Anesthesia for Cesarean Section in Patient with Spondylometaphyseal Dysplasia

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Spondylometaphyseal dysplasia (SMED) is a very rare form of dwarfism that represents an entity distinct from the heterogeneous group of skeletal dysplasias known as spondyloepiphyseal and spondylometaphyseal dysplasias. Less than 20 patients have been identified with this type of skeletal dysplasia. 1

Prominent features of this disease consist of severe skeletal deformities, including short limb short stature, kyphoscoliosis with spinal cord compression, markedly contracted pelvis with narrowing of sacrosciatic notches, coxa vara deformity of both hips, pectus carinatum, and odontoid hypoplasia. Other nonspecific congenital abnormalities, including cleft palate, hemangiomas, inguinal hernias, congenital hydrourephrosis, and mitral valve prolapse, also may be associated. Anesthetic management for patients with the more common types of dwarfism has been described. 2–3 We describe the successful anesthetic management of cesarean section for a patient with SMED and marginal placenta previa.

REPORT OF A CASE

A 29-year-old primigravida with SMED (fig. 1) was admitted for vaginal spotting and mild uterine contractions at 34 weeks gestation. Her height was 101.5 cm, weight 29.5 kg. Physical examination was remarkable for limited neck and jaw mobility, very high anterior larynx, and severe thoracolumbar scoliosis. Her family history was unknown because she was adopted. Medical history and gestational history had been essentially unremarkable. Admission laboratory revealed serum sodium 135 mEq/l, serum potassium 3.9 mEq/l, blood glucose 79 mg/dl, and hemoglobin 11.8 g/dl. Ultrasound examination revealed marginal placenta previa and estimated fetal gestational age of 35 weeks. Ritodrine, a beta-sympathomimetic amine, was infused at 0.05 mg/min (0.03% solution; 10 ml/h) in an attempt to stop uterine contractions. A maintenance infusion of 5% dextrose in lactated Ringer’s solution at 125 ml/h was also begun.

Sixty hours after admission, the patient began to have moderate vaginal bleeding and strong uterine contractions every 2–3 min. The obstetrician then decided to perform emergency cesarean section. The ritodrine drip was discontinued at that time. The patient was transported to the operating room. Left uterine displacement and inhalation of oxygen via a facemask were employed. The patient was positioned on the operating table with left uterine displacement and inhalation of oxygen maintained. Physical examination revealed an arterial blood pressure of 120/80 mmHg, heart rate 165 beats/min, respiratory rate of 44 breaths/min, inspiratory rates to midscapulae in both lung fields, a prominent fourth heart sound, and moderate vaginal bleeding. Laboratory tests obtained earlier that day revealed serum sodium 137 mEq/l, serum potassium 2.9 mEq/l, serum glucose 149 mg/dl, and hemoglobin 11.5 g/dl. Continuous monitoring of the patient’s electrocardiogram, lead II, in the operating room showed sinus tachycardia and no ST-segment changes. Fetal heart rate in the operating room remained between 120–140 beats/min with good variability. A difficult endotracheal intubation was anticipated, so the mouth and hypopharynx were anesthetized with nebulized 4% lidocaine and laryngoscopy was performed. Since the epiglottis and posterior arytenoids were visualized, an iv anesthetic rapid sequence induction was chosen instead of an awake endotracheal intubation. Inhalation of oxygen via a face mask was reinstituted after laryngoscopy and i.v. succinylcholine 1.5 mg was administered iv. The patient was prepped and draped. During this time increased vaginal bleeding was noted.

Anesthesia then was induced with ketamine 30 mg and succinylcholine 45 mg iv, employing cricoid pressure. Despite rapid intubation of the trachea with a cuffed 6.0 mm endotracheal tube and stylet in place, mild cyanosis and bradycardia to 55 beats/min resulted, which quickly resolved by hyperventilation with 100% oxygen. Breath sounds were bilaterally equal, and the endotracheal tube was secured with the 12 cm mark at the teeth. Anesthesia subsequently was maintained with 50% nitrous oxide, 70% oxygen, 0.5% halothane, and 0.2% succinylcholine infusion. Eight minutes after induction, a 1,850-g female infant was delivered. Presentation of the infant was frank breech, and uterine incision to delivery time was 110 s. The Apgar score was 2 at 1 min. The trachea of the infant was immediately intubated, and hyperventilation was instituted with 100% oxygen. Heart rate, however, remained less than 100 beats/min. Epinephrine 0.05 mg was administered via endotracheal tube. Heart rate increased to 160 beats/min, and the infant was transferred to the neonatal ICU with a 5-min Apgar score of 5. Morphine sulfate, 5 mg, was given iv to the mother and the halothane concentration was reduced to 0.25% for the remainder of the case. The mother’s arterial blood gases drawn shortly after delivery were pH 7.30, PaCO2 40 mmHg, PaO2 230 mmHg (FiO2 0.70), and the hemoglobin was 8.7 g/dl. Estimated blood loss at that time was 800 ml. Since surgical hemostasis had not yet been attained, 1 unit of whole blood was given. The surgical procedure and subsequent anesthetic course were uneventful with stable vital signs, and the trachea was extubated after meeting standard extubation criteria. Estimated blood loss at the end of the procedure was 1,000 ml. Laboratory results in the recovery room revealed a hemoglobin of 10.6 g/dl, pH 7.46, PaCO2 33 mmHg, and PaO2 183 mmHg (FiO2 0.40 by face mask). Chest roentgenograph obtained in recovery room revealed perihilar infiltrates suggestive of pulmonary edema. The subsequent postoperative course of the mother was uneventful. The infant required mechanical ventilation for 36 h, and the subsequent postnatal course was without further complication.

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DISCUSSION

The cause of the neonatal depression may be attributed to several factors. The presence of placenta previa, which was noted to be actively bleeding at the time of induction, and the episode of maternal hypoxemia on induction likely resulted in decreased uteroplacental blood flow and oxygenation. In addition, the prematurity of the infant and delivery by cesarean section, which lacks the advantage of vaginal squeeze to assist in clearing ultrafiltrate from the infant's lungs, all likely contributed to the neonatal depression.

The selection and management of anesthesia for the dwarf patient presents a unique challenge. Several features of these patients present common problems. Included among these are the following: 1) potential for respiratory embarrassment, 2) difficulty with airway management, 3) potential for cervical cord compression and neurologic problems, and 4) difficulty with conduct anesthesia.

Oxygen uptake is markedly increased in pregnancy because of increased metabolic consumption. In addition, the lowering of FRC to levels approximating closing volume when near-term patients assume the supine position further lowers oxygen reserve. The usual decrease in FRC is even more pronounced in the dwarf patient. This is because the xiphoid process to symphysis pubis distance is less than usual, causing enhanced encroachment of the uterus on the diaphragm. Kyphoscoliosis may further compromise respiratory function. Additionally, engagement of the fetal head usually is prevented by the combination of contracted maternal pelvis severe lumbar lordosis. This maintains the uterus a total abdominal organ, further decreasing FRC. The addition of pulmonary edema, which may have been present in our patient, further decreases oxygen reserve. We feel the combination of these changes resulted in hypoxemia during induction of anesthesia as evidenced by cyanosis and bradycardia. The cause of pulmonary edema may have been overzealous hydration by the obstetric team or ritodrine therapy. Left ventricular failure is thought to be the mechanism by which ritodrine produces pulmonary edema. The left ventricular failure is the result of a 50-60% increase in cardiac output caused by the beta-adrenergic effects of ritodrine. This increase in cardiac output is added on to an existing 35-40% increase in cardiac output, which is a normal physiologic change associated with pregnancy. Other possible contributing factors include the marked increase in blood volume, which is also a normal physiologic change associated with pregnancy, and excessive fluid therapy in an attempt to maintain arterial blood pressure in the face of peripheral vasodilation from ritodrine therapy.

Other complications from ritodrine therapy are related to its unwanted beta-adrenergic activity and include maternal hypotension from peripheral vasodilation, widened pulse pressure, tachycardia, angina, anxiety, and restlessness. Metabolic disturbances arise from activation of glycogen phosphorylase and produce increases in blood glucose and lactate levels. There is also an intracellular movement of potassium ions causing hypokalemia, which was noted in our patient. Frequently, hematocrit decreases during ritodrine therapy because an increased amount of IV fluid is administered in order to maintain arterial blood pressure resulting in hemodilution. Although hemodilution may have been present, active bleeding from the placenta previa certainly contributed to the decrease in hemoglobin level in this patient.

Evaluation of the patient before institution of ritodrine therapy includes obtaining a careful medical history with emphasis on the cardiovascular system, an electrocardiogram, and measurements of blood glucose, electrolytes, and hematocrit. Evaluation during ritodrine therapy includes frequent monitoring of heart rate and arterial blood pressure, close observation for early signs of pul

FIG. 1. Patient with SMED 2 days postpartum. Note short stature, short neck.
monary edema, accurate fluid input and output recording, and serial measurements of hematocrit, blood glucose, and electrolytes. Additional preoperative evaluation of the patient with SMED should include arterial blood gases to assess oxygenation, pulmonary function tests in the patient with a preoperative history of dyspnea that limits activity, and cervical spine roentgenography in the patient with neurologic symptoms.

General anesthesia has been recommended as the technique of choice for dwarf patients, although difficulty with endotracheal intubation is a concern. Ketamine, which may aggravate ritodrine-induced tachycardia, was chosen over sodium thiopental as the induction agent in our patient because of the questionable intravascular volume status. In choosing an endotracheal tube size, Waite found that weight was generally a better predictor of tube size in children with proportionate small stature, while in nonachondroplastic dwarfs, such as our patient, neither age nor weight seemed to be the ideal predictor, and the tube size selected was generally smaller than would be anticipated. The trachea of our patient was intubated with a 6.0-mm endotracheal tube, and multiple tube sizes were readily available on induction. Precautions to avoid overdistention flexion also should be employed because odontoid hypoplasia, foramen magnum insufficiency, and atlantoaxial instability may occur in as many as 75% of non-achondroplastic dwarfs.

The inherent problems with conduction anesthesia are mainly due to skeletal abnormalities, which include severe lumbar lordosis, kyphoscoliosis, prolapsed intervertebral disc, deformed vertebral bodies, and a relatively narrow spinal canal containing a normal-sized spinal cord. These abnormalities produce technical difficulties in performing spinal or epidural anesthesia. Epidural anesthesia has theoretic advantages over spinal anesthesia because it allows titration of local anesthetic to appropriate block levels. Conduction anesthesia was not a consideration in this patient because of her questionable intravascular volume status caused by the acute blood loss from the placenta previa and the urgency of the procedure. If conduction anesthesia is considered, these patients should be fully informed of the possible technical difficulties and potential for general anesthesia in the event of block failure.

In summary, we present a case of a patient with SMED and marginal placenta previa who underwent anesthesia for cesarean section after failed tocolysis. Problems and methods of anesthetic management in the patient with SMED have been discussed.

REFERENCES