Ultrasound-guided axillary brachial plexus blocks for pseudo-syndactyly surgeries in a patient with epidermolysis bullosa: a case report

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Abstract: Epidermolysis bullosa (EB) is a rare genetic disease characterized by recurrent blister formation following injuries or traumas. In patients with EB, general anesthesia may result in potential airway obstruction, aspiration and prolonged hospital stay due to airway instrumentation-associated new bullae formation and scarring. On the other hand, regional anesthesia has been shown to be efficient and safe. Ultrasound (US)-guidance particularly provides additional benefits to this clinical situation by minimizing skin contacts, improving block success and preventing complications. We describe a patient with EB, whose bilateral pseudo-syndactyly surgical treatment was successfully managed by the use of US-guided axillary brachial plexus blocks.

Key words:

INTRODUCTION

Epidermolysis bullosa (EB) is a group of inherited skin and mucous membrane disorders, characterized by non-inflammatory recurrent blister, healing and scar formation that follows mechanical injury or trauma. The blisters may appear after heat, rubbing, scratching, electrodes, adhesive tapes, blood pressure cuff, facemask, airway instrumentation, or alcohol wipes. They may occur on the skin as well as the eyes and mucosal surfaces, including oral cavity, oropharynx, larynx, esophagus, stomach, intestines, lungs, bladder, and genital region (1-4). Scarring may result in airway obstruction or chewing and swallowing difficulties, leading to chronic malnutrition, slow growth, and cachexia. Avoiding mechanical injury of skin and mucous membranes, and opting for minimally invasive techniques are prerequisites for these patients during any anesthesia management.

Brachial plexus block has been used as a regional anesthesia technique for decades. It has become a mainstay of the anesthesiologist’s armamentarium, particularly following the introduction of ultrasound (US)-guidance to clinical practice, improving success and reducing complication rates (5-8). Preference of a proper regional anesthesia technique over general anesthesia, and meticulous performance under US-guidance may be associated with lower peri-/post-operative morbidity and mortality rates, particularly in subgroups of patients with co-morbidities.

We report the case of an EB patient, whose bilateral pseudo-syndactyly surgical treatment was successfully managed using US-guided axillary brachial plexus blocks (ABPB).

CASE REPORT

After obtaining written informed consent, a 24 year-old male (176 cm and 40 kg), ASA physical status III patient with EB was scheduled twice for contracture release of his both mitten (pseudo-syndactyly) hands at 6-month intervals, in the Department of Plastic Surgery, Division of Hand Surgery. He was diagnosed with EB when he was a newborn with widespread blistering. Complications in the form of progressive scarring into the oropharynx and esophagus, chronic malnutrition, cachexia, fusion of fingers and toes, cicatricial tissue formation, loss of fingernails and toenails, joint contractures restricting movement, and recurrent skin infections occurred by time. His physical examination revealed numerous blistering and disfiguring scars on his body, and hypodontia. The Mallampati score was III, and the patient had restricted mouth opening (Figure 1). He had no additional disease in his past medical history.

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After an inspection of the axillary region to exclude an EB-related lesion and/or infection, and after discussion with the surgical team, we decided to perform an US-guided ABPB to minimize the risk of new bullae formation on the face and/or in the oral cavity by mask ventilation, direct laryngoscopy, and also to avoid a difficult intubation scenario.

Once in the operating room, pulse-oximetry was applied for monitoring. A 22-gauge intravenous (IV) cannula was placed in a vein of the contralateral hand, and an infusion of normal saline was started. Once having checked for normal vital signs, an intravenous sedation was induced using 1 mg of midazolam and 25 µg of fentanyl. Supplemental oxygen was administered through a facemask at a 2 L/min flow.

With the patient supine, the head was turned away from the side to be blocked. The arm was abducted to 90°, to allow for US probe and needle placement. After disinfection of the skin with chlorhexidine gluconate 0.5% in 70% denatured ethanol, the linear US probe (5-13 MHz, GE Healthcare, Wauwatosa, USA) covered with a sterile US sleeve was positioned in the short axis orientation. The musculo-cutaneous nerve between the biceps and the coraco-brachialis muscles, as well as the hyper-echoic median, ulnar and radial nerves were identified in the vicinity of the axillary artery. Following a 2 mL of 2% lidocaine skin infiltration, a 22-gauge 50 mm insulated needle (Stimuplex A, B Braun, Melsungen, Germany) was inserted using an in-plane technique. It was directed toward the musculo-cutaneous nerve, and posterior and anterior aspects of the axillary artery, respectively. Then, a mixture of 2% lidocaine and 0.5% bupivacaine (50/50 volume/volume) was injected in fractioned doses of 4 mL, 8 mL and 8 mL after repetitive negative aspirations for blood. The local anesthetic (LA) spread was verified. The patient did not report any paresthesia or discomfort during the procedure. Sensory (7/8 sensory loss to pinprick test on the skin for four terminal brachial plexus nerves [0: normal sensation, 2: no sensation]) and motor (10/12 motor blockade for four terminal brachial plexus nerves [0: normal motor function, 3: complete motor block, unable to move the arm, elbow, or fingers]) blocks were both assessed, and considered successful at 30 minutes. Thereafter, intravenous sedation using 1 mg of midazolam and 25 µg of fentanyl was repeated.

Surgery involved a transverse incision on the distal palmar crease, and vertical incisions on the web spaces. Fingers were extended, and K-wires were applied to all of them (Figure 2). Surgery was completed uneventfully within approximately 2 hours. This procedure was initially performed on the right side, and 6 months later on the left side. The patient was very satisfied with the anesthetic technique and management of acute postoperative pain. Hence, the second procedure used the same sedation protocol and loco-regional anesthesia technique. After surgery on the right hand, the patient was pain-free for 16 hours [numeric rating scale ≤3 (NRS: 0: no pain, 10: worst pain imaginable)]. Postoperative analgesia was insured using 1g of intravenous paracetamol twice daily only. Following the second surgery, the patient described pain (NRS ≥4) at post-operative hour 14. The same analgesic order was applied. He did not require any supplemental analgesic medications. No complications are to be mentioned until discharge on post-operative day 2 after both interventions.

**Discussion**

Airway instrumentation is generally required during general anesthesia (GA). However, in patients with EB, any attempt to insert a laryngeal mask airway or a tracheal tube can cause new bullae formation, and scarring in the oral cavity (9-11). This can lead to laryngospasm, airway obstruction, and aspiration. As a consequence, lengthening
of hospital- and/or intensive care unit stay may occur, with increased post-operative morbidity and mortality. Another potential hazard in these patients is the presence of restricted mouth opening, possibly leading to difficult intubation (3). They may also have an increased sensitivity to non-depolarizing muscle relaxants (12). Hence, general anesthesia should not be the first choice, whenever possible.

The classical regional anesthesia advantages, including minimized stress, simplicity, efficacy, good surgical conditions, early post-operative analgesia, and outpatient comfort, are also valid for EB diagnosed patients (11-14). As reported in Chevaleraud et al. (14), prevention from blistering is essential. In comparison with GA, regional anesthesia does not raise the problem of airway management. Therefore, anesthesia and analgesia techniques that insure spontaneous breathing, and avoid adverse events/complications are preferable for these patients. This must, however, be adapted as a function of the concerned procedure.

The use of a brachial plexus block approach in these patients has already been reported for several upper extremity procedures (3,11,13-16). For example, Meola et al. (3) performed a successful axillary brachial plexus block with catheter placement, and using a continuous intravenous ketamine and remifentanil sedation. Their patient had EB, as well as xeroderma pigmentosa. After failed attempts of oro-tracheal intubation and laryngeal mask airway insertion, loco-regional anesthesia achieved adequate pain control for the major orthopedic surgery to be performed. Other authors (13) have described the use of a bilateral axillary and midhumeral nerve block in two patients with an anticipated difficult airway due to EB. Based on their experience of 185 procedures in 157 cases, Chevaleraud et al. (14) emphasized the value of first-line regional anesthesia, particularly in the upper limb, even in very young children (≤3 years). For cases requiring hand surgeries, they advocated the use of regional plexus blocks as the sole anesthetic techniques.

US-guidance provides additional efficiency and safety for regional techniques, as the anesthesiologists are then able to view the image of the target nerve directly, insert the needle through the skin only once, guide it under real-time observation, monitor the spread of LA, and avoid complications by visualizing the hazardous structures (17). US-guidance helps shortening the time needed to perform the nerve block, reduces the number of needle passes, resulting in faster onset and longer duration times. US-guidance also

![Image](image-url)

Fig. 2. — Pseudo-syndactyly in our patient with Epidermolysis Bullosa. A. Pre-operative right hand, B. Post-operative right hand, C. Pre-operative left hand, D. Post-operative left hand.
improves the success rate, decreases the incidence of complications, as well as perioperative morbidity (18-20). US-guided nerve blocks are progressively becoming the gold standard, particularly in the high-risk patient demanding a wider choice of surgical and anesthetic options, as to ensure optimal clinical care and decreased risk of complications (17, 21). The use of a single shot ABPB was already recommended in 1995 for EB patients (14, 15). To our knowledge, only one case of EB patient undergoing surgery under US-guided ABPB has been reported so far in the literature (11). The authors shared their successful experience of ABPB combined with sedation/anaesthesia in a pediatric patient. As anticipated, their anesthesia plan provided good surgical conditions and postoperative analgesia, minimized stress, and avoided the manipulation of the airway.

In our case, guidance by neuro-stimulation to seek for and identify nerves was not helpful. Indeed, contractures and pseudo-syndactyly were confounding the obtained responses. After discussing with the surgeon, and balancing the risks and benefits for the patient, we chose to perform an US-guided ABPB under sedation for both anesthesia and post-operative analgesia. Besides achieving successful anesthesia and analgesia, US-guidance also added the advantage of minimizing possible contacts with the skin, and prevented from excessive palpations to identify anatomical landmarks.

In EB patients, infection or inflammation may accompany blistering and scarring. Close attention has to be paid to block preparation and performance, that must be strictly sterile. Significant risk for contamination and colonization of needles and catheters has been demonstrated following preparation with 10% povidone-iodine (22, 23). We therefore preferred to apply a chlorhexidine/ethanol solution, which has been reported to be efficient at disinfecting the skin before regional anesthesia (24). A sterile US sleeve to cover the probe (25), and sterile gel were both used during the ABPB procedure.

In our patient, US-guided ABPB provided excellent surgical anesthesia and post-operative analgesia without new bullae formation or scarring. This occurred for the two pseudo-syndactyly surgeries at a 6-month interval. US-guidance also provided greater confidence in block success, with minimal mechanic injury. As a further recommendation, anesthesiologists should clarify EB patient history with regard to frequently associated co-morbidities and clinical features, particularly in terms of mucosal involvements.

The anesthesia/analgesia technique should also be planned according to the concerned surgical procedure.

References


