Case Series

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Clinical characteristics and anaesthetic management of severe scoliosis patients with spinal muscular atrophy: case series

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Introduction and importance: There is no expert consensus or guidance on perioperative anaesthesia management for spinal surgery of spinal muscular atrophy (SMA) patients with severe scoliosis (Cobb≧90°). We provide a comprehensive summary of the perioperative characteristics observed in patients with SMA and propose an optimized perioperative management strategy for anaesthesia.

Methods: This study is a retrospective single-centre research. Twenty-six SMA patients with severe scoliosis underwent posterior spinal fusion surgery from September 2019 to September 2022 were enroled. The main outcomes were to show the patients' characteristics in anaesthesia, intra- and post-operative periods.

Outcomes: Nineteen patients underwent awake transnasal/transairway intubation. The median anaesthesia time of 25 patients treated under total intravenous anaesthesia was 425 min. After operation, the Cobb angle and correction rate in the coronal plane were median 54.0° and 54.4%. The length of mechanical ventilation with endotracheal intubation in ICU was median 17.5 h in 8 patients. The ICU length of stay of postoperative hospital was median 19 days. Postoperative pneumonia developed in nine patients, atelectasis in two patients, and pleural effusion in six patients. All patients did not need special oxygen therapy after discharge.

Conclusion: Multidisciplinary consultation, lung-protective ventilation strategy, appropriate anaesthetic drugs and reasonable blood transfusion scheme and postoperative monitoring were important in anaesthesia, intraoperative and postoperative periods in the patients of severe scoliosis with spinal muscular atrophy.

Keywords: anaesthetic management, severe scoliosis, spinal muscular atrophy

Introduction

Spinal muscular atrophy (SMA) is a rare genetic disease characterized by progressive muscle weakness and atrophy caused by degeneration of spinal cord anterior horn cells. The first case was described by Austrian pathologist Guido Werdnig in 1891[1]. Studies have shown that SMA is related to the deletion or mutation of the motor neuron survival (SMN) 1 gene at 5q13.2 on chromosome 5. The decreased expression of SMN protein leads to progressive skeletal muscle hypotonia, muscular atrophy[2]. According to the onset age and the degree of motor nerve damage, SMA is divided into subtypes I–IV[3]. The earlier the onset of SMA, the more serious the condition and the higher the mortality will be. In recent years, with the emergence of specific drugs and the development of spinal orthopaedics, the survival period of SMA patients has been extended[4]. The clinical manifestations of SMA patients are diverse, especially for patients with severe scoliosis and thoracic deformity, they usually exhibited very obvious restrictive ventilation dysfunction. For such patients, posterior spinal correction is often required to alleviate their symptoms and improve their quality of life. However, most of the current literature focuses on the description of surgical operations, and there is no case series report on perioperative anaesthesia management of SMA patients with severe scoliosis (especially Cobb≧90°) for spinal orthopaedics. The Cobb Angle

HIGHLIGHTS

- The anaesthetic management of spinal muscular atrophy (SMA) patients with severe scoliosis has a great risk.
- SMA patients need a multidisciplinary consultation for adequate preoperative evaluation.
- In anaesthesia, lung-protective ventilation strategy, appropriate anaesthetic drugs and reasonable blood transfusion scheme are used to optimize intraoperative anaesthesia management.
- Individualized anaesthesia management is the key to ensuring the perioperative safety of SMA patients with severe scoliosis.

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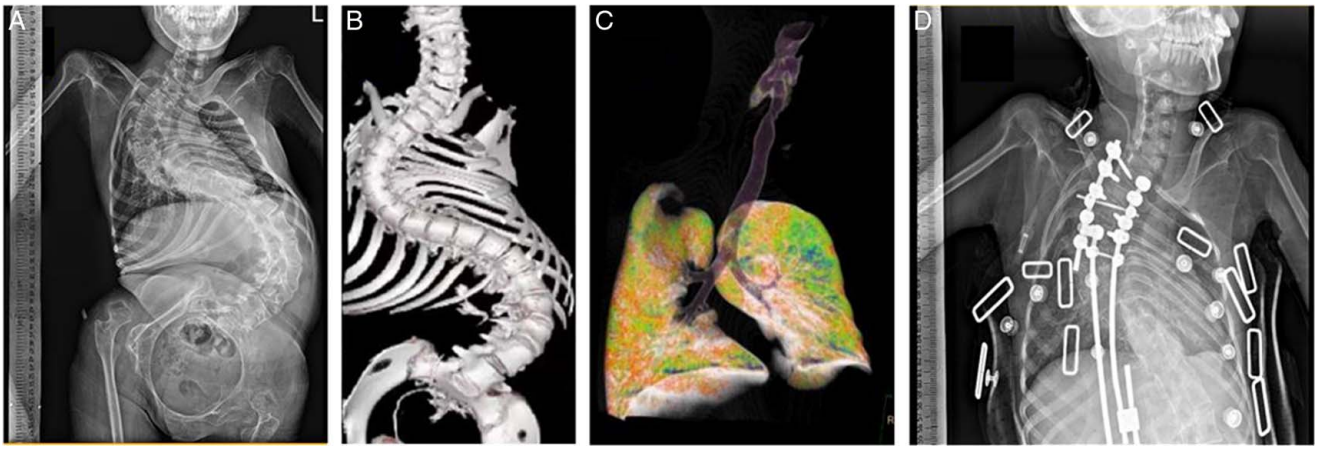

Figure 1. Typical case 1 A 12-year-old female spinal muscular atrophy patient with type II, whose cobb's angle (coronal plane) was 145° for lumbar spine and 136° for thoracic spine (A, B). Preoperative three-dimensional airway reconstruction showed a little atelectasis in the right upper lobe and left lower lobe (C). Pulmonary function showed a severe restrictive ventilatory dysfunction (FVC was 32% of predicted and FEV1 was 25.2% of predicted). She underwent posterior scoliosis correction under general anaesthesia and admitted to ICU with endotracheal tube and extubated at 21 h. After extubation, chest radiograph showed significant improvement of thoracic deformity (D).

is defined as the angle formed by the extension line of the upper edge of the parietal vertebra and the extension line of the lower edge of the terminal vertebra. In this series of cases, anaesthesiologists usually face great challenges and the considerations mainly come from two aspects: (1) The influence of pathophysiological changes on anaesthesia. For instance, preoperative pulmonary dysfunction, thoracic malformations and airway difficult due to limited movement of the cervical spine pose and Mouth opening decreased pose challenges in anaesthesia management; (2) How to maximize the protection of patients' spinal cord function and prevent pulmonary complications. From September 2019 to September 2022, the Spine Center of our institution performed surgery on 26 SMA patients with severe scoliosis (Cobb≧90°), of which Figures 1 and 2 are two typical cases. All patients had no severe adverse events during the perioperative period. According to our experience and other suggestions in the literature[5–7], we then provided a simple comprehensive checklist summarising our management strategy.

Methods

We reviewed and analyzed the electronic medical records of 26 SMA patients with severe scoliosis (Cobb≧90°) underwent posterior spinal fusion surgery from September 2019 to September 2022. All patients were surgically treated by a team of seasoned surgeons. Medical records were reviewed to validate and retrieve information on the patients' characteristics (age, sex, body mass index, American Society of Anaesthesiologists grade, preoperative lung function, comorbid history, baseline BP, preoperative Cobb angle, scoliosis type, orthopaedic rate, vertebral fusion segments, pedicle screws, and surgical osteotomy) and intraoperative data. Postoperative data inclusion incidence of postoperative complications, postoperative ICU/hospital length of stay and oxygen therapy on the first day after extubation.

This retrospective single-centre data review followed the Strengthening the Reporting of Observational Studies in Epidemiology (STROBE) statement guidelines and was conducted in accordance with the principles of the Declaration of Helsinki. Ethical approval for this study (******) was provided by Medical Ethics Committee of (******), on 15 June 2023. The study was registered at the Chinese Clinical Trial Registry (http://www.chictr.org.cn) with registration number ChiCTR ****** on 6 July 2023. This work has been reported in line with the PROCESS criteria[8] and all data were obtained from the hospital's electronic medical records system.

Results

The clinical data of 26 SMA patients with ASA grade III–IV (type I $n = 1$ and type II $n = 25$) undergoing posterior spinal fusion were collected. The median BMI was 16.35 kg/m^2 [interquartile range (IQR): 14.63–18.45].Twenty-five patients completed the preoperative pulmonary function test except one due to inability to cooperate, 9 patients (34.6%) had mild to moderate restrictive ventilatory dysfunction, 11 patients (42.3%) had severe restrictive ventilatory dysfunction and 5 patients had extremely severe restrictive ventilatory dysfunction, and one patient needed noninvasive positive pressure ventilation (NIPPV) at the night before operation. The results also showed that median FVC% of pred and FEV1% of pred were 31.6 (IQR: 25.7–47.7) and 31.0 (IQR: 25.1–50.8), respectively. The median Cobb angle of all patients before operation was 117.0 degree (IQR: 98.8–140.0) in coronal plane and 105.5 degree (IQR: 93.0–121.25) in sagittal plane (Table 1).

Data related to anaesthesia, intra- and post-operative was shown in Table 2. Nineteen patients (73.1%) underwent Oral or nasal intubation under the guidance of FOB. The mean time from the induction of anaesthesia to the start of surgery was 74.6 min. The median anaesthesia time of 25 patients treated under total intravenous anaesthesia was 425 minutes (IQR: 358–512). Only one patient administered intraoperative sevoflurane inhalation anaesthesia.

The median operation time of all patients was 338 minutes (IQR: 295–364), the median blood loss and the amount of red

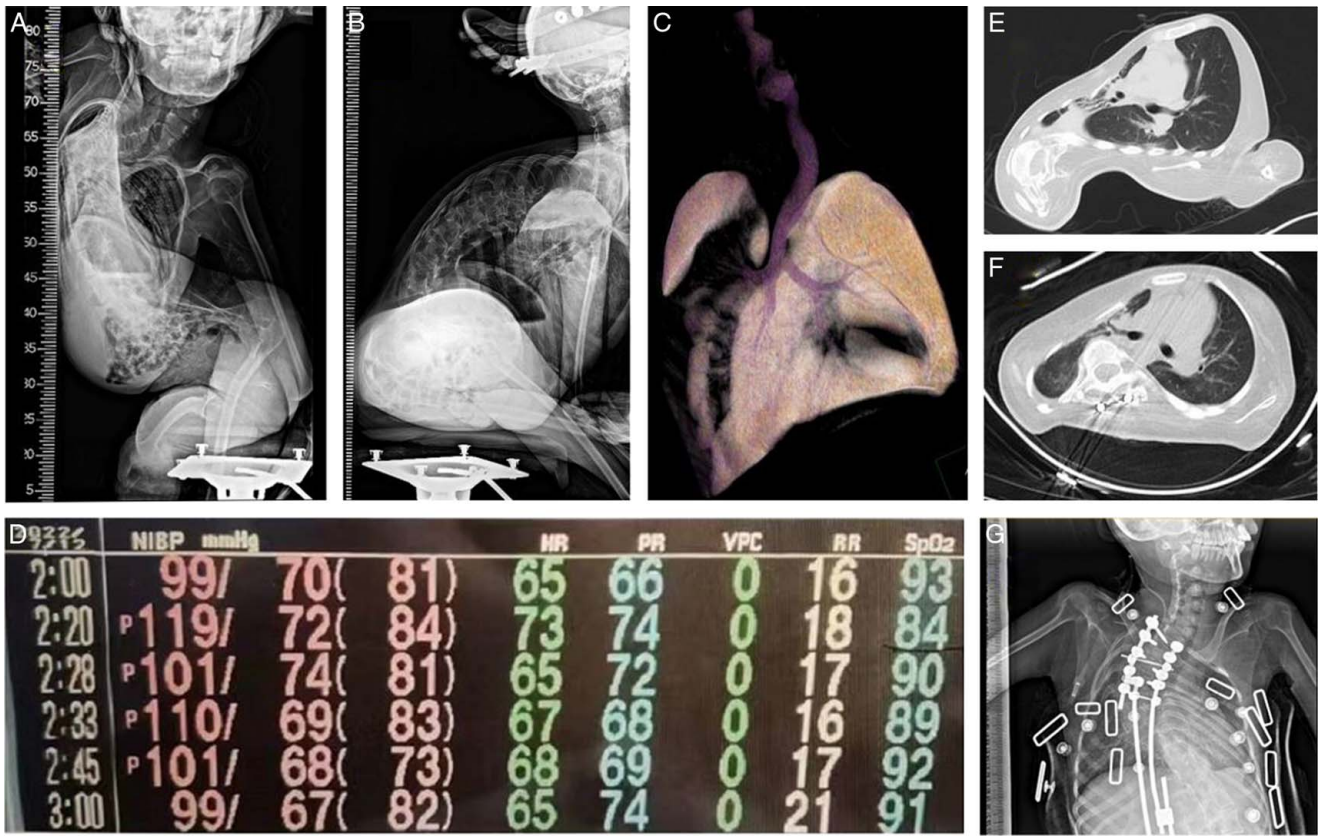

Figure 2. Typical case 2 A 16-year-old male spinal muscular atrophy patient with type II, whose cobb's angle was 163° for scoliosis (A, E) and 128° for kyphosis (B). Preoperative three-dimensional Airway reconstruction showed thoracic and spinal deformity with right lung destruction (C). Pulmonary function showed a severe restrictive ventilatory dysfunction (FVC was14% of predicted and FEV1 was 14.3% of predicted). Noninvasive ventilator was used at night and SPO_2 was less than 90% on average (D). He underwent posterior scoliosis correction under general anaesthesia and extubated when the operation is over. Then he was admitted to ICU for intensive care. Re-examination of chest X-ray showed that the thoracic deformity was significantly improved (F, G).

blood cell suspension during the operation were 1200 ml (IQR: 600–2125) and 392 ml (IQR: 112–600), respectively. The median amount of autologous blood transfusion and crystal rehydration were 375 ml (IQR: 125–500) and 2000 ml (1538–2500), respectively. The median number of vertebral fusion segments was 18 (IQR: 13.0–18.3), and 4 patients underwent intraoperative osteotomy.

After operation, the Cobb angle and correction rate in the coronal plane were median 54.0 degrees (IQR: 41.5–67.3) and 54.4% (IQR: 48.2–61.3), and the Cobb angle and correction rate in the sagittal plane were median 41.5 degrees (IQR: 35.0–54.3) and 56.6% (IQR: 48.9–66.3), respectively. Eighteen patients (69.2%) were extubated in the operating room postoperatively. Eight patients were admitted to the ICU due to inadequate spontaneous respiration for maintaining oxygenation, potentially attributed to preoperative respiratory dysfunction, a significant Cobb Angle and excessive intraoperative bleeding. All patients were sent to the intensive care unit (ICU) for treatment after surgery. The length of mechanical ventilation with endotracheal intubation in ICU was median 17.5 hours (IQR: 8.3–21.0) in 8 patients. The ICU length of stays averaged 2 days (IQR: 1–3), and the length of stay of postoperative hospital was median 19 days (IQR: 13–24). At the first day after operation or removing the endotracheal tube, 10 patients (38.5%) needed inhale oxygen via nasal high flow, 12 patients (46.2%) needed inhale oxygen via nasal catheter, and only 4 patients (15.4%) did not need oxygen therapy. Postoperative pneumonia developed in 9 patients (34.6%), atelectasis in 2 patients (19.2%), and pleural effusion in 6 patients (23.1%). All patients did not need special oxygen therapy after discharge.

Discussion

As the absence of evidence-based guidelines, our protocol is developed from the perioperative management of a series of SMA type I and II patients (Table 3). The table described the experiences gained and is the first to concentrate on anaesthetic management of severe scoliosis (Cobb $\geqq$ 90°) patients with spinal muscular atrophy undergoing posterior scoliosis correction. With few similar studies in the literature, it is difficult to compare the 'effectiveness' of our strategy with others. However, with no postoperative re-intubation, no deaths, and no patient required more respiratory support than they had on admission, it demonstrates that our plan based on common considerations is safe and effective. All that is due to our MDT team's thorough preoperative evaluation, adequate intraoperative preparation and careful postoperative monitoring.

Table 1

Demographic and preoperative characteristics of the patients

Variable	
Sex (female/male)	16/10
Age, year, median (IQR)	18 (13–22)
Height, cm, median (IQR)	148 (140–160)
Weight	
Actual body weight, kg, median (IQR)	36.7 (31.4–42.5)
BMI, kg/m^2, median (IQR)	16.35 (14.63–18.45)
SMA type, *n* (%)	
I	1 (3.8)
II	25 (96.2)
III	0
IV	0
Anaesthesia ASA class, *n* (%)	
I	1 (3.9)
II	2 (7.7)
III	18 (69.2)
IV	5 (19.2)
Preoperative respiratory function, *n* (%)	
Untested	1 (3.9)
Normal	0
Mild restrictive	2 (7.7)
Moderate restrictive	7 (26.9)
Severe restrictive	11 (42.3)
Extremely severe restrictive	5 (19.2)
FVC % of predicted	31.6 (25.7–47.7)
FEV1% of predicted	31.0 (25.1–50.8)
Oxygen therapy at admission, *n* (%)	
NIPPV support	1 (3.8)
High-flow oxygen therapy	0
Nasal cannula oxygen therapy	0
None	25 (96.2)
History of congenital heart disease	0
Preoperative Cobb's angle	
Coronal plane, degree, median (IQR)	117.0 (98.8–140.0)
Sagittal plane, degree, median (IQR)	105.5 (93.0–121.25)
90°–119°, *n* (%)	15 (57.7)
120° –149°, *n* (%)	9 (34.6)
≥ 150°, *n* (%)	2 (7.7)

ASA, American Society of Anesthesiologists; IQR, interquartile range; NIPPV, noninvasive positive pressure ventilation; SMA, spinal muscular atrophy.

Pre-anaesthesia evaluation

Airway evaluation: Some SMA patients have difficulty in opening their mouth because of the stiffness of the lower frontal joint. It is reported that the incidence of difficulty in opening mouth in SMA type II children aged 6–10 years is about 23%, and the incidence will increase significantly to 46–78% after adulthood[9]. In addition, due to severe scoliosis and joint contracture, the cervical spine activity of such children is often limited, which may be related to many difficult intubations or intubation failures previously reported[10]. In our research, the majority (73.1%) of the patients presented with a challenging airway and required intubation under the guidance of bronchoscopy.

Pulmonary function evaluation: Severe scoliosis often leads to pulmonary restrictive disease and affects respiratory function. Therefore, preoperative pulmonary function assessment is required before anaesthesia, especially to assess the patient's dependence on oxygen, so as to provide a baseline reference for the target value of postoperative pulmonary rehabilitation.

Table 2

Intraoperative and postoperative data in the 26 operated patients

Anaesthesia-related and surgery-related data	
Induction of GA	
Time from induction of GA to start of surgery (min)	74.6 ± 17.4
Midazolam IV (mg/kg)	0.078 ± 0.003
Propofol IV (mg/kg)	2.0 ± 0.8
Fentanyl IV (μg/kg)	2.0 ± 0.2
Rocuronium IV (mg/kg)	0.6 ± 0.3
Difficult intubation, *n* (%)	19 (73.1)
Oral or nasal intubation under the guidance of FOB, *n* (%)	19 (73.1)
During GA and surgery	
Length of sevoflurane administration (in 1 patient having GA with volatile anaesthetic), min	362
Length of total intravenous anaesthesia (in 25 patients having GA with Propofol and Remifentanil), min, median (IQR)	425 (358–512)
Tidal volume (VT), ml/kg, median (IQR)	6.8 (6.0–8.0)
Length of surgery, min, median (IQR)	338 (295–364)
Intraoperative blood loss, ml, median (IQR)	1200(600–2125)
Intraoperative fluids, ml, median (IQR)	
Ringer's acetate IV	2000(1538–2500)
HES	125 (0–500)
CRBC	392 (112–600)
Plasma	200 (0–400)
Autotransfusion	375 (125–500)
Diuresis, ml, median (IQR)	950 (688–1200)
No. vertebral bone fusion segments, individual, median (IQR)	18 (13.0–18.3)
Number of pedicle screw, individual, median (IQR)	25 (19–29)
Intraoperative osteotomy, *n* (%)	4 (15.4)
Postoperative Cobb's angle	
Coronal plane, degree, median (IQR)	54.0(41.5–67.3)
correction rate, %	54.4(48.2–61.3)
Sagittal plane, degree, median (IQR)	41.5(35.0–54.3)
correction rate, %	56.6(48.9–66.3)
Preoperative blood glucose, mmol/l, median (IQR)	5.9 (5.6–6.2)
Postoperative blood glucose, mmol/l, median (IQR)	8.4 (7.4–9.6)
Post-anaesthesia intubation PH levels, median (IQR)	7.40(7.37–7.43)
PH levels after surgery, median (IQR)	7.34(7.31–7.38)
Post-anaesthesia intubation PCO_2 levels, mmHg, median (IQR)	33.9(31.8–38.0)
PCO_2 levels after surgery, mmHg, median (IQR)	39.7(35.3–42.8)
Post-anaesthesia intubation PO_2 levels, mmHg, median (IQR)	227.0(199.5–334.5)
PO_2 levels after surgery, mmHg, median (IQR)	218.0(200.5–284.5)
Status of post-GA	
Extubation, *n* (%)	18 (69.2)
Time from end of surgery to extubation, min, median (IQR)	20 (15–21)
Controlled ventilation via endotracheal tube, *n* (%)	8 (30.8)
Postoperative data	
Admitted to ICU, *n* (%)	26 (100)
Length of mechanical ventilation with endotracheal intubation, hours, median (IQR)	17.5 (8.3–21.0)
ICU Length of Stay, days, median (IQR)	2 (1–3)
Oxygen therapy on the 1st day after extubation, *n* (%)	
NIPPV support	0
High-flow oxygen therapy	10 (38.5)
Nasal cannula oxygen therapy	12 (46.2)
None	4 (15.4)
Post-op length of stay, days, median (IQR)	19 (13–24)
Post-op pulmonary complications, *n* (%)	
Pneumonia	9 (34.6)
Atelectasis	2 (19.2)
Pleural fluid	6 (23.1)
Re-intubation	0
Oxygen therapy at discharge, *n* (%)	
NIPPV support	0
High-flow oxygen therapy	0
Nasal cannula oxygen therapy	0
None	26 (100)

CRBC, concentrated red blood cells; FOB, fiberoptic bronchoscope; GA, general anaesthesia; IQR, interquartile range; NIPPV, noninvasive positive pressure ventilation op, operation; OR, operating room.

Table 3

Anesthetic management strategy for severe scoliosis patients with SMA in Xinhua Hospital

	Preoperative	Intraoperative	Postoperative
Respiratory system	1. Airway evaluation; 2. Pulmonary function evaluation 3. Oxygen-dependence evaluation	1. Intubation: trachea was intubated orally or nasally under the guidance of fiberoptic bronchoscope according to treatment of difficult airway; 2. Individualized lung-protective ventilation strategies: VT: 6 ml/kg; PEEP: driving pressure (△P)-guided PEEP titration.	1. Rehabilitation therapist guided pulmonary rehabilitation exercise immediately after entering the ICU if the patient cooperates no matter whether the extubation is completed. 2. Sequential oxygen therapy after extubation targeting the level of oxygen-dependence before operation: high-flow oxygen inhalation —Nasal cannula oxygen therapy
Cardiac system	Bedside echocardiography to re-evaluate cardiac function (LVEF and LVFS)	1. Restrictive liquid management and goal-directed fluid therapy strategy (Goals: MAP≧80 mmHg, urine volume≧1 ml/kg/h; Blood lactic acid < 2 mmol/l, Scvo2 ≧65%); 2. Blood transfusion scheme: transfusion when Hct is ≦30% - 35%	Continue Restrictive liquid management and goal-directed fluid therapy strategy to avoid pulmonary oedema caused by excessive liquid.
Anaesthetics	None	TIVA without muscle relaxants	Sufficient analgesia and proper sedation
Other	MDT assessment[a] Blood glucose monitoring Nutrition support[b]	Blood glucose monitoring Body temperature protection	Blood glucose monitoring Body temperature protection Nutrition support etc.

Hct, haematocrit; LVEF, Left ventricular ejection fraction; LVFS, Left Ventricular Fraction Shortening; MAP, Mean arterial pressure; $Scvo_2$, systemic central venous oxygen saturation; TIVA, total intravenous anaesthesia; VT, Tidal volume.

[a]MDT is usually organized by surgeons, including Anaesthesia department, ICU, Imaging department, Respiratory department, Gastroenterology department, Nutrition department and Rehabilitation department, etc.

[b]The nutrition support program is implemented by the Nutrition department.

Cardiac system evaluation: SMA type I patients often have organic heart disease. Although the probability of organic heart disease in SMA II–IV children is low, they may have different degrees of arrhythmias. In addition, due to thoracic malformation and scoliosis, children often have restrictive ventilation disorder, resulting in long-term hypoxia and pulmonary hypertension, which will lead to the decline of cardiac function (especially right cardiac function)[11]. Therefore, we should re-evaluate the cardiac function of these children before anaesthesia.

Blood glucose monitoring: Skeletal muscle and fat are important sources of gluconeogenesis during fasting period before operation. However, SMA patients have low muscle and fat content and even a short fasting time may increase the risk of hypoglycemia during the perioperative period[12]. Therefore, blood glucose monitoring should be runned through the whole perioperative period.

Anaesthesia procedure

Preoperative fasting: Fasting was 6 h, water was forbidden for 2 h. Additionally, 5% glucose was given intravenously before operation.

Anaesthesia method: The patients were routinely intubated orally or nasally under the guidance of fiberoptic bronchoscope according to the reference of difficult airways. Total intravenous anaesthesia (TIVA) was used throughout the whole operation.

Intraoperative monitoring: The intraoperative monitoring included routine invasive arterial blood pressure measurement, depth of anaesthesia assessment using entropy index /BIS, neuroelectrophysiological monitoring and hourly blood gas analysis during the operation.

Anaesthetics selection: Reviewing previous research reports, SMA children have used almost all anaesthetics, and no special contraindications and complications have been found so far[13].

Inhalation anaesthetics: At present, it is generally believed that the probability of malignant hyperthermia in SMA children with no family history is not higher than that in the general population. Several retrospective studies on inhalation anaesthetics for SMA children have not reported the occurrence of malignant hyperthermia or rhabdomyolysis[13]. At the beginning in our department, we used sevoflurane inhalation anaesthesia for one patient, and no adverse events were found. In the subsequent anaesthesia scheme, we use total intravenous anaesthesia mainly based on the consideration that inhalation anaesthesia will affect intraoperative motor evoked potential (MEP) monitoring. Almost all inhalation anaesthetics can inhibit the excitation conduction of MEP in the motor cortex, spinal cord anterior horn and neuromuscular junction, etc., resulting in decreased amplitude and prolonged latency of MEP. While TIVA have little impact on MEP and can improve the sensitivity of MEP monitoring[14,15].

Intravenous anaesthetics and opioids: We mainly use sedation-analgesia with propofol and remifentanil for intravenous anaesthesia. Fentanyl is only given in the induction of anaesthesia, and remifentanil, a short-acting opioid, is mainly used for analgesia in the maintenance of anaesthesia. The main reason we considered is that fentanyl overdose can result in respiratory depression, which affects the extubation after surgery.

Muscle relaxants: In this case series, only 0.6 mg/kg rocuronium was given during anaesthesia induction, and no muscle relaxants were used during anaesthesia maintenance, mainly based on three considerations: (1) Scoliosis correction surgery requires less muscle relaxation and will not affect the process of surgery under the condition of sufficient analgesia; (2) Avoid interfering with intraoperative MEP monitoring, affecting arousal effect and avoiding respiratory depression caused by residual muscle relaxants after surgery. (3) In view of the significant increase in the sensitivity of SMA children to nondepolarizing muscle relaxant and the potential risks of rhabdomyolysis and hyperkalemia[16,17], if necessary, we recommend strict titration administration under continuous muscle relaxant monitoring to minimize the dosage of muscle relaxants and routine use of antagonists.

Analgesia protocol: Analgesia protocol in postoperative period are postoperative incision local anaesthesia, patient-controlled intravenous analgesia and oral NSAID/tramadol in the ward.

Fluid infusion and blood transfusion management

In order to avoid pulmonary oedema caused by excessive liquid, we adopted restrictive fluid management and goal-directed fluid therapy strategy. Specifically, the goal is to maintain MAP≧80 mmHg, urine volume≧1 ml/kg/h, Blood lactic acid < 2 mmol/L and Scvo2 ≧65%(obtained through an internal jugular central venous catheter). The blood pressure is set at MAP≧80 mmHg due to the spinal cord can maintain good blood perfusion at this level of MAP[18]. The blood transfusion scheme depends on the situation of haemorrhage and erythrocyte suspension are given when Hct is lower than 30–35%.

Individualized lung-protective ventilation strategies and extubation

The success of extubation depends on two factors:

Reduce the damage of anaesthesia to lung function: Studies have shown that mechanical ventilation during operation will cause atelectasis[19]. Although a small amount of atelectasis is not enough to affect the pulmonary function of people with normal pulmonary function, A small amount of atelectasis can aggravate the hypoxia symptoms of SMA patient with severe scoliosis due to their pulmonary function reserve is almost lost. Therefore, we use lung-protective ventilation strategy during anaesthesia. A previous study in our department found that compared with the standard intraoperative ventilation, the method using driving pressure (△P)-guided PEEP titration (tidal volume of 6 ml/kg body weight plus individualized PEEP for minimizing △P) can significantly reduce the incidence and severity of postoperative pulmonary complications (PPCs), improved pulmonary function without increasing the incidence of other complications[20]. Although these findings were found during abdominal surgery, we used the strategy for SAM patients with severe scoliosis who are ventilated in prone position due to there are currently no relevant ventilation guidelines for such patients. In addition, it should be noted that the patient's tidal volume was calculated based on the actual body weight rather than predicted body weight. The thoracic deformity leads to varying degrees of lung volume reduction, making it difficult to assess the predicted body weight of such patients. Briefly, the inhaled oxygen concentration is usually set at 60–80% at the beginning of anaesthesia to keep the oxygen saturation at 95–100%, so as to avoid atelectasis caused by high concentration oxygen absorption to the greatest extent. Tidal volume was set at 6 ml/kg and an incremental PEEP titration protocol (an increment of 2 cm H_2O for every 8 min from 0 to 14 cm H_2O) was performed to identify the optimal individualized PEEP that resulted in minimum driving pressure (calculated as plateau pressure – PEEP), indicating maximum respiratory compliance. Subsequently, this optimal individualized PEEP was maintained throughout the procedure. Because spinal surgical correction may change the patient's pulmonary compliance, we corrected the best PEEP again with the method described above after completing spinal correction. The driving pressure above 15 cm H_2O believed to be associated with PPCs. In our clinical practice, the minimum driving pressure corresponds to the PEEP setting value, and we have not encountered any instances of driving pressure exceeding 15 cm H_2O.

The effect of operation in the cardiopulmonary function: Theoretically, surgical correction can improve the patients' restrictive ventilation dysfunction. However, this series of patients are all patients with severe scoliosis, and the change of bilateral lung volume caused by obvious thoracic malformation may aggravate hypoxia, because the pulmonary function reserve of these patients has basically been lost. And if the spine is overcorrected, mediastinal displacement may cause a drop in blood pressure. The spontaneous breathing test (SBT) was performed after the patient was fully awake and his/her muscle strength recovered.When the withdrawal criteria were met(Respiratory rate < 25 times/min and SPO_2 > 90% when 60% oxygen concentration is given), the extubation was carried out.

Postoperative monitoring in ICU

All patients need to enter the ICU for continuous cardiopulmonary function monitoring for at least 24 h. Although the lung-protective ventilation strategy is adopted during the operation, mechanical ventilation and surgery will affect the cardiopulmonary function to varying degrees. In addition to the impact of postoperative pain and wearing protective equipment, patients' dependence on oxygen will increase temporarily. Therefore, we adopted the sequential oxygen therapy method, that is, if need (Respiratory rate ≧25 times/min and SPO_2 ≦ 90%), high-flow oxygen inhalation was given for 24 h to provide higher PEEP after extubation, and then the nasal oxygen inhalation was used as appropriate. In addition, airway suction and secretion clearance are also very important. These monitoring measures and treatment may be fully guaranteed in the ICU.

In short, the biggest challenge for such patients during the perioperative period is the impact of surgical procedures and anaesthesia on their cardiopulmonary function. According to our experience, there are three main points: (1) Displacement of the mediastinum can lead to a decrease in blood pressure and even heart rate if the spinal correction is overcorrected during the operation; (2) Inappropriate ventilation strategies can lead to increased pulmonary atelectasis, resulting in extubation failure; (3) Postoperative thoracolumbar braces may cause difficulty in expectoration and restrictive dysfunction, which may lead to re-intubation. The main protective measures we take to overcome the above problems are (1) anesthesiologists need to closely observe the impact of surgical correction on circulation and maintain communication with surgeons; (2) Adopt individualized lung-protective ventilation strategies to avoid pulmonary atelectasis; (3) Rehabilitation training and high-flow oxygen inhalation—Nasal cannula Sequential oxygen therapy after extubation to avoid re-intubation. It should be noted that such patients are very fragile, with limited compensatory function of the heart and lungs, and small negative measures may have significant clinical manifestations. Negligence in any aspect can lead to serious consequences. Therefore, we emphasize that multidisciplinary participation in the treatment of patients is necessary to ensure smooth progress through the perioperative period.

Ethical approval

Ethical approval for this study (XHEC-C-2023-067-1) was provided by Medical Ethics Committee of Xinhua Hospital Affiliated

to Shanghai Jiao Tong University School of Medicine, Shanghai, China, on 15 June 2023.

Consent

Written informed consent was obtained from the patients or the patient's parents/legal guardian (for minors) for publication of this case series and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contribution

L.J. and Y.M. conceived the study, administered the project, conducted formal analysis, and wrote the first draft of the manuscript. L.W., Y.D. and N.H. contributed in the methodology, formal analysis and writing original draft. L.W., Y.D., N.Y. and J.D. performed data collection. J.Y. supervised the whole study. All authors reviewed and approved the final version of the manuscript.

Conflicts of interest disclosure

The authors declare no conflict of interest.

Research registration unique identifying number (UIN)

1. Name of the registry: Chinese Clinical Trial Registry.
2. Unique Identifying number or registration ID: ChiCTR 2300072561.
3. Hyperlink to your specific registration (must be publicly accessible and will be checked): https://www.chictr.org.cn/showproj.html?proj=199660.

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Availability of data and materials

All data can be made available upon request to the corresponding authors.

Provenance and peer review

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