(7):341–344. [Medline]
11. Benumof JL. ASA difficult airway algorithm. New thoughts and considerations. In: Hagberg CA, editor. Handbook of difficult airway management. Philadelphia: Churchill Livingstone; 2000. pp. 31–48.
12. Yang D, Deng XM, Tong SY, et al. Fiberoptic intubation through Cookgas intubating laryngeal airway in two children. Anaesthesia 2009; 64:1148–1149. [Medline] doi: 10.1111/j.1365-2044.2009.06097.x.
13. Jagannathan N, Roth AG, Sohn LE, et al. The new air-Q intubating laryngeal airway for tracheal intubation in children with anticipated difficult airway: a case series. Pediatr Anaesth 2009; 19:618–622. [Medline]
14. Fiadjoe JE, Stricker PA. The air-Q intubating laryngeal airway in neonates with difficult airways. Pediatr Anesth 2011;(21):702-3 [Medline] doi: 10.1111/j.1460-9592.2011.03558.x.
15. Erlacher W, Tiefenbrunner H, Ka¨ stenbauer T, Schwarz S, Fitzgerald RD. CobraPLUS and Cookgas air-Q versus Fastrach for blind endotracheal intubation: a randomised controlled trial. Eur J Anaesthesiol 2011;28:181–186 [Medline] doi: 10.1097/EJA.0b013e328340c352.
16. Turan A, KayaG, KoyuncuO, et al. Comparison of the laryngeal mask (LMA) and laryngeal tube (LT) with the new perilaryngeal airway (CobraPLA) in short surgical procedures. Eur J Anaesthesiol 2006; 23:234–238. [Medline]
17. Akca O, Wadhwa A, Sengupta P, Durrani J, Hanni K, Wenke M, et al. The new perilaryngeal airway (CobraPLUS™) is as efficient as the laryngeal mask airway (LMA™) but provides better airway sealing pressures. Anesth Analg 2004; 99:272–278. [Medline]
18. van Zundert A, Al-Shaikh B, Brimacombe J, Koster J, Koning D, Mortier EP. Comparison of three disposable extraglottic airway devices in spontaneously breathing adults: the LAM-Unique, the Soft Seal Laryngeal Mask, and the Cobra perilaryngeal airway. Anesthesiology 2006; 104:1165–1169. [Medline]
19. Joffe AM, Liew EC, Galgon RE, Viernes D, Treggiari MM. The second-generation air-Q intubating laryngeal mask for airway maintenance during anaesthesia in adults: a report of the first 70 uses. Anaesth Intensive Care. 2011 Jan;39(1):40-5. [Medline]
20. Dorfman DW, Ciminello FS, Wong GB. Tongue suture placement after cleft palate repair. J Craniofac Surg. 2010 Sep;21(5):1601-3. [Medline] doi: 10.1097/SCS.0b013e3181ebccb1.
21. Bath AP, Bull PD. Management of upper airway obstruction in Pierre Robin sequence. J Laryngol Otol. 1997 Dec;111(12):1155-7. [Medline]
22. Meyer AC, Lidsky ME, Sampson DE, Lander TA, Liu M, Sidman JD. Airway interventions in children with Pierre Robin Sequence. Otolaryngol Head Neck Surg. 2008 Jun;138(6):782-7. [Medline] doi: 10.1016/j.otohns.2008.03.002.

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