

Role of regional anaesthesia in a patient with epidermolysis bullosa dystrophica for emergency surgery

Sir,

Epidermolysis bullosa is a confluence of dermatological disorders causing easy bruising of skin and mucosa. It has a predilection to affect stratified squamous epithelium, thereby affecting the skin, oral cavity, pharynx, oesophagus and genitourinary system [Table 1]. Extracutaneous manifestations like laryngotracheal stenosis, oesophageal strictures, infections, hypermetabolic state and cardiomyopathy pose a great challenge to anaesthesiologists.

An 8-year-old male child weighing 12 kg had dystrophic nails with erosive desquamative lesions at birth which was followed by the development of blisters over palm, soles and scalp during the neonatal period. He was diagnosed with epidermolysis bullosa dystrophica. Complete haemogram and renal function tests were normal. The patient presented with acute urinary retention and meatal stenosis for which emergency meatoplasty was planned. On examination, healed atrophic scars were observed all over the body. Oral cavity examination revealed multiple hypoplastic teeth, multiple palatal and buccal mucosal erosions and an active bleeding ulcer on the dorsum of tongue with an external appearance of a difficult airway. Anaesthetic plan was intravenous sedation with caudal analgesia. Intravenous cannula was secured after a eutectic mixture of local anaesthetics cream application without occlusive dressing. The patient was premedicated with midazolam 0.5 mg and glycopyrrolate 120 µg intravenously.

Electrocardiography electrodes with adhesive removed were applied. A cotton roll was wrapped around the arm before blood pressure (BP) cuff application and clip sensor for pulse oximetry.

Intravenous sedation was provided with ketofol (ketamine + propofol) in 1:1 ratio at 0.5 mg/kg bolus dose. After lubrication of face mask with lignocaine jelly, oxygen supplementation on spontaneous breathing was maintained. Using landmark technique, after chlorhexidine disinfection, caudal block was performed in left lateral position with 7 mL of 0.25% bupivacaine. All pressure points were adequately cotton padded. Intraoperative and postoperative periods were uneventful.

Regional anaesthesia was preferred over general anaesthesia to minimise airway manipulation and trauma. We chose regional anaesthesia as meatoplasty is a short duration procedure and does not require absolute loss of muscle tone. We chose intravenous sedation over inhalational because the administration of short-acting inhalational agents requires a tight mask seal for end-tidal carbon dioxide monitoring, oxygen supplementation and anaesthetic agent delivery. A tight fit mask can cause skin disruption and blisters formation which could be disfiguring. The use of caudal block provided the added advantage of intraoperative and postoperative analgesia, and prevented adverse airway reflexes. Without caudal block, postoperative pain relief would have been difficult to manage and would lead to higher intraoperative sedative dose requirements which would predispose to transient apnoea requiring mask ventilation. Avoiding airway manipulation and instrumentation prevented the spontaneous blisters development in the airway tract, thereby reducing morbidity.^[2] Adequate analgesia is mandatory as children tend to roll around the table leading to skin trauma and blisters development.^[3] Caudal block made the procedure opioid-free as it can

Table 1: Epidermolysis bullosa clinical presentation^[1]

| Type | Inheritance | Defective Gene | Areas affected and Manifestations | Airway involvement | Special features |
|-----------------------------------|------------------------------|-------------------------------------|--|--|---|
| Epidermolysis bullosa simplex | Autosomal dominant | Keratin | Oral ulcers, nail shedding, hair loss, hyperkeratosis of the palms and soles | Rarely laryngeal stenosis | Blisters heal without scarring |
| Junctional epidermolysis bullosa | Autosomal recessive | Laminin | All areas lined with stratified squamous epithelium – skin, gastrointestinal mucosa, respiratory mucosa and genitourinary mucosa | Scarring and obstruction | Manifests since birth and carries a high mortality rate |
| Epidermolysis bullosa dystrophica | Autosomal dominant/recessive | Alpha-7 chain integrity in collagen | Oral, oesophageal and anal mucosa. Dilated cardiomyopathy, pseudo-syndactyly, oesophageal strictures leading to malnutrition and anaemia | Limited mouth opening and laryngotracheal stenosis | Multiple blister formations all over the body even with minimal mechanical insult |

lead to pruritus. Pruritus is deleterious as scratching makes things worse. Other measures that we undertook to avoid shear stress and formation of new blisters were lubrication of face mask with lignocaine jelly, adequate cotton padding over pressure points, cotton wrapping before application of BP cuff and clip type pulse oximeters.^[4] Non-adhesives can be used for fixing intravenous cannulas to avoid medical adhesive-related skin injuries.^[5]

If these patients require general anaesthesia, then it is better to avoid supraglottic airway and oropharyngeal airways as they can worsen oral mucosal integrity. Nasopharyngeal airways and nasal intubation can be used as the columnar epithelium of the nasal cavity is slightly more resistant to disruption.^[6] All airway equipment should be lubricated well to minimise airway trauma.^[7] Other intraoperative complications observed in epidermolysis bullosa include fluid loss, electrolyte disturbances, anaemia, hypoproteinaemia and nutritional disturbances.^[1]

To conclude, in the present case of epidermolysis bullosa, the use of caudal anaesthesia with sedation during the surgical procedure provided a successful outcome.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

**Thirumurugan Arikrishnan, Deepak Chakravarthy,
Vijayanthi Vijayan, Gnanasekaran Srinivasan**

Department of Anaesthesiology and Critical Care, Jawaharlal Institute of Postgraduate Medical Education and Research, Puducherry, India

Address for correspondence:

Dr. Thirumurugan Arikrishnan,
No. 75, Mariamman Koil Street, Thattanchavady,
Puducherry - 605 009, India.
E-mail: a.thiru1990t@gmail.com

Submitted: 04-Apr-2021

Revised: 18-Nov-2021

Accepted: 06-Dec-2021

Published: 22-Dec-2021

REFERENCES

1. Wittkugel E, Kandil A. Anesthesia for epidermolysis bullosa. In: Goudra BG, Duggan M, Chidambaram V, Venkata HPK, Duggan E, Powell M, Singh PM, editors. *Anesthesiology: A Practical Approach*. 1st ed. Cham: Springer; 2018. p. 421–8.
2. Aikawa K, Tanaka N, Morimoto Y. Anesthetic management with subcostal transversus abdominis plane block in recessive dystrophic epidermolysis bullosa for peritoneal dialysis catheter replacement: A case report. *JA Clin Rep* 2018;4:37.
3. Bowen L, Burtonwood MT. Anaesthetic management of children with epidermolysis bullosa. *BJA Educ* 2018;18:41–5.
4. Lin AN, Lateef F, Kelly R, Rothaus KO, Carter DM. Anesthetic management in epidermolysis bullosa: Review of 129 anesthetic episodes in 32 patients. *J Am Acad Dermatol* 1994;30:412–6.
5. Bloria S, Chauhan R, Luthra A, Bloria P. Medical adhesive-related skin injuries caused by taping of the eye using acrylic-based adhesive tapes in prone surgery: A case report. *Indian J Anaesth* 2020;64:345–6.
6. Saraf SV, Mandawade NJ, Gore SK, Padhye UD, Pereira CS. Epidermolysis bullosa: Careful monitoring and no touch principle for anesthesia management. *J Anaesthesiol Clin Pharmacol* 2013;29:390–3.
7. Singh S, Ray A, Gupta N, Sethi N. Anaesthetic management of a rare case of paediatric epidermolysis bullosa. *Indian J Anaesth* 2019;63:73–4.

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

| Access this article online | |
|--|--------------------------------|
| Quick response code | Website: www.ijaweb.org |
|  | DOI: 10.4103/ija.ija_287_21 |

How to cite this article: Arikrishnan T, Chakravarthy D, Vijayan V, Srinivasan G. Role of regional anaesthesia in a patient with epidermolysis bullosa dystrophica for emergency surgery. *Indian J Anaesth* 2021;65:906–7.

© 2021 Indian Journal of Anaesthesia | Published by Wolters Kluwer - Medknow