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Original Contribution

Peripheral nerve block in patients with Ehlers-Danlos syndrome, hypermobility type: a case series☆

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Abstract

Study Objective: Ehlers-Danlos syndrome (EDS) is an inherited disease characterized by defects in various collagens or their post translational modification, with an incidence estimated at 1 in 5000. Performance of peripheral nerve block in patients with EDS is controversial, due to easy bruising and hematoma formation after injections as well as reports of reduced block efficacy. The objective of this study was to review the charts of EDS patients who had received peripheral nerve block for any evidence of complications or reduced efficacy.

Design: Case series, chart review.

Setting: Academic medical center.

Patients: Patients with a confirmed or probable diagnosis of EDS who had received a peripheral nerve block in the last 3 years were identified by searching our institutions electronic medical record system.

Interventions: The patients were classified by their subtype of EDS. Patients with no diagnosed subtype were given a probable subtype based on a chart review of the patient's symptoms.

Measurements: Patient charts were reviewed for any evidence of complications or reduced block efficacy.

Main Results: A total of 21 regional anesthetics, on 16 unique patients were identified, 10 of which had a EDS subtype diagnosis. The majority of these patients had a diagnosis of hypermobility-type EDS. No block complications were noted in any patients. Two block failures requiring repeat block were noted, and four patients reported uncontrolled pain on postoperative day one despite successful placement of a peripheral nerve catheter. Additionally, blocks were performed without incident in patients with classical-type and vascular-type EDS although the number was so small that no conclusions can be drawn about relative safety of regional anesthesia in these groups.

Conclusions: This series fails to show an increased risk of complications of peripheral nerve blockade in patients with hypermobility-type EDS.

1. Introduction

Ehlers-Danlos syndrome (EDS) is a group of connective tissue disorders that manifest in the skin and joints, yielding a

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clinical presentation of extreme elasticity of the skin, easy bruising and joint hypermobility, with incidence estimated at 1:5000 [1,2]. Six subtypes of EDS are currently recognized: classical, hypermobility, vascular, kyphoscoliosis, arthrochalasia, dermatosparaxis and other; this replaces the traditional classification designated by types I-XI. Hypermobility-subtype EDS (previously known as type III) has the highest prevalence affecting 1 in every 10,000 to 15,000 individuals [3]. Additionally, chronic joint pain is associated with hypermobility-type EDS and frequently requires orthopedic surgical intervention. Because of this, hypermobility-type EDS patients are often candidates for peripheral nerve block.

EDS has been associated with airway complications, making regional anesthesia an attractive anesthetic technique [2]. However, the use of regional anesthesia in EDS patients is controversial [2,4]. EDS patients, particularly those with the vascular subtype, may be prone to hematoma formation due to vascular fragility. Additionally, it has been postulated that tissue scarring or some other unidentified mechanism may inhibit the spread of local anesthesia and reduce block efficacy [2].

Large case series of regional anesthesia in EDS patients have not been reported. Individual cases of successful labor epidural analgesia and spinal anesthesia in patients with EDS have been described [5,6]. Skin infiltration of local anesthesia has also been used successfully in a series of patients with hypermobility type EDS, however, it was noted that duration of analgesia was much less for EDS patients compared to controls [7,8]. In contrast, a successful peripheral nerve block in a patient with hypermobility type EDS has been reported and no reduction in block duration was noted [9]. Apart from this case report, data on the safety and efficacy of single injection peripheral nerve blocks and continuous peripheral nerve blocks in EDS is lacking. Here we present a series of patients with EDS who received single injection peripheral nerve blocks or continuous peripheral nerve catheters in an attempt to quantify success rates and associated complications.

2. Materials and methods

Institutional review board approval to perform a retrospective case series analysis using an anesthesia electronic health record database was obtained, and a requirement for written informed consent was waived. Although there are minor variations in practice, at our institution both the performance of regional anesthesia and the associated documentation are relatively standardized. This allowed us to systematically search for any regional anesthetic complications or failures. The standard practice and documentation for a regional anesthetic is described below.

Regional anesthesia is generally performed in a preoperative holding area and the block is assessed and documented prior to the surgery. Ultrasound guidance is generally used for block placement, and 10-30 mL of 0.5% ropivicaine, 0.5% bupivacaine or 1.5% mepivacaine are used for the initial bolus. Patients who receive peripheral nerve catheters generally receive infusion rates of 6 mL/hr of 0.2% ropivicaine for 1-3 postoperative days. Single shot blocks are performed using Stimuplex needles (B. Braun USA, Bethlehem, PA). The Arrow StimuCath system is used for peripheral nerve catheters (Teleflex, Research Triangle Park, NC). If the patient is discharged home with an infusion pump, the On-Q pump was used for infusion (I-Flow, Irvine, CA). Blocks are usually performed by personally supervised resident physicians; however some blocks may be personally performed by the attending anesthesiologist.

Immediately after the block is performed, regional anesthesia procedure notes are generated by the provider, documenting the type of block, techniques used, medications given, and any immediate complications. After the patient's surgery has concluded, the patient is seen by a member of the anesthesia staff and any complications of anesthesia (either regional or general) are documented in a postoperative note. If the patient's pain is poorly controlled, the regional anesthesia team is called to re-assess the block, and replace the block if necessary; if the regional team needs to replace the block a new note is written.

Inpatients with peripheral nerve catheters are followed postoperatively by an acute pain service with visits documented in the electronic record. If the patient has a peripheral nerve catheter placed and is discharged home with an infusion pump, a member of the anesthesia staff calls the patient daily and writes a note regarding the patient's pain control and any complications noted with the peripheral nerve block. If the patient has a peripheral nerve catheter placed and is discharged home with an infusion pump, a member of the anesthesia staff calls the patient daily and documents the contact in the medical record.

An automated query of our institution's electronic health record system (Epic, Verona, WI) identified patients who carried a diagnosis of EDS and had a peripheral nerve block performed. EDS patients were identified by searching for *ICD-9* codes associated with EDS in the patient's medical record, and by a text search for 'Ehlers-Danlos' in the patient's preoperative evaluation. Peripheral nerve blocks were identified by searching patient records for a regional anesthesia note associated with a surgical encounter. After these patients had been identified, the charts were manually examined to confirm that the patients carried a diagnosis of EDS and had received a peripheral nerve block.

The blocks that were identified with this query included femoral, sciatic, saphenous, lumbar plexus, paravertebral, and brachial plexus blocks; all of these were included in the case series. Blocks that are generally performed by the surgeon at our institution, such as digital blocks, were not identified by this query. Patient's charts were manually reviewed to determine whether an EDS subtype diagnosis had been made by a physician. If no subtype diagnosis had been made, the chart was reviewed by the authors for any