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Spinal Anesthesia in an Infant with Epidermolysis Bullosa

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EPIDERMOLYSIS bullosa is a rare group of genetic disorders of the skin with dominant and recessive modes of transmission. The dominant simplex form of epidermolysis bullosa is characterized by vesicles at sites of friction or trauma. The anesthetic concerns and difficulties have been described previously.[1–5] A variety of general, regional, and local anesthetic techniques have been used successfully in adults with epidermolysis bullosa.[6–9] In infants and children with this disorder, although anesthetic management usually consists of general anesthesia delivered by mask or endotracheal tube, or intravenous or intramuscular anesthesia, these techniques may pose formidable problems. Regional brachial plexus anesthesia in children[5,10] and the use of caudal anesthesia in an infant with epidermolysis bullosa[11] have been reported. We describe the successful use of spinal anesthesia in a pediatric patient with epidermolysis bullosa that obviated the need for either face-mask or endotracheal intubation.

Case Report

A 3.1-kg, 12-week-old male infant with a history of failure to thrive presented for placement of a gastrostomy tube. He was the 2.1-kg

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product of a 38-week pregnancy complicated by intrauterine growth retardation. The diagnosis of epidermolysis bullosa simplex was made by biopsy shortly after birth when the patient was observed to have numerous denuded areas of skin and bullae lesions throughout his oral cavity. His hospital course was significant for several episodes of third-degree heart block and an echocardiogram showing only a small patent foramen ovale. His skin developed widespread denuded, weeping areas and he had several occurrences of septicemia requiring antibiotics. With the use of intramuscular ketamine and supplemental halothane administration by gentle mask, he underwent central venous catheterization. This procedure was tolerated well without complications; however, the patient's dermatologic condition continued to worsen.

The patient became unable to tolerate oral feedings; an upper gastroesophagram showed a normal esophagus and ability to swallow, and he began receiving parenteral nutrition and intralipids. After an increase in respiratory effort, including audible stridor and tachypnea, flexible bronchoscopy was performed, revealing large arytenoids, laryngomalacia, and thickened vocal cords without tracheal problems. During this procedure, the patient was given intravenous propofol for sedation and an endotracheal tube was placed in his nasopharynx to provide a nasopharyngeal airway. This procedure was tolerated well, but nasal mucosal and intraoral blistering increased in subsequent days. Because of the patient's inability to be fed orally, a decision was made to place a gastrostomy tube to provide nutritional support. A preoperative physical examination was significant for numerous blistering lesions on the patient's face, extremities, and trunk, some of which were weeping with purulent fluid. Chest auscultation revealed rhonchi and inspiratory stridor.

The operating room was prepared with radiant warming lights and the operating table was covered with sheepskin. The patient was placed in a sitting position on the operating table and a providone-iodine (betadine) spray was applied to a lesion-free area at L4-5. A 23-G, 1.5-inch spinal needle was placed and clear cerebrospinal fluid (CSF) was obtained. After an epinephrine wash, a solution of 3 mg tetracaine (1 mg/kg) and an equal volume (0.3 ml) of 10% dextrose was injected. The patient was then quickly and carefully brought to a supine position to rest on a four-lead foam electrocardiogram electrode set for sensitive skin (Sentry Medical Products, Irving, CA) with no adhesive backing, and a satisfactory trace was obtained. Propofol infusion was started at 50 $\mu g \cdot kg^{-1} \cdot min^{-1}$ after incremental intravenous injections to 2 mg/kg to achieve sedation and prevent excessive upper extremity movement. The patient's arms were secured with well-lubricated Xeroform gauze (Baxter, Deerfield, IL). Xeroform gauze was gently wrapped around the arm, on which a blood pressure cuff was placed. The precordial stethoscope also rested on lubricated gauze with a hole cut to the stethoscope aperture. A pulse oximeter probe (Nelcor, Pleasanton, CA) was placed on the foot and gently secured with gauze wrap. No tape or adhesive dressings were used. The surgical field was gently blotted with betadine prep solution and the drapes were sutured in place. The gastrostomy insertion proceeded uneventfully without changes in vital signs. Heart rate was maintained at 160–170 beats/min, blood pressure at 80–100/40–55 mmHg, and respiratory rate at 24–32 breaths/min. Oxygen saturation was maintained at 96–100% with oxygen from a nearby mask (no skin contact). Surgical time was 35 min, after which the patient was returned to the neonatal intensive care unit. The patient had approximately 70 min of motor block from the spinal anesthesia and appeared to be very comfortable postoperatively without requiring additional analgesia until acetaminophen was given after 2 h, followed 4 h later by intravenous morphine. No surgical or anesthetic complications arose as a result of the procedure and no new lesions were observed the following day.

Discussion

Epidermolysis bullosa may result in blistering, fusion, and scarring of the lips and oropharyngeal structures. These lesions, which develop in response to trauma, friction, or pressure, may pose significant difficulties in airway manipulation during general anesthesia. Although relatively rare, there have been reports of laryngeal involvement.[12–14] The patient in this report had no laryngeal bullous lesions; however, he did have oropharyngeal blisters, and thickened vocal cords with laryngomalacia, making intubation potentially difficult and early postoperative extubation uncertain. Face mask application, oral airways, laryngoscopy, and tracheal intubation may cause progression of facial, labial, or oral lesions at sites of pressure or even finger contact.[6,15,16] In addition, 13 of 33 (40%) patients with epidermolysis bullosa had restricted mouth opening and 7 were difficult to intubate.[6] Thus, when possible, anesthetic techniques involving airway manipulations should be avoided in these patients.[8,10,16] The patient described in the current report did not require a face mask for supplemental oxygen or anesthetic.

There are now several reports on the utility of spinal and epidural anesthesia for adult patients with epidermolysis bullosa.[7–9] Initial fears that regional anesthesia would result in an increase in infections or tissue sloughing after anesthetic infiltration and skin preparation have not been realized. No intraoperative or postoperative complications were associated with major regional anesthesia. Kaplan[5] and Kelly[10] demonstrated the success of brachial plexus anesthesia in pediatric patients with epidermolysis bullosa. In addition, Yee *et al.*[11] showed that caudal epidural anesthesia may be used safely for circumcision in an infant with epidermolysis bullosa. Many similarities exist between caudal and spinal anesthesia, and both are relatively easy to perform. However, spinal anesthesia may provide more reliable intraoperative anesthesia, especially for mid/upper abdominal surgery in which large volumes of local anesthesia would be necessary *via* the caudal approach. Several infants undergoing lower abdominal or lower extremity procedures received caudal anesthesia and required sedative supplementation, resulting, in one patient, in the need for oxygen.[17] Also, one patient in that study required supplemental local

anesthesia infiltration, which should be avoided in epidermolysis patients. Spinal anesthesia has been shown to be safe, efficacious, and, indeed, advantageous for high-risk neonates and infants.[18,19] The use of spinal anesthesia without supplemental inhalational, intravenous, or intramuscular anesthesia has become popular for high-risk infants, to decrease the occurrence of postoperative apnea. However, this technique has not been previously described in infants with epidermolysis bullosa. We have demonstrated that the spinal anesthetic technique may be applied to these patients with minimal use of adjunct intravenous anesthetic agent to decrease upper body movement. The dose of tetracaine used in this patient was selected to produce a satisfactory anesthetic level while not exceeding T2-T4.[20] The infant in the current report had excellent anesthesia throughout the perioperative course, and experienced no new dermatologic lesions postoperatively. It is important to note that local anesthesia was not infiltrated subcutaneously before the spinal anesthetic to avoid tissue sloughing at the site of infiltration.

Although spinal anesthesia provided complete surgical analgesia, it did not provide adequate surgical conditions, because the infant was still moving his upper body. In other patients, such as high-risk infants, arm restraints or swaddling of the upper body is sufficient to restrict arm movement. However, in patients with epidermolysis bullosa, this maneuver may, of course, result in an augmentation of lesions. This was successfully prevented by the administration of propofol intravenously for sedation during surgery. The use of intravenous or intramuscular ketamine has become quite popular for epidermolysis bullosa patients[7,21,22] because it produces sedation/unconsciousness in addition to analgesia, while somewhat sparing respiratory drive. However, ketamine may also result in significant problems, including hypertension, tachycardia, and agitated movement, which may be distressing in patients with epidermolysis bullosa. Hamann and Cohen[4] report lip and palate desquamation from the use of a mouth gag in a patient receiving ketamine.

In summary, rigorous maneuvers are necessary to decrease exacerbation of existing lesions and to prevent the formation of new lesions in patients with epidermolysis bullosa. In addition to obvious dermatologic concerns, deliberate preoperative evaluation should be performed with respect to malnutrition, dehydration, chronic infections, anemia, and contractions, which are prevalent in these patients. We describe the successful use of spinal anesthesia in an infant with epidermolysis bullosa. This technique was not associated with any perioperative complications. Careful preoperative planning among anesthesiologists, surgeons, and dermatologists is crucial to providing a favorable anesthetic and postoperative course for patients with epidermolysis bullosa. Airway manipulation should be avoided if possible, and spinal anesthetic, as described in this report, represents a valuable alternative.

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