Case Study: Anesthesia and perioperative care for a patient with Beckwith-Weidemann syndrome

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Introduction: Beckwith-Wiedemann Syndrome is an overgrowth disorder. The syndrome is usually sporadic, but may be inherited. These children are at risk for developing hypoglycemia and various types of tumors. The clinical picture of this syndrome can vary from patients being mildly to greatly affected. We report our clinical experience of a patient with Beckwith-Weidemann syndrome.

Case Report: An 18-month-old male patient with a history of Beckwith-Wiedemann syndrome was scheduled for hepatic resection of tumor. He was a product of an in vitro fertilization and twin delivery. His twin sister is reported to be completely healthy. He had history of neonatal hypoglycemia and corrected omphalocele at birth. Apnea and bradycardia were resolved after a hemiglossectomy at three months of age.

After proper pre-oxygenation, the anesthesia was induced with intravenous thiopental and cis-atracurium. The anesthesia was maintained with isoflurane. A thoracic epidural was placed for postoperative analgesia. The patient was extubated immediately after a seven hour long resection of the hepatoma. He was made comfortable postoperatively utilizing epidural anlagesia. No peri-operative complications were noted for this patient.

Discussion: Beckwith-Wiedemann syndrome is a common genetic overgrowth syndrome that is associated with visceromegaly, macroglossia, abdominal wall defects, pre- and postnatal overgrowth, and neonatal hypoglycemia. The incidence of Beckwith-Wiedemann syndrome has been reported as approximately 1:15,000 births, with approximately equal incidence in males and females.

The syndrome is thought to be due to an alteration in the genes or can also be an autosomal dominant inheritance. A recent series of observations has suggested, a link between in vitro fertilization and imprinting disorders, such as Beckwith-Wiedemann syndrome might be suspected. Prenatal ultrasounds have occasionally been helpful in the diagnosis when omphalocele and other characteristics are present. The patients’ birth weights and lengths are usually above average. Macroglossia can effects on the child’s ability to eat, breathe, or speak. In severe cases, surgical correction of macroglossia is necessary. In this case, the patient’s apnea and bradycardia were resolved after hemiglossectomy. Hypoglycemia occurs shortly after birth. Abdominal wall defects can occur, which include omphalocele, diastasis recti or umbilical hernia. Hemihippertrophy may not present at birth but can become apparent later in childhood. The incidence of malignant tumor in reported cases is about 5-10 %. These include Wilms’ tumor, hepatoblastoma, and neuroblastoma. Children with hemihypertrophy seem to have a higher risk of developing tumors. Their intelligence is usually normal.

Anesthetic management for patients with Beckwith-Wiedmann syndrome may be complicated by macroglossia and underlying associated illnesses. Difficulty in tracheal intubation, refractory hypoglycemia, and tracheomalacia have been reported in neonatal Beckwith-Wiedmann syndrome. Thorough preoperative evaluation, including evaluation of the cardiac and genitourinary systems, perioperative monitoring, and adequate postoperative analgesia are essential for a successful outcome.

References