

## Thoracic epidural anaesthesia for laparoscopic cholecystectomy in patient with type Iv spinal muscular dystrophy

Tariq Torkey Atta <sup>1</sup> MBChB DA CABA&IC ,  
 Saad Muhammad Wajih <sup>1</sup> MBChB DA CABA&IC ,  
 Zinah Majid Mnati <sup>1</sup> MBChB DA FICMS A&IC.

<sup>1</sup>Dept. of Anaesthesia & Intensive Care Unit, Al-Yarmouk Teaching Hospital.

Date Submitted: 2/1/2015  
 Date Accepted: 21/5/2015

Address for Correspondence:  
 Dr. Zinah Majid Mnati,  
 E-mail:  
 zeena\_majid@yahoo.com

### Abstract

Spinal muscular atrophy (SMA) is a rare genetic neurodegenerative disease characterized by degeneration of spinal cord lower motor neurons, which results in atrophy of skeletal muscles, hypotonia and muscle weakness. Patients with type IV SMA often have onset of weakness at adulthood.

Anesthetic management is often difficult in these patients as a result of muscle weakness and hypersensitivity to neuromuscular blocking agents also succinylcholine induced hyperkalemia, postoperative intensive care unit admission and long hospital stay.

Laparoscopic surgery is normally performed under general anesthesia, but regional techniques have been found beneficial, usually in the management of patients with major medical problems. The goals for anesthetic management of these patients include satisfactory anesthesia during surgery, excellent postoperative analgesia with minimal compromise of respiratory function without exacerbation of neurologic signs and symptoms.

We describe a 37-year-old male patient with type IV SMA scheduled for laparoscopic cholecystectomy. We decided to use an epidural technique to avoid muscle relaxants, tracheal intubation and to evaluate the efficacy and feasibility of thoracic epidural anesthesia for laparoscopic cholecystectomy, so that it can be used later as Anesthetic technique in patients when general anesthesia is not feasible. After operation, there was no exacerbation of neurologic signs and symptoms or development of respiratory complications neither a need for intensive care unit admission.

**Keywords:** Spinal muscular atrophy, Epidural anesthesia, laparoscopic cholecystectomy.

### INTRODUCTION

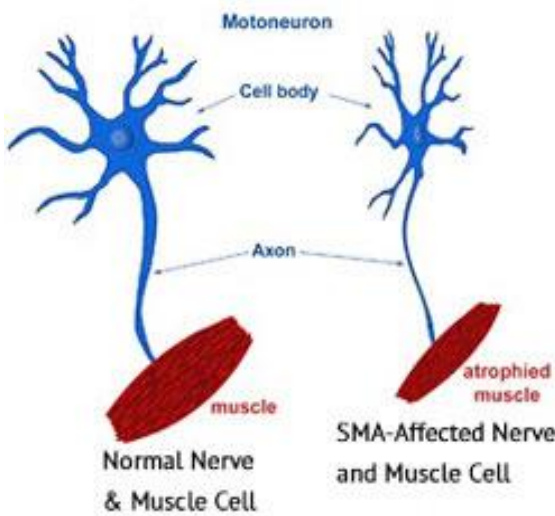
Spinal muscular atrophy (SMA) is a rare lower motor neuron disease that occurs due to degeneration of anterior horn cell. SMA patients require particular attention for narcosis management since there is a possibility for mechanical ventilation when a delayed recovery of spontaneous respiration occurs after an operation using general anesthesia as a sensitive reaction against skeletal muscle relaxants and weakened respiratory root <sup>[1]</sup>.

In over 95% of cases of SMA, a genetic autosomal recessive disorder causes a functional loss of the

survival motor neuron1 (SMN1) gene on chromosome V. In 2% of the cases the transmission appears to be either autosomal dominant, X-linked or sporadic <sup>[2]</sup>. SMN1 encodes a protein, SMN, which is crucial to the assembly of the spliceosome, a structure involved in processing mRNA <sup>[3]</sup>. It may also mediate mRNA trafficking in axons. SMA is thought to result from insufficient levels of SMN protein in spinal anterior motor neurons. Absence of the gene SMN1 leads to atrophy and death of spinal anterior motor neurons. A second gene, SMN2, located in the centromeric region of the same chromosome, in part modulates the clinical symptoms of SMA. SMA is usually diagnosed by

quantifying the presence of the SMN1 gene by polymerase chain reaction (PCR), replacing muscle biopsy or electromyography. The copy number of this weakly expressed homolog may be an important determinant of the severity of the disease [4].

Consequently; SMA can display various degrees of skeletal muscle atrophy with progressive muscle weakness of the limbs and trunk, pulmonary insufficiency due to scoliosis and chest wall muscle weakness, autonomic dysfunction, and dysphagia, resulting in severe physical disability. As shown in Figure (1).



**Figure (1) Spinal Muscular Atrophy Muscle and Nerve Appearance**

Clinical classification criteria for spinal muscular atrophy shown in table (1) [5].

**Table 1.**

SMD Types	Age of onset	Highest function achieved
Type I (Werdnig-Hoffmann disease)	0-6 months	Never sit
Type II (intermediate)	7-18 months	Sit never stand
Type III (mild, Kugelberg-Welander disease) in adulthood	> 18 months	Stand and Walk during adulthood
Type IV (adult)	2°-3° decade	Walk unaided

Laparoscopic cholecystectomy is normally performed under general anesthesia, but regional techniques, such as low thoracic epidural and lumbar spinal, have been used, usually to manage patients with significant medical problems[6][7]. However, this anesthetic approach requires a relaxed and cooperative patient, low intra-abdominal pressure (IAP) to reduce shoulder pain,

patient discomfort and ventilation disturbances, reduced table tilt, and precise with gentle surgical technique [8].

This study was aimed to assess the efficacy and the expected side effects of thoracic epidural anesthesia for elective laparoscopic cholecystectomy in patient with type IV spinal muscular dystrophy, so that later it can be used as anesthetic technique in patients who are at risk for general anesthesia.

### Case Report

A 37-year-old male (height 1.60 m; weight 60kg) with SMA type IV was scheduled for laparoscopic cholecystectomy. The diagnosis of SMA type IV was based on clinical progressive symptoms of proximal muscular weakness especially his both lower limbs that began since he was 10 years. He was on wheelchair for one year ago. At present, he could not lift his arms up against resistance only, but had no difficulty in swallowing or breathing, no dyspnea, his EMG study reveal the diagnosis of moderate to severe spinal muscular dystrophy and no evidence of peripheral polyneuropathy, He had a family history of SMA, his two sisters (still alive with also moderate to severe muscle weakness). Physical examination revealed that he had muscular strength 3–4 grading (with grade 3 in the upper limb and grade 4 in the lower limb), decreases of tendon reflex in lower extremities, no extrapyramidal tract signs, no particular cranial nerves involvement, no paraesthesia and no other associated medical problems. Chest X-ray, pulmonary function test and ECG were normal. The laboratory examinations were normal.

On the day of surgery, premedication was given to the patient (ranitidine 50 mg IV, dexamethasone 8 mg IV, metoclopramide 10 mg IV) and an intravenous infusion of 500 mL of Ringer lactate was administered. Electrocardiogram, non-invasive blood pressure, respiratory rate/minute and SpO<sub>2</sub> were monitored.

Preoperative vital signs were blood pressure: 150/100 mmHg; respiratory rate per minute was: 16 breathes/min; pulse rate 90 beats/min; ECG: normal sinus rhythm; and pulse oxymetry revealed: 98% oxygen saturation. From a manual muscle examination before operation (use modified versions of the Oxford scale of muscle strength in clinical practice)[9], both legs were at the 4<sup>th</sup> grade of muscle strength.

At the sitting position, under all aseptic technique, local infiltration of 4ml of 1% lignocaine was used to anaesthetize the puncture site(T<sub>8</sub>-T<sub>9</sub> intervertebral space) and an 18-gauge Touhy needle was introduced in midline at T<sub>8</sub>-T<sub>9</sub> thoracic epidural space using loss of air resistant technique and identification of the epidural

space was done, epidural catheter was inserted about 4 cm in the epidural space.

After a test dose of 3 ml of 1.5% lidocaine with 1:200,000 epinephrine, neither increase of heart rate, nor spinal anesthesia symptom was detected after 10 min. monitoring. Then injection of 9 ml of 0.5% bupivacaine hydrochloride through epidural catheter. To measure the level of motor block after anesthesia, a modified Bromage scale<sup>[10]</sup> (0: One can stretch and raise his/her leg; 1: One cannot raise his/her leg and cannot bend his/her knee; 2: One cannot his/her knee but can bend his/her ankle; 3: One cannot bend his/her ankle) was applied.

The level of sensory block, was assessed by pin-prick stimulus, covering the area of the proposed incision (T<sub>4</sub>-L<sub>3</sub>) dermatome level was confirmed before surgery, which was achieved after about 20 minutes.

Pneumoperitoneum was established after about 35 min. from time of catheter insertion with low pressure carbon dioxide of 10 mm Hg at a flow rate of 2 L/min. antitrendenburg position with left lateral tilt of the operating table was employed as minimal as possible in order to minimize diaphragmatic irritation. Vital signs during operation were recorded every 5 minutes, blood pressure: 120-160/60-90 mmHg and pulse rate was: 90-100 beats/minute.

A nasal cannula with 5 liter/min. of oxygen in inspired air was used, SpO<sub>2</sub> was 100%, respiratory rate was around 18-20 breathes/min. Only 50 µg fentanyl IV in 2 increment doses was given to the patient (1<sup>st</sup> dose was given at time of reaching the gall bladder bed, the other dose 10 min before the end of the surgery), 2 liter of IV fluid (2ringer lactate and 2 glucose saline) and 15 mg IV ephedrine in 3 increment doses to maintain blood pressure within the base line readings. Operation time was about one hour, and the epidural catheter was removed at the end of the operation.

After finishing the operation, the level of modified Bromage scale was 2 and the sensory nerve block was decreased up to the T<sub>6</sub> segment dermatoma. Three hours after the operation, the level of modified Bromage scale was dropped to 1 and the sensory nerve block to the T<sub>11</sub> segment dermatoma; 5 hours after the operation, the stage of modified Bromage scale was 0, no significant changes was found during examination of his vital signs, and the sensory nerve block was declined to the L<sub>1</sub> segment. The patient did not show adverse effects including nausea, vomiting, and dizziness in the ward. Twelve hours after finishing the operation, sensory nerve was recovered to the condition before operation, and also for the manual muscle examination, both legs were maintained at the 4th stage. A physical exam of

motor and neurological system was conducted 24 hours after finishing surgery did not show any new disability.

## DISCUSSION

Epidural anesthesia was considered safe for laparoscopic cholecystectomy without associated respiratory depression as the respiratory control mechanism remains intact to allow the patients to adjust their minute ventilation.

Moreover, the respiratory changes are less evident in awoken patients under regional anesthesia<sup>[11]</sup>.

The central neuraxial anesthesia has been found beneficial usually in patients with significant medical diseases when low intra-abdominal pressure and less degree of patient tilt during surgical procedure is used<sup>[12]</sup>.

SMA was first described by Guido Werdnig at the end of the nineteenth century and was divided into four types according to age at onset and severity<sup>[13]</sup> SMA is characterized by degeneration of motor neurons of the spinal cord, which results in hypotonia, muscle weakness and atrophy (especially of the proximal muscles of the shoulder and pelvic girdle), and weak of deep tendon reflexes, with little or no impairment of sensory systems and the diaphragm and extraocular muscles remaining unaffected until the late stages of the disease. The diagnosis of SMA needs to be confirmed by electromyography and muscle biopsy<sup>[13]</sup>.

The management of anesthesia in patients with SMA is often difficult because of muscle weakness, anesthesia-induced respiratory complications, hypersensitivity to non-depolarizing muscle relaxants, and succinylcholine-induced hyperkalemia. Also worries of neuraxial (epidural or spinal) blocks could worsen the weakness. Very little information is available in the anesthetic textbooks regarding the management of such cases although it was indicated that muscle relaxants, opioids, and thiopental could all have a prolonged duration of action<sup>[14]</sup>.

However, Hebl et al.<sup>[15]</sup> argue that neurological damages after operation occur from the combination of various factors including the lifestyle of patients, damages during operation, pressure of the tourniquet, posture of the patients during operation, dystocia, and anesthesia methods, and for SMA diseases with progressive neurological damages, neurologic symptoms can be worsened regardless of methods of anesthesia and operations.

Moreover, Hebl et al.<sup>[15]</sup> said there was no case in which new or worsened neurologic damages occurred among 136 patients who had diseases showing neurologic

dysfunction and who underwent regional anesthesia and pain control. Weston and DiFazio<sup>[16]</sup> reported A case that conducted epidural pain control during delivery for a patient with type III spinal muscular atrophy, and Buettner<sup>[17]</sup> reported A case that conducted Cesarean delivery with epidural anesthesia for a patient with type III spinal muscular atrophy.

Meanwhile, Choi et al.<sup>[18]</sup> conducted total intravenous anesthesia without muscle relaxants for patients with type II spinal muscular atrophy, while Hong et al.<sup>[19]</sup> also use total intravenous anesthesia without muscle relaxant for patients with type III spinal muscular atrophy. Both cases were for head and neck surgeries, and it is considered that general anesthesia was conducted for children patients.

Kitson et al.<sup>[20]</sup> reported a 38 years old type III SMA parturient undergoing Caesarean Section who had a history of tracheostomy during previous pregnancy. So she had a known failed intubation (hence the previous tracheostomy) and awake fiberoptic intubation (FOI) was performed. Anesthesia was induced with alfentanil and propofol and maintained with isoflurane, nitrous oxide, and oxygen without muscle relaxants.

Regarding the use of regional anesthesia for cholecystectomy, Bejarano et al.<sup>[21]</sup> used spinal anesthesia with midazolam sedation for laparoscopic ventral hernia repair and concluded that spinal anesthesia is feasible and well tolerated.

Van Zundert et al.<sup>[22]</sup> stated that segmental spinal anesthesia can be used safely for patients with impaired organ function.

Lau et al.<sup>[23]</sup> also quoted that laparoscopic hernia can be performed successfully under spinal anesthesia his study was conducted on healthy patients.

Gupta, et al,<sup>[11]</sup> 48 adult patients of ASA grade I and II scheduled for elective laparoscopic cholecystectomy were enrolled for thoracic epidural anesthesia with 15 ml of 0.75% ropivacaine and 50 µg fentanyl and conclude that it was efficacious method and has preserved ventilation and hemodynamic changes within physiological limits during pneumoperitoneum with minimal treatable side effects.

In our case report and according to the previous mentioned cases reports we tried to use the technique of thoracic epidural anesthesia for patient with type IV SMD, our patient already complained from motor weakness in both upper and lower limbs with preserved respiratory function, trying to avoid the use of muscle relaxant, and associated problem of delay recovery, we notice that when use low intra-abdominal pressure during laparoscopic cholecystectomy was efficacious

method, surgeon and patient were satisfied, no significant side effects were reported, so it had been considered that thoracic epidural anesthesia is a good alternative choice from general anaesthesia for patients with spinal muscular atrophy for laparoscopic surgery. Further evaluation of the method is required with larger numbers is desirable.

**Acknowledgments:**

The authors were grateful to the surgical team, nursing staff of Al-Yarmouk teaching Hospital for their valuable assistance and thanks to the patient and his family for their cooperation.

**Declaration of interest:**

Availability of other techniques rather than general anesthesia when it can't be feasible as usual technique for patient complains from comorbid illness.

**Author contributions:**

All authors were involved in drafting the article and revising it critically for important intellectual content, and all authors approved the final version to be published had full access to all of the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.

Study conception and design. All authors.

Acquisition, Analysis and Interpretation of data all authors.

All authors read and approved the final manuscript.

- **Funding:**

Al-Yarmouk Teaching Hospital.

- **List of Abbreviations**

Symbol	Meaning
SMD	Spinal Muscular Dystrophy
IV	Intravenous
µg	Micro gram
Mg	Milligram
cm	Centimeter
Min	Minute
sec	Second
T <sub>8</sub> – T <sub>9</sub>	Eighth and ninth Thoracic epidural level
T <sub>4</sub> – L <sub>3</sub>	Fourth Thoracic epidural level to third lumbar epidural level

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