Anesthetic management of tongue reduction in a case of Beckwith-Wiedemann syndrome

Meenu Batra, Umesh K. Valecha
Departments of Anesthesiology and Critical Care, BLK Superspeciality Hospital, New Delhi, India

Abstract
Anesthesia for partial glossectomy in a premature child with Beckwith-Wiedemann syndrome presents as a unique challenge to the Anesthesiologist. Airway management in patients presenting with macroglossia is especially significant and requires meticulous preparation and pre-operative assessment. This report delineates the anesthetic concerns such as an anticipated difficult airway due to a large tongue, prematurity, hypoglycemia and an oral cavity surgery and their management.

Key words: Anesthesia, Beckwith-Wiedemann syndrome, hypoglycemia, macroglossia, prematurity

Introduction
Beckwith-Wiedemann syndrome (BWS) is a congenital disorder which comprises a wide constellation of features namely macrosomia, macroglossia, abdominal wall defects (exomphalos/omphalocele, umbilical hernia and diastasis recti), ear creases or pits, hemihypertrophy (of one side of the body) and neonatal hypoglycemia. Furthermore, associated are nevus flammeus, hepatomegaly, nephromegaly, exophthalmos, embryonic tumors, psychomotor retardation, microcephaly, cardiac anomalies, musculoskeletal abnormalities and hearing loss.[1] This entity was previously known as Exomphalos-Macroglossia-Gigantism syndrome.

Cases are mostly sporadic, but may have an autosomal dominant transmission with incidence of 1:15,000 births. About 1% of cases may demonstrate chromosomal abnormalities at 11p15.5, 5q35.[2]

Patients with BWS may require surgery for omphalocele, inguinal hernias, macroglossia and less often for correction of cleft lip and palate.[3]

Anesthetic considerations in these patients include risks and concerns of prematurity, difficult airway due to craniofacial involvement and macroglossia and systemic co-morbidities such as hypoglycemia and cardiopathies (atrial and ventricular septal defects, patent ductus arteriosus, tetralogy of Fallot, left ventricular hypoplasia and idiopathic cardiomegaly).

We hereby present a case of BWS with macroglossia undergoing tongue reduction surgery.

Case Report
We describe a male child born to a gravida seven para two live births two abortions four mother through a non-consanguineous marriage at 32 weeks of gestation with a birth weight of 3.2 kg. The baby cried immediately after birth. This neonate was diagnosed with BWS in view of large for gestational age, macroglossia, umbilical hernia and hypoglycemia. There was no history of a similar illness in the family.

The child was kept in the nursery for 15 days during which he underwent various investigations. Ultrasonography of cranium was normal while that of abdomen demonstrated mild hepatomegaly and right nephromegaly. Echocardiography confirmed normal cardiac findings.

At 2 months of age and weight of 4 kg, the mother complained of difficulty in feeding and multiple episodes of seizures for which the infant was admitted to pediatric intensive care unit (PICU). His vital signs were normal. In addition to the physical findings at birth, microcephaly and right hemihypertrophy were noted. On central nervous system (CNS) examination,
child was irritable with normal tone and power. Routine blood work illustrated hypoglycemia to the level of 41 mg% for which intravenous (IV) glucose infusion in the form of N/2 + 12.5% dextrose + 1:100 potassium chloride (KCl) was administered @ 16 ml/h. Random blood sugars were monitored every hour for 24 h followed by 2 h screening. The blood sugar level remained less than 50 mg% with the above mentioned concentration. Hence, the glucose infusion was increased to 20% dextrose in N/2 with 1:100 KCl @ 16 ml/h. The blood sugar levels rose to 50-100 mg%. Nasogastric feeding was also started at this time. Once the child started to accept feeds well, 40-60 ml milk was administered through the nasogastric tube with 10 g sugar added to it. At this time, the IV infusion rate was gradually reduced from 16 ml/h to 4 ml/h over 4 days and finally discontinued as the child maintained blood sugar levels between 100 and 150 mg%. CNS irritability decreased over 2 days. There was no recurrence of seizures, hence, attributing the initial seizures to hypoglycemia. The child did not show any spike of temperature which remained between 98°F and 98.6°F. Arterial blood gases were also monitored daily during the stay in PICU with pH in the range of 7.36-7.44, pO2 of 90-140 mmHg, pCO2 of 35-40 mmHg and no evidence of metabolic acidosis or alkalosis. The serum sodium remained in the range of 135-140 mmol/L, serum potassium of 3.5-4.2 mmol/L and serum chloride of 100-105 mmol/L.

To improve oral feeding, a subtotal glossectomy was planned for the patient at 11 weeks and 3 days of age. After 6 h of nil per oral status, patient was taken in a pre-warmed operation theatre and covered with a warming blanket. Routine non-invasive monitors were applied in the form of electrocardiogram, non-invasive blood pressure, pulse oximetry (SpO2). A 24G IV cannula was already in situ for elective ventilation. Airway equipment such as size 2.0-3.5 mm ID uncuffed endotracheal tubes (ETT), curved and straight laryngoscope blades, Guedel airways of multiple sizes, pediatric bougie, pediatric flexible fiberoptic bronchoscope, laryngeal mask airway and various face masks were kept ready to handle the anticipated difficult airway.

Nasogastric tube which was already in place was aspirated for any residual feed. Pre-oxygenation was done using a size 2 face mask to accommodate the large protruding tongue. Induction was performed with increasing concentration of sevoflurane with 100% oxygen followed by direct laryngoscopy with curved blade #2. Glottis was easily visualized and intubation was tried with a size 3 uncuffed ETT through the nose. Failure to negotiate this tube through the vocal cords led to tracheal intubation with a size 2.5 ETT. Muscle relaxation was achieved with atracurium 1 mg and 0.5 mg top-ups as and when required. The throat was packed with saline soaked guaze. Anesthesia was maintained with 50% oxygen with nitrous oxide and sevoflurane to maintain a minimum alveolar concentration of 1.0-1.3. Ventilation was achieved through pressure control mode (PCV) at inspiratory pressure ($P_{\text{insp}}$) of 10-12 cm H2O and respiratory rate (RR) of 22-23/min to attain a tidal volume of 30-40 ml with end tidal carbon dioxide maintained in the range of 37-42 mmHg.

Dexmethylone 0.2 mg/kg was administered to reduce surgery related airway edema. Analgesia was attained with paracetamol suppository 80 mg per rectal and IV fentanyl 2 mcg as per requirement.

Intraoperative vitals and blood sugar levels were within the normal limits in the range of 145-215 mg%. Anterior two-third wedge resection of the tongue was done and the estimated blood loss was approximately 10 ml. The procedure was uneventful and the patient was shifted to the PICU with ETT in situ for elective ventilation.

The child was put on PCV mode with inspired oxygen concentration ($\text{FiO}_2$) of 0.5, inspiratory pressure ($P_{\text{insp}}$) 20 cm H2O, positive end expiratory pressure 5 cm H2O, maximum airway pressure ($P_{\text{max}}$) 30 cm H2O, RR 22/min, inspiratory expiratory ratio 1:2. Sedation was maintained with fentanyl and midazolam. After the 1st post-operative day, sedation was tapered and patient was put on pressure support ventilation. $\text{FiO}_2$ and pressure support were gradually reduced and trachea was extubated on the 3rd post-operative day. A test dose of 60 ml blood was transfused on the post-operative day 2 for a hemoglobin<7 g%. Vital signs remained stable in the post-operative period. Glycemic control was achieved by 2 h blood sugar monitoring and continuation of IV glucose infusion.

**Discussion**

BWS is an overgrowth disorder, classically characterized by omphalocele, macroglossia and gigantism along with a myriad of anatomical and metabolic abnormalities.\(^{[1]}\) It has an incidence rate of 1: 15,000 with 85% of cases being sporadic and a demonstrable karyotype abnormality on chromosome 11 in fewer than 1% cases.\(^{[2]}\)

BWS also increases the risk of prematurity by 50% and its associated complications.\(^{[4]}\) Macroglossia along with anterior abdominal wall defects constitute the most common features of BWS.\(^{[5]}\)
Macroglossia causes major functional problems in early infancy. The enlarged tongue interferes with breast feeding, causes airway obstruction and sialorrhea. Limitation in tongue movement leads to impaired speech and dental alveolar protrusion. These have major developmental and psychological impact. Hence, it becomes imperative that the tongue size be reduced surgically during the 1st year of life.

A patient of BWS presents as an anesthetic management challenge. The areas of concern are prematurity, macroglossia, hypoglycemia and associated cardiac defects, if any.[6]

The age of prematurity directly influences the procedure related risk. Macroglossia in particular may cause difficulty in airway management. Therefore, a thorough pre-operative assessment of the airway is essential. A neck X-ray lateral view may be helpful but the best assessment in this regard is by direct laryngoscopy or fiberoptic laryngoscopy under sedation. It is mandatory that all difficult airway apparatus including cricothyrodotomy and tracheostomy sets be available before initiation of anesthesia.

Naso-tracheal intubation is the preferred technique of airway management for glossectomy as was used in our case.[7] We used an uncuffed endotracheal tube and packed the pharynx with saline soaked gauze, although cuffed tubes have also been used for such cases.[8] Tracheal intubation is preferred under sedation with inhalational agents while the patient is breathing spontaneously as in our case. Failure to negotiate size 3 ETT could have been due to a hypertrophied subglottis as in this hypertrophy disorder or a narrow subglottis in accordance with the age of prematurity of this child.

In surgeries of the oral cavity, it is our preference and also recommended in the literature to electively ventilate these patients for at least 24 h in view of any unanticipated bleed and airway edema.[6] Furthermore, in our case the child was pre-term with post-gestational age being only 43.5 weeks, which may have increased risk of post-operative apnea.

A vigilant blood sugar monitoring can avoid episodes of hypoglycemia and consequent neurological sequelae in these patients.[9]

Management of BWS patients is a multi-disciplinary effort including psychological and genetic counseling along with long-term follow-up and rehabilitation. A patient of BWS can be successfully managed by thorough pre-operative planning and awareness of the possible complications.

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