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CASE REPORT

Airway Management for Neonates with Beckwith-Wiedemann Syndrome (Bws): Case Report

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ABSTRACT

Introduction: Beckwith-Wiedemann syndrome (BWS) is a congenital disorder with abnormalities such as macroglossia and abdominal wall defect. Children with BWS predicted to have difficult airway due to macroglossia in perioperative airway management. Management : 4 days old male neonates with macroglossia on BWS is schedule for umbilical cord hernia revision. Patient was assessed as ASA 3 based on his physical status. First patient was pre-oxygenated and ventilation using size 1 mask, and after ventilation archived, induction started with 4 vol% Sevoflurane followed by intravenous agent Fentanyl 7.5 mcg and Atracurium. The initial intubation attempt with direct laryngoscope failed due to unclear visualization of vocal cord because obstruction of the tongue. The second attempt was made using VL, vocal folds visualized according to Cormack–Lehane grade IIb, and neonate bougie with a kinking, size 3 uncuffed ETT. Postoperatively, patient was extubated before admitted back to the NICU. Conclusion: Macroglossia causes anatomical airway abnormalities in children with BWS. It was predicted that might cause difficult ventilation, intubation and extubation. Airway management with tongue traction and awake tracheal intubation are reported to facilitate the intubation. Other emergency equipment including bougie, FOB, cricothyroidotomy set, and tracheostomy set needs to be ensured before anesthesia administration begins.

Keywords: Airway management, Beckwith-Wiedemann Syndrome, macroglossia, neonate

INTRODUCTION

Beckwith-Wiedemann syndrome (BWS) is a congenital abnormalities overgrowth involving a predisposition to tumor development with estimated incident 1 in 13.700 birth.[1] BWS feature with organomegaly, hemihypertrophy, macroglossia, hypoglycemia, abdominal wall defect, and tumorigenesis like nephroblastoma and hepatoblastoma.[2,3] Macroglossia itself causes of upper airway obstruction and difficult, and has been predicted with complicated perioperative anesthetic management. We report the perioperative airway management of a neonates with Beckwith-Wiedemann syndrome who underwent umbilical cord hernia revision.

CASE

A 4-day-old male neonate with Beckwith-Wiedemann syndrome who underwent umbilical cord hernia revision. Patient with respiratory distress due to early onset neonatal sepsis and airway obstruction due to macroglossia. Patient base ventilation with HFNC with flow 5 L/min and FiO2 21%, patient more often positioned prone or on their side. Through preoperative examination patient with history of chest wall retraction, and body temperature instability, with the highest temperature 38.7°C. Local examination revealed enlarged tongue, short muscular neck and receding chin, making it protruding and keeping the mouth permanently open, with the result difficult to assess mallampati score, and mouth opening. All other congenital abnormalities were looked for and ruled out by the pediatrician. Preoperative patient with 2 times prolong APPT, with minimal pericardial effusion from echocardiography.

Before patient transfer to the operating room, all the preparation for difficult ventilation/ intubation like video laryngoscope and fiber optic bronchoscopy were kept ready. to anticipate difficult airways in this patient, we have prepared several plans. Plan A: the patient will be induced using sevoflurane inhalation anesthesia, if ventilation cannot be carried out then the patient will be intubated using sleep non-apnea, but if ventilation can be controlled the induction will be given Fentanyl 1-2 mcg/ kgBW and muscle relaxant. The patient was then subjected to oral intubation using a direct laryngoscope with neonate bougie preparation. Plan B: Intubation using a video laryngoscope. Plan C: Intubation using fiberoptic bronchoscopy. In realization, patient was induced using sevoflurane 4 vol%, ventilation could be controlled using mask number 2 with the 2 person C-E ventilation technique and the tongue was pulled using gauze. Then intubation was assisted with Fentanyl 7.5 mcg and Atracurium 0.6 mg/kgBW intravenously. The first intubation attempt using a direct laryngoscope, the vocal cords were difficult to identify. Then the patient is revisualized using a video laryngoscope. The vocal folds were highly Glottic view was CL grade IIb, successful intubation was carried out using a neonate bougie and ETT number 3.5 uncuffed. After that, ETT was connected to a ventilator with pressure control settings with a target tidal volume of 6-8 ml/kg.

During intraoperative maintenance anesthesia using sevoflurane with a MAC target of 0.8 and intermittent Fentanyl. The operation lasted 1 hour 30 minutes with anesthesia duration of 2 hours 15 minutes. Because the operation wasn't carried out in the airway area and the hemodynamics were stable intraoperatively, the patient was planned to undergo a trial extubation to return the patient to the initial modality before surgery. The patient underwent awake extubation. After extubation the patient appeared stable, with minimal snoring, minimal chest retraction, and adequate chest expansion. After surgery, the patient was transported back to the NICU using a simple mask of 2 lpm and returned with HFNC flow of 5 liters per minute and FiO2 of 21%. Vital signs were found to be stable with a heart rate of 112 times per minute, respiratory rate of 35 times per minute and saturation of 97% with improvement in chest wall retractions.

DISCUSSION

Airway management in neonates is high-risk procedure, often in challenging and stressful situations. Even for elective surgery, it carries a higher risk of complications compared to older pediatric population.[4,5] Because of unique anatomical dan physiological airway in

neonates, obstruction is relatively common. Macroglossia is one of anatomical anomalies in supraglottic- oropharynx can cause upper airway obstruction and difficult airway management and it's commonly appeared in 80% cases with BWS. [3,5]

Macroglossia have been present since birth and tongue enlargement can be seen in terms of length and thickness. Microscopic and histochemical examination of tongue specimens from patients with Beckwith-Wiedemann syndrome did not reveal any tissue abnormalities other than hyperplasia and hypertrophy. Patient's oral cavity was found to be normal according to age so that only the tongue was enlarged.[6,7] Macroglossia causes upper airway obstruction and complicates mask ventilation and intubation procedures.[8] Because the tongue is larger than the oral cavity, patients with macroglossia have a higher prevalence of difficulty mask ventilation, difficult intubation which causes multiple attempts intubation and the high incidence of hypoxia. Therefore, it is important for anesthesiologist to understand airway management in neonate patients with Beckwith-Wiedemann syndrome.

To maintain airway patency during surgical procedures, tracheal intubation is required in all operations. Generally, the tongue can be removed during laryngoscopy for tracheal intubation.[7] If necessary, assistance by pulling and holding the tongue can be provided to facilitate the intubation procedure and improve ventilation by performing tongue traction maneuvers forward and downward.[6,7]

Deep anesthesia or muscle relaxant, will cause the tongue to fall into the retro lingual cavity.[7,8] Other research to overcome this problem is to inspect the awake vocal folds after intravenous sedation.[7] If the glottis can be visualized well with laryngoscopy then rapid induction can be carried out on the patient. Other case reports also suggested carrying out awake tracheal intubation or inspection of the awake vocal folds to visualize the glottis before intravenous induction or inhalation is given to the patient.[6] To aid this process, topical anesthesia may be considered. This method is reported to facilitate the endotracheal intubation process. Other reseacrh also recommend conducting a preoperative airway assessment in patients with Beckwith Wiedemann syndrome, one of which is by taking a lateral view neck x-ray. However, for best visualization, direct laryngoscopy or FOB laryngoscopy can be done under sedation before intubation is performed.[9]

Different from the other, research gave muscle relaxant, Rocuronium, after preoxygenation and ventilation. This administration was base on increasing difficulty in the intubation procedure without muscle relaxants, with the preparation of Sugammadex as a reversal agent for muscle relaxants if intubation was unsuccessful. Muscle relaxants also help reduce the incidence of laryngospasm during laryngoscopy.[10]

In this case, muscle relaxant was carried out during the induction process. This is in line with the aim to reduce resistance while intubation. The muscle relaxant given based on ventilation that can be controlled.[10] Mask ventilation and preoxygenation difficulties have been reported, where insertion of an oropharyngeal airway (OPA) was required to maintain adequate ventilation.[7] In other reports, a nasopharyngeal airway (NPA) fashioned from an endotracheal tube (ETT) was used to relieve airway obstruction during ventilation under anesthesia.6 Besides reducing obstruction, an ETT-based NPA can be connected to the breathing circuit to facilitate oxygen delivery and administration of inhalational anesthetics during attempts at intubation .

In the present case, neither an NPA nor an OPA was inserted after induction because ventilation and oxygenation were adequately maintained using a size 2 face mask combined with the tongue-pull technique. However, NPA placement remains a useful alternative when mask ventilation becomes difficult. In managing the airway of pediatric patients with macroglossia due to Beckwith-Wiedemann syndrome, preparation of other tools such as bougie, pediatric fiberoptic bronchoscopy (FOB), laryngeal mask airway (LMA), and various mask sizes need to be prepared for anticipation. Preparation for cricothyroidotomy and tracheostomy procedures before anesthesia

procedure as a last resort if the airway cannot be achieved by intubation or the use of other assistive devices. In this case report, extubation was carried out immediately after surgery, this was based on the consideration that the operation was not in the airway area and the patient had the initial modality of spontaneous breathing with HFNC. Immediate postoperative extubation is considered to have advantages in terms of reducing damage to laryngeal structures, preventing aspiration, and reducing the duration of stay in the NICU.[13]

CONCLUSION

Airway management in the neonates with Beckwith-Wiedemann syndrome is a high-risk procedure and it is often performed under pressure. In addition, prepare techniques to help manage the airway such as tongue traction, awake tracheal intubation, use of a bougie, video laryngoscope, and fiberoptic bronchoscopy, we should also prepare for crash airway situation with cricothyroidotomy and tracheostomy. This situation is best managed by a multi disciplinary team with approach to training, equipment, planning and management will help to reduce errors, morbidity and stress for those involved.

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