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Caesarean delivery in a pregnant woman with epidermolysis bullosa: anaesthetic challenges

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ABSTRACT
Epidermolysis bullosa is a heterogeneous group of hereditary diseases characterised by extreme fragility of skin and mucosa, with blister and lesion formation spontaneously or in response to trauma. Anaesthetic management of these patients is challenging with respect to positioning, monitoring, use of medical devices and airway management. These challenges are increased when managing labour. We report an elective caesarean delivery in a nulliparous woman with autosomal recessive dystrophic epidermolysis bullosa, managed successfully with spinal anaesthesia.
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Keywords: Epidermolysis bullosa; Regional anaesthesia; Caesarean delivery

Introduction

Epidermolysis bullosa (EB) is a heterogeneous group of hereditary disorders in which extreme fragility of the skin and mucous membranes leads to the formation of blisters and ulcers, spontaneously or following minor trauma.[1,2] Extracutaneous involvement may also occur, dependent on the type of EB, with manifestations in skin, adnexa, teeth, the gastrointestinal and urinary tract and pulmonary epithelia.[1]

The incidence of EB is 1 in 17 000–300 000 live births, with an estimated 500 000 cases worldwide. It is not affected by race or ethnic group, and affects both sexes equally.[1,3]

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Although more than 20 subtypes of EB are recognized, the three main types are simplex EB, dystrophic EB and junctional EB. The classification is based on how deep in the skin layers the blisters form, and multiple subtypes are recognised.[1,2]

Simplex EB is the most common presentation of the disease (92% of cases) and is mainly inherited as a dominant trait. Blisters appear at birth, develop in response to trauma, and often primarily affect the palms of the hands and soles of the feet. Usually their frequency diminishes with age.[1]

Dystrophic EB (5% of cases) is caused by mutations that affect collagen VII and can be inherited as a dominant or recessive trait; the rarer recessive forms are generally more severe. Blistering is present from birth and depending on the subtype, this may be localized or generalised; the lesions leave dystrophic scars on healing.[1]

Junctional EB (1% of cases) has an autosomal recessive pattern and lesions can be either localised or generalised. All variants of junctional EB with one exception manifest at birth.[1]

In all EB cases, the repetitive process of blistering and subsequent resolution can result in severe scarring, mutilation, atypical cell growth, debilitating conditions and can be life-threatening.[2,4]

Anaesthetic management of EB requires careful planning to avoid trauma to the skin and mucosa, which can lead to new lesions or deterioration of pre-existing lesions, resulting in significant intra- and post-operative complications. Particular care is required with patient positioning, preparation of the skin aseptically and the use of routine monitoring and medical devices to deliver anaesthesia.[2,5–10]

Labour management further increases the challenges. Reports published about management of these patients are sparse but all stress the need for a multidisciplinary approach between anaesthetists, obstetricians and neonatologists, to plan the delivery.[11–14] Different anaesthetic techniques have been used successfully in patients with EB.[11–17] More important than the technique used is the careful management of these patients and the "no touch principle".[4]

Case report

A 26-year-old nulliparous woman (46 kg, 1.56 m), with dystrophic EB (autosomal recessive), presented to the delivery unit at 36 weeks' gestation. The patient had a history of oesophageal atresia, with multiple general anaesthetics for oesophageal dilatation. No airway management issues were reported. At presentation she reported only some minor difficulties with solid food intake, but denied gastro-oesophageal reflux or heartburn.

She had a long history of generalised blister formation, including in the oral cavity, occurring spontaneously or after minimal friction. In early pregnancy a squamous cell carcinoma was diagnosed on her right hand, requiring amputation of the hand. This procedure was performed under a brachial plexus block, without complication. No chemotherapy or radiotherapy was needed and the patient decided to proceed with the pregnancy.

The patient underwent anaesthetic review at 30 weeks' gestation. She presented with bullae and cicatricial lesions all over the body, including on the back. Some of these lesions had an inflammatory pattern. Airway evaluation revealed limited mouth opening (~3 cm), a Mallampati score of 3, with poor dentition and inability to protrude her tongue (Fig. 1a). Neck

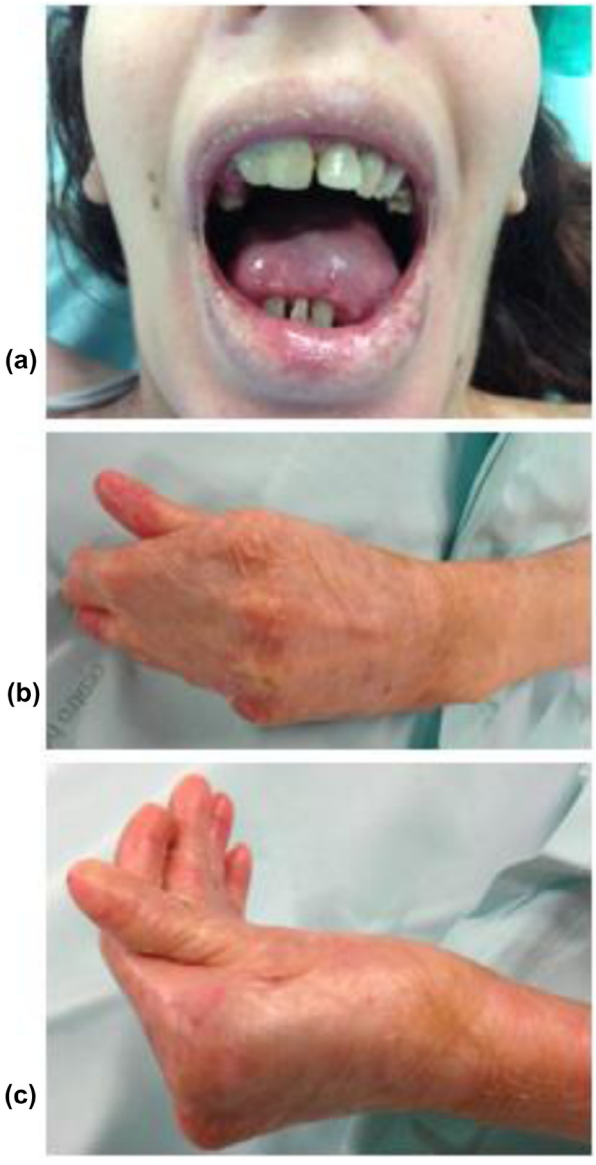

Fig. 1 Patient physical examination. (a) Limited mouth opening, with no ability to protrude the tongue. (b, c) Pseudosyndactyly and poor peripheral venous access as a result of repeated blister formation

mobility and thyromental distance were normal. Due to pseudosyndactyly in the left hand (Fig. 1b, c) and the scaring lesions throughout the body, peripheral venous access was expected to be difficult.

She was found to be anaemic (haemoglobin 8.4 g/dL) and was transfused with one unit of packed red cells. Platelet count, coagulation, albumin and biochemistry tests were normal.

After a multidisciplinary team discussion including the obstetrician, anaesthetist, neonatologist and patient, a scheduled caesarean delivery at 38 weeks' gestation and under spinal anaesthesia was planned.

The patient was advised to avoid friction on her back and to apply moisturising cream on the skin to minimise the effects of friction and skin stretching on pre-existing lesions and the development of new lesions.

Caesarean delivery was performed at 36 weeks' gestation, two weeks earlier than planned, due to the appearance of a right axillary swelling that required urgent investigation and treatment, being a suspected metastasis of the squamous cell carcinoma. She was admitted to hospital on the day before surgery. Blood was collected for blood count, coagulation status and for holding of red blood cells.

On the day of surgery no premedication was given. The perioperative team took a variety of extra precautions. The surgical table was padded with a standard gel mattress. The patient positioned herself in a wedged supine position on the surgical table, avoiding aorto-caval compression, and was monitored with an ear probe pulse oximeter, non-invasive blood pressure with underlying skin wrapped in gauze (Fig. 2a) and 5-lead ECG with a silicone surface between skin and electrodes (Fig. 2b). Peripheral venous access (20 gauge cannula) was achieved in the left arm and the cannula secured with gauze (Fig. 3) due to the lesions present in both superior and inferior limbs (Fig. 4a). Intravenous pantoprazole 40 mg and metoclopramide 10 mg were administered. An anaesthetist with experience in difficult airway management was present in the operating room and devices for difficult intubation (different size blades,

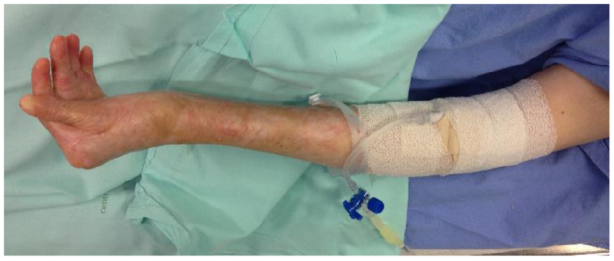

Fig. 3 Peripheral venous access, properly secured with gauze

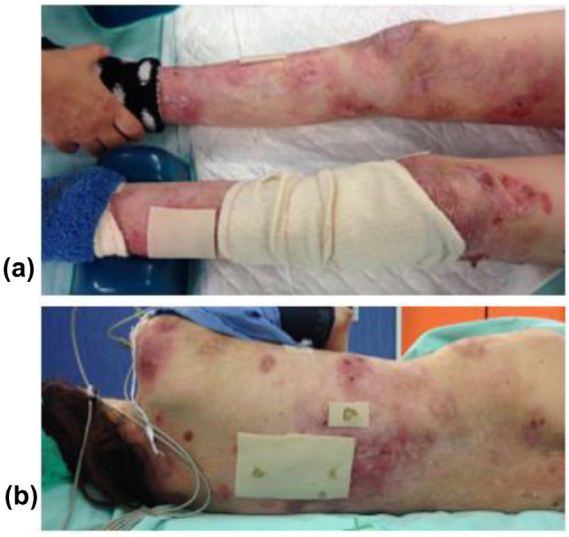

Fig. 4 Skin lesions in both lower limbs (a) and back (b) on the day of delivery

McCoy blade, C-Mac® videolaryngoscope and a fibrescope) were available.

The patient then turned herself to the left lateral decubitus position. Some areas of her back were free of lesions (Fig. 4b) and the lumbar skin was gently prepared with povidone iodine solution; friction on the skin was avoided and the skin preparation was allowed to dry spontaneously (Fig. 5a). Sterile lubricant gel on

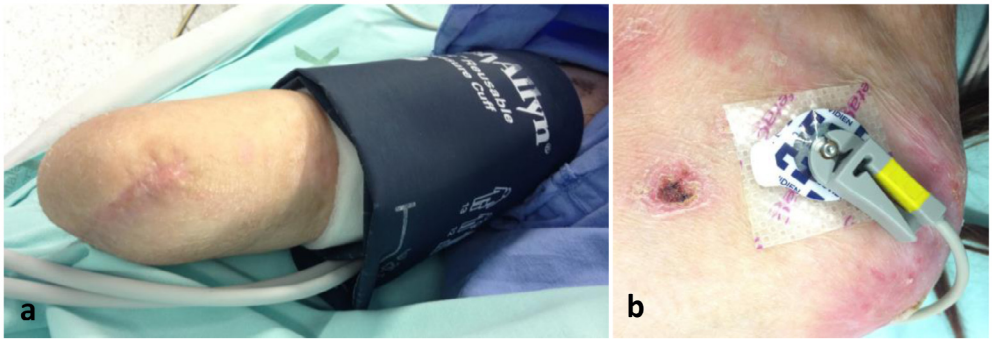

Fig. 2 Special features of standard monitoring. (a) Non-invasive blood pressure measurement, with underlying skin wrapped in gauze. (b) Silicone surface between skin and ECG electrodes

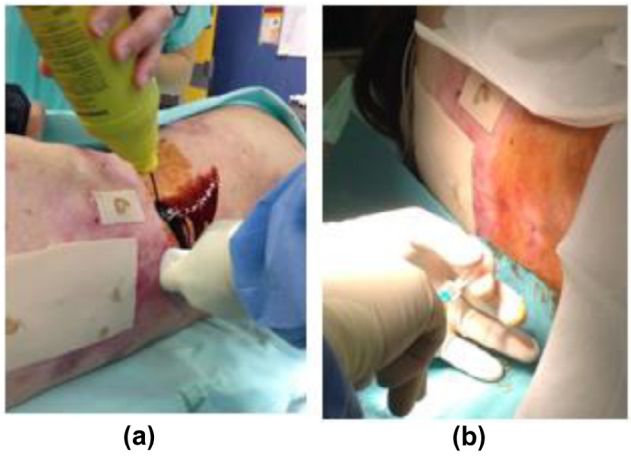

Fig. 5 Gentle aseptic preparation of skin, without friction (a), preparing to perform the spinal block (b)

the gloves was used to aid atraumatic palpation and puncture. The skin over the L3–4 interspace was infiltrated with lignocaine 1% and a single puncture was performed with a 27 gauge Whitacre spinal needle. Heavy bupivacaine 0.5% (7.5 mg) and sufentanil 2.5 µg were administered (Fig. 5b). The puncture site was then protected with a non-adhesive dressing and the patient turned herself back into the wedged supine position.

A T6 level sensory block was achieved. Initial hypotension due to sympathetic blockade was managed with two fractionated boluses of phenylephrine 100 µg. The patient complained of pruritus and this was managed with intravenous ondansetron 4 mg. Antibiotic prophylaxis (cephazolin 2 g) was administered before the surgical incision. Surgical skin disinfection was performed with povidone iodine, without friction, and non-adhesive surgical field drapes were used.

The procedure was performed without monopolar diathermy and lasted 40 minutes, with no complications. Oxygen was given via a face mask placed next to the patient's face, to avoid direct contact; oxygen saturation remained above 97% throughout. Haemodynamic stability was achieved after the initial hypotension. Estimated blood loss was 300 mL.

Intravenous paracetamol 1 g and ketorolac 30 mg were given for post-operative analgesia. At the end of surgery, the patient was moved carefully to the bed and transferred to the recovery room, where she was monitored for a further two hours without complications.

She was discharged home four days later, without complications. The surgical wound was healing well.

Discussion

Patients with EB present a challenge to the anaesthetist. Difficulties increase when managing a pregnant woman with EB for delivery due to the physiological changes of pregnancy and the features of EB.[12–17] A multidisciplinary approach to plan the mode of delivery and the anaesthetic technique is essential.[16]

Previous studies have shown no difference in outcome between caesarean and vaginal delivery.[11] In our case the decision to perform a caesarean delivery was based on obstetric data. Lesions in the vaginal canal and the risk of developing new blisters during a long labour were of concern. Caesarean delivery also permitted a more controlled delivery at which appropriately trained staff and equipment were available.

Table 1 Management points when caring for patients with epidermolysis bullosa

Standard monitoring	Pulse oximeter: no concerns
	Non-invasive blood pressure: underlying skin wrapped in gauze
	Electrocardiography: silicone surface between the patient and the electrodes
Positioning	Encourage auto-positioning
	Positioning without friction
	Gel mattress for body and limbs
Peripheral venous canulation	Can be difficult
	Secured with gauze
Surgical fields, dressings, protections	Non-adhesive
Skin asepsis	Without friction
	Left to dry spontaneously
Airway management	Nasal cannula/face mask lubricated
	Other devices (laryngoscope blade, tracheal tube, laryngeal mask) lubricated
	Avoid friction with facial skin
	Difficult airway management can occur
	Trauma, bleeding, new lesions in oral mucosa
Regional anaesthesia	Avoid puncture in areas with skin lesions
	Neuraxial anaesthesia and peripheral nerve blocks should be performed
	Catheters for intra- and postoperative analgesia can be used (tunnelled or fixed with non-adhesive dressings)
Surgical concerns	Avoid the use of monopolar diathermy
	Wound healing can be problematic

The choice of anaesthetic technique was based on consideration of airway management. Studies have reported airway issues in EB patients, with 23% incidence of difficult intubation.[3,4,18–20] The physiological changes of pregnancy can exacerbate this problem.

Reports describe both spinal and epidural anaesthesia for caesarean delivery, and epidural analgesia for vaginal delivery, without complication.[11–17] Some studies report the tunnelling of the epidural catheter, with similar results to when non-adhesive dressings are used. Our choice of spinal anaesthesia, rather than epidural or spinal-epidural anaesthesia, was based on the use of a simpler technique that might minimise trauma and pressure on the skin. Moreover, an epidural catheter during the intra- and postoperative period would increase the risk of the appearance of new lesions.[9]

A particular concern was that of intravenous access, which is likely to be difficult for patients with EB. A peripheral intravenous cannula was used; this was a considered choice, but equipment for central venous access was available had peripheral access failed or been inadequate. The possible need for general anaesthesia was considered, for example if skin lesions on the back precluded neuraxial anaesthesia or if spinal anaesthesia itself proved inadequate. Difficult airway equipment and trained personnel were available to manage this.

Knowledge of EB is indispensable in order to avoid worsening the clinical manifestations of the disease.[7–10] Table 1 summarises points of care inside and outside the operating room.

In summary, this case report describes the successful management of a caesarean delivery under spinal anaesthesia in a patient with dystrophic EB. Anaesthetic and obstetric management of patients with EB is a challenge. A multidisciplinary approach is essential and plans should be formulated at an early stage.

The method of delivery and provision of anaesthesia must take into account patient positioning, the use and positioning of monitors, and careful anaesthetic, surgical and nursing care. Prevention of new lesions is a primary aim.

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