Case Report

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Tailored Anaesthesia in Paediatric Beckwith-Wiedemann Syndrome: A Case Report

Anaesthesia Section

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ABSTRACT

Beckwith-Wiedemann Syndrome (BWS) is a complex overgrowth disorder characterised primarily by macroglossia, omphalocele, and gigantism, along with a range of anatomical and metabolic abnormalities such as neonatal hypoglycaemia, embryonal tumours, visceromegaly, naevus flammeus, and anal dimple. Airway management in paediatric patients poses a significant challenge for anaesthesiologists. Anaesthetic management in patients with BWS is demanding because macroglossia may cause difficult mask ventilation, intubation, and extubation. These patients often require repeated anaesthetic exposure for the correction of macroglossia and associated abnormalities, and their perioperative management can be taxing for anaesthesiologists. Therefore, the anaesthetic management of children with BWS is particularly challenging due to the combination of age-related physiological limitations and disease-specific abnormalities. Here, the authors present the case of a five-month-old female infant diagnosed with BWS, with macroglossia, naevus flammeus, and an anal dimple, who underwent surgical reduction of macroglossia. The patient also demonstrated additional systemic findings including non-obstructing hydrocephalus, syringohydromyelia, hepatosplenomegaly, and enlarged kidneys on imaging, further increasing the complexity of anaesthetic care. Given the likelihood of a difficult airway, comprehensive preoperative evaluation, meticulous preparation, and well-structured backup strategies for airway management, endotracheal intubation, and intraoperative haemodynamic management are critical to navigating the anaesthetic complexities in patients with BWS. In addition, vigilant postoperative monitoring is essential due to the risk of airway obstruction and hypoglycaemia. The present case underscores the importance of multidisciplinary coordination and individualised anaesthetic planning to ensure safe perioperative outcomes.

Keywords: Airway management, Difficult airway, Macroglossia, Paediatric anaesthesia

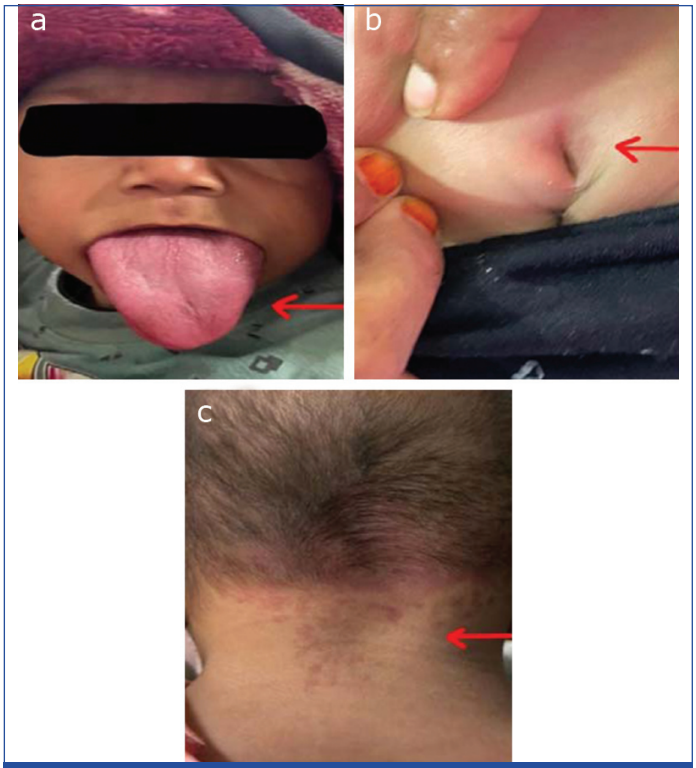
CASE REPORT

The authors describe a five-month-old female infant, born to a gravida 3, para 2, live births 2 mother through a non-consanguineous marriage at 38 weeks of gestation via normal vaginal delivery, with a birth weight of 3.6 kg. The baby cried immediately after birth. She was diagnosed with BWS on the basis of macroglossia [Table/Fig-1a], an anal dimple [Table/Fig-1b], and naevus flammeus [Table/Fig-1c]. Earlobe creases and pits were absent. There was no family history of a similar illness.

At 20 days of age, the mother reported difficulty in feeding and a history of snoring, for which the neonate was evaluated. Genetic testing was normal. In 10-15% of clinically diagnosed BWS cases, genetic testing may be normal because mosaicism can limit detection (the abnormality may be present only in some tissues and not in blood, where testing is usually performed), and current molecular methods may not detect subtle or rare abnormalities.

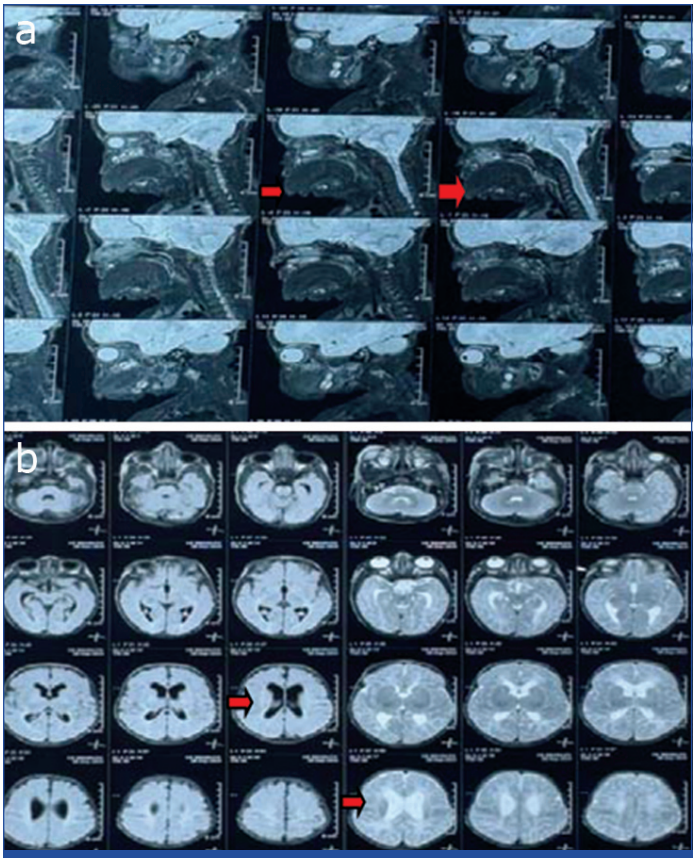
Contrast-Enhanced Magnetic Resonance Imaging (CE-MRI) of the face demonstrated macroglossia [Table/Fig-2a]. On clinical examination, head size was normal, while CE-MRI of the brain revealed non-obstructing hydrocephalus [Table/Fig-2b]. CE-MRI of the whole spine showed syringohydromyelia, along with enlarged kidneys and hepatosplenomegaly. Echocardiography confirmed normal cardiac findings.

There was no history of neonatal pneumonia, jaundice, seizures, cyanotic spells, blood transfusion, drug allergy, or recent upper respiratory tract infection. Immunisation was complete for age, and developmental milestones were appropriate. Airway examination revealed an enlarged, protruding tongue with inability to approximate the lips. On chest auscultation, air entry was equal bilaterally, and heart sounds were normal. Haematological and biochemical investigations were within normal limits.



[Table/Fig-1]: Image depicting a BWS child with notable clinical features including a) Macroglossia b) an anal dimple; c) nevus flammeus.

Informed written high-risk consent was obtained, including potential complications such as significant bleeding, postoperative airway obstruction due to swelling, injury to nerves controlling tongue sensation and movement, and the possible need for mechanical ventilation.



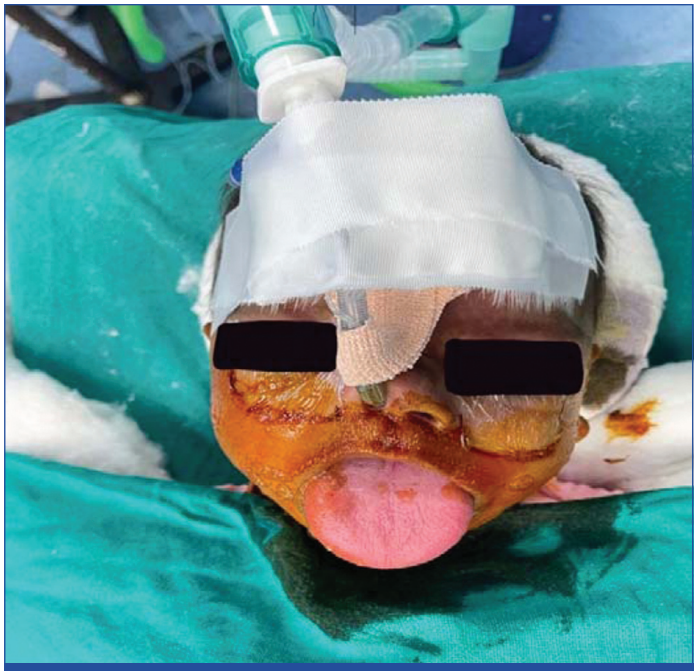
[Table/Fig-2]: a) CE-MRI face depicting macroglossia; b) CE-MRI brain depicting non-obstructing hydrocephalus.

To improve oral feeding, support adequate development, and achieve cosmetic correction and subsequent speech improvement, surgical debulking of the tongue was planned at five months of age (weight: 6 kg). After confirming Nil Per Os - {Nothing by mouth (NPO)} status, the patient was shifted to a prewarmed operating theatre with the surgical safety checklist completed. A difficult airway cart was prepared, including 3.0-3.5 cuffed endotracheal tubes and flexometallic tubes, curved and straight laryngoscope blades, oropharyngeal and nasopharyngeal airways of various sizes, a paediatric stylet and bougie, a paediatric flexible fibreoptic bronchoscope, laryngeal mask airways, face masks of multiple sizes, and an Artificial Manual Breathing Unit (AMBU) bag to manage the anticipated difficult airway.

Standard American Society of Anesthesiologists' (ASA) monitoring was applied. Baseline heart rate was 152/min and oxygen saturation was 100% on room air. Preoxygenation was performed using a size 2 Rendell Baker Soucek (RBS) face mask to accommodate the large protruding tongue. Inhalational induction was carried out with incremental concentrations of sevoflurane in 100% oxygen. Intravenous cannulation was secured in the right hand with a 24G cannula and IV fluids {(Deviated Nasal Septum (DNS)} were started. Injection glycopyrrolate 5 mcg/kg Body Weight (BW) and fentanyl 2 mcg/kg BW were administered.

After loss of consciousness with maintained spontaneous respiration, a check laryngoscopy using a curved blade was performed following optimal positioning. The Cormack-Lehane grade was IIIa, which improved to grade II with Backward, Upward, Rightward Pressure (BURP). Muscle relaxation was then achieved using succinylcholine 1.5 mg/kg BW. Nasal intubation was performed using a 3.5 mm internal diameter flexometallic oral endotracheal tube guided into the glottis with Magill's forceps [Table/Fig-3].

A loading dose of atracurium 0.5 mg/kg and maintenance doses of 0.1 mg/kg were given throughout surgery. Anaesthesia was maintained with Nitrous Oxide:Oxygen (N_2O:O_2) (1:1) and sevoflurane. Dexamethasone 0.15 mg/kg was administered to reduce surgery-related airway oedema. Adequate analgesia was provided using



[Table/Fig-3]: Infant with BWS intubated nasally using flexometallic tube due to macroglossia.

IV paracetamol 7.5 mg/kg BW and supplemental fentanyl 2 mcg as required. Intraoperative vital signs remained stable, and blood glucose levels were within the range of 110-240 mg/dL throughout the procedure. Tongue resection was completed, with an estimated blood loss of approximately 40 mL. The intraoperative period was uneventful.

At the end of surgery, the patient was shifted to the Paediatric Intensive Care Unit (PICU) for monitoring of tongue oedema and to prevent postoperative airway obstruction. She was kept sedated on Synchronised Intermittent Mandatory Ventilation (SIMV) mode for 24 hours. Strict glycaemic control was maintained in the PICU with two-hourly blood glucose monitoring, and adequate postoperative analgesia was ensured. After discontinuation of sedation and return of adequate spontaneous respiration, the patient was extubated and observed for six hours before being shifted to the ward.

DISCUSSION

BWS is a congenital overgrowth disorder with an incidence of approximately 1:13,700 to 1:17,000 live births, with equal sex distribution. About 85% of cases are sporadic, while the remaining 15% are familial, arising from recurrent chromosomal alterations in the imprinted 11p15.5 region [1]. The most common anomaly in BWS is macroglossia, often accompanied by anterior abdominal wall defects. Macroglossia is present in approximately 95% of patients with BWS [2] and presents notable airway challenges for anaesthesiologists.

Macroglossia may lead to malfunction of the stomatognathic system, breathing and speech difficulties, mandibular enlargement, diastema, and other orthodontic abnormalities [3]. These can significantly impact development and psychosocial well-being, making surgical reduction of tongue size necessary during the first year of life.

Airway management in these patients is particularly critical and requires meticulous preoperative assessment and preparation. Key concerns include prematurity, macroglossia, hypoglycaemia, associated cardiac anomalies, airway oedema, and postoperative airway obstruction.

Although laryngoscopy in non-paralysed paediatric patients is challenging, we opted for inhalational induction with sevoflurane in 100% oxygen to allow for an initial check laryngoscopy. After establishing adequate mask ventilation using a larger RBS mask

to accommodate the protruding tongue, succinylcholine was administered for neuromuscular blockade. Maintaining spontaneous breathing during induction is reported to be important in patients with potentially difficult upper airways [4].

Nasotracheal intubation was successfully performed using a 3.5 mm ID flexometallic endotracheal tube, which is the preferred technique for airway management during glossectomy [5].

Batra M et al., reported a similar case in which sevoflurane inhalational induction followed by nasotracheal intubation was used successfully, mirroring our strategy of preserving spontaneous respiration before securing the airway to mitigate the risk of failed ventilation [4]. In another case described by Tsukamoto M et al., orotracheal intubation was performed following induction with sevoflurane and rocuronium. Their findings further support the cautious use of muscle relaxants only after confirming adequate mask ventilation-an essential step in anticipated difficult airway scenarios such as ours [2].

Airway obstruction due to postoperative tongue oedema is a well-recognised complication. Therefore, the authors elected to ventilate the patient for 24 hours with vigilant blood glucose monitoring to prevent neurological sequelae. This approach aligns closely with recommendations from Celiker V et al., who also advised 24-hour elective ventilation post-glossectomy to prevent airway compromise [6]. In contrast, Bicer C et al. reported a case of an infant with BWS in whom nasotracheal intubation was performed following induction with thiopental sodium and succinylcholine, and immediate postoperative extubation was undertaken [7].

In the present case, a larger RBS face mask was used to accommodate the protruding tongue. The necessity of a fully equipped difficult airway cart has been emphasised consistently across similar case reports. These recurrent observations underscore the critical importance of thorough preoperative planning and simulation-based preparedness in ensuring safe and effective anaesthetic management of high-risk paediatric patients such as those with BWS.

CONCLUSION(S)

BWS presents significant anaesthetic challenges, primarily due to macroglossia and its implications for airway management. This case highlights the importance of meticulous preoperative planning, including readiness with a difficult airway cart and anticipation of postoperative complications such as airway oedema. The use of inhalational induction with preserved spontaneous respiration, followed by controlled intubation, ensured a safe approach to securing the airway. Elective postoperative ventilation played a crucial role in preventing airway obstruction and ensuring patient safety. Vigilant glycaemic control and adequate analgesia are also essential to optimise outcomes in such complex paediatric cases. An individualised, multidisciplinary approach is vital for the safe and effective anaesthetic management of children with BWS.

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