

The Anesthetic Management of a Case of Beckwith-Weidemann Syndrome Presenting With a Large Tongue

Beckwith-Weidemann Sendromlu Büyük Dili Olan Vakada Anestezi Yönetimi

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Abstract

The Beckwith-Wiedemann syndrome is characterized by macroglossia; pre- or postnatal gigantism; abdominal wall defects; hypoglycaemia; umbilical hernia or diastasis recti; ear creases or posterior helical ear pits; facial nevus flammeus; renal anomalies; neoplasms; hemihypertrophy; congenital cardiac malformations; intestinal malrotation; mental retardation; polydactyly; and cleft palate. Anesthetic considerations of 7 months old girl with Beckwith-Wiedemann syndrome during reduction of macroglossia are presented and discussed.

Key words: **Anesthesia; Beckwith-Wiedemann syndrome; Macroglossia.**

Özet

Beckwith-Weidemann sendromu makroglossi; pre veya postnatal jigantizm; abdominal duvar defekti; hipoglisemi; umbilikal herni veya diastazis rekti; kulak keçesinde kıvrılmalar veya posterior helical pitler; fasial nevus flammeus; renal anomaliler; neoplasmlar; hemihipertrofi; konjenital kardiyak malformasyonlar; intestinal malrotasyon; mental retardasyon; polidaktili ve yarık damak ile karakterizedir. Bu yazıda Beckwith-Weidemann sendrom'lu, 7 aylık kız çocukta, makroglossi küçültme operasyonu için verilen anestezi uygulaması bildirilmiştir.

Anahtar Kelimeler: **Anestezi; Beckwith-Wiedemann sendromu; Makroglossi.**

Introduction

The Beckwith-Wiedemann syndrome is the most well-known and most common of the overgrowth syndromes which are less common than syndromes with growth deficiency. An early estimate of birth prevalence of syndrome was 1 in 13,700, although more recent estimates have suggested 1 in 12,000. The molecular basis of Beckwith-Wiedemann syndrome is complex, involving deregulation of imprinted genes found in 2 domains within the 11p15 region (1).

The Beckwith-Wiedemann syndrome is characterized by macroglossia; pre- or postnatal gigantism; abdominal wall defects such as omphalocele, umbilical hernia or diastasis recti; ear creases or posterior helical ear pits; facial nevus flammeus; renal anomalies such as nephromegaly, multiple calyceal cysts or hydronephrosis; hemihypertrophy; congenital cardiac malformations; intestinal malrotation; polydactyly; and cleft palate. Neoplasms consist of Wilms tumor, hepatoblastoma, adrenocortical tumor and rhabdomyosarcoma, may occur during the early childhood years. Rarely, mental retardation has been reported (2). Severe neonatal hypoglycaemia may be present due to hyperinsulinism (3).

The abdominal wall defects, malignancies, cleft palate and macroglossia seen in Beckwith-Wiedemann syndrome require surgical interventions. The anaesthetic implications of this disease relate to airway obstruction as a result of macroglossia, circulatory and hemodynamic problems due to congenital cardiac malformations and perioperative hypoglycaemia because of hyperinsulinism.

Case report

The girl was delivered to a 32-year-old mother at 34 week by cesarean section after an uncomplicated prenatal course with a birth weight of 2490 g. Both parents were healthy and there was no family history of medical disorders. At birth, the infant was noted to have an omphalocele, large open fontanels, hypoglycemia and macroglossia. Her serum glucose level was 49 mg/dl at 2 h after birth. An echocardiogram revealed ventricular septal defect and aneurysmatic interatrial septum. She was diagnosed with Beckwith-Wiedemann syndrome, without chromosome banding analysis. Hypoglycemia was treated with a continuous infusion of 10% dextrose solution. The omphalocele was surgically repaired at 1 day after birth.

At 7 months of age, the patient was scheduled for reduction of macroglossia under general anesthesia. She was 72 cm

tall (between 50th-75th percentile) and weighed 8300 g (between 25th-50th percentile). Physical examination revealed a large protruding tongue (Picture 1). Other findings of physical examination were unremarkable and all routine laboratory tests, including serum glucose levels, were within normal ranges. Neither chest X-ray nor electrocardiogram showed any abnormalities. The patient was not pre-medicated; an intravenous infusion of 1/3 saline-dextrose solution was given during the fasting period. In the operating room, electrocardiogram, pulse oximetry and noninvasive arterial blood pressure were monitored. Following intravenous sedation with 2 mg/kg of thiopental sodium, easy visualization of the glottis was assured by laryngoscopy. Afterwards, anesthesia was induced with another 2 mg/kg of thiopental sodium, 2 µg/kg of fentanyl and 1 mg/kg of succinylcholine. The mask ventilation of the patient was not easy so an oropharyngeal airway was inserted for adequate ventilation. Nasotracheal intubation was performed with 4.5 mm tracheal cuffed tube with the help of an assistant who hold the tongue with a clamp. Anesthesia was maintained with 70% nitrous oxide in oxygen and 2% sevoflurane and further muscle paralysis was maintained by 0.1 mg/kg of vecuronium. No remarkable hemodynamic changes were observed throughout the surgical procedure. Plasma glucose levels were measured throughout the operation at 30 minutes intervals. At the end of the surgery, anesthesia was terminated and the patient was extubated smoothly after the reversal of muscle relaxants. No airway obstruction was observed. The patient was discharged 3 days after the operation without any complication occurred during the postoperative course.



Picture 1. Macroglossia and nasal endotracheal intubation.

Discussion

Macroglossia is the most common feature of Beckwith-Wiedemann syndrome (2). Macroglossia is typically present at birth and involves increased length and thickness of the tongue. Depending upon the degree of severity, macroglossia can lead to complications involving feeding and respiration in infancy. Later, macroglossia can impede speech articulation and lead to malocclusion of the mandible (4). The primary cause of distress during anaesthesia for patients with Beckwith-Wiedemann syndrome is the large, protuberant tongue. The key to airway management in patients with macroglossia is not to attempt mask ventilation which requires keeping the tongue in the mouth. Tracheal intubation is required in all cases, and nasotracheal intubation is required for glossectomy. Most of the tongue can be moved out of the way during laryngoscopy for tracheal intubation, although it may be necessary for an assistant to help hold the tongue (5). The administration of high concentration intravenous anaesthetics or muscle relaxant to the patient may cause the tongue to fall into the retrolingual space. Thus, in the case reported here, following intravenous sedation, awake vocal cord inspection was attempted and after the visualization of the glottis was assured by laryngoscopy, rapid induction was applied. Although bag/mask ventilation was not found to be easy, endotracheal intubation was performed without difficulty.

In the present case we preferred to use cuffed tracheal tube for endotracheal intubation although the patient was 7 months old. For many years, tracheal tubes without a cuff were routinely recommended for children. This was based on the anatomy of the infant and small child's airway. Another concern in children was that the cuff might cause damage at the cricoid ring and, because this is a complete ring of cartilage, even a small amount of edema will result in significant narrowing and very large increases in resistance to flow. This latter concern was largely based on experience with older type tubes that used high-pressure cuffs made from irritant materials. Modern implant-tested tubes with high-volume, low-pressure cuffs have much less propensity to cause harm (6). Also, authors recommend using a cuffed tube in patients with Beckwith-Wiedemann 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 (7).

In the immediate postoperative period after glossectomy, the tongue can swell to greater than the preoperative size,

so tracheal intubation may be continued for several days after surgery, until swelling subsides (5). However some anaesthetists extubate trachea just after surgery and report that a nasopharyngeal airway is useful in relieving postoperative airway obstruction (8). In the presented case, the patient was extubated just after the surgery because the tongue was not swelled and it was smaller after glossectomy when compared with the preoperative size. Although we did not use a nasopharyngeal airway, a respiratory complication including postoperative airway obstruction was not observed.

Beckwith-Wiedemann syndrome is an uncommon cause of hyperinsulinaemic hypoglycaemia in the neonatal period. The hypoglycaemia, although usually mild or asymptomatic, may be severe, requiring medical an occasionally surgical treatment (9). In the presented case, although the preoperative, glucose values were in the normal ranges, for prevention from hypoglycaemia, intravenous solution with glucose was infused throughout the operation. Plasma glucose levels were measured throughout the operation at 30 minutes intervals and at 2 hours intervals postoperatively in the first 24 hours.

In conclusion, we have described the anesthetic management of a child with Beckwith-Wiedemann syndrome and discussed the anesthetic problems associated with the disease. Difficulties in endotracheal intubation due to macroglossia and hypoglycaemia during the perioperative period are critical problems for anaesthetists.

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