

Caudal epidural block instead of general anesthesia in an adult with Duchenne muscular dystrophy

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Abstract: Duchenne muscular dystrophy (DMD), first described in 1834, is an X-linked dystrophinopathy, leading to early onset skeletal muscle weakness. Life expectancy is reduced to early adulthood as a result of involvement of voluntary skeletal muscles with respiratory failure, orthopedic deformities, and associated cardiomyopathy. Given its multisystem involvement, surgical intervention may be required to address the sequelae of the disease process. We present a 36-year-old adult with DMD, who required anesthetic care during surgical debridement of an ischial pressure sore. Given his significant respiratory muscle involvement, ultrasound-guided caudal epidural anesthesia was used instead of general during the surgical procedure. The technique and its applications are discussed, with particular emphasis on the feasibility and safety of using regional anesthetic techniques in patients with DMD.

Keywords: Duchenne's muscular dystrophy, caudal epidural, regional anesthesia

Introduction

Duchenne muscular dystrophy (DMD), an X-linked disorder, is the most common form of muscular dystrophy. It results from mutations in the dystrophin gene, located on chromosome Xp21.1.¹ Dystrophin is an integral protein in regulation of the integrity of sarcolemma. Mutations resulting in either dysfunction or total absence of this protein, result in interruption of the sarcolemma with progressive myofibril atrophy, necrosis, and fibrosis.^{2,3} DMD patients develop progressive neuromuscular weakness, skeletal deformities, and cardiopulmonary complications, with cardiac or respiratory failure being the primary cause of death during the second and third decades of life.⁴ Patients with DMD may present for surgical procedures to correct progressive orthopedic deformities resulting from the disease process. Given the associated respiratory and cardiac involvement, anesthetic care remains challenging.^{2,5} Patients with DMD are especially susceptible to the adverse effects of general anesthesia and procedural sedation. Specific concerns that may arise include difficult endotracheal intubation, prolonged neuromuscular blockade, the need for postoperative ventilation, and hyperkalemia, rhabdomyolysis, and cardiac arrhythmias resulting from prolonged exposure to volatile anesthetic agents.^{4,5} To avoid the aforementioned perioperative risks, regional anesthetic techniques may be considered as an alternative to general anesthesia. We present the use of caudal epidural anesthesia as an alternative to general anesthesia in a 36-year-old man with DMD who presented for debridement of an ischial pressure injury. The use of regional anesthesia in this patient population is discussed, and techniques for catheter placement reviewed.

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Case report

Written, informed consent was obtained from the patient for publication of this case report. Institutional Review Board (Nationwide Children's Hospital, Columbus, OH, USA) approval is not required for publication of isolated case reports. The patient was a 36-year-old, 61.1 kg adult, scheduled for surgical debridement of a right ischial pressure ulcer at an unspecified stage (ICD code: L89.319). His past history was positive for DMD; restrictive lung disease with chronic respiratory failure requiring Bilevel Positive Airway Pressure (BiPAP) support at night and Sipper ventilator support during the day. He also had a history of dysphagia and swallowing dysfunction with pulmonary aspiration resulting in gastric tube dependence. Echocardiography showed normal cardiac anatomy with mild left ventricular systolic dysfunction and an ejection fraction of 53%. His past surgical history was significant for posterior spinal fusion and gastrostomy, during which there was no history of problems with general anesthesia other than difficult vascular access. He had no known allergies. Medication included ranitidine (150 mg twice a day), sertraline (100 mg, once a day), metoprolol tartrate (25 mg, twice a day), enalapril (10 mg, twice a day), sennosides-docusate (once a day), polyethylene glycol (17 g, once a day), and multivitamin supplements. Physical examination revealed a thyromental distance of more than three fingerbreadths and a Mallampati Class I airway. There was limited range of motion (flexion and extension) of the neck. A thoracolumbar surgical scar from the previous posterior spinal fusion was noted on his back, and the spinous processes from T₂ to L₅ could not be palpated. The remainder of the physical examination and preoperative vital signs were unremarkable. After review of the patient's current status of a difficult airway and significant restrictive lung disease requiring ventilator dependence, previous history of associated comorbid conditions, and after discussion with consulting services, it was decided to offer regional instead of general anesthesia. This patient had difficult anatomy for lumbar epidural of spinal anesthesia because of the fused spinous processes from his prior spinal fusion surgery and dense scar tissue on the back. The spinous processes from T₂ to L₅ were not felt and appropriate landmark palpation was not possible due to adipose tissue over the sacral area. Given the patient's previous history of posterior spinal fusion and his difficult anatomy, it was decided that the best option for neuraxial anesthesia would be caudal epidural anesthesia with ultrasound guidance. The anesthetic plan, risks, benefits, and options including caudal epidural anesthesia were discussed with the patient and informed consent obtained. On the day of surgery, the patient

was held nil per os and gastric tube feeding were held for 8 hours. His usual morning doses of sertraline and ranitidine were administered through the gastric tube while the enalapril was held. He was transferred to the operating room with his BiPAP machine using his usual night-time settings (20/4 cmH₂O). He was positioned in left lateral decubitus position, standard American Society of Anesthesiologists monitors were placed, and ultrasound-guided peripheral intravenous access was achieved. Midazolam (a total of 4 mg in divided doses) was administered intravenously for anxiolysis. After sterile preparation, superficial anesthesia of the skin and subcutaneous tissue was achieved with 1% lidocaine. Ultrasound imaging (GE 12L-RS linear transducer, GE Medical Systems Co., Jiangsu, People's Republic of China) of the sacral hiatus was performed in the transverse and longitudinal views (Figures 1 and 2). Ultrasound imaging of the sacral area also demonstrated the sacral hiatus displaced ~4–5 cm toward the right from the midline. On the first attempt, a 3.5 inch, 17-gauge Tuohy needle (Epimed Spirol® Epidural Set, Epimed International Inc., Dallas, TX, USA) was advanced under ultrasound guidance toward and through the sacrococcygeal ligament into the caudal epidural space (Figure 3). A 19-gauge epidural catheter was advanced through the needle and into the epidural space. The Tuohy needle was removed and 10 cm of the catheter left in the caudal epidural space and secured in place. An initial bolus dose of 5 mL of 3% chloroprocaine was administered. After 5 minutes, sensory changes were noted to pin prick and temperature test in the lower extremities with no systemic complaints. An additional 10 mL of 3% chloroprocaine was administered and adequate surgical anesthesia was achieved over the sacral dermatomes. During the procedure, the patient received one additional bolus of 5 mL of 3% chloroprocaine, 15 minutes after the start of the procedure. No significant changes were noted in heart rate or blood pressure with the administration of the epidural medications. The only other intraoperative medication was intravenous acetaminophen (1,000 mg) for postoperative analgesia. The surgical procedure, which lasted ~30 minutes, was completed with minimal blood loss and no intraoperative complications. The patient remained on nasal BiPAP until the end of the procedure. Following completion of the surgical procedure, the caudal epidural catheter was removed in the operating room. The patient's postoperative course was unremarkable. He remained as an inpatient for wound care and underwent a second brief procedure, which was performed with the administration of local anesthetic to the surgical site, 1 week after the first procedure. He was discharged home the day following the second procedure.

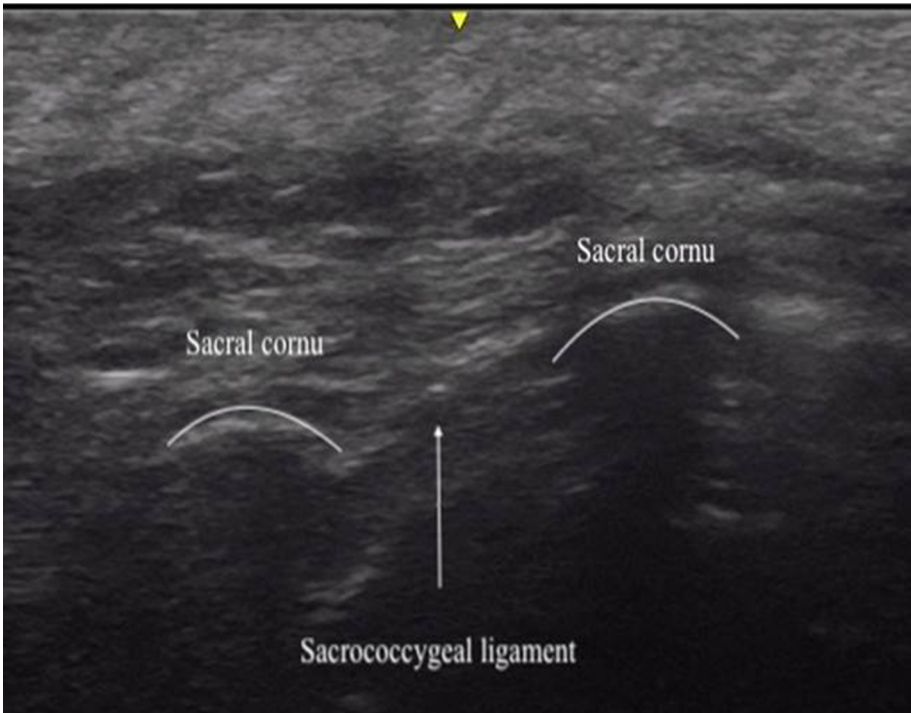


Figure 1 Transverse ultrasound image of the sacrum at the level of the sacrococcygeal ligament showing the ligament and the sacral cornu.

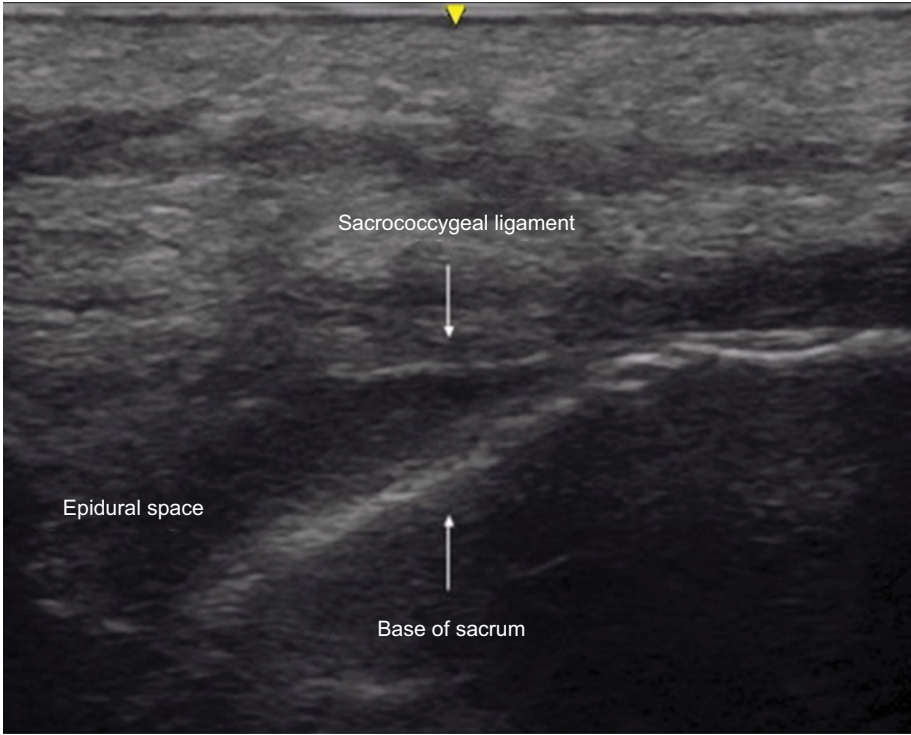


Figure 2 Longitudinal image of the sacrum showing the sacrococcygeal ligament, the base of the sacrum, and the caudal epidural space.

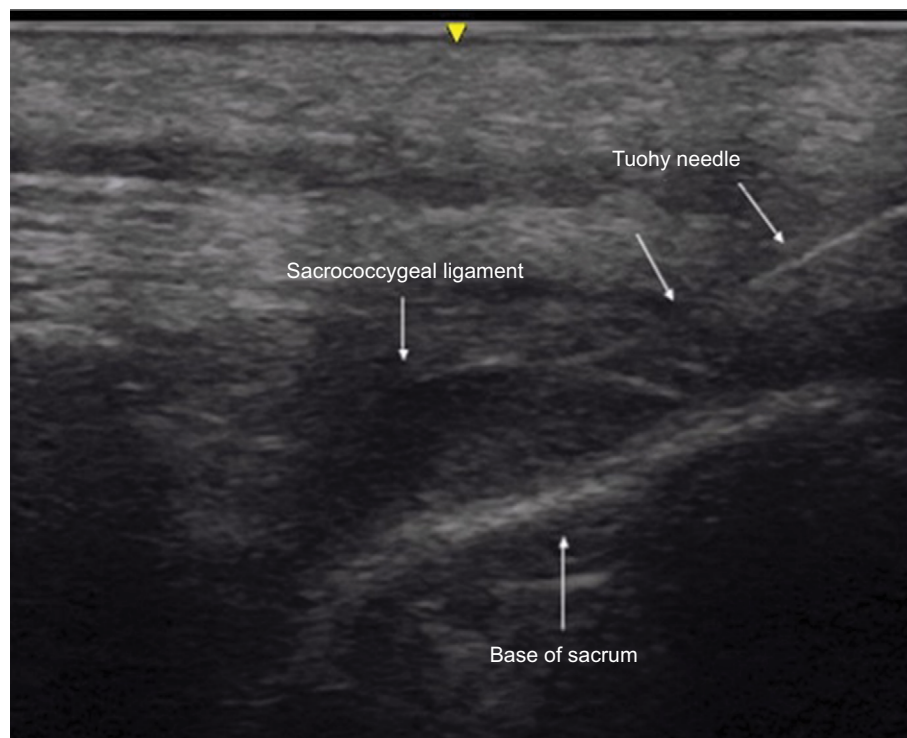


Figure 3 Longitudinal image of the sacrum showing the advancement of the Tuohy needle toward the sacrococcygeal ligament.

Discussion

Caudal epidural anesthesia, with the injection of a local anesthetic agent into the epidural space through the sacral hiatus, is commonly used to provide postoperative analgesia in pediatric patients.^{6,7} The technique may also be used instead of general anesthesia to avoid potential perioperative complications in patients with comorbid conditions.⁸ Although the lumbar approach to the epidural space is frequently chosen for surgical anesthesia in older patients, the caudal approach may be used when previous surgical procedures or anatomical deformities preclude the lumbar approach. Such was the case in our patient, as he had previously undergone posterior spinal fusion with instrumentation which limited access to the lumbar epidural space.

Needle or catheter placement for caudal epidural anesthesia may be achieved using superficial anatomical landmarks or fluoroscopy-guided or ultrasound-guided techniques.^{9–11} Klocke et al¹⁰ first described the use of ultrasound to achieve caudal epidural access for the administration of epidural corticosteroids. Although frequently performed in children using a blind approach with palpation of bony landmarks on the sacrum, given our patient's body habitus, his previous surgical procedure on the lumbosacral spine, and difficulties with palpation of sacral bony landmarks, we chose to use ultrasound to identify the site of needle entry and guide insertion of the needle into the caudal epidural space.

Anesthesia-related concerns in patients with DMD include potential difficulties with airway management and endotracheal intubation, restrictive pulmonary disease, cardiac muscle involvement with cardiomyopathy, and the potential for adverse effects associated with the prolonged administration of inhalational anesthetic agents.^{5,12–15} Given these concerns, regional anesthesia has been used as a means of avoiding general anesthesia and its potential complications in patients with DMD (Table 1).^{16–20} As with our patient, these anecdotal reports demonstrate the feasibility of using various regional anesthetic techniques instead of general anesthesia in this patient population.

Caudal epidural anesthesia is achieved by the injection of a local anesthetic agent into the epidural space. Potential complications associated with caudal epidural anesthesia include those related to needle placement and those from the local anesthetic agent.^{21–23} Complications related to needle placement include dural puncture, bleeding, infection, and trauma to neural elements or the spinal cord. Coagulation parameters including platelet count should be evaluated prior to needle placement. Given our patient's previous surgical procedure involving the lumbar spine and his anatomical challenges, we chose the caudal approach to the epidural space with ultrasound guidance to limit the potential for complications related to needle placement. Complications related to the local anesthetic agent include local anesthetic

Table 1 Reports of regional instead of general in DMD patients

Study	Patient demographics	Outcome
Molyneux et al ¹⁶	A 36-year-old woman with DMD	Combined spinal–epidural anesthesia for Cesarean section. Successful intraoperative care with uneventful postoperative course
Bang et al ¹⁷	A 22-year-old man with DMD	The patient was scheduled to undergo reduction and internal fixation of a left distal femur fracture. Under ultrasound guidance, separate injections were performed to provide anesthesia of the femoral and lateral femoral cutaneous nerves as well as the sacral plexus. The surgical procedure was performed using the regional anesthetic technique and his postoperative period was uneventful
So et al ¹⁸	Two patients (2 and 14 years of age) with presumed DMD	Muscle biopsy to confirm the diagnosis of DMD was performed after peripheral blockade (interscalene and femoral) with ultrasound guidance and nerve stimulation
Vandepitte et al ¹⁹	A 27-year-old male patient with DMD	Drainage of a thoracic wall hematoma was performed after intercostal nerve blockade under ultrasound guidance
Büget et al ²⁰	A 17-year-old male patient with DMD	Echocardiography demonstrated dilated cardiomyopathy, mitral regurgitation, and an ejection fraction of 23%. Supraclavicular block provided anesthesia for resection of a rhabdomyosarcoma and left arm amputation

Abbreviation: DMD, Duchenne muscular dystrophy.

systemic toxicity, cardiovascular effects from sympathetic blockade, and high motor blockade with respiratory effects. Slow incremental injection is recommended especially in patients with comorbid cardiac involvement to allow for monitoring of potential hemodynamic effects (hypotension and bradycardia). Judicious fluid administration or administration of a vasoactive agent may be needed to treat hemodynamic changes. To achieve the rapid onset of epidural anesthesia and limit the potential for systemic toxicity, given its rapid systemic metabolism, we chose to use chloroprocaine.²⁴ An additional concern regarding placement of a caudal epidural catheter vs lumbar or thoracic placement is the potential risk of infection given its location in the sacral dermatomes and risk of fecal soiling and contamination. As with all placement techniques, adequate skin preparation and coverage with a bio-occlusive dressing may help limit the potential for such concerns. Furthermore, subcutaneous tunneling has been suggested as an option when caudal epidural catheters are left in place for prolonged postoperative use.²⁵ This approach was not utilized in our patient as short-term intraoperative use of the catheter was planned.

We present the use of caudal epidural anesthesia in a patient with DMD and significant respiratory involvement to avoid the potential risks and perioperative complications associated with general anesthesia. Given previous spinal surgery with fusion and instrumentation, we chose a caudal approach to the epidural space. Ultrasound guidance was used to guide needle and catheter placement due to limited ability to identify anatomical landmarks. Our report adds additional anecdotal experience to that previously reported demonstrating the potential efficacy of using regional anesthesia instead of general anesthesia in this challenging patient population.

Disclosure

The authors report no conflicts of interest in this work.

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