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CASE REPORT

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Anesthetic management of a patient with spinal muscular atrophy type II for scoliosis surgery: a case report

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Abstract

Background Spinal Muscular Atrophy (SMA) is a rare autosomal recessive genetic disorder characterized by degeneration of motor neurons in the spinal cord, resulting in progressive limb muscle weakness, atrophy, and severe scoliosis. Clinically, it is divided into four types according to age at onset and severity. There are few cases reported in the literature presently, especially type II cases, and no expert consensus or guideline for the anesthetic management of spinal muscular atrophy (SMA) with scoliosis. This article discusses anesthesia management and intraoperative considerations for this patient, as well as how to help the patient reduce perioperative complications. To the best of our knowledge, this is the first case of continuous thoracolumbar dorsal ramus nerve block for pain relief after scoliosis surgery in a patient with spinal muscular atrophy type II.

Case presentation We described a 17-year-old patient with spinal muscular atrophy scoliosis (SMA type II) who underwent posterior scoliosis osteotomy and orthopedic laminectomy and fusion under general anesthesia without muscle relaxants, A series of optimized anesthesia management measures were successfully implemented, aiming to reduce perioperative related complications. After the operation, continuous thoracolumbar dorsal ramus nerve block was carried out and achieved a good analgesic effect. The patient was discharged 33 days after hospitalization. It is indicated that anesthesia management for patients with SMA is a real challenge for all anesthesia providers.

Conclusions For patients with SMA undergoing scoliosis surgery, total intravenous anesthesia without muscle relaxants and continuous thoracolumbar dorsal ramus nerve block after surgery have been proven to be both efficient and safe. It is also crucial to implement preoperative multidisciplinary consultation, lung-protective ventilation strategy, appropriate anesthetic drugs, reasonable blood transfusion scheme, as well as strengthened postoperative monitoring and multimodal analgesia.

Keywords Spinal muscular atrophy, Anesthetic management, Severe scoliosis, Thoracolumbar dorsal ramus nerve block

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Background

Spinal muscular atrophy (SMA) is an autosomal recessive lower motor neuron disease that can lead to progressive proximal muscle weakness and skeletal muscle atrophy. The incidence of SMA is approximately 1/10,000–20,000 live births, while the frequency of carrying the gene in the general population is 1/40 – 1/70 [1]. Patients with SMA are often accompanied by severe scoliosis and require surgical treatment. Anesthetic management for these patients could be challenging for prolonged duration, potential massive haemorrhage, as well as problems such as hypersensitivity to neuromuscular blocking agents, muscle weakness, poor pulmonary function, etc. This article focuses on the perioperative characteristics of a patient with neuromuscular scoliosis and SMA type II who underwent posterior serious scoliosis correction surgery and proposes an optimized anesthetic management strategy, as well as how to help the patient reduce perioperative complications. To the best of our knowledge, this is the first case of continuous thoracolumbar dorsal ramus nerve block for pain relief after scoliosis surgery in a patient with spinal muscular atrophy type II.

Case presentation

We report the case of a 17-year-old male patient with spinal muscular atrophy type II underwent scoliosis correction surgery under general anesthesia. The patient had a bodyweight of.

41.5 kg and a body height of 165 cm. He was diagnosed with Spinal Muscular Atrophy (SMA) type II by genetic testing at the age of 5 ~ 6 months. Scoliosis started at 9yrs and gradually got worse. Intrathecal injection of nusinersen was started when he was 15yrs and rehabilitation training was carried out, but he still could not sit up independently (Fig. 1). There was no need for ventilator-assisted breathing at night, but monitoring report showed that he was on moderate sleep apnea-hypopnea syndrome, mainly obstructive apnea, with moderate nocturnal hypoxemia, and the lowest blood oxygen saturation reaching 81%. In terms of family history, his brothers are normal, and one of them is an SMA gene carrier.

After admission, relevant examinations were further completed. The full-spine anteroposterior radiograph of the patient showed that in the sitting position, T_3 and T_5 thoracic vertebrae were convex to the right, with a Cobb angle of approximately 45°, a Cobb angle of

Fig. 1 The clinical photos of the patient in the anteroposterior and lateral positions. The patient's spine protrudes significantly to the right, with a notable deformity of posterior concave-convex scoliosis. There is muscle atrophy and deformity in the upper and lower extremities, and the patient is unable to sit independently

approximately 69.1° on the right side (R-side bending), a Cobb angle of approximately 54.3° on the left side (L-side bending), and a Cobb angle of 41.9° during traction. The T_5 and T_3 thoracolumbar vertebrae had a leftward convexity at the end of the cone, with a sitting Cobb angle of approximately 83.9°, a Cobb angle of approximately 55.3° on the right side, a Cobb angle of approximately 101.1° on the left side, and a Cobb angle of approximately 57.7° during traction (Fig. 2). CT scan showed an S-shaped scoliosis of the spine, accompanied by vertebral rotation and abnormal thoracic cage morphology (Fig. 3). Pulmonary function test reveals poor pulmonary function while forced vital capacity (FVC) accounted for 29.6% of the predicted value, forced expiratory volume in the first second (FEV_1) accounted for 34.6%. No obvious abnormalities were found in the urinary system, hepatobiliary system and cardiovascular system by using doppler ultrasound. The preoperative anesthesia evaluation showed that there was a suspected difficult airway, with Mallampati III and a mouth opening of less than 2 cm.

After preoperative multidisciplinary consultation, the patient was sent to the operating theater.

A venous access was established on the right upper extremity. Routine monitoring of vital signs, including heart rate (HR), blood pressure (BP), peripheral capillary oxygen saturation (SpO_2), and electrocardiogram (ECG), was carried out. The HR was recorded at 70 bpm, the BP at 100/60 mmHg, and the SpO_2 at 98%. Following sufficient oxygen inhalation and denitrogenation via a face mask, 20 μg of fentanyl was administered, propofol was administered in a target-controlled manner(TCI) at a concentration of 3.0 μg/ml, and 20 mg of lidocaine was intravenously injected for the induction. Once the patient was asleep, an endotracheal tube with an inner diameter of 7.0 mm was inserted under the guidance of a fiberoptic bronchoscope and laryngoscope. Intermittent Positive Pressure Ventilation, and the ventilation parameters were set as follows: tidal volume (VT) of 350 ml, a respiratory rate (f) of 13 bpm, and a positive end-expiratory pressure (PEEP) value of 5cmH_2O. Subsequently, the right radial artery was punctured for invasive arterial blood pressure monitoring.

For the anesthesia maintenance, the TCI of propofol was at 3.0 ~ 3.5 μg/ml, remifentanil was at 2–4 ng/ml, and dexmedetomidine was administered at a rate of 0.5 $\mu g \cdot kg^{-1} \cdot h^{-1}$.The bispectral index (BIS) was monitored and maintained within the range of 40 ~ 60. Body temperature and spinal cord electrophysiology were monitored, and an autologous blood transfusion device was activated. To manage bleeding, tranexamic acid was administered during the operation with an initial loading dose of 10 $mg \cdot kg^{-1}$ followed by a maintenance dose of 2 $mg \cdot kg^{-1} \cdot h^{-1}$ to mitigate the risk of bleeding. Simultaneously, deliberate hypotension was implemented to

Cobb angle	Sitting	R-side bending	L-side bending	Traction
C3- T5	45	69.1	54.3	41.9
T5- L3	83.9	55.3	101.1	57.7

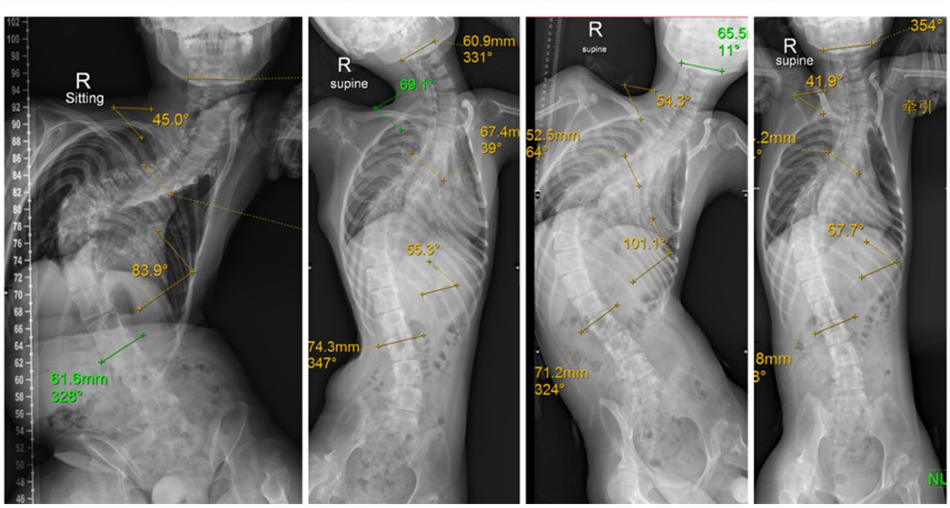

Fig. 2 The anteroposterior radiograph of the patient's entire spine. When in the sitting position, the thoracic vertebrae T3 and T5 protrude to the right, with the Cobb angle being approximately 45°, about 69.1° when bending to the right (R-side bending), about 54.3° when bending to the left (L-side bending), and 41.9° when traction (Traction); the cone ends of the thoracolumbar vertebrae T5 and T3 protrude to the left, with the Cobb angle being approximately 83.9° in the sitting position, about 55.3° when bending to the right, about 101.1° when bending to the left, and about 57.7° when traction

Fig. 3 CT shows an S-shaped scoliosis of the spine, accompanied by vertebral rotation and abnormal thoracic cage morphology. The cervical spine segment is convex to the left, the thoracolumbar spine is significantly convex to the right with the thoracolumbar segment as the apex, some vertebral bodies have rotational deformity, the physiological curvature of the cervical spine becomes straight, and the thoracic spine is kyphotic

maintain the mean arterial pressure between 70 ~ 75 mmHg. Intermittent blood gas monitoring was also performed during the procedure. Additionally, measures were taken to protect the patient's body temperature, ensuring that hypothermia did not occur throughout the operation. Intraoperative nerve monitoring indicated that somatosensory evoked potentials (SEPs) were essentially normal, and motor evoked potentials (MEPs) in both lower limbs remained unchanged from the preoperative state. The operation lasted for 8.5 h, No muscle relaxants were utilized during the entire surgical procedure.378 ml of autologous blood was transfused. The final estimated blood loss was approximately 400 ml, no blood products were administered to the patient.

We used continuous thoracolumbar dorsal ramus nerve block (TDRNB) for postoperative analgesia. Before wound closure, multiorifice catheters were placed at the discretion of the attending orthopaedic surgeon. The catheters were placed laterally adjacent to the implanted pedicle screws under direct visualization (Fig. 4B) to target the TDRNs at multiple vertebral levels. the L_4 approach was used, and the nerve block catheter was indwelled at the left T_8 position. And the L_1 approach was used, and the nerve block catheter was indwelled at the right T_{12} position (Fig. 4A**).** Each side of the catheter was continuously pumped with 0.15% ropivacaine (3 ml/hr) after operation. The patient was sent to the PICU after extubation. Comprehensive motor and sensory examinations were performed, and the patient-reported pain score was recorded as well. The results showed that the postoperative analgesic effect was good, with an NRS score of 0/2, and no opioids were used for analgesia within 48 h after the operation, and no obvious complications were observed. The patient recovered and was discharged, with a total hospital stay of 33 days.

Discussion

The SMA was first reported by Austrian neurologist Guido Werdnig in 1891 [2]. According to the age of symptom onset and the degree of motor nerve damage, it can be classified into I-IV subtypes clinically [3]. The earlier the onset, the more severe the disease, and the higher the mortality rate. This case is of SMA type II, which is also called Dubowitz disease. It generally starts to display symptoms between 6 and 18 months of age. In 1964, Dubowitz detailed the phenotypic characteristics of intermediate SMA [4], generalized weakness of

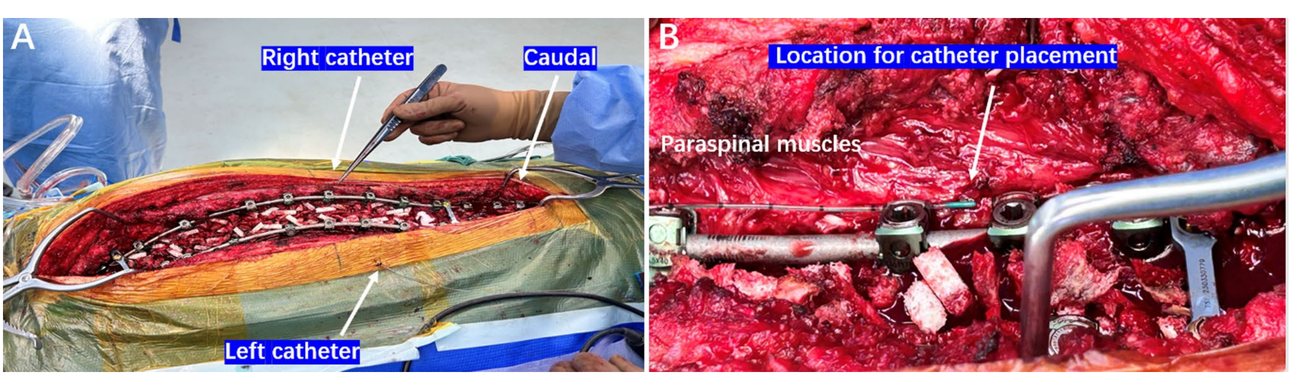

Fig. 4 Thoracolumbar dorsal ramus nerve block for postoperative analgesia. Figure A shows the dorsal ramus nerve catheters prior to closure. On the left side, the approach is at L4, and the nerve block catheter is indwelled at the T8 position. On the right side, the approach is at L1, and the nerve block catheter is indwelled at the T12 position. Figure B shows the location for catheter placement

the trunk and limb muscles, the ability to sit independently, inability to stand and walk, progressive scoliosis, hip and knee contractures, and laxity of the hand and finger joints. Respiratory dysfunction is the most common complication of SMA and one of the main causes of death. Patients may have difficulty in clearing lower respiratory tract secretions due to weak cough, and some patients with severe scoliosis may have restrictive ventilatory dysfunction due to severe chest wall deformity [5]. The severity of scoliosis is usually evaluated by measuring the angle of lateral curvature, and the Cobb angle measurement method is the most commonly used. The Cobb angle is named after the orthopedic surgeon John Robert Cobb and is defined as the angle formed by the extension lines of the upper edge of the apical vertebra and the lower edge of the terminal vertebra [6]. When the Cobb angle reaches 40°, it is considered as severe scoliosis, and surgical treatment should be considered. For the pediatric patients with SMA and severe scoliosis (Cobb ≧ 90°),intraoperative anesthetic management for patients with SMA is a real challenge for anesthesia providers. There is no evidence-based consensus or guideline for the management of anesthesia in this group of patients currently. We have proposed an optimized perioperative anesthesia management strategy based on the perioperative characteristics of SMA type II patients.

Firstly, a multidisciplinary consultation should be recommended before the operation. It's important to evaluate the patient's cardiopulmonary function and assess whether the patient has indications of a difficult airway. We should consider the impact of its pathophysiological changes, including restrictive ventilatory dysfunction caused by preoperative thoracic deformity, difficult airway due to limited cervical spine movement and mouth opening, large surgical trauma and more bleeding. The spinal cord function of the patient should be protected to the greatest extent to prevent pulmonary complications Intraoperatively. Matthew et al. reported a study that included 40 SMA type I and II patients, and found that the incidence of postoperative pneumonia in type II patients was 4.5%, and the incidence of postoperative atelectasis was 31.8% [7]. Lai Wang et al. reported 26 cases of severe scoliosis with SMA patients (type I $n = 1$; type II $n = 25$), in which the incidence of difficult intubation was 73.1%, the incidence of postoperative pneumonia 34.6%, the incidence of atelectasis 19.2%, the incidence of pleural effusion 23.1%, and the rate of postoperative ICU admission was 26% [8]. Therefore, adequate preoperative evaluation and preoperative multidisciplinary discussion are necessary.

Secondly, appropriate drugs and anesthetic methods are of great importance for the patient.

We recommend using total intravenous general anesthesia (TIVA) without muscle relaxants during the operation, even when performing endotracheal intubation. Patients with SMA may display increased sensitivity to and prolonged effect of nondepolarizing neuromuscular blockers, even after reversal. neuromuscular monitoring can be unreliable in patients with SMA [9]. Succinylcholine should be avoided because of possible risk of inducing rhabdomyolysis and hyperkalemia in the presence of lower motor neuron denervation hypersensitivity. There are reports of patients who required prolonged intubation and respiratory support due to muscle weakness despite reversal of neuromuscular blockade and four equal twitches on train-of-four (TOF) stimulation [10]. The impact of residual neuromuscular blocking drugs or high-level neural axis block on chest wall dynamics should be minimized as much as possible to avoid excessive suppression of central respiratory drive. The residual effects of anesthetic drugs, pain stress, and increased postoperative metabolic demands can all lead to acute respiratory failure, which in turn requires increased mechanical respiratory support. Opioids have been used in perioperative care of patients with SMA, with the major concern being respiratory depression. Because of this, short acting opioids are better suited for intra-operative use, along with careful titration and constant monitoring. In terms of fluid infusion and blood transfusion management, in order to avoid pulmonary edema caused by excessive fluid, we adopted restrictive fluid management and goal-directed fluid therapy strategies, and timely initiated autologous blood transfusion devices. Intra - operative monitoring of body temperature, bleeding, and depth of anesthesia such as Bispectral Index (BIS) monitoring is of great importance.In patients with SMA, anesthetic technics were flexible according to existing reports, but no consensus was ever achieved, which might be perfect for this kind of patients [11, 12].

Lastly, postoperative pain management after multilevel thoracolumbar scoliosis surgery remains challenging. The ideal nerve - block technique is supposed to meet the following requirements: During the operation, it should not interfere with the nerve - monitoring signals, nor impede the process of tracheal extubation. In the postoperative period, it must not obscure the results of postoperative nerve monitoring, nor have any adverse impacts on the motor and sensory functions of the lower limbs. Additionally, it should not induce urinary retention, block the phrenic nerve, or trigger high - level spinal anesthesia.

It has been reported that the use of erector spinae block in multimodal analgesia may improve the outcome of the enhanced recovery after surgery (ERAS) program, alleviate pain for up to 24 h after thoracolumbar decompression spine surgery [13]. Oezel [14] et al. proposed that bilateral ESP catheters were placed by spine surgeons under direct visualization during the operation.

A new method of multi-level lumbar fusion, in which the surgeon places the erector spinae plane (ESP) catheter under direct visualization and compares it with single erector spinae plane block, found that the pain scores in both groups were lower, and the opioid consumption was moderate, improving pain after lumbar spine surgery and minimizing opioid consumption, promoting postoperative recovery.Another new analgesic method was first proposed and reported by Professor JEFF XU in 2020 [15]. Jeff L Xu et al. reported the use of Continuous Multiorifice Infusion Catheter for thoracolumbar dorsal ramus nerve block for postoperative analgesia in scoliosis surgery, Intraoperatively, 4 multiorifice catheters were placed lateral to the implanted pedicle screws. Two catheters were placed on each side, and the results showed that it significantly improved postoperative comfort and pain perception. Patients who received TDRN block catheters required the least opioids postoperatively, avoiding blockade distribution to the ventral ramus nerves and sympathetic nerve chains, making the multimodal analgesia regimen easier to implement.

For the above two postoperative analgesic techniques, we recommend thoracolumbar dorsal ramus nerve block and have made improvements to its methodology. Two multiorifice catheters were placed on each side lateral to the implanted pedicle screws. The results showed that the postoperative analgesic effect was good, with an NRS score of 0/2, and no opioids were used for analgesia within 48 h after the operation, and no obvious complications were observed. Postoperative oral painkillers can choose acetaminophen, pregabalin, etc. If the patient has a history of kidney damage or gastrointestinal bleeding, Non-Steroidal Anti-Inflammatory Drug (NSAID) should be used with caution. Therefore, we recommend multimodal analgesia to achieve a good postoperative analgesic effect.

Conclusion

In this case, for the patient with neuromuscular scoliosis and SMA type II undergoing posterior scoliosis correction surgery, total intravenous anesthesia without muscle relaxants and continuous thoracolumbar dorsal ramus nerve block after surgery was proved to be both efficient and safe. It is also crucial to implement preoperative multidisciplinary consultation, a lung-protective ventilation strategy, appropriate anesthetic drugs, a reasonable blood transfusion plan, as well as strengthened postoperative monitoring and multimodal analgesia.

Author contributions

Chen zy completed the patient's surgical anesthesia, wrote the main manuscript, provided original data such as pictures, and discussed and analyzed the case. Chen xw, Liang xy, Niu q, all authors reviewed the manuscript. Dr. Chan yw, Xu xb, also helped review and revise the article. Therefore, I have added their names in my article .

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None.

Data availability

No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate

In this case report, ethics approval and consent to participate are not applicable. This is because this case is a retrospective analysis. The data used are all data routinely collected during the clinical diagnosis and treatment process and have been anonymized. It does not involve additional interventions or specific research operations on patients. Clinical trial number is not applicable. We confirm that this manuscript has not been published previously and is not under consideration elsewhere. All authors have approved the manuscript and are in agreement with its submission to BMC Anesthesiology. The patient provided written informed consent for the publication of this case report, and all ethical guidelines have been followed.

Consent to participate

Not applicable.

Competing interests

The authors declare no competing interests.

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