Difficult Airway Management in Pediatric Patient with Smith-Lemli-Opitz Syndrome

Praetel Claudia, MD, David A. Paulus, MD

Department of Anesthesiology, University of Florida College of Medicine, Gainesville, FL, USA

Introduction: Difficult airway management with various airway devices has been extensively described in the literature for children with congenital malformation syndromes and associated craniofacial abnormalities (1,2,3). We report a case of a pediatric patient with Smith-Lemli-Opitz Syndrome and multiple predictors for a difficult airway.

Report of a Case: A 4-year-old girl with Smith-Lemli-Opitz syndrome (SLOS) (Figure) presented for tympanostomy tubes and adenotonsillectomy the morning of surgery. She had uneventfully undergone 4 surgeries approximately 3 years ago. Clinical characteristics of this syndrome, caused by an abnormal cholesterol biosynthesis (deficiency of 7DHC-reductase with resultant low plasma cholesterol levels), were evident in this child: microcephaly with micrognathia, anteverted nares, low set ears, short first metacarpals, mental and growth retardation (body weight 10 kg). Her mother reported the presence of an abnormally small trachea, requiring a smaller sized ETT than predicted, bifid uvula, high arched palate, microgastria, and severely delayed gastric emptying. The child had food adversity and received tube feeds. Blepharoptosis and ambiguous genitalia had been surgically treated in the past. Premedication was not needed in this extremely playful and happy child. Intraoperative monitoring included ECG, noninvasive blood pressure, pulse oximetry and precordial stethoscope. The stomach was gently suctioned via gastric tube. She accepted the mask and we induced general anesthesia with sevoflurane, and nitrous oxide with oxygen. Intravenous access was obtained and adequate anesthetic depth was subsequently achieved with propofol. Mask ventilation proved easy without an oral airway. Despite two operators (senior resident and attending) and multiple blades (Wis-Hip 1.5, Mac 2, Mac 3 and Miller 2) we could not obtain a view of the vocal cords. Oxygen saturation (SpO₂) was maintained at 100% due to uncompromised mask ventilation. A blind intubation attempt around the epiglottis resulted in an esophageal intubation. The decision was made to use the pediatric fiberoptic Bullard laryngoscope. This was not successful because glottic structures could not be seen due to insufficient length of the pediatric Bullard laryngoscope in this patient. We returned to mask ventilation and then used the adult Bullard laryngoscope (Circon-ACMI) without the extender, which afforded the view of the epiglottis and with cephalad motioning we were able to intubate the trachea atraumatically with an uncuffed 4.0 ETT. A leak at 18-cm H₂O demonstrated a correct size in this particular patient. The remainder of the anesthetic went uneventfully. Upon completion of the procedure the child was extubated once fully awake.

Discussion: Although there are many alternative strategies to successfully manage a difficult airway, namely with a laryngeal mask airway (LMA) and sequential intubation, flexible fiberoptic, tube exchanger, light wand, retrograde intubation etc. we decided to initially perform a routine direct laryngoscopy to assess the particular anatomy in this child. Previous experience with older children with distinct craniofacial abnormalities had been quite encouraging. Despite the failure of the pediatric Bullard scope we felt that because of the size of this child’s airway with a particularly long, high arched palate we could utilize the added size of the adult Bullard scope. The anticipated small sized pediatric ETT (4.0) was no obstacle, because the adult Bullard will allow a 3.5 ETT to slide over the attached guide wire. In summary, this case report demonstrates the versatility and reliability of the adult Bullard laryngoscope for difficult intubations. Especially because the alternative approach (flexible fiberoptic bronchoscope) used in a series of 14 patients with SLO (4) requires a pediatric bronchoscope to fit the small ETT size, which may not be available at all institutions. This case also emphasizes the need for good preoperative evaluation and preparation, a specific plan with alternatives, avoidance of muscle relaxation until demonstration of adequate mask ventilation, and availability of a wide array of airway devices in children with congenital malformation syndromes affecting the airway.

References:

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