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CASE REPORT

A case report of the successful use of regional anaesthesia and mixed sedative techniques in an adolescent with Duchenne muscular dystrophy

R Tladi, R Swart

Department of Anaesthesiology, Faculty of Health Sciences, University of the Free State, South Africa
Corresponding author, email: *reitu.tladi@gmail.com*

Summary
In this case report, we present a 12-year-old male who presented with clubbed feet for Achilles tenotomy with comorbid Duchenne muscular dystrophy and obesity. Owing to his prior surgical history and requirement for a prolonged intensive care unit stay with mechanical ventilation, we opted for a regional anaesthetic technique with mixed pharmacological and non-pharmacological sedation. The patient tolerated the nerve blocks with minimal sedation with midazolam and local anaesthesia and the surgery was performed successfully with light sedation and the help of his favourite song.

Keywords: Duchenne muscular dystrophy, regional anaesthesia, sciatic nerve block, sedation, non-pharmacological sedation

Case report

A 12-year-old male was booked for an elective bilateral Achilles tenotomy for clubbed feet. On history the patient's father reported that his son had Duchenne muscular dystrophy. The patient was on chronic prednisone 25 mg daily, captopril 25 mg three times daily, calcium supplements and multivitamin syrup daily. The patient was also intellectually impaired and attended a local school for learners with special needs.

The patient had undergone an open reduction and internal fixation (ORIF) of his femur four years previously. This was completed under general anaesthesia and complicated with postoperative respiratory weakness. The patient did not meet extubation criteria and was admitted to the intensive care unit (ICU) for 13 days of postoperative ventilatory support.

On clinical examination the patient was alert with limited responses on questioning. He was in a wheelchair and unable to stand. He had a high blood pressure of 145/84 mmHg and a pulse rate of 96 beats per minute. On airway examination, he had a short, thick neck and receding chin. Cardiovascular and respiratory system examination showed no abnormalities.

Laboratory investigations showed a haemoglobin level of 16.1 g/dL and a platelet count of 231 x 10^9/L on full blood count. Kidney functions, calcium, magnesium and phosphate levels were all within normal limits.

Considerations of potential complications, such as rhabdomyolysis following general anaesthesia and technical difficulties performing a regional block in an obese patient, were discussed with the surgical team. The decision for the Achilles tenotomy to be done under regional nerve blocks and mild sedation was taken, following a detailed discussion with the patient's father. Written consent was obtained from the primary caregiver for the case to be published.

On presentation to theatre, the patient was clinically well, but unable to sit without assistance. A 22G intravenous line was inserted in the anaesthesia procedure room. Blood pressure and pulse oximetry monitors were connected, face mask oxygen at 40% was provided and 0.5 mg midazolam was administered prior to doing the nerve blocks. The patient was supine with both knees flexed at 45 degrees. Aseptic technique was maintained, the skin was localised with 5 ml of 1% lignocaine and ultrasound-guided bilateral popliteal sciatic nerve blocks were done, as shown in Figures 1 and 2, using 20 ml of 0.5% bupivicaine approximately 1 hour before surgery. The patient displayed minimal discomfort and cooperated well during the nerve block procedure.

On commencement of surgery, the patient displayed discomfort and anxiety as the surgical incision was made. A further

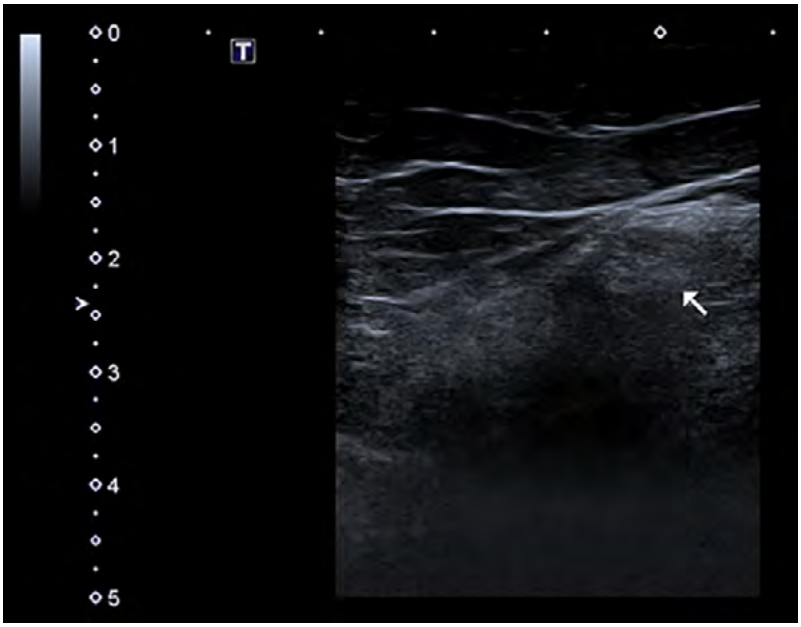

Figure 1: Ultrasound image of the patient's left popliteal sciatic nerve (white arrow)

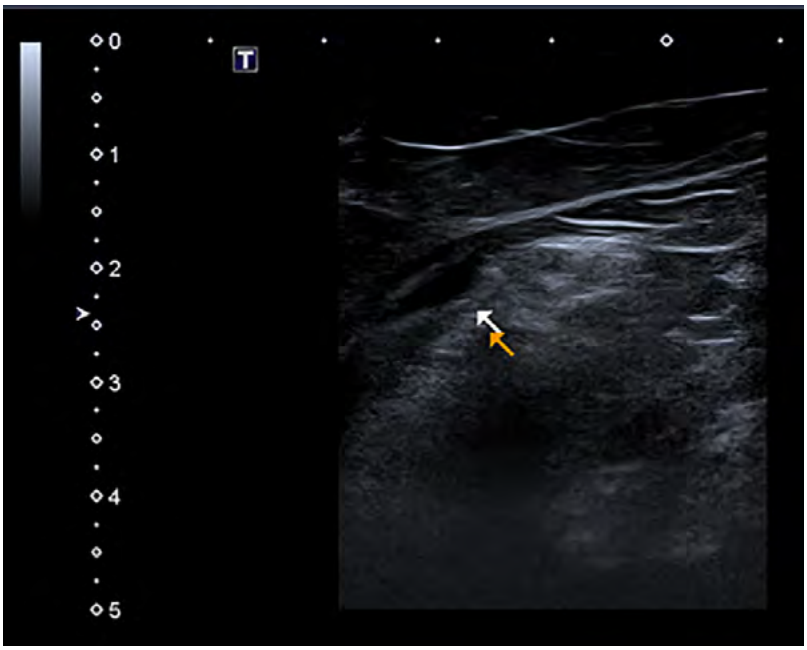

Figure 2: Needle adjacent to sciatic nerve (white arrow) with local anaesthetic spread above and below (yellow arrow) the nerve

0.5 mg of midazolam and 15 mg ketamine were administered intravenously. He showed no further pain response to surgical stimulus but was still anxious. A smartphone was used to play cartoons to distract him, but he requested that his favourite song be played. His anxiety settled and he was comfortable for the remainder of the procedure which lasted approximately 40 minutes in total. The patient had a short, uncomplicated recovery room stay and was discharged home the following morning.

Discussion

Subsequent to Conte first describing neuromuscular diseases in 1836, the link with anaesthetic-related morbidity and mortality would not be established until 1962.[1] Denborough et al. described the association between halogenated hydrocarbons and a family of patients who suffered from spasmodic complications, yet it was not until Guillaume Armand Duchenne du Boulogne described a pseudohypertrophic muscular paralysis in children that the most common form of childhood muscular dystrophy would come to be named after him.[2,3]

Duchenne muscular dystrophy (DMD) is an X-linked recessive hereditary disorder.[3] It has an incidence in the general population of 1:3 500 live male births. The disorder is characterised by abnormal or absent dystrophin.[3] The dystrophin-glycoprotein complex is a series of proteins that bind the myofibrils to the cellular matrix and stabilise the sarcolemma during muscular activity.[4] Deficiencies in this complex leads to fatty infiltration and progressive fibrosis of the muscles, complicated by worsening weakness. Full innervation of muscles is maintained, but the sarcolemma becomes increasingly permeable to creatine kinase and electrolytes, which may precede the onset of muscle weakness.[4] The increases in creatine kinase may also be seen in asymptomatic female carriers of the gene.[3] The dystrophin-glycoprotein complexes of the respiratory and cardiac muscles are also involved and tend to become increasingly defective with age.[4] Pharyngeal and gastric muscles may also become involved, with an increased risk for aspiration.[4]

Early signs and symptoms of the disorder include a waddling gait with falls, difficulty climbing stairs and pseudohypertrophy of muscles.[4] Intellectual disability is also common, with 30% of affected boys testing less than 70 of the full-scale intellectual quotient (FSIQ) test in a Serbian population.[5] The disease generally becomes apparent in boys aged 2–5 years.[4] These patients are generally wheelchair-bound by the age of 8–10 years and life expectancy is limited by cardiomyopathies and pulmonary complications in their late twenties to early thirties.[3] These patients are also at risk for anaesthesia-induced rhabdomyolysis.[6]

The anaesthetic considerations for patients with DMD are therefore their age (and potential natural history of the disease), avoiding the triggers of anaesthetic-induced rhabdomyolysis (succinylcholine and halogenated hydrocarbons), the potential for respiratory compromise postoperatively, baseline cardiac function, intellectual ability (with regards to issues of consent and cooperation) and the potentially increased risk of aspiration.[1,3,4,6]

Kyphoscoliosis may also develop leading to impaired respiratory mechanics and the potential for further cardiac compromise.[4] Regional anaesthesia has been suggested as a potential strategy to avoid complications associated with general anaesthesia. This approach has been promoted as a preferred technique by Schieren et al., where this may be feasible, keeping in mind the possibility of respiratory compromise with a high spinal where neuraxial anaesthesia is chosen.[6] In patients with advanced disease, premedication and sedation may also be problematic, but judicious use of anxiolytics (in a monitored setting) and non-pharmacological sedative techniques may be useful, as stress and anxiety can aggravate myopathic symptoms.[6]

With the main objectives for the case being the provision of effective analgesia while avoiding complications and adverse outcomes, our anaesthetic technique proved successful. The use of a regional nerve block and both pharmacological and non-pharmacological sedation allowed for the Achilles tenotomy to be performed while sparing an ICU admission in a hospital setting with limited ICU resources. Subsequent to the patient's postoperative critical care admission four years prior to this surgery, the caregiver expressed considerable concern and anxiety regarding the anaesthetic plan, which was also successfully alleviated by the outcome of this admission.

An approach to a general anaesthetic still needs to be considered when planning regional anaesthesia in preparation for failed or incomplete nerve blocks, and the patient being uncooperative or anxious despite being sedated. Although there

is no association between DMD and malignant hyperthermia (MH), the risk of anaesthesia-induced rhabdomyolysis (AIR) must be considered.[3] Patients with AIR can develop hyperkalaemia and rhabdomyolysis following exposure to volatile anaesthetic agents. Depolarising neuromuscular blocking agents are also contraindicated in these patients as cardiac arrest may occur by the same mechanism. Patients with neuromuscular disease also exhibit an increased sensitivity to non-depolarising neuromuscular blockers.[3] If muscle relaxants are required, the use of short-acting non-depolarising muscle relaxants is advisable. Together with standard blood pressure, pulse oximetry, electrocardiography (ECG), capnography and temperature monitoring, neuromuscular blockade monitoring is essential.[3,7] Total intravenous anaesthesia with an anaesthetic circuit that is not contaminated by volatile agents is the recommended general technique for patients with DMD and other disorders putting them at risk for AIR or MH.

For this procedure the patient's legal guardian, his father, gave consent. The patient's mother had passed away, therefore his father had become his legal guardian. As this patient was unable to assent due to impaired decisional capacity, the legal guardian granted full consent to the publication of this case report, as governed by the Declaration of Helsinki.[8,9]

Cardiomyopathies, conduction abnormalities and respiratory complications in patients with DMD can lead to severe morbidity and mortality in the perioperative period,[1] making regional nerve block techniques advantageous. Despite this patient's body habitus and increased body mass index (BMI), the nerve block was performed without great difficulty and caused minimal discomfort to the patient. Regional anaesthesia may be the preferred method of anaesthesia, depending on the extent of the surgery and potential technical challenges such as the requirement for akinesia, abnormal spinal curvature and the patient's ability to cooperate. Cases of myotonic contractures have been reported secondary to incomplete neuraxial blockade, likely due to impairment of descending inhibitory pathways.[7]

The American Society of Anesthesiologists (ASA) defines sedation as a continuum of states of consciousness ranging from anxiolysis to general anaesthesia.[10] As commonly used sedatives cause varying degrees of respiratory, cardiovascular and central nervous system suppression, caution is key when administered to patients with neuromuscular disease. Close patient monitoring and emergency airway equipment are mandatory when using sedation techniques. Low dose ketamine and midazolam have been reported as safe sedation-analgesia options.[7] Opioids should be used sparingly. In patients with compromised respiratory function opioids should be avoided.[3,7] Non-pharmacological adjuvant therapies to sedating children are becoming increasingly popular and may be useful in this patient population. These mainly include distracting the patient's attention for the duration of the procedure.[11] Objects, such a toys, stationery and electronics can be used to provide visual or auditory distractions. Our patient sang along to his favourite song while surgery was being performed. The presence of a family member can also reduce the patient's anxiety. Role-playing or telling the child to close his or her eyes and pretend to be somewhere else are other useful methods. Hypnosis by a trained provider is also described as an adjunct to sedation.[11]

In conclusion, the combination of regional anaesthesia and a balanced pharmacological and non-pharmacological sedation technique proved to be a safe alternative to general anaesthesia for patients with DMD. Our approach was effective and we suggest it be considered when managing patients at risk of AIR and MH.

Conflict of interest

The authors declare no conflict of interest.

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Ethical approval

Ethics Committee approval was obtained from the University of the Free State Health Sciences Research Ethics Committee (UFS-HSD2020/0655/3006).

ORCID

R Tladi https://orcid.org/0000-0002-9219-4134
R Swart https://orcid.org/0000-0003-3873-4424

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