Original Article

Peri-operative management of children with spinal muscular atrophy

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ABSTRACT

Background and Aims: Current multi-disciplinary management of children with spinal muscular atrophy (SMA) often requires the surgical management of spinal deformities. We present the outcomes of our peri-operative experience around the time of their spinal surgery and share our neuromuscular perioperative protocol. **Methods:** A single-centre retrospective chart review was performed to evaluate all children with SMA types I and II that underwent thoracolumbar spinal deformity correction (posterior spinal fusion or growing rod insertion) from 1990 to 2015. Electronic medical records were reviewed to assess pre-operative, intraoperative, and postoperative variables. T-tests, Wilcoxon Rank Sum, Fisher's Exact tests were performed as appropriate. **Results:** Twelve SMA I and twenty-two SMA II patients were included. Type I patients tended to be smaller and had a higher percentage (36.4% vs 4.5%) of American Society of Anesthesiologists (ASA) class 4 patients. Preoperative total parenteral nutrition (TPN) was utilised in 75.0% of type I and 18.2% type II patients. A difficult intubation was experienced in around 25% of the patients (20.0% SMA I, 27.3% SMA II). Approximately two hours of anaesthetic time was required in addition to the actual surgical time in both types. The intensive care unit (ICU) length of stay averaged 6 (4.0-7.5) days for type I and 3 (3-5) days for type II ($p = 0.144$). Average post-operative length of stay was (8 (7-9) vs. 7 (6-8)) $P = 1.0$. **Conclusion:** Children with type I and II SMA have similar hospital courses. The surgical and anaesthesia team should consider perioperative TPN and NIPPV (non-invasive positive-pressure ventilation), anticipate difficult intubations, longer than usual anaesthetic times, and potentially longer ICU stays in both SMA type I and II.

Key words: Anaesthesia, management, perioperative, spinal muscular atrophy

INTRODUCTION

Spinal Muscular Atrophy (SMA) is a rare disease that has devastating effects on the neuromuscular system resulting in progressive weakness. This disease often leads to shortened lifespan due to progressive respiratory failure,[1,2] especially in the most severe subtypes (Type I and II). While exciting new treatments including nusinersen934[3] are changing disease progression, many young non-ambulatory children with SMA types I and II have progressive spinal deformities requiring treatment. In addition to the introduction of disease modifying therapies, aggressive multidisciplinary medical management has drastically improved their overall survival.[2] Orthopaedic management of the spinal deformities either with spinal fusions or posterior (distraction type) growing rods may help to slow pulmonary decline.[4]

With improved survival, there will likely be increasing numbers of severely affected children that may require spinal stabilisation. These children may be

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cared for at institutions that are not familiar with the peri-operative care of these frail children undergoing such major surgeries. While much has been written about the orthopaedic aspects of treating these spinal deformities,[5] current literature lacks comprehensive, SMA specific, peri-operative management strategies, focusing on major surgery (such as spine surgery). Most of the previous reports are expert opinion,[6] case studies,[7,8] combine diagnoses,[7] and focus only on the respiratory system. In this manuscript, we describe the results of our 25 year history managing spine deformity in these children, with particular focus on the most severe SMA type I and type II children, undergoing either posterior spinal fusion or limited fusion with placement of posterior distraction type spinal growing rods. We then provide a simple comprehensive checklist summarising our management strategy based on our experience and other recommendations in the literature.

METHODS

This study was approved by the Institutional Review Board at the University of Wisconsin-Madison (IRB #2017-0008). A single centre retrospective chart review was performed to include all children with SMA types I and II undergoing thoracolumbar spinal surgery including either posterior spinal fusion or growing rod insertion from July 27th 1990 to August 11th 2015. 34 total (12 SMA type I, 22 SMA type II) patients who underwent spinal surgery at our institution were identified. Children with SMA who were undergoing other surgical procedures that were not thoracolumbar spinal surgery were excluded from the study. Anaesthesia management proceeded with either intravenous (IV) propofol (21 patients) or inhalational induction with nitrous oxide and sevoflurane (13 patients). Fiberoptic bronchoscopy was most often used as mouth opening becomes limited often prohibiting the use of videolaryngoscopy (which was only used in 4 cases). Then, anaesthetic maintenance proceeded with a total IV anaesthetic with propofol and remifentanil (29 patients) or maintenance with sevoflurane (5 patients). Perioperatively, institutional practice was use of remifentanil if using a total IV anaesthetic with neuromonitoring. Neuromuscular blockade was not routinely used especially when neuromonitoring was performed. In rare cases, rocuronium or cisatricurium was used for neuromuscular blockade and reversed at the conclusion of surgery. Intraoperatively, Standard American Society of Anesthesiologists (ASA)monitors, invasive blood pressure monitoring with arterial line, and in 26 patients somatosensory evoked potentials were monitored and in 22 patients motor evoked potentials (MEP) were monitored. Additionally, fentanyl and either morphine or hydromorphone was used for post-operative pain control in 12/34 patients. Postoperatively, 16 patients received an epidural catheter with an infusion of local anaesthetic. Electronic medical records of 12 SMA I and 22 SMA II patients were reviewed to assess pre-operative, intraoperative and postoperative variables.

T-tests, Wilcoxon Rank Sum, Fisher's Exact tests were performed as appropriate. P values <0.05 were considered significant, however P values were also adjusted using the Benjamini-Hochberg adjustment to control any false discovery.

RESULTS

Thirty-four type I and type II SMA patients who underwent spinal surgery at our institution were identified. Twelve were SMA type I and twenty-two were SMA type II patients. Type I patients tended to be smaller and had a higher percentage (36% vs 4.5%) of ASA class 4 patients. Pre-operative total parenteral nutrition (TPN) was utilised in 75% of type I and 18% of type II patients. A difficult intubation was experienced in 20.0% SMA I and 27.3% SMA II cases. In our retrospective review, we looked for documentation of difficult intubation. No intraoperative differences were found in surgical time, estimated blood loss (EBL), or transfusion volumes. Approximately two hours of anaesthetic time was required in addition to the actual surgical time regardless of SMA type. Postoperative courses were also similar for both types with all patients initially admitted to the intensive care unit (ICU) and ICU length of stays averaged 6 (4.0-7.5) days for type I and 3 (3-5) days for type II patients ($p=0.144$). Average post-operative length of stay was 8 days for type I (7-9) vs. 7 days for type II (6-8)) patients. $P = 1.0$. Atelectasis was common post-operatively in both type I (25%) and type II (32%) patients. Only one type I and one type II patient had post-operative pneumonia. No patients in either cohort required reintubation or had respiratory failure requiring support. No perioperative deaths or conversions to permanent controlled ventilation via tracheotomy were required during the perioperative stay [Table 1].

Table 1: Pre-operative, intra-operative and post-operative variables in children with SMA types I and II

Variable	SMA type I (*n*=12)	SMA type II (*n*=22)	*P*	Adj *P*	Overall
Pre-Operative					
Weight - kg	18.0 (3.4)	29.2 (12.7)	0.001	0.026	28.6 (17.0)
ASA Class			0.033	0.254	
2	0 (0.0%)	0 (0.0%)			0 (0.0%)
3	7 (63.6%)	21 (95.5%)			31 (86.1%)
4	4 (36.4%)	1 (4.5%)			5 (13.9%)
Pre-operative NIPPV			0.043	0.262	
None	0 (0.0%)	4 (18.2%)			7 (18.9%)
Nocturnal	10 (83.3%)	18 (81.8%)			28 (75.7%)
Continuous	2 (16.7%)	0 (0.0%)			2 (5.4%)
Pre-operative TPN					
Parental Nutrition Initiated Pre-op	9 (75.0%)	4 (18.2%)	0.002	0.046	13 (35.1%)
Intra-Operative					
Difficult Intubation	2 (20.0%)	6 (27.3%)	1	1	8 (22.9%)
Direct Laryngoscopy Technique - Yes	9 (90.0%)	17 (77.3%)	0.637	0.801	29 (82.9%)
TPN Intra-op	5 (41.7%)	4 (18.2%)	0.224	0.514	9 (24.3%)
Crystalloid Saline or Lactated Ringers - ml	900.0 (555.0-1950.0)	2150.0 (1425.0-3325.0)	0.047	0.262	2000.0 (922.5-3311.8)
Crystalloid Saline or Lactated Ringers - ml/kg	45.0 (34.2-99.5)	76.7 (57.4-97.1)	0.252		
Urine Output - ml	350.0 (110.0-537.5)	400.0 (237.5-712.5)	0.366	0.595	400.0 (220.0-925.0)
Urine Output - ml/kg	19.2 (8.2-28.6)	11.9 (7.8-22.2)	0.651		
Estimated Blood Loss - ml	325.0 (150.0-440.0)	400.0 (300.0-862.5)	0.187	0.457	400.0 (250.0-750.0)
Estimated Blood Loss - ml/kg	17.9 (8.8-25.0)	17.6 (11.2-24.2)	0.589		
Transfusion	9 (75.0%)	16 (72.7%)	1	1	28 (75.7%)
Packed Red Blood Cells - ml	350.0 (187.5-425.0)	325.0 (12.5-612.5)	0.796	0.941	350.0 (50.0-700.0)
Packed Red Blood Cells - ml/kg	18.4 (10.7-22.3)	10.6 (1.1-18.3)	0.244		
Time with Temp <36	0.0 (0.0-90.0)	90.0 (15.0-157.5)	0.068	0.264	75.0 (0.0-135.0)
Anaesthesia Total Time - hr	7.1 (2.1)	7.5 (2.1)	0.605	0.801	7.7 (2.2)
Surgery Total Time - hr	4.9 (2.0)	5.6 (1.6)	0.308	0.541	5.6 (2.0)
Spinal Deformity					
Cobb Angle Pre	64.7 (19.7)	59.3 (26.8)	0.55		61.4 (24.0)
Cobb Angle Post	21.0 (11.5)	25.3 (24.3)	0.502		23.7 (20.4)
Cobb Angle Difference	-43.1 (18.5)	-37.8 (32.8)	0.6		-40.0 (27.5)
Pulmonary Status Post-op			0.319	0.541	
Controlled ventilation via endotracheal tube or trach	9 (75.0%)	20 (90.9%)			31 (83.8%)
NIPPV	3 (25.0%)	2 (9.1%)			6 (16.2%)
Spontaneous	0 (0.0%)	0 (0.0%)			0 (0.0%)
Post-Operative Course					
Admitted to ICU - Yes	11 (100.0%)	21 (100.0%)	1	1	35 (100.0%)
ICU Length of Stay - nights	6.0 (4.0-7.5)	3.0 (3.0-5.0)	0.023	0.227	4.0 (3.0-6.0)
Total Length of Stay - nights	8.5 (7.8-10.0)	8.0 (6.0-9.0)	0.059	0.263	8.0 (6.0-9.0)
Post-op Length of Stay - nights	8.0 (7.0-9.0)	7.0 (6.0-8.0)	0.175	0.457	8.0 (6.0-9.0)
NIPPV at Discharge			0.179	0.457	
None	0 (0.0%)	4 (18.2%)			7 (18.9%)
Nocturnal	10 (83.3%)	17 (77.3%)			27 (73.0%)
Continuous	2 (16.7%)	1 (4.5%)			3 (8.1%)
Post-op Pulmonary Complications			1	1	
Pneumonia	1 (8.3%)	1 (4.5%)			2 (5.4%)
Respiratory Failure Requiring Support	0 (0.0%)	0 (0.0%)			0 (0.0%)
Re-intubation	0 (0.0%)	0 (0.0%)			0 (0.0%)
Atelectasis	3 (25.0%)	7 (31.8%)			10 (27.0%)
None	8 (66.7%)	14 (63.6%)			25 (67.6%)

ASA-American Society of Anesthesiologists, TPN-Total parenteral nutrition, NIPPV-Non-invasive positive pressure ventilation, ICU-Intensive care unit

DISCUSSION

This study demonstrates that SMA children, prior to disease modifying treatments, independent of disease type can safely be managed through very major orthopaedic surgery. Previous studies have focused on the orthopaedic outcomes of spinal surgery.[5,7,9-11] In many of these previous spine studies, SMA patients are

Table 2: Perioperative management strategy for children with SMA[23-25]

	Pre-operative consultation	Intraoperative	Post-operative
Pulmonary	Pulmonary consultation for all patients prior to surgery Conduct pulmonary function testing if patient is able to perform Consider preoperative training on non-invasive positive pressure ventilation (NIPPV) if not currently managed with respiratory support during sleep due to increased risk for hypoventilation post operatively secondary to pain management and anaesthesia Consider preoperative training with mechanical insufflation-exsufflation if history of ineffective cough, recurrent pneumonia or low MEP (i.e., MEP <60 cm H_2O)	Respiratory support per endotracheal intubation with positive pressure ventilation Neuromuscular patients with chronic respiratory insufficiency require ventilatory support and not oxygenation alone, during induction and recovery. Patients should be kept intubated post operatively or extubated to NIPPV Monitoring during anaesthesia should include capnography to complement oximetry to avoid hypercarbia and hypoxemia.[25]	Implement airway clearance post operatively with secretion mobilisation technique preferably intrapulmonary percussive ventilation (IPV, Metaneb) followed by mechanical insufflation-exsufflation and airway suctioning within 1 hour of admission to post operative care unit and every 4 hours post operatively via ETT, tracheostomy tube or orally. Consider delaying extubation until respiratory secretions well controlled, weaned to room air and pain control is optimised. Optimize ventilation before adding oxygen as the most likely cause of hypoxemia is hypoventilation. Hypoventilation is exacerbated by narcotic pain use. Extubate patients to NIPPV and work toward use during sleep only. NIPPV may be needed following hospital discharge and recovery.
Anaesthesia/ Pain	Anaesthesia consultation for all patients prior to surgery with general anaesthesia Evaluate airway carefully and plan for intubation difficulties given patients' physical limitations and potential abnormalities (e.g., decreased mouth opening, enlarged tongue) Consider indirect visualisation of the larynx including the possible fiberoptic intubation (i.e., fiber optic visualisation is often necessary)	Consider use of total IV anaesthesia technique for induction and maintenance of general anaesthesia Avoid depolarising muscle relaxants (i.e., succinylcholine) and non depolarising muscle relaxants If non depolarising muscle relaxants used, titrate dose and continuously monitor neuromuscular function Avoid inhaled anaesthesics in DMD patients; these agents may be considered in SMA patients Intubation without neuromuscular blockade is preferred when possible Short acting opioids are suitable for intraoperative use in SMA patients	Sedating analgesia may have additive effect on baseline respiratory insufficiency however postoperative pain control should not be compromised because of respiratory suppression concerns.[26,27] Opiate-based analgesia in combination with acetaminophen and NSAID should be considered as part of routine post-procedural management with anticipation of providing appropriate NIV and cough assistance. Patients may also benefit from benzodiazepines for muscle spasm and discomfort.
Nutrition	Nutritional status assessment for all patients prior to surgery with goal to optimise nutrition and plan nutrition support during hospitalisation. Swallow evaluation may be helpful to determine ideal post-operative feeding strategy Pre-admit patients for total parenteral nutrition (TPN) if they require overnight enteral feeds. Patients with SMA have a fatty acid oxidation metabolic disorder as part of having SMA and do not tolerate prolong periods of time without nutrition.	Consider continuing TPN infusion if start preoperatively. Monitor glucose status intraoperatively to avoid hypoglycemia.	Initiate bowel regime to avoid and treat constipation, with consideration or prokinetic GI medications as ileus is common Any patient who cannot achieve adequate oral nutrition within 24-48 h after surgery should receive enteral feeding with small-diameter nasoduodenal tube or parenteral nutrition Consider gastric decompression with nasogastric tube in patients with GI dysmotility If patient is receiving TPN, continue TPN until patient tolerates receiving at least 50% of goal enteral feed.
Cardiac	Cardiac consultation for all DMD patients prior to surgery For patients with history of symptoms suggestive of cardiac involvement or at risk for cardiac dysfunction, cardiac evaluation should be completed within 3-6 months prior to anaesthesia.	Close cardiac monitoring	Monitor cardiac and fluid status postoperative For DMD patients, obtain postoperative cardiac consultation Use telemetry monitoring
Other	Discuss goals of care, tracheostomy potential, prolonged dependency on mechanical ventilation and advance directives with patient/family		Consult Palliative care for goals of care planning, pain management considerations

SMA-Spinal muscular atrophy; PDPH-Postdural puncture headache; NIPPV-Noninvasive positive pressure ventilation; ETT-Endotracheal tube; MEP-Maximal expiratory pressure; NIV-Non invasive ventilation; DMD-Duchenne Muscular Dystrophy; NSAID-Nonsteroidal anti-inflammatory drugs; TPN-Total parenteral nutrition; GI-Gastrointestinal

combined with other conditions (Duchenne Muscular Dystrophy or Cerebral Palsy) during the analysis[5,12-16] and often only SMA type II or type III children are studied.[11,17] Some of these studies have focused on the use of non-invasive positive pressure ventilation (NIPPV) in these mixed patients,[18,19] while others focused on the long-term pulmonary effects of spine surgery.[4,16,17] Other studies have focused more on the anaesthetic side of treating only SMA children. However, these are often case reports,[8,20] studies combining different anaesthetics with different surgical procedures on all types of SMA children,[21] or focus on anaesthetics given during very minor procedures such as intra-thecal injections.[22] This manuscript is the first to concentrate on severely affected children (Types I and II evaluated separately) undergoing similar operative treatments and compare their perioperative courses. With the dearth of similar studies in the literature, it is difficult to make direct comparisons with the 'effectiveness' of our perioperative management strategy [Table 2].[23-25] However, with only two pneumonias (6%), no post-operative re-intubation or unplanned tracheostomies, and no deaths, the serious complications appear quite low. Only one patient out of the 34 was discharged home post-operatively requiring more respiratory support (continuous non-invasive positive-pressure ventilation (NIPV)) than they had on admission (nocturnal NIPV). Although study populations differ, our results appear in line or better than previous reports,[12,26] despite our population including many more frail type I children. While around 30% of the children experienced atelectasis, this appeared independent of disease severity and consistent to other studies of patients with abnormal baseline lung function follow spine surgery.[27] However, it should be noted that the atelectasis appeared to resolve rather quickly as the average lengths of stay was one week.

Several interesting findings became apparent in our review. First, despite significant experience at our institution in dealing with children with SMA, an additional two hours of operating room time was required for safe anaesthetic management and neuromonitoring in addition to the actual operative time to safely anaesthetise and wake these children. This time could be longer at less experienced institutions. This knowledge may be useful for arranging Operating Room (OR) times, staff assignments, and anticipated Paediatric Intensive Care Unit (PICU) hand-offs. Possibly playing into that extra anaesthetic time is that nearly 25% of the children had difficult intubations. Pre-operative knowledge of this by screening may ensure proper equipment and experienced staff available for these cases. Finally, while not statistically significant, it was interesting that type I children typically had a longer PICU course but a very similar hospital stay, indicating that many were almost good enough for hospital discharge when transferred out of the PICU.

The limitations should be identified and discussed. Perhaps the greatest limitation to this study is that the peri-operative plan used today and presented in this manuscript was developed over the 25 year period caring for these children. Thus, children today may receive slightly different care than they did at the beginning of the review period, due to increased experience and overall knowledge in caring for these children. The authors feel that while having an exact standard protocol to follow from the start would have been ideal from methodology standpoint, the organic nature in which the protocol was developed does not negate our results. Moreover, over 25 years, the change from paper records to electronic medical records complicated the review process. The results presented perhaps represent the 'worst case scenario' as they include outcomes during our learning curve. The smaller sample size may be seen as another limitation. Whereas one of the limitations in many of the previous studies has been their heterogeneous population,[27] we chose to restrict our findings to a much more homogeneous population, with both approaches having their strengths and weaknesses. The purpose of the current study was to focus on the perioperative hospital stay associated with their spinal surgery, thus it may be seen as a limitation that we did not specifically report on the follow-up outcomes and complications associated with the orthopaedic procedures per se; however, we have previously reported on these outcomes in other studies[4,28] and have found them to have minimal complications.

CONCLUSION

In conclusion, this study demonstrates that with appropriate multi-disciplinary care, children with SMA undergoing spine surgery can be safely managed throughout the perioperative period. Children with type I and II SMA have similar hospital courses and can be surgically treated with proper perioperative management. The surgical and anaesthesia team should consider perioperative TPN and NIPPV, anticipate difficult intubations, longer anaesthetic times, and longer ICU stay in caring for these children. This data demonstrates few serious peri-operative complications in these fragile children undergoing major spinal surgery. The summary of our perioperative

treatment strategy may serve as a useful reference for other centres caring for these children.

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Conflicts of interest

There are no conflicts of interest.

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