

Utilization of paravertebral nerve blocks as part of a multimodal analgesic regimen in a patient with Bernard-Soulier syndrome undergoing a Nuss procedure

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Abstract

We present a case of regional analgesia utilized in a 43-year-old woman with Bernard-Soulier syndrome (BSS) undergoing a Nuss procedure for the treatment of pectus excavatum. BSS is an extremely rare bleeding disorder (1:1,000,000) associated with prolonged bleeding times, giant platelets, and thrombocytopenia. Due to the rare incidence and heterogeneity in bleeding predisposition due to BSS, there is no clear consensus in management of such cases, and to our knowledge, utilization of regional analgesic techniques have not been described in the literature.

The Nuss procedure is considered “minimally” invasive, and epidural analgesia is frequently utilized at our institution. Due to our patient’s heterozygous presentation of BSS and mild history of bleeding, a modified perioperative multimodal analgesic plan was chosen which included bilateral single injection paravertebral nerve blocks (PVBs).

Our report describes successful utilization of PVBs in a patient with BSS and our approach to this rare hereditary condition.

Keywords: paravertebral nerve block, Bernard-Soulier syndrome, Nuss procedure, coagulopathy

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Introduction

Bernard-Soulier syndrome (BSS) is an extremely rare bleeding disorder with an incidence of 1:1,000,000 patients [1]. Patients with BSS have thrombocytopenia and prolonged bleeding times due to an inheritable platelet disorder. This autosomal recessive coagulopathy has been attributed to mutations affecting the membrane-bound glycoprotein Ib/V/IX found on platelets. Bleeding can be spontaneous or life threatening in patients who carry both recessive copies of the gene;

however, due to incomplete penetrance, heterozygous patients may still have a mild-to-moderate bleeding risk [2].

Perioperative pain management of patients without coagulopathies undergoing the Nuss procedure remains a challenge [3], and thoracic epidural analgesia has been frequently utilized with good results [4]. Due to the rare but potentially devastating neurologic sequelae due to an epidural hematoma, there is a high level of caution when considering a neuroaxial analgesic technique in these patients. However, as clinical manifestation and bleeding risk vary between patients with BSS, particularly a heterozygous patient, we propose that a peripheral regional technique might be utilized while considering the patient’s clinical presentation of the coagulopathy.

We describe the successful performance of bilateral thoracic paravertebral nerve blocks (PVBs) for peri-

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operative analgesia in a patient with BSS undergoing the Nuss procedure.

Case report

A 43-year-old, 64 kg woman with a history of BSS, presented for the Nuss procedure for treatment of symptomatic pectus excavatum. In addition to the Nuss procedure, bilateral breast implants were to be removed and re-implanted by the end of the surgery.

Prior to her diagnosis of BSS, the patient had a history of gum bleeding, bruising, and menorrhagia. However, she had never developed life-threatening episodes of bleeding. Three years previous, the patient experienced prolonged bleeding from a dental procedure which led to the evaluation of her coagulopathy and subsequent diagnosis of BSS. A peripheral blood smear revealed large platelets with an otherwise normal platelet count and coagulation panel. The patient was found to have a heterozygous presentation of the disorder, but due to incomplete penetrance, she may still have manifested a degree of coagulopathy and bleeding risk with surgical procedures. An uneventful past surgical history included bilateral breast augmentation and further dental procedures for which she received prophylactic platelet transfusions. Her preoperative platelet count was 209,000/ μ L. Complete blood count, prothrombin time, and renal profile were within normal limits. The patient's hematologist recommended 1 unit of single-donor apheresis platelets to be transfused prior to surgery or any procedure being performed. No specific platelet function tests were recommended or performed in preparation for the surgical procedure. Additional platelets were made available for potential intraoperative use if clinically significant bleeding or poor clot formation were noted.

Risk and benefits of a regional anesthetic technique as an adjunct to the perioperative analgesic plan were discussed with the patient and surgical team. Although the preoperative clinical presentation of the patient coagulation status was benign, a continuous epidural was not considered due to potential unknown manifestations or exacerbation of the coagulopathy with the surgery. After platelet transfusion and achievement of moderate sedation, single-injection PVBs were performed bilaterally at the T3-T6 nerve roots. Three mL of 0.5% ropivacaine were injected at each site (24 mL total) through a 22-gauge Tuohy needle. No immediate complications were noted. The block was performed in the preoperative holding room, and sensory deficit to ice over bilateral T3-T6 dermatomes was confirmed prior to proceeding to the operating room (OR). The perioperative analgesic regimen, initiated prior to the surgery, also included scheduled doses of 1,000 mg of oral acetaminophen every 8 hours

and 900 mg of oral gabapentin every 8 hours. Non-steroidal anti-inflammatory drugs (NSAIDs) were not used due to the potential effect on the platelet function.

After an uneventful induction of general anesthesia and tracheal intubation, the operation proceeded as planned and included bilateral breast implant removal and replacement. The surgical incision included 5 cm curvilinear incisions bilaterally along the mammary crease for the breast implant removal. For the Nuss procedure, a right-sided 5 mm port was made at the 7th intercostal level in the midaxillary line for the placement of a camera and chest insufflation. Two bars were utilized to correct the chest wall deformity with bilateral 1 cm incisions made at the 5th and 6th intercostal level. At the end of the procedure, subcutaneous catheters were placed bilaterally along the midaxillary line lateral to the incisions, and set to deliver 4 mL/hour of 0.5% ropivacaine per side via an elastomeric pump (connected prior to emergence). The patient's trachea was extubated in the OR 10 minutes after the conclusion of the operation, and emergence was uneventful. Excessive bleeding was not noted during the surgery and estimated blood loss was 50 mL. In the OR, the patient received a total of 50 μ g sufentanil and 50 mg ketamine intravenously for the 4 hours surgical procedures.

The patient was evaluated by the regional anesthesia team 10 minutes after the patient's arrival in the recovery room. She was awake, alert, oriented, breathing without distress, and satisfied with her initial pain control. Her pain reported via the 0 to 10 Numerical Rating Scale (NRS) was 4 at rest and 7 with deep inspiration and coughing. Fentanyl patient-controlled analgesia (PCA) was started with a 20 μ g bolus every 8 minutes and a continuous ketamine infusion at 10 mg/hour. The patient reported feeling comfortable, and her pain scores were less than 3 before being discharged from the recovery room.

On the first postoperative day (POD), re-evaluation of the block demonstrated complete resolution, and small areas of decreased thermal sensation were only noted along the subcutaneous catheters. Overall, the patient reported her pain was well-controlled, and she was satisfied with her postoperative pain management. The rest of the hospital stay was uneventful. Ketamine infusion was stopped on POD 2, at which time 10 mg of oral oxycodone hydrochloride extended release every 12 hours and 5 mg of oxycodone immediate release every 4 hours, as needed, were initiated. Subcutaneous incisional catheters were discontinued on POD 3. With decreasing use of the fentanyl PCA, IV opioids were weaned and discontinued by POD 4, and the patient was discharged home on POD 5. The patient's postoperative overall median pain scores from arrival in the postanesthesia case unit (PACU) to

hospital discharge are summarized in Figure 1 in 12 to 24 hour intervals. Dynamic pain was assessed with deep inspiration and cough.

Discussion

Rare bleeding disorders pose unique challenges to anesthesiologists in managing patients in the perioperative period. First described by Bernard and Soulier in 1948 [5], BSS is an extremely rare bleeding disorder characterized by prolonged bleeding time, giant platelets, and thrombocytopenia. Thrombocytopenia is not always present, and studies suggest that the low platelet count is due to decreased platelet survival and/or decreased platelet production [1]. Clinical presentation and bleeding risk are also variable, and most heterozygous patients are at a mild to moderate risk of bleeding [2]. Besides monitoring the degree of thrombocytopenia, clinical presentation is primarily used to guide the need for therapy. Platelet transfusion is the main treatment of uncontrolled bleeding; however, activated factor VIIa and desmopressin have also been used as adjuncts to transfusion [1, 2].

There is a lack of consensus about the perioperative management of patients with BSS due to its rare and variable presentation. Case reports involving patients with BSS include off-pump cardiac bypass [6], breast surgery [7], endodontic surgery [8], total hip arthroplasty [9], laparotomy [10], and mandibular osteotomy [11]. In most cases, preoperative thrombocytopenia is present and corrected via platelet transfusion prior to the surgery. Further intraoperative transfusions are used based on the clinical impression of the patient's hematologic status and extent of blood loss related to

the surgery [6, 7, 9, 10]. There is a lack of evidence guiding a perioperative plan in patients presenting with a history of mild symptoms associated with BSS and normal platelet count, but prophylactic transfusion of platelets has been utilized [8, 11]. NSAIDs are often avoided in these patients, and there is also a lack of information regarding management of postoperative pain beyond the use of opioids.

First described by Dr. Donald Nuss in 1987, the Nuss procedure is used routinely for correction of pectus excavatum. Anesthetic concerns with the Nuss procedure include significant postoperative pain of the anterior chest wall, which may impede the patient's respiratory efforts. A thoracic epidural is routinely placed for patients undergoing the procedure for perioperative pain control, and such practice is widely supported [3, 12]. Other regional analgesic techniques included bilateral PVB catheters, which have been shown to be non-inferior to a thoracic epidural for postoperative analgesia in pediatric patients undergoing the Nuss procedure [13].

Epidural analgesia is frequently utilized at our institution in patients undergoing the Nuss procedure; nevertheless, pain often remains difficult to control in this surgical population, and multiple analgesic strategies are often used. It should also be noted that our case also involved re-implementation of breast implants. Innervation of the pectoralis muscle arises from the brachial plexus (medial and lateral pectoral nerves), thus anesthetizing only the thoracic nerve roots, either via PVBs or an epidural, may not have been adequate. In spite of the initial high dynamic pain score, the patient reported feeling comfortable overall, and satisfied with the management of her pain. Overall, we feel that

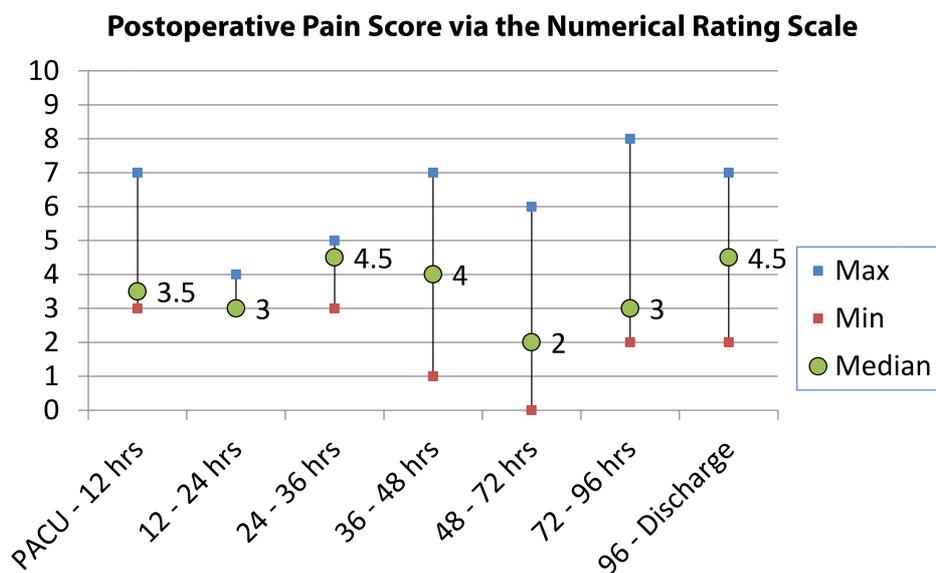


Fig. 1. Postoperative pain scores via the Numerical Rating Scale. PACU: postanesthesia case unit

PVBs were an essential component of her multimodal therapy since it would anesthetize most of the anterior chest wall, and wound catheters alone often only cover superficial incisional pain.

Utilization of a regional analgesic technique in the presence of a coagulopathy accrues additional concerns. The placement of a thoracic epidural carries a rare but significant risk of epidural hematoma. It is challenging to come to a consensus on the risks and benefits profile of a regional neuroaxial technique in the presence of a rare coagulopathy as clinical evidence is often scarce. Therefore, neuroaxial techniques are often entirely avoided in patients who have bleeding tendencies in order to avoid the risk of an epidural hematoma and ensuing neurological complications [14].

The unique presentation of our patient includes a heterozygous presentation of BSS, mild past medical history of prolonged bleeding, no recent history of bleeding or bruising, normal platelet count, and prophylactic transfusion of platelets prior to the procedure. As single injections PVBs only provide analgesia for up to 8 hours [15], continuous PVB catheters are often utilized at our institution and the option was considered. It is possible that placement of PVB catheters, or an epidural catheter, may have been uneventful. However, the patient only underwent minor surgical procedures in the past, and it was unknown if exacerbation of the coagulopathy were to occur peri- or postoperatively with a more extensive operation. There was also the concern of clot dislodgment in the postoperative period with the removal of (in the case of PVBs) multiple catheters.

Currently, the evidence is lacking and conflicting in comparing the risk and/or sequela of bleeding due to PVBs versus epidural catheter placement as per guidelines by the American Society of Regional Anesthesia (ASRA) [16]. Unfortunately, there is a lack of evidence available to make definitive recommendations in regards to thoracic PVBs. ASRA guidelines note that bleeding, rather than neurologic sequelae, is more of a concern in patients undergoing a peripheral anesthetic technique, and cautious recommendations generalize all deep peripheral nerve blocks to follow the same guidelines as neuroaxial [16].

However, clinicians find some of the guidelines too restrictive and not reflective of current clinical practice. Utilization of PVBs has been described as an alternative to an epidural technique in patients with marginal anticoagulation status, such as enoxaparin thromboprophylaxis [17, 18], and in altered coagulation states, such as in open liver resection [19] or pulmonary resection [20]. Though a risk of paravertebral extrapleural hematoma (PEH) exists, it is assumed that a major hemorrhage is less likely and neurologic complications from a PEH are less disastrous compared to an extra-

dural hematoma. Nevertheless additional studies need to be undertaken to evaluate the safety profile of PVBs: a lack of evidence of poor outcomes does not constitute proof safety.

Conclusion

From a case report, it is difficult to make conclusions on the safety and efficacy of PVBs compared to epidural or opioid analgesia alone in patients undergoing the Nuss procedure with a rare inheritable platelet disorder. Currently, clinical evidence addressing PVBs in patients with varying coagulation states is limited and, at times, conflicting. Future studies are required to address the risks and benefits of PVBs in patients with a marginal coagulation status. However, clinical manifestation and the severity of coagulation disorder should also be considered before precluding all regional techniques and their analgesic benefits.

Conflict of interest

Nothing to declare

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Utilizarea blocului paravertebral în analgezia multimodală la un pacient cu sindrom Bernard-Soulier supus procedurii Nuss

Rezumat

Am prezentat utilizarea analgeziei regionale în cazul unei paciente de 43 de ani cu sindrom Bernard-Soulier (BSS), supusă procedurii Nuss pentru tratamentul toracelui excavat. BSS reprezintă o afecțiune hemoragică extrem de rară (1:1 000 000) asociată cu prelungirea timpului de sângerare, trombocite gigante și trombocitopenie. Nu există un consens clar de tratament al acestor cazuri, datorită incidenței reduse și heterogenității în predispoziția de sângerare din BSS, iar, din câte știm, utilizarea tehnicilor de anestezie regională nu au fost descrise în literatură pentru aceste cazuri.

Procedura Nuss este considerată minim invazivă, iar analgezia peridurală este frecvent utilizată în instituția noastră. Datorită formei heterozigote de BSS prezentată de pacienta noastră, cu antecedente medii de sângerare, s-a utilizat un plan de analgezie multimodală perioperator modificat, care a inclus câte un bloc paravertebral (PVB) bilateral.

Prezentarea noastră descrie utilizarea cu succes a PVB în cazul unei paciente cu BSS și abordarea noastră în fața acestei rare maladii ereditare.

Cuvinte cheie: bloc paravertebral, sindrom Bernard-Soulier, procedură Nuss, coagulopatie