Clinical Reports

Subarachnoid and epidural anaesthesia for patients with epidermolysis bullosa

Fred J. Spielman MD, Elizabeth S. Mann MD

Successful anaesthetic management of two patients with severe epidermolysis bullosa was accomplished using subarachnoid and epidural blockade. In order to avoid complications of regional anaesthesia, antiseptic preparation of the skin should not involve scrubbing nor should adhesive tape be used to secure catheters. In addition local infiltration of the skin is best avoided. The potential complications of skin and oral mucosal damage associated with mask or endotracheal inhalation anaesthesia was avoided.

Key words

COMPLICATIONS: epidermolysis bullosa;

ANAESTHETIC TECHNIQUE: subarachnoid, epidural.

Recessive epidermolysis bullosa (EB) is an uncommon but severe skin disorder characterized by disintegration of the subepidermal connective tissue in response to trauma, friction, or even pressure. The separation of the epidermis from the underlying dermis leads to bullae and scar formation. Dental defects and mucosal scarring in the oral cavity, pharynx and esophagus are often present.¹⁻³ Administration of anaesthesia to such patients is

challenging, particularly with regard to airway management. Excessive trauma from masks, airways or endotracheal tubes must be avoided in order to prevent new lesions and airway obstruction.⁴ Many patients have limited ability to open their mouths and immobility of the tongue, making intubation and airway control difficult. Regional anaesthesia has not been employed because of fear of infection and tissue sloughing secondary to local infiltration and antiseptic preparation of the skin. This case report describes the successful use of subarachnoid and epidural anaesthesia in two patients with severe epidermolysis bullosa.

Case one

A 155 cm 48-kg 20-year-old woman with recessive EB was scheduled for surgical treatment of vaginal agglutination. Her past medical history included several oesophageal dilatations with general anaesthesia for strictures. Physical examination revealed bullae and scarring on multiple areas of her body. Her mouth was small due to scarring at the lateral margins. Preoperative laboratory data were within normal limits except for haemoglobin of 85 g/l, for which she received two units of packed red blood cells prior to surgery. After prehydration with lactated Ringers solution 500 cc, providone-iodine (betadine) solution was poured on a lesion-free area at L₂₋₃, local anaesthesia was not used and a 25-gauge spinal needle was placed without difficulty. A hyperbaric solution of 10 mg tetracaine and ten per cent dextrose produced satisfactory analgesia to T₁₂ bilaterally. The 40-minute surgical procedure was uneventful. The patient was dis-

From the Department of Anaesthesiology, University of North Carolina at Chapel Hill, School of Medicine, Chapel Hill, North Carolina

Address correspondence to: Dr. F.J. Spielman, Department of Anesthesiology, University of North Carolina, School of Medicine, Chapel Hill, North Carolina 27514.

charged from the hospital on the second postoperative day without evidence of systemic or local infection or skin trauma at the site of the lumbar puncture.

Case two

A 152 cm 28-kg 30-year-old man with severe recessive EB was scheduled for establishment of a feeding gastrostomy. Past medical history included multiple oesophageal dilatations. However, the strictures progressed such that he was unable to chew, swallow, and pass enough food to maintain adequate nutrition. Physical exam revealed bullae and scar formation over most of his body. There was syndactyly and contractures of his fingers and toes. Many of his teeth were absent or in poor repair. He had limited ability to move his tongue or open his mouth. Preoperative laboratory data were within normal limits except for sinus bradycardia and a haemoglobin concentration of 11.5 g/dl. After prehydration with lactated Ringer's solution 500 cc, providone-iodine (betadine) solution was poured on a lesion-free area at L₂₋₃, and infiltration of the supraspinous and interspinous ligaments was performed with one per cent lidocaine. A 17-gauge needle was placed in the epidural space and a 20-gauge epidural catheter was inserted 3 cm without difficulty. It was not secured in place by adhesive tape. A total of 32 ml of 3.0 per cent 2-chloroprocaine produced excellent analgesia to T₅ bilaterally. The patient tolerated the operation well and was discharged from the hospital on the sixth postoperative day. There were no apparent anaesthesia-related sequelae.

In addition, six months prior to the above operation, this patient underwent debridement of ulcers on his left lower extremity under a subarachnoid block using a hyperbaric 5.0 per cent lidocaine solution. The anaesthetic was successful and without complications.

Discussion

Epidermolysis bullosa (EB) is a cutaneous disease in which slight trauma disrupts the cohesion between the epidermis and dermis, resulting in the formation of vesicles, bullae, and ulcers.⁵ Prior to surgery a concerted effort should be made to treat the malnutrition, severe anaemia and chronic infections that are common to patients with EB.⁶ Certain technical precautions decrease the hazard of creat-

ing new lesions. Adhesive tape should never be used in securing intravenous catheters, electrocardiogram leads and precordial stethoscopes. Blood pressure cuffs must be adequately padded and inflated to a minimal pressure above systolic blood pressure to obtain a reading. Careful positioning should be performed in order to minimize pressure on affected areas.⁷

No one anaesthetic technique has been shown to have an advantage over any other; however, ketamine has been quite popular for patients with EB.⁸⁻¹¹ It is felt to be an excellent technique because of its ability to provide rapid and adequate analgesia while maintaining intact pharyngeal-laryngeal reflexes.¹⁰ However, airway protection with ketamine is not absolute and aspiration can occur.¹² Ketamine does little to block visceral pain; it provides no muscle relaxation, and, in fact, may increase muscle tone. It is contraindicated in patients with hypertension, myocardial insufficiency, psychiatric disorders and increased intracranial pressure. Recovery is sometimes complicated by unpleasant dreams.¹³

There is considerable controversy as to whether general anaesthesia should be managed with a mask or intubation; however, both techniques may pose formidable problems. In severe cases, scarring and contractures of the mouth in addition to gross dental problems make intubation difficult. Immobility of the tongue due to bullae and ulcers may also complicate intubation. James¹⁴ found that 13 of 33 (40 per cent) of the patients he studied had restricted mouth opening, seven were difficult to intubate and two (six per cent) could not be intubated. Laryngoscopy and tracheal intubation can cause facial, labial, and oral bullae. A face mask for induction or maintenance of anaesthesia may cause bullae at the site of contact and under the chin due to the supporting fingers.¹⁵ Oropharyngeal airways may produce bleeding from the buccal mucosa and soft palate. Esophageal strictures, common in patients with EB, may increase the risk of regurgitation and aspiration.¹¹ When possible, all airway manipulations should be avoided in patients with EB.¹⁶

Although the literature on the anaesthetic management of EB contains statements such as "selected cases might qualify for a brachial plexus block"⁴ or "in the older patients, regional blocks are preferable to general anaesthesia,"³ we are aware of only one reported instance of regional anaesthesia

used in patients with EB,¹⁷ an axillary block in a four-year-old for hand surgery. The principal advantage of regional anaesthesia is the avoidance of airway manipulation. We have found adult patients with EB to be good candidates for regional anaesthesia. They are aware of and concerned about the hazards of general anaesthesia and acceptance of the mild discomforts associated with regional anaesthesia. Because of poor caloric intake and consequent thin habitus anatomic landmarks can be readily defined. Special considerations must be adhered to when performing regional anaesthesia. Antiseptic preparation of the skin should be done by pouring the solution over the area; scrubbing is contraindicated.⁷ Adhesive tape should never be used to secure epidural catheters. Local infiltration of the skin may produce bullae formation; but infiltration of the ligaments and muscles is safe. As in other patients, coagulopathy, local or systemic infection or patient refusal are contraindications to regional anaesthesia.

In conclusion, we have employed subarachnoid and epidural blockade in two EB patients, with excellent patient acceptance, and have observed no complications. The use of regional anaesthesia avoids the potentially serious complication of airway manipulation in these patients.

References

- 1 Thompson JW, Ahmed AR, Dudley JP. Epidermolysis bullosa dystrophica of the larynx and trachea, acute airway obstruction. *Ann Otol* 1980; 89: 428-9.
- 2 Marshall BE. A comment on epidermolysis bullosa and its anaesthetic management for dental operations. *Br J Anaesth* 1963; 35: 724-7.
- 3 Zarem HA, Pearson RW, Leaf N. Surgical management of hand deformities in recessive dystrophic epidermolysis bullosa. *Br J Plast Surg* 1974; 27: 176-81.
- 4 Hubbert CH, Adams JG. Anesthetic management of patients with epidermolysis bullosa. *South Med J* 1977; 70: 1375-6.
- 5 Nix TE Jr, Christianson HB. Epidermolysis bullosa of the esophagus: report of two cases and review of the literature. *South Med J* 1965; 58: 612-20.
- 6 Yonfa AE, Thomas JB, Labbe R, et al. Epidermolysis bullosa, a protocol for general anesthesia and successful dental rehabilitation. *Anesthesiol Rev* 1982; 9: 20-5.
- 7 Yonfa AE. Hand surgery and epidermolysis bullosa. In: *Common Problems in Pediatric Anesthesia*. Stehling LC (ed.). Chicago: Year Book Medical Publishers (1982) pp 219-23.
- 8 Hamann RA, Cohen PJ. Anesthetic management of a patient with epidermolysis bullosa dystrophica. *Anesthesiology* 1971; 34: 389-91.
- 9 Lee C, Magel EL. Anesthetic management of a patient with recessive epidermolysis bullosa dystrophica. *Anesthesiology* 1975; 43: 122-4.
- 10 LoVerme SR, Oropollo AT. Ketamine anesthesia in dermolytic bullous dermatosis (epidermolysis bullosa). *Anesth Analg* 1977; 56: 398-401.
- 11 Milne B, Rosales JK. Anaesthesia for correction of oesophageal stricture in a patient with recessive epidermolysis bullosa dystrophica: case report. *Can Anaesth Soc J* 1980; 27: 169-71.
- 12 Wood M. Intravenous anesthetic agents, drugs and anesthesia. In: *Pharmacology for Anesthesiologists*. Edited by Wood M, Wood AJJ. Baltimore: Williams and Wilkins (1982) pp 219-23.
- 13 Prevostnik SJ. Intravenous anesthesia. In: *Introduction to Anesthesia, The Principles of Safe Practice*. Edited by Dripps RD, Eckenhoff JE, Vandam LD. Philadelphia: WB Saunders (1982) pp 154-6.
- 14 James I, Wark H. Airway management during anesthesia in patients with epidermolysis bullosa dystrophica. *Anesthesiology* 1982; 56: 323-6.
- 15 Wilson F. Epidermolysis bullosa: a rare disease of anaesthetic interest. *Br J Anaesth* 1959; 31: 26-31.
- 16 Berryhill RE. Skin and bone disorders. In: *Anesthesia and Uncommon Diseases*. Edited by Katz J, Benumof J, Kadis LB. Philadelphia: WB Saunders (1981) pp 562-4.
- 17 Rowlingson JC, Rosenblum SM. Successful regional anesthesia in a patient with epidermolysis bullosa. *Regional Anesth* 1983; 8: 81-3.

Résumé

On a eu recours à la rachi-anesthésie et au bloc péri-dural pour l'anesthésie de deux patients affligés d'épidermolyse bulleuse. Pour prévenir les complications d'ordre dermatologique, le badigeonnage de la peau doit se faire sans frotage et on doit éviter l'emploi de tout pansement adhésif ou ruban gommé. Il est aussi préférable de ne pas infiltrer la peau localement. En procédant sous anesthésie régionale, les dommages à la peau ou aux muqueuses par les contacts du masque ou du tube endotrachéal ont été évités.