

Multimodal Analgesia Using Dexmedetomidine and No Narcotics in an Epidermolysis Bullosa Patient Undergoing Unilateral Syndactyly Releases and Full Thickness Skin Grafting

C S Soria, M Greenberg

Citation

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Abstract

The patient is an 11-year-old girl with severe epidermolysis bullosa dystrophica with chronic sloughing, bilateral hand contractures, and syndactyly, who was scheduled for outpatient syndactyly releases and full thickness skin grafting. Pain control posed a significant challenge because the patient had a poor tolerance for opioid analgesics, with side effects including severe pruritus and constipation. The sole analgesics used perioperatively included dexmedetomidine, acetaminophen, ketorolac, and local anesthetic injection, with complete avoidance of opioids.

INTRODUCTION

Epidermolysis bullosa (EB) is a rare genetic mechanobullous disorder characterized by excessive fragility of the skin and mucous membranes. Most patients are children who do not survive past early adulthood due to failure to thrive, sepsis, skin cancer, and respiratory failure. This is especially profound in the EB population, where patients have lifelong pain and high narcotic tolerance. Current literature on anesthetic management of EB patients advocates for multimodal analgesia, including systemic opiate and non-opiate analgesics, local infiltration of the surgical field, peripheral nerve blocks, and neuraxial techniques. Anesthesiologists are advised to use narcotics carefully, as narcotic usage can precipitate postoperative nausea, vomiting, and pruritus. These side effects can have devastating consequences for EB patients, leading to painful new skin lesions and esophageal trauma requiring prolonged hospitalization for feeding, pain management, and dressing changes (1, 2). In this manuscript, we demonstrate that dexmedetomidine is a major analgesic that can be used to minimize narcotic requirements for chronic pain conditions as severe as EB. There is no description in current literature of a major surgery in an EB patient where pain was well-controlled without the use of any narcotic.

CASE REPORT

The patient is an 11-year-old girl, 1.17 meters tall, 17 kg, with severe epidermolysis bullosa (EB) dystrophica, with chronic sloughing, bilateral hand contractures, and syndactyly. She was scheduled to undergo left hand skin excision, de-cocooning with first web space deepening, syndactyly releases, and full thickness skin grafting. At home, her chronic pain regimen included modest amounts of oral morphine and ibuprofen, and spreading cannabidiol oils over her skin. The cannabinoid was most effective and her mainstay of therapy. With narcotics, she would develop significant pruritus and constipation. In the past, the pruritus led to such severe scratching of her skin that she required readmission for management of resulting skin ulcers. She also had presented for multiple surgeries and wound care treatments in the intensive care unit throughout her lifetimes. Over time, she had developed an increased tolerance to narcotic analgesics.

The patient and her family requested avoiding narcotics as much as possible. We discussed with the patient and surgeon the possibility of performing a regional block such as a brachial plexus block under general anesthesia to assist with pain control. A regional technique would decrease anesthetic

and systemic analgesic requirements. The family elected not to receive a regional block. Instead, the plan was for us to use primarily dexmedetomidine for systemic analgesia and for the surgeon to perform local anesthetic infiltration of the surgical field.

The patient walked herself to the operating room (OR) and positioned herself on the OR table. Routine monitors were placed with caution to minimize unnecessary pressure, avoid adhesives, and provide extra lubrication of all equipment in contact with the patient's skin. Due to the long duration and extensive nature of the surgery, which would require careful positioning and periods of high stimulation, we elected to perform the surgery under general anesthesia with an endotracheal tube. A laryngeal mask airway was not used to minimize the risk of trauma to the oropharynx and larynx from sliding a relatively large silicone structure down the mucosa.

A gentle inhalational mask induction was performed with 8% sevoflurane. The anesthesia resident provided gentle chin lift to assist with ventilation, and the patient was maintained on spontaneous ventilation. The anesthesia attending established IV access without a tourniquet using a 22 G catheter. The patient was induced with propofol 3 mg/kg and vecuronium 0.1 mg/kg. Gentle direct laryngoscopy with a Miller 2 blade was performed and a 4.5 cuffed endotracheal tube. Anesthesia was maintained with a 0.5% sevoflurane and a propofol infusion of 200 mcg/kg/min. The low concentration of sevoflurane was enough to ensure lack of awareness. The anesthetic based primarily on a propofol infusion was intended to minimize postoperative nausea and vomiting, which would cause new esophageal and pharyngeal lesions.

Dexmedetomidine boluses of 0.5-1 mcg/kg were provided throughout the case, for a total of 25 mcg of dexmedetomidine. Additionally, weight-based dosing of acetaminophen 15 mg/kg and ketorolac 0.5 mg/kg were given just prior to emergence from anesthesia. The plastic surgeon first used a knife and tenotomy scissors to perform a syndactyly release of the second, third, and fourth web spaces. A full thickness skin graft was then harvested from the left groin and sutured over the web spaces and volar aspects of the fingers. The wounds were dressed with bacitracin ointment and covered with xerform and kling wrap, and secured in a long arm cast to pad bony prominences. Prior to dressing application, the surgeon injected a total of 10 cc of 0.25% bupivacaine for local

anesthetic. The patient was extubated in the operating room while deeply anesthetized to decrease the risk of airway trauma from coughing and bucking. In the recovery room, the patient was comfortable and free of pain and pruritus. She was discharged to home the same day.

DISCUSSION

EB is a rare genetic mechanobullous disorder characterized by excessive fragility of the skin and mucous membranes (1). Patients will undergo multiple surgeries in their lifetime, most commonly for wound care and extensive dressing changes, and repair of syndactyly, joint contractures, esophageal stricture, and tracheal stenosis. The importance of not creating new lesions cannot be overemphasized. New lesions may have debilitating consequences, including the inability to eat, prolonged hospitalization, respiratory difficulty, and infection.

It is important to provide adequate postoperative analgesia. This enables speedy recovery from surgery and discharge to home. In the immediate postoperative period, uncontrolled pain may result in emotional and physical distress, sleep disturbance, impaired respiratory function, increased metabolic requirements, and impaired gastrointestinal mobility. In particular, EB patients who emerge from anesthesia in significant pain may thrash and scratch their skin, creating new traumatic lesions. In the long term, poor analgesia can lead to chronic pain, behavioral changes such as difficulty sleeping, and poor wound healing.

Multimodal analgesia employs opioid and non-opioid analgesics, as well as neuraxial techniques and regional blockade. Dexmedetomidine in particular is an important non-opiate sedative analgesic. It acts centrally as an alpha-2 agonist at the local ceruleus to cause sedation and anxiolysis. Peripherally it acts at alpha-2 receptors in the dorsal horn of the spinal cord and inhibits substance P release for anti-nociceptive and opioid synergistic effects. Consequences of large bolus, rapid administration of dexmedetomidine include refractory bradycardia and hypotension.

Administration as an infusion or small boluses over time can mitigate this risk. With a duration of action of 60-120 minutes, dexmedetomidine can be given intraoperatively and still provide postoperative analgesia in the immediate recovery period. Unlike opioids, dexmedetomidine does not act at opioid receptors and does not cause constipation or pruritus (2-4).

Until now, there has been no published case report of a

patient with epidermolysis bullosa undergoing major surgery and receiving dexmedetomidine as the primary analgesic with absolutely no use of narcotics for perioperative pain control. Avoidance of narcotics in this patient population highlights the potential role for dexmedetomidine as a potent analgesic. EB patients suffer from chronic pain and undergo painful procedures. They can develop considerable tolerance to opioids over time. Minimization of narcotics decreases side effects such as sedation, postoperative nausea and vomiting, and pruritus (5). This is in line with the American Academy of Pediatrics (AAP), Center for Disease Control, and Food and Drug Administration's goals of reducing narcotics in the United States. Poisoning from opioid analgesics has become the most frequent type of drug poisoning and correlates with an increase in prescription of opioid analgesics. The AAP has formed the "Committee on Substance Use and Prevention" to develop guidelines to treat opioid use disorders among adolescents in particular and ensure that patients with severe refractory pain receive appropriate analgesics.

KEY CLINICAL MESSAGE

Goals of safe anesthetic management for EB patients include preventing new lesions, optimizing pain control, and avoiding airway trauma, especially with emergence and induction. Multimodal analgesia decreases narcotic use, thereby reducing sedation, constipation, and pruritus. Dexmedetomidine is an important non-opioid analgesic for chronic pain patients undergoing major surgeries.

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Author Information

Claire S. Soria

Department of Anesthesiology, University of California, San Diego
San Diego, California, United States

Mark Greenberg

Department of Anesthesiology, University of California, San Diego
San Diego, California, United States