Regional Anesthesia in a Child with Epidermolysis Bullosa

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Epidermolysis bullosa (EB) is an hereditary disorder with dominant and recessive modes of genetic transmission.\(^1\) It is characterized by bullous formation in strati-
CASE REPORT

A 4-yr-old, 89-cm girl, weighing 10 kg, with autosomal recessive type EB, was scheduled for release of flexion contractures and co-occurring of the left hand, by skin grafting and local skin flaps. In addition to skin manifestations, she had recurrent oral bullae and brief periods of dysphagia. A trial of phenotoin failed to improve her condition. She was followed medically on a regular basis, and had a very stable and supportive family. Examination revealed cutaneous and intra-oral blistering and scarring, in various stages of formation and resolution. Her hand deformities were bilateral.

The child received no premedication the morning of surgery. Anesthesia was induced with 250 mg of thiopental rectally. As the thiopental effect dissipated, 40 mg of ketamine was given im, and another 40 mg was administered im 10 min later, to facilitate securing an intravenous line and monitoring devices. The latter consisted of a precordial stethoscope and an automatic blood pressure measuring unit. Xeroform® and Webril® were used to wrap the arm for both blood pressure and tourniquet cuffs. The stethoscope rested on Xeroform®, with a hole cut to the size of the stethoscope aperture. A skin temperature probe was placed in the right axilla. Oxygen was insufflated at 5 l/min over the mouth and nose.

Another 30 mg of ketamine iv was given to facilitate performing a left brachial plexus block, using an axillary approach. The axilla was bloomed with a povodine iodine solution, with care taken not to rub or abrade the skin. A nerve stimulator capable of 2 Hz and a current output of 0.5–1.5 ma was used to localize the plexus. The ground electrode was connected to a saline-soaked sponge placed under the patient's back, and the other electrode to an all metal 25-gauge 4-cm needle. The needle was placed as high in the axilla as possible, and directed toward the axillary artery. The needle direction was adjusted until maximal wrist flexions occurred at the lowest current possible. At this point, following negative aspiration for blood, 8 ml of 0.25% bupivacaine was injected, with complete cessation of hand movement.

During the first hour, surgery on the hand caused no alteration in heart rate, arterial blood pressure, ventilation, or movement by the patient to indicate inadequate anesthesia. Prior to harvesting of skin from the left thigh, 20 mg of ketamine and 7.5 µg of fentanyl were administered iv. Nevertheless, the patient moved and grimaced, and the blood pressure, heart rate, and respiratory rate increased as the incision continued. An additional 20 µg of fentanyl iv was given for harvesting of the skin.

Twelve hours passed before the patient complained of pain in the hand. The mother reported that the patient recalled awakening with pain during the procedure. Although the child could not tell the parent the location of the "hurt," it was presumed to have been in the leg at the time of the skin graft harvesting. In addition, the child told the mother of feeling herself being moved. We were unable to determine if this was related to intraoperative or recovery room events.

Eight months later, the patient returned for a similar procedure on the right hand. She now weighed 11.5 kg. The anesthetic was managed similarly to that used for the left hand procedure, except that, in addition to a right axillary block, 2.5 mg of diazepam was administered iv to reduce recall and 2.0 ml of 0.25% bupivacaine was injected as the needle was advanced toward the right anterior iliac spine from an insertion just inferior and medial to this structure. This was performed to block the right lateral femoral cutaneous nerve. There was no abrupt change in the patient's vital signs or activity to indicate inadequate anesthesia during the surgical manipulations. Following administration of the regional blocks until the end of each surgery, the patient received average hourly doses of 1 mg/kg of ketamine and 1.3 µg/kg of fentanyl during the first procedure (3 h of surgery), and 1.5 mg/kg of ketamine and 1.5 µg/kg of fentanyl during the second procedure (3.5 h of surgery).

DISCUSSION

Intraoperative anesthetic problems in the patient with EB relate primarily to bullae formation from trauma to squamous epithelium. This includes cutaneous, oral, pharyngeal, laryngeal, and esophageal surfaces. There is particular concern about lesions developing from face masks, oral Airways, laryngoscopes, and endotracheal tubes. Although the incidence of severe reactions is low when appropriate precautions are observed,2,3 ketamine and/or insufflation of inhaled volatile anesthetics have been the methods recommended, in order to minimize risks to the airway during surgery.4–6

Although regional anesthesia has been suggested,7 its use has not been described for EB, either as supplements to or as the main anesthetic. Reports that have addressed regional techniques have discouraged its use out of concern about physical and chemical trauma to skin, because contractures may make the technique difficult, the planned surgery was too extensive, and infected skin lesions were present or might occur at the site of needle placement.7–11 There may also be concern about performing regional anesthesia in pediatric patients because of the uncooperative nature of children when manipulated for regional techniques, the inability to be certain about needle placement near neural structures, and lack of knowledge and experience with regional techniques and local anesthetic doses in children. These problems can be readily addressed,12,15 and there is no reason why regional techniques cannot be considered and used for procedures such as the one described in this report.

Regardless of the anesthetic chosen, the anesthesiologist may be required to support the airway, because of respiratory obstruction or depression from any of the agents used. A soft, padded dressing should be used to protect the face during mask placement. Oral Airways, endotracheal tubes, and laryngoscope blades should be generously lubricated. The head and endotracheal tube should be secured to keep movement to a minimum. The tube can be kept in place with umbilical tape tied to it and wrapped around the head, using a soft dressing or padding between the tape and skin.

In summary, a child with EB was given two brachial plexus regional anesthetics, one of which was combined with a lateral femoral cutaneous nerve block. The authors maintain that EB should not be viewed as an absolute contraindication to regional anesthesia, but that this modality can be considered when it is judged to be an appropriate anesthetic technique for a patient with this condition.

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Hyperkalemia after Succinylcholine Administration in a Patient with Closed Head Injury without Paresis

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Succinylcholine-induced hyperkalemia can occur in many clinical conditions, both with and without skeletal muscle paralysis. The report by Stevenson and Birch describing hyperkalemia following succinylcholine administration to a patient recovering from a closed head injury is unique. As pointed out by Miller and Savarese, one should be hesitant in concluding that succinylcholine should not be administered to patients with a closed head injury on the basis of a single case report. This report presents another case of hyperkalemia associated with succinylcholine administration in a patient recovering from a closed head injury, and further justifies concern when administering succinylcholine to this group of patients.

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