UDK: 616-056.7:575.224.23]-089.5-053.2
DOI: https://www.doi.org/10.55302/MJA2373085d

CASE REPORT

PERIOPERATIVE ANESTHETIC MANAGEMENT IN A CHILD WITH BECKWITH-WIEDMAN SYNDROME

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Abstract:

Beckwith-Wiedemann Syndrome (BWS), a rare genetic disorder, poses distinctive challenges for anesthesiologists due to its associated physical anomalies and potential complications. This case report details the anesthetic management of a 2-year-old boy with BWS undergoing a procedure for agenesis testis bilateral. Key considerations include macrosomia, hypoglycemia risk, airway abnormalities, cardiovascular anomalies, and abdominal wall defects. The patient successful perioperative care involved meticulous preoperative assessment, careful planning, and collaboration with specialized medical teams. Anesthesia comprised a combination of medications, monitoring, and airway management techniques tailored to the patient's unique needs. The discussion emphasizes the importance of considering macrosomia, hypoglycemia, anatomical abnormalities, and other factors in anesthesia planning for BWS patients. Vigilant postoperative monitoring is highlighted. The conclusion underscores the necessity for experienced anesthesiologists familiar with the challenges associated with BWS, emphasizing the significance of tailored approaches, comprehensive assessments, and collaborative care for the safety and well-being of these patients during surgical interventions.

***Key Words**: Beckwith-Wiedemann Syndrome, macrosomia, airway management, perioperative care*.

Introduction

Beckwith-Wiedemann syndrome (BWS) is a complex overgrowth disorder with an estimated incidence of 1:13,700 live births. It is caused by variety of genetic or epigenetic alterations within two domains of imprinted gene on chromosome 11p15. It is mainly characterized by macroglossia, omphalocele and gigantism.

Perioperative anesthetic management might be complicated by anatomical airway abnormalities, recurrent hypoglycemia, electrolyte imbalance and possible cardiovascular anomalies. Prior preparation, preoperative assessment and strategies to manage airway obstruction is essential for

the successful administration of anesthesia. Hereby we present the airway management and anesthesiologic considerations of a 2-years old boy who underwent a procedure for “agenesio testis billateralis”.

Case Report

The patient is a 2-years old boy and weighting 11kg. His medical documentation included results from magnet resonance and there was described “agenesio testis billateralis” and enlarged lymph nodes in 10mm in inguinal region. He was operativelytreated for omphalocele 2 years ago. The Mallampati grade was III, the blood oxygen saturation was 98% in room air and there were no abnormalities in laboratory parameters and cardiac function. Prior to the OR admission, premedication was administered. We had monitoring of electrocardiogram (ECG), NIBP, heart rate and blood saturation. Infusion with 0,9% NaCl was started on peripheral vain, and we administrated Fentanyl, Propofol, Suxamethonium chloride, and after successfully intubation Rocuronium. We placed a 4mm cuffed endotracheal tube using video-laryngoscope (Storz) with the Macintosh blade size 2. Anesthesia was maintained with Sevoflurane 2vol% in air and oxygen (50% - 50% concentration). Heart rate was 126-128bpm, blood saturation 100% and end-tidal carbon dioxide was 40 - 42mmHg. The surgical procedure was completed in 2 hours and 15 minutes without any complications. Neuromuscular blockade was reversed with Neostigmine and Atropine. After extubation his respiratory and hemodynamic conditions were stable.

Discussion

Beckwith-Wiedmann syndrome is caused by mutation in the genes and this patient’s diagnose was based on a chromosomal analysis postpartum. Anesthesia management in BWS is a challenge because of the abnormal airway anatomy, gigantism, visceromegalia, cardiac defects and endocrine abnormalities (hypothyroidism, hypoglycemia). Nephro-urological anomalies in patients with BWS is 28% - 61% including cortical and medullary cysts and higher incidence of hypercalciuria and nephrolithiasis. The importance of hypercalciuria is related to perioperative renal dysfunction. Cardiac defects occur in up to 13% -20% of patients. It’s necessary to have careful pre-operative evaluation. Besides for routine evaluation tests and clinical exams, a preoperative chest radiograph is required, not only to diagnose some evidence of cardiac anomaly, but also to exclude thoracic neuroblastoma. Polycythemia and hypothyroidism, though less common, should be ruled out before surgery due to their adverse effect on perioperative events in terms of bleeding and delayed weaning from mechanical ventilation. Preparation before the induction of anesthesia is especially important in cases that may show potential for difficult ventilation and intubation. In the operating room, different-sized masks, tracheal tubes, nasal and/ or oral airways, a stylet, laryngeal masks, fiberscope and video laryngoscope, as well as a tracheostomy set should be kept available (1,2). Premedication is usually avoided if there is a suspicion of possible airway compromise following sedation, although it was used in our case without any complications. Inhalational induction is comparatively safe because of the fact that during mask ventilation overdose of sedatives may cause tongue to fall back into the retro lingual space leading to severe airway obstruction. Awake intubation is an alternative, but it is avoided due to discomfort and pain, and it may increase in intracranial pressure (2). Comprehensive cardiac evaluation including ECG, echocardiography and CT angiography is only necessary when cardiac anomaly is suspected during clinical examination. Abdominal ultrasound is required to access for organomegaly, nephrocalcinosis, medullary sponge kidney

and other structural abnormality (3). In our case, the following strategy was followed for the anesthetic administration: induction with fentanyl and propofol, ventilation via a mask and performing a rapid sequence intubation with use of Suxamethonium chloride for shorter need of ventilation, and a video laryngoscope which is a tool of choice for suspected difficult intubations. Cuffed endotracheal tube is preferred because the size of trachea is not easy to predict in BWS patients (larger size trachea in BWS), further, to avoid risks incurred during changing of tracheal tube (in situation of difficult endotracheal intubation). After achieving intubation, rocuronium was administered for neuromuscular blockade. There is no contraindication for any of the anesthetic agents. Opiates should be used cautiously to avoid postoperative airway obstruction and apnea. All the neuromuscular blocking agents can be safely used unless there is a general contraindication (renal or hepatic insufficiency). Reversal of neuromuscular blockade is safe with neostigmine. Other considerations are that BWS patients are more prone for metabolic stress response and electrolyte shift during perioperative period. Patients with BWS can be at risk for pre-operative hypoglycemia, but mainly in the neonatal period, and our case was a 2- years child with no such problem (4,5). The postoperative care depends upon the age of the child, type of surgery performed, underlying cardiac problem and presence of hyaline membrane disease. Careful management of glucose and electrolyte homeostasis is mandatory in selected cases.

Conclusion

Planning and preparation for difficult ventilation or intubation include different-sized masks, tracheal tubes, nasal and/ or oral airways, a stylet, laryngeal masks, fiberscope and video laryngoscope, as well as a tracheostomy set. It’s necessary to have knowledge how to manage complications if you have during the treatment (hypoglycemia, low cardiac output, slow metabolism of anesthetics and difficult ventilation).

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