CASE REPORT

Successful intubation in a child with Lowe syndrome using fiberscope and Glidescope®

Dheeraj Kapoor, MD, FCCP, FACEE, FCCS*, Nitin Ahuja, MBBS**, Meghana Srivastava, DNB***

**Assistant Professor, **Post Graduate Trainee, ***Senior Resident*

Department of Anesthesiology & Intensive Care, Govt. Medical College & Hospital, Chandigarh-160030 (India).

Correspondence: Dheeraj Kapoor, 1207, Sector 32 B, Chandigarh-160030 (India); Phone: 911724622549, 919646121549; E-mail: kapoor.dheeraj72@gmail.com; dheeraj.kapoor72@gmail.com

ABSTRACT

We report the airway management in anticipated difficult airway of a two year old male child with diagnosis of Lowe syndrome, employing two airway management devices. Fiberscope assisted Glidescope® intubation was employed to manage the difficult airway. Lowe syndrome is a rare inherited metabolic disorder with hypotonia, delayed motor and mental milestones, renal dysfunction and hypokalemia. The child had an anticipated difficult airway by virtue of large head circumference with frontal bossing, retrognathia and high arched palate. A careful preanesthetic evaluation and discrete attention to the distinctive components of the syndrome are the essence of successful perioperative management. Airway management in these patients requires standard algorithmic approach to difficult airway with careful selection of ventilation and intubation techniques and aids suitable in these situations to prevent any catastrophes.

Keywords: Lowe syndrome; Oculocerebrorenal syndrome; Glidescope®; Fiberscope; Stylet; Glottic aperture; Optimum external laryngeal maneuver

Citation: Kapoor D, Ahuja N, Srivastava M. Successful intubation in a child with Lowe syndrome using fiberscope and Glidescope®. Anaesth Pain & Intensive Care. 2012;16(3):283-286

INTRODUCTION

Oculocerebrorenal syndrome of Lowe (OCRL) is a rare X-linked recessive multi-system disorder.[1] The classic triad of this syndrome consists of congenital cataract, neonatal or infantile hypotonia accompanied with subsequent mental impairment and renal tubular dysfunction.[1] Renal Fanconi's syndrome was found to be associated with Lowe syndrome.[1] It is signified by low molecular weight proteinuria, proximal renal tubular acidosis, renal phosphate wasting leading to rickets, osteomalacia resulting in pathological fractures, bicarbonate wasting, hypercalciuria, aminoaciduria and hypokalemia.[2] Lowe syndrome is caused by mutation of the OCRL gene, located on chromosome Xq24-26 which provides coding for enzyme phosphatidyl inositol biphosphatase leading to the accumulation of phosphatidyl inositol and mutual disequilibrium of phosphoinositides causing the characteristic clinical features.[2-4,7] Anesthetic management for such patients undergoing surgical procedures can be challenging, due to problems like high intraocular pressure, mental retardation, metabolic abnormalities (due to renal tubular dysfunction) and difficult airway. We report the airway management of a two year old male child with Lowe syndrome, posted for congenital cataract surgery having anticipated difficult airway, by employing two airway devices simultaneously including fiberscope and Glidescope® (Verathon Inc. Bothell, WA, USA) for endotracheal intubation.

CASE REPORT

A 2-year-old boy, diagnosed with Lowe's syndrome, was scheduled for bilateral cataract extraction. Preanesthetic evaluation of the child was done. The child was born out of a full-term normal vaginal delivery in a

hospital and was observed to have cataract in both eyes at birth. There was no history suggestive of any perinatal complications in the mother, who stated that the child had delayed motor milestones like neck holding and sitting with support. The child had suffered from recurrent lower respiratory tract infections. There was no history of difficulty in feeding or past medical or surgical treatment. Family history was insignificant.

On physical examination, the child had a large head circumference, 54.1 cm (more than 97th percentile), frontal bossing, severe retrognathia and high arched palate. He had decreased muscle tone and sluggish deep tendon reflexes. His body weight was 8 kg (less than 3rd percentile). He had a skin tag above his left tragus. His vital signs were within normal limits and no other gross abnormality was detected on systemic examination. His blood investigations showed hemoglobin (14gm/dl), hypokalemia and raised alkaline phosphatase (284 IU/L). Albumin was detected on urine analysis. His serum sodium, phosphate, magnesium and calcium levels were normal. He was further investigated for suspected congenital abnormalities. Renal function tests, blood sugar, arterial blood gas analysis, chest radiograph and thyroid function test were ordered and the results were found within normal limits. Ultrasonography (USG) of whole abdomen was done to rule out any gross congenital abnormality with no positive findings.

He was planned for scheduled cataract surgery. Oral potassium supplementation was given preoperatively to correct hypokalemia. On the day of surgery, no pre-medication was given. After taking written parental consent, the child was taken in the operating room (OR). In the OR, continuous electrocardiogram (ECG), non-invasive blood pressure (NIBP) and pulse oximeter (SPO2) were monitored. After preoxygenation with 100% oxygen for 3 min, anesthesia was induced with sevoflurane (2-4%) in 6 l/min of oxygen. An intravenous access with 22G IV cannula was placed on the dorsum of the left hand. Subsequently, injection glycopyrrolate 50 μg (6μg/kg), fentanyl 15 μg (2μg/kg) were administered intravenously. On confirmation of adequate mask ventilation we planned for visualization of glottic aperture using reusable Glidescope®, using pediatric size blade, in anticipation of difficult intubation. We were able to visualize only the posterior region of the glottic aperture (arytenoids and the epiglottis only, Cormack-Lehane grade 2B) even with application of optimum external laryngeal manipulation. We introduced the uncuffed endotracheal tube (4.5 mm internal diameter), premounted on the preformed stylet (Gliderite®) supplied with the Glidescope® using the technique described by Ron M Walls,[7] but failed to introduce the distal tip of the ETT in front of the glottic aperture which was present too anterior. We than decided to introduce ETT, mounted on a flexible fiberscope, while visualizing the glottic aperture by Glidescope®. We mounted the 4.5 mm internal diameter uncuffed ETT on flexible fiberscope (PENTAX® Europe GmbH, slim Fl-10P2 intubation fiberscope, distal tip diameter 3.4 mm) and introduced it through oral route. The child's trachea was successfully intubated using this technique. After ensuring the correct placement of the ETT, the child was paralyzed with atracurium besylate 5 mg (0.6mg/kg). Anesthesia was maintained with sevoflurane (0.8-1 MAC) in oxygen and nitrous oxide (50-50%) with intermittent doses of atracurium for muscle relaxation. Intravenous paracetamol 150 mg (20mg/kg) was administered towards the end of surgical intervention. Intra-operative ABG analysis and blood sugar values were unremarkable. Duration of the surgery was about 60 minutes and a total of 50 ml of normal saline was administered. On conclusion of the surgery, once spontaneous breathing returned, anesthesia was reversed with neostigmine (0.4 mg) and glycopyrrolate (80 μg) and extubation of the trachea was performed. Post-operatively, oxygen was given by venturi mask (FiO_2 0.5). He was observed in the post-anesthesia care unit (PACU) for 3 hours before being transferred to the ward. The post-operative course was uneventful.

DISCUSSION

Oculocerebrorenal syndrome of Lowe or Lowe-Terry-Mac Lachlan syndrome is reported as a multi-system X-linked recessive disorder, mainly affecting males, with prevalence of 1 in 500,000 in general population.[2,5] In India, however, the incidence and prevalence of this syndrome is not known.[4] The diagnosis is made in individuals showing typical clinical features along with demonstration of reduced activity of inositol polyphosphate 5-phosphatase enzyme, in cultured skin fibroblasts.[10] Low molecular weight proteinuria may be the most sensitive marker of renal dysfunction occurring in this disorder, as it can be seen early in life even in the absence of clinically significant aminoaciduria or any other renal abnormalities.[6,11,12]

The disease is manifested usually in three stages, where cataracts and glaucoma associated with mental disability, are evident during the neonatal period.[1,2] Fanconi's type of proximal renal tubular dysfunction occurs till the mid childhood period.[8,9] The third phase may be complicated by chronic renal failure during second decade of life.[2,3,8,9] Clinical features include a prominent forehead (frontal bossing) with thin and sparse hair, short stature, hypotonia, protruding ears, decreased

deep tendon reflexes, failure to thrive, maladaptive repetitive behavior, cryptorchidism, gastroesophogeal reflux, aspiration pneumonitis, poor cough reflex, hypermobile joints, delayed puberty, dental malformations and coagulopathies.[8,9,12] Alkaline phosphatase (ALP) is commonly found elevated in Lowe syndrome.[6] Anesthesia may be required in these children for various surgical procedures like cataract extraction, strabismus correction, scoliosis reconstruction, herniotomy and orchidopexy.

Our case presented with bilateral congenital cataract, hypotonia, delayed milestones, history of frequent respiratory tract infection and low body weight (failure to thrive). He was diagnosed as Lowe syndrome on basis of these clinical entities along with evidence of proximal renal tubular dysfunction resulting in proteinuria, hypokalemia and raised ALP. We also noticed a skin tag over his left tragus, a distinct feature of this syndrome as observed by Scaricaoglu et al.[1] We anticipated a difficult airway due to the presence of large head circumference, frontal bossing, retrognathia and high arched palate which is typically seen in this syndrome.[2,4,5,8]

Our anesthetic concerns in this patient were primarily difficult airway, hypokalemia, hypotonia, positioning and risk of hypoglycemia. Oral potassium supplements and glucose-containing fluids were given pre-operatively for management of hypokalemia and for prevention of episodes of hypoglycemia or dehydration. No premedication was allowed to prevent exacerbation of existing hypotonia. Airway management in this patient needed special concern in view of distinct craniofacial abnormalities. We anticipated difficult ventilation in this child attributing to large head and retroganthia with propensity of airway obstruction and therefore preferred inhalational induction with sevoflurane, preserving the spontaneous breathing of the patient. After ensuring adequate mask ventilation, we decided for check laryngoscopy with Glidescope® without administering a muscle relaxant. GlideScope® videolaryngoscope is a contemporary system for tracheal intubation that employs a video camera enclosed into a plastic laryngoscope blade. We selected Glidescope® because it provides a superior view than obtained with a conventional laryngoscope.[13,24] Scaricaoglu et al encountered a difficult airway due to limited neck extension and intubated their patient with aid of guidewire.[1] Pandey R et al reported success to intubate their patient only in the fourth attempt.[5] Both the case reports had a child with craniofacial abnormalities similar to this child. We, therefore, did not use conventional laryngoscope in this case to avoid further complications. However, we were only able to visualize the posterior region of the glottic aperture (Cormack-Lehane grade 2B) with the Glidescope® even after applying optimum external laryngeal manipulation.We tried to maneuver the ETT into the laryngeal inlet using the dedicated stylet (Gliderite®) by Ronn M Wall technique[7] but were unsuccessful due to anteriorly placed larynx. Maneuvering the ETT towards the vocal cords is a known drawback of the Glidescope® despite using stylet owing to the acute curvature of the blade[15] and can even cause airway trauma.[16,17] We than planned for using flexible fiberscope to function as a rescue stylet without using its optics, as its distal tip can be easily manipulated. We mounted the ETT on the flexible fiberscope and directed the tip along the curvature of the Glidescope® blade and than through the vocal cords visualized on the Glidescope® screen.

On search of literature we found few reports successfully employing the same technique in adults, although it has seldom been practiced till now in pediatric group.[18-20] Fiberoptic intubation, as a sole airway device could have been the preferred option by many anesthesiologist in similar cases with difficult airway but it lacks the advantage of panoramic view of glottic aperture offered by Glidescope® which averts the problems such as airway trauma caused by the obtrusion of the ETT on the glottic structures during tube passage over the fiberscope.[21,22]

Lowe syndrome is the rare clinical entity with multisystem involvement. Anesthesia care providers should always have a clinical suspicion of difficult airway in these patients and should delineate a focused plan to manage the airway with multiple airway techniques and devices. In this case, the incorporation of flexible fiberscope as a rescue stylet provided an advantage of superior maneuverability for ETT insertion, along with the Glidescope® empowering better visualization of the glottis, hence avoiding trauma to the glottic structures.

REFERENCES

1. Saricaoglu F, Demirtas F, Aypar U. Preoperative and perioperative Management of a patient with Lowe syndrome diagnosed to have Fanconi's syndrome. Paediatr Anaesth 2004;14:530–532.
2. Loi M. Lowe syndrome. Orphanet J Rare Dis. 2006 May 18;1:16. [Medline]
3. Kim S.M., Lee J.H., Cho S.H., Kim S.H., Chae W.S., Jin H.C. Anesthetic management of a patient with Lowe syndrome accompanied by end stage renal disease: A case report. Anesth Pain Med. 2012;7:200-202. [KoreaMed]
4. Singh A.R., Singh J.R., Kaur H., Sachdeva G.S., Kaur A., Singh A.D. Lowe Syndrome Oculocerebrorenal Syndrome of Lowe): A Case Report of Two Brothers from India. Int J Hum Genet. 2002;2:243-249.
5. Pandey R, Garg R, Chakravarty C, Darlong V, Punj J, Chandralekha. Lowe's syndrome with Fanconi syndrome for ocular surgery: perioperative anesthetic considerations. J Clin Anesth. 2010;22:635-7. [Medline]
6. Ramanathan G, Patil S. A case of Lowe syndrome (oculocerebrorenal syndrome): Clinical implications and anaesthetic management. SAJAA. 2009;15:29-31. [AJOL]
7. Walls RM, Samuels-Kalow M, Perkins A. A new maneuver for endotracheal tube insertion during difficult GlideScope intubation. J Emerg Med. 2010 Jul;39(1):86-8. Epub 2010 Jan 25. [Medline]
8. Santos M.T.B.R., Watanabe M.M., Manzano F.S., Lopes C.H., Masiero D. Oculocerebrorenal Lowe syndrome: a literature review and two case reports. Spec Care Dentist. 2007;27:108-111. [Medline]
9. Harrison M, Odell W, Sheehy EC. Dental findings in Lowe syndrome. Pediatr Dent. 1999;21:425-28. [Medline]
10. Zhang X, Hartz PA, Philip E, ET al.Cell lines from kidney proximal tubules of a patient with Lowe syndrome lack OCRL inositol polyphosphatase and accumulate phosphatidylinositol 4, 5- biphosphatase. J Biol Chem.1998;273:1574- 82. [Medline]
11. Laube GF, Russell-Eggitt IM, van't Hoff WG. Early proximal tubular dysfunction in Lowe's syndrome. Arch Dis Child. 2004;89:479–80.
12. Lewis R.A., Nussbaum R.L., Brewer E.D. Lowe syndrome. 2001 [Updated 2012 Feb 23]. In: Pagon R.A., Bird T.D., Dolan C.R., et al., editors. Gene Reviews [Internet]. Seatle (WA): University of Washington, Seatle;1993.
13. Griesdale DE, Liu D, McKinney J, Choi PT. Glidescope® video-laryngoscopy versus direct laryngoscopy for endotracheal intubation: a systematic review and meta-analysis. Can J Anaesth. 2012;59:41-52. Epub 2011 Nov 1.
14. Su YC, Chen CC, Lee YK, Lee JY, Lin KJ. Comparison of video laryngoscopes with direct laryngoscopy for tracheal intubation: a meta-analysis of randomised trials. Eur J Anaesthesiol. 2011;28:788-95.
15. Doyle DJ: The GlideScope video laryngoscope. Anesthesia. 2005;60:414 –5
16. Cooper RM: Complications associated with the use of the GlideScope videolaryngoscope. Can J Anaesth. 2007;54:54–7.
17. Hsu WT, Hsu SC, Lee YL, Huang JS, Chen CL: Penetrating injury of the soft palate during GlideScope intubation. Anesth Analg. 2007;104:1609 –10
18. Moore MS, Wong AB. GlideScope intubation assisted by fiberoptic scope. Anesthesiology. 2007;106:885.
19. Morillas Sendín P, del Olmo Rodríguez C, de Diego Isasa P, Rouco Gil R. Combined use of the Glidescope and a fiberoptic bronchoscope in a case of unexpected difficult intubation. Rev Esp Anestesiol Reanim. 2008;55:454-5.
20. Sharma D, Kim LJ, Ghodke B. Successful airway management with combined use of Glidescope videolaryngoscope and fiberoptic bronchoscope in a patient with Cowden syndrome. Anesthesiology. 2010;113:253-5.
21. Johnson DM, From AM, Smith RB, From RP, Maktabi MA: Endoscopic study of mechanisms of failure of endotracheal tube advancement into the trachea during awake fiberoptic orotracheal intubation. Anesthesiology. 2005;102:910–4.
22. Maktabi MA, Hoffman H, Funk G, From RP: Laryngeal trauma during awake fiberoptic intubation. Anesth Analg. 2002;95:1112–4.

★★★★★