Anesthetic Considerations for Beckwith-Wiedemann Syndrome

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Beckwith-Wiedemann Syndrome occurs in the neonatal period at a frequency of approximately one in 13,700 live births. The syndrome consists of macroglossia, omphalocele, visceromegaly, and neonatal hypoglycemia. Therefore, these infants frequently require operative procedures in the newborn period. Very little has been published regarding anesthetic management of these infants, although abnormal airway anatomy, as well as cardiomyopathy and hypoglycemia, may complicate perioperative management. We present the preoperative evaluation and intraoperative management of a 29-day-old infant with Beckwith-Wiedemann Syndrome who presented for subtotal pancreatectomy to treat severe refractory neonatal hypoglycemia.

CASE REPORT

A 2.9-kg term male infant was born by cesarean section after an uncomplicated prenatal course, except for the presence of polyhydramnios, which had been diagnosed predelivery by sonogram. Apgars scores at 1 and 5 min were 6 and 8, respectively, due to decreased respiratory effort and poor muscle tone.

Initial physical exam revealed a large fluctuant bell-shaped abdomen, undescended testes, small phallus, subluxation of the hips, and a small omphalocele. Tachypnea (RR 60-80 breaths/min) and a systolic ejection murmur grade II/VI heard at the lower left sternal border were noted. Airway examination revealed a slightly enlarged tongue, but the infant's lungs could be easily ventilated with bag and mask. Chest x-ray showed mild cardiomegaly with clear lung fields. The hematocrit was 58%, and serum glucose was 20 mg/dl (normal ≥ 40 mg/dl) with an insulin level of 174 μU/ml (normal 4-24 μU/ml).

The infant continued to have episodes of symptomatic hypoglycemia (glucose < 50 mg/dl) during the first 4 weeks of life. The hypoglycemia was initially treated with dextrose 12.5% in water, which had to be increased stepwise to dextrose 25% in water during the first week of life. Dextrose and cortisone acetate were added to help control the hypoglycemia. However, by the end of the fourth week, the infant was requiring dextrose 50% in water at a rate of twice maintenance fluids to keep serum glucose in the 40-60 mg/dl range.

Abdominal sonogram showed normal kidneys, liver, and spleen, but an abdominal mass was visualized. CT scan defined cystic adrenals and enlargement of the pancreas. Echocardiogram showed a PDA, bicuspid aortic valve, and biventricular hypertrophy with mild depression of ventricular contractility.

The infant developed symptomatic congestive heart failure that was treated with furosemide, and fluid restriction. Hypoglycemia persisted, necessitating a subtotal pancreatectomy for presumed neodioblastosis.

Preoperative medication included an extra dose of cortisone acetate (15 mg/m²), antibiotics, and his scheduled dose of furosemide. In the operating room, monitors consisted of an EKG, both transcutaneous and end-tidal P CO₂, precordial stethoscope, oscillometric blood pressure device, and a pulse oximeter. Awake laryngoscopy revealed easy visualization of the glottis. Following oxygenation, anesthesia was induced with fentanyl (10 μg/kg) and pancuronium (1 mg/kg). Tracheal intubation was accomplished without difficulty and a catheter was inserted into the right radial artery for monitoring of electrolytes, glucose, and arterial blood pressure.

Anesthetic maintenance was accomplished with fentanyl totaling 50 μg/kg over 5 h, pancuronium bromide, and an air-oxygen mixture to maintain oxygen saturation between 94-98%. Maintenance fluids of dextrose 30% in 25% normal saline were administered through a broviac catheter and hourly glucose levels were measured. Blood glucose concentration remained between 80-110 mg/dl during the procedure and rose to 150 mg/dl approximately 1 h postoperatively. The trachea remained intubated and the infant was transported to the neonatal intensive care unit, where mechanical ventilation was required for the next 4 days. Microscopic examination of the resected portion of pancreas showed islet cell hyperplasia consistent with the presumed diagnosis of neodioblastosis.

DISCUSSION

Beckwith-Wiedemann Syndrome was first described in 1963 by Beckwith in three related necropsy cases consisting of macroglossia, omphalocele, and visceromegaly (pancreatic islet cell hyperplasia, renomegaly, and adrenal cortex hyperplasia).† In 1964, Wiedemann added two more cases in living children with these same clinical features, but also with severe hypoglycemia and postnatal gigantism.‡

Macroglossia is the most common feature. It occurs in approximately 95% of patients.¹ Microscopic and histochemical studies of the tongue have failed to demonstrate tissue anomaly other than hyperplasia or hypertrophy of intrinsic muscles of the tongue.² The oral cavity is of nor-

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† Beckwith JB: Extreme caymegaly of the adrenal fetal cortex, omphalocele, hyperplasia of kidneys and pancreas and Leydig cell hyperplasia. Another syndrome? Presented at The Annual Meeting of the Western Society for Pediatric Research, Los Angeles, California, November 11, 1963 [unpublished].

nal size for age, only the tongue is enlarged. Early treatment, such as glossectomy or tracheostomy, may be required to prevent respiratory difficulty. Chronic respiratory obstruction can lead to pulmonary hypertension and resultant cor pulmonale. With growth, protuberance of the tongue lessens due to oral cavity enlargement and repositioning of the hyoid bone posteriorly and inferiorly, permitting gradual accommodation of the tongue.

Hypoglycemia is usually severe, requiring glucose administration and hyperglycemic medications. Prompt treatment is important to prevent the neurologic sequelae of seizures and mental retardation. Addition of diazoxide, corticosteroids, glucagon, or epinephrine in various combinations may be necessary. Insulin levels are above normal even when the blood glucose level is below 60 mg/dl. Hyperinsulinism is due to pancreatic islet cell hyperplasia or nesidioblastosis. Hypoglycemia will usually spontaneously subside by age 4 months. If hypoglycemia is uncontrollable, a partial pancreatectomy may be performed.

In the late 1970s, a joint retrospective study found 13 cases of Beckwith-Wiedemann Syndrome. Twelve of these were found to have cardiovascular abnormalities. There were congenital cardiac defects in seven and idiopathic cardiomegaly in five. Congenital lesions included TOF, ASD, VSD, PDA, and hypoplastic left ventricle. Idiopathic cardiomegaly, which was mild to moderate, was not associated with cardiorespiratory symptoms, and disappeared by age 6 months. Cardiomegaly was attributed to hypoglycemia or visceromegaly.

With regard to anesthetic management, many infants will have upper airway obstruction while awake. Infants with macroglossia will require either awake tracheal intubation or awake vocal cord inspection. This can be facilitated using topical anesthesia. If the glottis can be viewed, then either an intravenous induction or an inhalational induction may be undertaken. In our case, a high-dose narcotic technique was chosen because of the presence of congestive heart failure. As in the case of any child with pre-existing airway obstruction, if an inhalation induction is chosen, we avoid the use of nitrous oxide, because if obstruction or laryngospasm occur, delivery of an FIO2 of 1.0 affords more time for various maneuvers to correct the problem before severe hypoxemia occurs. A nasopharyngeal airway formed from an endotracheal tube is useful in relieving obstruction as the anesthetic level deepens. The breathing circuit may then be attached to the endotracheal tube being used as a nasopharyngeal airway for delivery of anesthetic gases and oxygen during intubation attempts. Another maneuver that can improve ventilation is forward and downward traction on the tongue. In all cases, a tracheostomy tray should be ready and a surgeon skilled in pediatric tracheostomy present.

Other anesthetic concerns include intraoperative monitoring of serum glucose, electrolytes, and hematocrit. As in this case, hypoglycemia can be severe and may be the reason for surgery. The electrolyte concentrations are usually normal, but may be abnormal if an adrenal tumor exists.

Preoperative steroids are required for those infants receiving chronic steroid therapy. A preoperative dose of either cortisone acetate or prednisone (15 mg/M2) the morning of surgery should be given, as well as an infusion of 15–25 mg/M2 of hydrocortisone given postinduction.

In summary, infants with Beckwith-Wiedemann Syndrome may have a varied clinical presentation. Infants with tumors, uncontrollable hypoglycemia, exophthalmos, and macroglossia may require surgical intervention. A comprehensive preoperative examination should include assessment of the airway, cardiovascular status, endocrine status, glucose, and electrolytes. Preparations for difficult airway management should be made. Anesthetic choice should be dictated primarily by the surgical procedure and preoperative cardiovascular status. Intraoperative serum glucose monitoring is important to prevent the neurologic sequelae of unrecognized hypoglycemia.

REFERENCES