Anesthetic management of two cases of Beckwith-Wiedemann syndrome

Yoshinobu Kimura[1], Yasuhiro Kamada[2], and Saori Kimura[3]

[1] Department of Anesthesia, Takikawa Municipal Hospital, 2-34-2 Omachi, Takikawa 073-0022, Japan
[2] Department of Anesthesia, Nikko Memorial Hospital, Muroran, Japan
[3] Department of Anesthesiology, Sapporo Medical University, Sapporo, Japan

Abstract
Two cases of children with Beckwith-Wiedemann syndrome are presented. This syndrome consists of various abnormalities, including macroglossia, visceromegaly, omphalocele, and gigantism. These abnormalities frequently require operative correction during the neonatal period. We anesthesiologists should make plans for difficult airway managements in patients with this syndrome. Our two patients also showed larger sized tracheas than those estimated by their age and height. A cuffed tube, though still controversial, has recently been used in children. We recommend using a cuffed tube in patients with this syndrome, because the appropriate tracheal size may not be predictable, tracheal intubation might be difficult, and risks incurred during changing of a tracheal tube should be avoided.

Key words Beckwith-Wiedemann syndrome · Cuffed tracheal tube · Airway management

Introduction

Beckwith-Wiedemann syndrome (BWS) is characterized by macroglossia, omphalocele, visceromegaly, gigantism, and hypoglycemia, and the incidence of the syndrome is reported as approximately 1 in 13 700 live births [1,2]. We anethesiologists should consider several problems that arise in planning the anesthetic management of patients with this syndrome. Macroglossia causes upper airway stenosis and difficulty in tracheal intubation, and visceromegaly shifts the diaphragm upward, which shortens the distance from the lips to the tracheal carina and reduces the functional residual capacity of the lungs. We managed two patients with this syndrome whose tracheas were larger than the sizes estimated by their age and height. We recommend the use of a cuffed tracheal tube (CTT) rather than an uncuffed tracheal tube (UTT) for children with BWS.

Case reports

Case 1

A 4190-g male infant was born by vaginal delivery at week 37. Apgar scores at 1 and 5 min were 4 and 9, respectively, due to neonatal asphyxia because of shoulder dystocia. Neonatal asphyxia was immediately solved with bag-and-mask ventilation. Abnormalities shown at birth were macroglossia, omphalocele, hypoglycemia, and bilateral renomegaly. His serum glucose level was 47 mg·dl^{-1} at 1 h after birth. He was diagnosed with BWS, without chromosome banding analysis. The omphalocele was repaired at birth and hypoglycemia was treated with a continuous infusion of 10% glucose in water.

At 14 months of age, he was scheduled for surgery for right inguinal hernia incarceration. His height was 73.2 cm and body weight was 12 kg. He did not receive any sedation before surgery. In the operating room, electrocardiogram, pulse oximetry, and noninvasive arterial blood pressure were monitored and peripheral venous access was established. His serum glucose level was 94 mg·dl^{-1}. After an assessment was made that there would be no difficulty in tracheal intubation by laryngoscopy in the awake state, anesthesia was induced with thiamylal sodium 50 mg and vecuronium 1 mg. Although bag-and-mask ventilation was difficult due to the enlarged tongue, tracheal intubation using a 4.0-mm UTT was performed smoothly, but the dimension of the UTT was too small for adequate ventilation. The tracheal tube was exchanged for a 4.5-mm UTT, but a large leakage was still found. Finally, re-intubation using a 5.0-mm UTT provided appropriate ventilation and leakage, with 20 cm H_2O airway pressure and the patient's head in a neutral position. Mild hypoxia

(peripheral oxygen saturation [$S_{P_{O_2}}$], 86%) was observed during the re-intubation procedure. The tip of the tube was placed 2 cm from the vocal cords. Anesthesia was maintained with sevoflurane 2.5% and an air-oxygen mixture (fraction of inspired oxygen [$F_{I_{O_2}}$], 0.35). No other hypoxemic episode was observed during the surgery. Plasma glucose level was kept within the normal range without any interventions. At the conclusion of the surgery, the patient was extubated smoothly and we did not require a nasal airway which had been prepared for the narrow pharynx caused by macroglossia. The patient was observed for 30 min before being taken out of the operating room. Neither remarkable changes nor complications were seen postoperatively.

Case 2

A 4226-g full-term male infant was born by vaginal delivery after an uncomplicated prenatal course. Apgar scores at 1 and 5 min were 9 and 9, respectively. At birth, omphalocele and macroglossia were found. The patient had neither visceromegaly nor tracheal abnormality. The patient, diagnosed with BWS at 3 months, underwent radical surgery for omphalocele at age 8 months at another hospital, but the record regarding the surgery was insufficient.

Plastic surgery for the omphalocele was scheduled at age 7 years. All routine tests showed results within normal ranges. Neither echocardiogram nor electrocardiogram showed any abnormalities. The patient's height was 136 cm (+2 SD), which was greater than the Japanese average height for the patient's age, and his body weight was 29.5 kg. The patient received no sedation before surgery. In the operating room, electrocardiogram, pulse oximetry, and noninvasive arterial blood pressure were monitored and peripheral venous access was established. Because macroglossia, and sleep apnea symptoms, had already been resolved, anesthesia was induced with propofol 50 mg and vecuronium 3 mg, with the preparation of a nasal airway. Ventilation with bag and mask was performed smoothly. The patient was intubated easily with a 6.0 mm UTT after sufficient muscle relaxation. Air leakage was excessive with various head positions, and was not changed with a 6.5 mm UTT. Changing to a 6.0 mm CTT provided adequate ventilation, with its cuff inflated by 1 ml, and appropriate leakage, with 20 cmH$_2$O airway pressure and the patient's head in a neutral position. The cuff was placed 0.5 cm below the cricoid level. The surgery was performed under general anesthesia maintained with sevoflurane 2% and an air-oxygen mixture ($F_{I_{O_2}}$, 0.30). No hypoxemia was seen during the surgery, which lasted for approximately 2 h. At the conclusion of the surgery, anesthesia was terminated and the patient was extubated smoothly after the reversal of muscle relaxants.

No airway obstruction was observed. No complications occurred during the postoperative course and the patient was discharged 2 days after the operation.

Discussion

BWS presents with variable anomalies which include omphalocele, macroglossia, visceromegaly, and severe congenital cardiac defects [1,2]. Hypoglycemia is often seen up to age 4 months. Surgery for these abnormalities is often indicated at an early stage following birth. Difficult airway management is one of the most important problems for anesthesiologists and there are no reports regarding the airway diameter in children with this syndrome.

The most common method for estimating appropriate UTT sizes for children is the modified Cole's formula (MCF): ID in mm = (age in years/4) + 4 [3,4] but it has been reported that this rule is not applicable for children under 2 years of age [5]. In contrast is the length-based method of Broselow's resuscitation tape (BRT), which was developed in 1992 to help resuscitators estimate dosages of resuscitation drugs and choose appropriate equipment [5,6]. BRT was found to be accurate in 77% of the cases versus 47% for MCF [6]. Daugherty et al. [7] have claimed that BRT is as accurate as MCF in both normal height and pathologically short children. Other proposed methods include estimates based on weight, fifth finger width or diameter, fifth finger nail width, diameter of the distal phalanx of the third finger, and arm span [5,8–11]. However, the outer diameter of tracheal tubes varies among the different brands and manufacturers and we cannot use these formulas automatically [12]. We used tubes Portex made by Smiths Medical Japan, Tokyo, Japan. Some studies state that BRT tends to underestimate ETT size and MCF overestimates the size [7,13].

Tracheal tube sizes calculated by length-based BRT and by MCF, and the tube sizes finally used in our two patients are shown in Table 1. These calculated values were underestimated partly because this syndrome involves gigantism. These models are derived from populations of healthy North American children based on the normal fiftieth percentiles for these parameters [6,8–11]. In addition, the length-based calculation could also be used in Korean children as a helpful adjunct [14].

In recent years, CTTs have been used in children because they have various advantages, such as a reduced tube exchange rate, sealed airway without the use of an oversized tube, reliable lung function monitoring and capnography, and reduced contamination of the operating room environment with anesthetic gas [15]. The

Table 1. Uncuffed tracheal tube (UTT) sizes used compared with predictions using the modified Cole's formula (MCF) and length-based Broselow's resuscitation tape (BRT)

	Age (years)	Height (cm)	Weight (kg)	MCF (mm)	BRT (mm)	Size used (mm)
Patient 1	1.2	73.2	12	4	4.3	UTT 5
Patient 2	7	136	29.5	6	5.75	CTT 6

incidence of multiple intubation attempts was approximately 19 times greater with a UTT than with a CTT [16]. Using a CTT is regarded as one way to avoid risky exchange or re-intubation in patients such as those with BWS who have a risk of technical difficulties with intubation.

In children with BWS, because visceromegaly shifts the lungs upward from a normal position, the possibility of bronchial intubation should be recognized. Evaluation of the appropriate length of tracheal tubes from the lips is difficult in these patients, but a CTT is useful for confirming an accurate position by palpating the cuff externally [17]. Special care should be taken with the adjustment of cuff pressure and it should be noted that some of the tube cuffs are poorly designed; being based on adult tracheal tubes, they are too long to be placed out of the larynx in children [12,15]. It has been said that cuffed tube intubation is not associated with an increased risk of postintubation stridor [18]. For pediatric basic and advanced life support it has also been confirmed that there are some advantages of CTTs over UTTs in children [19]. In children with BWS, where changing the tracheal tube is considered dangerous, one method is to choose a CTT using the Motoyama formula [20]: ID = (age/4) + 3.5 or the Khine formula [16]: ID = (age/4) + 3 from the start. We changed to a 6.0-mm CTT based upon the widely accepted assumption that a CTT would be approximately 0.5 mm larger in diameter than a UTT [6,7].

With only two cases experienced, it is difficult to determine whether or not the enlarged tracheas were simply an anomaly related to BWS; further studies are required.

In conclusion, we have described the airway management of two children with BWS. We recommend using a CTT in patients with BWS, because the appropriate tracheal size can not be predicted, tracheal intubation might be difficult, and risks incurred during changing of the tracheal tube should be avoided.

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