Osteogenesis Imperfecta: Anesthetic Management of a Patient for Cesarean Section: A Case Report


Osteogenesis imperfecta is a rare, autosomal dominant,¹ inherited disease of connective tissue with a reported incidence ranging from 1:21,000 to 1:60,000 births,² ³ with a higher incidence in females. Classically, the disease is manifested in two clinical forms: osteogenesis imperfecta congenitae (fetal type) and osteogenesis imperfecta tarda. In the congenita form, skeletal fractures occur in utero and death usually occurs in the perinatal period. The tarda form appears in childhood or early adolescence with the presence of blue sclera, multiple fractures after trivial trauma, the development of dwarfing syndrome with scoliosis and bowing of the femur and tibia, and the gradual onset of otosclerosis and deafness.⁴

Osteogenesis imperfecta tarda in pregnancy has been estimated to occur once every 25,000–30,000 pregnancies.⁵ Delivery by cesarean section is probably the method of choice⁶ ⁷ because of the increased risk of cephalopelvic disproportion, pelvic fractures, uterine rupture, and intrapartum or postpartum hemorrhage associated with this condition.

We report on the anesthetic management of cesarean section for a patient with extensive skeletal abnormalities and provide guidelines for the anesthetic management of patients with osteogenesis imperfecta.

REPORT OF A CASE

A 27-year-old primigravida, with osteogenesis imperfecta tarda dating from childhood, was admitted with a breech presentation at 38 weeks’ gestation for assessment. Her height was 154 cm, weight 52 kg. In the past she had sustained numerous fractures of her upper and lower extremities, a fractured pelvis, and a compression fracture of her sacrum. Her family history included a mother, brother, maternal aunt and uncle with short stature and histories of pathologic fractures. Positive physical findings included the presence of blue sclera, small brittle teeth, and receding mandible, mild pectus excavatum, thoraco-columbar kyphoscoliosis with decreased chest expansion and air entry bilaterally, and evidence of multiple previous fractures.

Hemoglobin, white blood count, serum electrolytes, calcium, phosphate, alkaline phosphatase, bleeding time (Ivy test), prothrombin time, partial thromboplastin time (PTT), and platelet count were all within normal limits. With an Fio₂ of 0.21, PaO₂ was 75.7 mmHg, PaCO₂ 31.7 mmHg, pH 7.4, standard bicarbonate 21.5 mEq/l. Pulmonary function testing revealed a moderate restrictive defect—vital capacity was 1.79 l or 61% of the predicted value. The forced expiratory value in 1 s FEV₁ was 79% of forced vital capacity. The electrocardiogram was normal.

After obtaining satisfactory fetal maturity studies, an elective cesarean section was scheduled. A continuous epidural anesthetic technique was chosen and informed consent obtained. Normal saline, 500 ml, was infused rapidly through a 16-gauge Teflon iv catheter. The epidural space was identified by the loss of resistance technique with a 16-gauge Tuohy needle, at L₃–₄ level, utilizing the left lateral position. Following a 2 ml test dose, a total of 12 ml 0.5% bupivacaine (Marcaine®; Breon Laboratories, New York, New York) in 2 ml bolus doses was infused over a 20-min period until a sensory block at the 6th thoracic dermatome was obtained. Following institution of the epidural block, no significant maternal heart rate and blood pressure changes from baseline values were obtained. In addition to the routine monitoring, an arterial line was inserted in the right radial artery for blood pressure monitoring and arterial blood–gas analysis. With supplemental oxygen given via a face mask, maternal PaO₂ was 105 mmHg intraoperatively. The patient was placed on a cooling blanket in the left lateral position. Temperature was monitored continuously by an axillary probe, and cold intravenous solutions were available if necessary. A 1.9-kg female infant was delivered through a standard midline vertical incision. The surgical procedure and subsequent anesthesia course was uneventful. The infant’s Apgar score was 7 at 1 min, 9 at 5 min. The infant had no apparent abnormalities—the sciera were of normal appearance and full body roentgenograms revealed no evidence of osseous or skeletal disease. Continuous epidural anesthesia was maintained for the first 24 h following surgery to avoid the need for systemic narcotics, with the potential for respiratory depression. The postoperative course was uneventful.

DISCUSSION

In addition to the usual pulmonary, circulatory, gastrointestinal, hepatic, and renal function changes associated with pregnancy, the skeletal and metabolic problems of osteogenesis imperfecta present potential anesthetic problems. Endotracheal intubation may be difficult because of the reduced range of cervical spine movement, the associated brittle teeth, and receding mandible. The associated kyphoscoliosis and pectus excavatum may cause significant pulmonary, mechanical, and gas exchange defects such as a reduced vital capacity,⁸ decreased chest wall compliance, maternal hypoxemia secondary to ventilation–perfusion mismatch,⁹ and in severe cases cor pul-
A mild hyperthermia with hyperhidrosis may occur in osteogenesis imperfecta, with features of abnormal cellular metabolism and central nervous system regulation, similar to patients with malignant hyperthermia. In addition, a mild bleeding tendency may be present due to decreased platelet adhesion, decreased platelet aggregation to adenosine diphosphate, and impaired platelet factor 3 release.

A major hazard of general anesthesia for cesarean section in this case was the anticipated difficulty or impossibility of endotracheal intubation following intravenous induction of anesthesia. To secure the airway prior to induction, awake intubation of the trachea with maternal sedation and topical anesthesia would have been required. This could be accomplished orally, blind nasally or with the aid of a fiberoptic bronchoscope. Alternative anesthetic techniques for cesarean section in this patient included local infiltration field block, spinal anesthesia, and epidural anesthesia.

Local infiltration field block for cesarean section generally is not recommended because in most cases supplemental anesthesia with narcotics and nitrous oxide is required, because of the difficulty of assuring adequate anesthesia and the need to use local anesthetics in large doses, with the inherent possibility of a toxic reaction. Spinal anesthesia would offer this patient the advantages of rapid onset of anesthesia, reliability, and minimal transplacental drug passage. The main disadvantages of spinal anesthesia in this case were the danger of intraoperative hypotension, despite prehydration, left uterine displacement and possible prophylactic ephedrine administration, and the danger of a high/total spinal block. A sensory level between the fourth and sixth thoracic dermatome is necessary for adequate surgical anesthesia. This level is achieved in the pregnant patient with doses of local anesthetic well below those required in the nonpregnant patient. The maternal short stature and kyphoscoliosis made the dose of local anesthetic required to achieve a fourth-sixth thoracic dermatome block difficult to predict.

We feel that a carefully administered lumbar epidural block, with meticulous technique, appropriate selection of local anesthetic dosage, adequate hydration, maternal hyperoxia, and avoidance of aorto-vena caval obstruction offered the best prospects for a successful maternal and fetal outcome. This technique allowed titration of the dose of local anesthetic to ensure a slow controlled onset of anesthesia, with less danger of intraoperative maternal hypotension. Over a 20-min period, 0.5% bupivacaine 12 ml was infused until a sensory block of the 6th thoracic dermatome was obtained. We titrated the local anesthetic because the short maternal stature and kyphoscoliosis made prediction of the dose required to achieve an adequate level of block difficult. The principal hazards associated with lumbar epidural anesthetic include convulsions from toxic blood levels of local anesthetics, excessively high dermatome level of block, or inadvertent subarachnoid injection. Had one of these untoward events occurred in this case, a means of instituting transtracheal ventilation rapidly would have been required. Hyperthermia in the parturient during regional anesthesia would be highly unlikely, despite the choice of an amide local anesthetic agent.

Before considering regional anesthesia for this patient, normal bleeding time, platelet count and clotting studies were confirmed. In osteogenesis imperfecta there may be technical difficulties inserting the epidural catheter due to the associated kyphoscoliosis. In this case no difficulty was experienced using the usual mid-line approach, but if difficulties arose, the lateral approach could have been tried. Skeletal abnormalities of the lumbar spine, as in osteogenesis imperfecta, are considered relative contraindications to regional anesthesia, but in this condition the inherent difficulties must be weighed up against the formidable risks involved in providing general anesthesia. Other contraindications to epidural anesthesia in this case would include the presence of infection at the site of needle injection, sepsis, hypovolemic shock, and patient refusal.

Based on the scant available literature and our experience with this case, guidelines for the anesthetic management of cesarean section for a patient with osteogenesis imperfecta should include ensuring that a bleeding diathesis does not exist; if cardiac lesions or thoracic cage deformities are present, careful preoperative cardiac and pulmonary function testing will be required; if no contraindications exist and if technically possible, a continuous epidural technique can be used for intraoperative anesthesia and postoperative analgesia; an arterial line can be inserted for blood pressure monitoring and arterial blood-gas analysis. Lastly, temperature should be monitored continuously, and a cooling blanket and cold intravenous fluids should be available if needed.

REFERENCES

Electrosurgical Burn at the Site of an Esophageal Temperature Probe

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The electrosurgical unit (ESU) has been used throughout operating rooms in the United States for almost 60 years.1 The hazards of stray radiofrequency (RF) current causing burns or other problems have been described.1-3 Electrical burns, not electrocution or fire, constitute the most common electrical hazard in operating rooms.4 Most ESU-associated burns occur at the attachment site of electrocardiogram (EKG) electrodes.3,4 Although a burn involving a rectal temperature probe has been reported,2 the internal nasal, oral, and rectal temperature monitors have not been reported as causes of electrical burns when used with the ESU. I describe a serious RF electrical burn at the site of an esophageal temperature probe.

REPORT OF A CASE

A 22-year-old man in good health was scheduled for right knee reconstruction under general anesthesia. After starting an IV infusion, monitors were placed, including the Ohio® 2100 Adult/Pediatric automated blood pressure monitor, the Physio-Control VSM 1/ESF Monitor® (for EKG monitoring), and a precordial stethoscope. Before preoxygenation, an uneventful induction of anesthesia and endotracheal intubation were performed, followed by the atrumatic insertion of an almost new (used less than 10 times) YSI® 702A Thermiliner Esophageal/Rectal Temperature Probe® (connected to the VSM 1/ESF monitor) through the nares, its full length, into the esophagus. Since electrocautery was to be used, the ESU dispersive electrode (Valley Lab Cohesive Dual Return Electrode®), compatible with the Valley Lab SSE2L ESU®, was properly applied to the patient's left thigh.

The operation, lasting slightly over 4 h, was devoid of any unusual anesthetic or surgical events. There were no arrhythmias, and the esophageal temperature ranged between 35.5 and 35.9°C. However, the temperature probe, after removal, was noted to have several smooth dark brown and black areas of discoloration on the white surface not noted prior to its insertion. After a routine emergence and extubation of the trachea in the operating room, the patient was admitted to the recovery room in a drowsy, easily arousable, stable condition, with blood pressure 150/80 mmHg, heart rate 84 beats/min, and respiratory rate 20 breaths/min while receiving 40% humidified oxygen via face mask.

In the recovery room, approximately 1 h after admission, the patient began spitting up copious amounts of blood-tinged saliva. Auscultation of the lungs revealed generalized congestion. An immediate chest radiograph revealed right hilar opacification. Vital signs remained

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