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Perioperative Optimization of Patients With Neuromuscular Disorders Undergoing Scoliosis Corrective Surgery: A Multidisciplinary Team Approach

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Abstract

Study Design: Narrative review.

Objective: The high rate of complications associated with the surgical management of neuromuscular spinal deformities is well documented in the literature. This is attributed to attenuated protective physiological responses in multiple organ systems.

Methods: Review and synthesis of the literature pertaining to optimization of patients with neuromuscular scoliosis undergoing surgery. Our institutional practice in the perioperative assessment and management of neuromuscular scoliosis is also described along with a clinical vignette.

Results: Respiratory complications are the most common to occur following surgery for neuromuscular disorders. Other categories include gastrointestinal, cardiac, genitourinary, blood loss, and wound complications. A multidisciplinary approach is required for perioperative optimization of these patients and numerous strategies are described, including respiratory management.

Conclusion: Perioperative optimization for patients with neuromuscular disorders undergoing corrective surgery for spinal deformity is multifaceted and complex. It requires a multidisciplinary evidence-based approach. Preadmission of patients in advance of surgery for assessment and optimization may be required in certain instances to identify key concerns and formulate a tailored treatment plan.

Keywords

perioperative, neuromuscular, scoliosis, multidisciplinary

Introduction

Neuromuscular disorders (NMD) constitute a diverse spectrum of pathologies that involve any part of the neural axis.[1] The precise manifestations of these disorders, present unique diagnostic challenges, and are dependent on the exact location along the neural axis that is affected.

When associated with spinal deformity, the morbidity associated with NMD has traditionally been thought to reflect the underlying neurological disorder rather than the structural spinal pathology and as such surgery has been obviated. Corrective surgery is increasingly recognized to improve patients' quality of life and reduce morbidity. This is reflected in the increasing volume of spinal surgery being performed for children with NMD over the past decade.[2] Even though surgical correction has been shown to improve an individual's quality of

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life, the surgery itself is associated with high morbidity. According to recent reports, 40% to 75% of patients with neuromuscular scoliosis experience an adverse event postoperatively.[2,3] As health systems begin to lean toward implementing value, understanding and creating appropriate pathways to preoperatively optimize these individuals will be an effective way of ensuring cost-effectiveness for the entire surgical episode of care. Herein we describe, with an illustrative case, our institutional perioperative multidisciplinary approach in the management of spinal deformity in NMD.

The etiology of spinal deformities in the presence of neuropathic or myopathic NMD is not fully understood. Muscle weakness and proprioceptive sensory loss have been hypothesized to be the propagators of spinal deformity.[4] Neuromuscular scoliosis (NMS) is characterized by early-onset progression, even after skeletal maturity, owing to persistent muscle weakness and altered tone. This early-onset nature coupled with limited mobility and secondary contractures results in stiff curve patterns and pulmonary compromise. Severe curves with associated pelvic obliquity also interfere with sitting position; causing costo-pelvic impingement, pain and may lead to pressure sores and skin breakdown.

Nonsurgical treatments are usually temporary in progressive cases. Surgery mainly involves spinal fusion, which is typically posterior.[5,6] The goals of treatment are to achieve and maintain a balanced spine, improve sitting position, reduce pain, maximize pulmonary function, and minimize the burden on caregivers.[6] A multidisciplinary approach may improve perioperative care and reduce the risk of complications. The team should include spinal surgeons, pediatricians, cardiologists, respiratory physicians, nutritionists, intensive care specialists, physiotherapists, and occupational therapists.

Methods

We conducted a review of the literature to identify articles relating to surgical optimization of patients with NMS using the PubMed, Web of Knowledge, Medline, and Cochrane Database of Systematic Reviews. Only studies using human subjects and those written in English were included. A broad search strategy was employed using the terms: "Neuromuscular" AND "Scoliosis." Two reviewers (RS and FS) evaluated the abstract of each article to determine the value of those articles to our review. The bibliographies of relevant articles were also evaluated to obtain further pertinent articles. Our institutional pathway will also be described along with a clinical vignette.

Results

The commonly reported risks of spinal surgery are more pronounced in patients with NMD who are inherently medically fragile. An appreciation of the potential difficulties and complications associated with treating these patients may facilitate the implementation "preemptive" pathways to reduce the perioperative burden on the patient. Broadly speaking, complications are been categorized into visceral (eg, respiratory, gastrointestinal, cardiac, neurological, genitourinary) and those relating to surgery (eg, blood loss, wound care, and pain).

Respiratory Complications

Postoperative respiratory complications are the most common following scoliosis corrective surgery in neuromuscular disorders.[7] It ranges between 19% and 50% in most reported series.[3] Respiratory muscle strength may be attenuated postoperatively due to pain and sedation, which may lead to retention of airway secretions and hypoventilation.[8] Aspiration, pneumonia, and atelectasis are the commonest complications postoperatively. Less frequently, pneumothorax, hemothorax, and chylothorax may occur.[3,9] Severe curves with associated muscular weakness predispose to restrictive lung disease.[10]

Appropriate planning may allow surgery to be carried out even in those with a background of respiratory problems. The British Thoracic Society have published consensus guidelines on the management of patients with NMD undergoing surgery.[8]

The first step is preoperative assessment to identify patients at risk of postoperative respiratory complications. These include daytime and or night-time hypoventilation, and patients with ineffective cough[8] and vital capacity <60% predicted.[11,12] The latter have increased risk of prolonged postoperative ventilation. Preoperative training of airway clearance techniques and the use of noninvasive ventilation may be useful. The latter is most effective when utilized immediately after extubation and may facilitate early extubation. Nutritional status is closely linked to respiratory function and should be reviewed preoperatively as poor nutrition may lead to diminished respiratory muscle function and early exhaustion. Involvement of the respiratory physicians and physiotherapists from the outset improves the preoperative pulmonary status and postoperative risk of complications.[8]

In the postoperative period, supplemental oxygen should be used with caution to avoid missing hypercapnia and the subsequent treatment of the underlying cause. Gastric distension and constipation may cause diaphragmatic splinting. Use of prokinetics and gastric decompression with nasogastric tubes should strongly be considered.

With particular reference to Duchenne muscular dystrophy (DMD), the American College of Chest Physicians (ACCP) recommends preoperative assessment of oxygen saturation as a measure of gas exchange and forced vital capacity (FVC) as a measure of lung volume.[13] The latter is a particular predictor of respiratory complications. Preoperative FVC <50% predicted is a marker of increased risk and FVC <30% predicted is considered high risk. These thresholds may be used to identify patients who may benefit from preoperative training in noninvasive positive pressure ventilation (NIPPV) to optimize the success of extubation.[14] Nocturnal ventilation has proved successful in reducing the postoperative complications in cases with DMD.[15]

The ACCP also advocates preoperative assessment of cough strength and effectiveness by measured maximum expiratory pressure (MEP) and peak cough flow (PCF).[16] For those patients with PCF less than 270/min or MEP <60 cm H_2O mechanically preoperative training in and postoperative use of assisted cough insufflation/exsufflation is advocated. Those patients with FVC <50% and FVC <30% should be strongly considered for postoperative NIPPV. Those who use NIPPV chronically should be extubated to NIPPV postoperatively. Khirani et al[14] have evaluated the use of both NIPPV and mechanical insufflation-exsufflation and observed no respiratory complications in their series of children with nonidiopathic scoliosis undergoing posterior spinal fusion.

Gastrointestinal Complications

A recent meta-analysis suggests that the third commonest complication involves the gastrointestinal system[9] and may occur in up to 13% of cases in some series.[17] Paralytic ileus, gastroparesis, and dysphagia are among the most common postoperative adverse events.[17] Pancreatitis and superior mesenteric artery syndromes have also been described.[18] Postoperative dysmotility is associated with correction of large rigid curves. It is hypothesized that neuropathy of the visceral nerves may be responsible for intestinal dysmotility. Prolonged administration of opioid medications postoperatively is also associated with reduced intestinal function.[9] Paralytic ileus has been noted to increase in incidence followings prolonged period of being in the prone position during surgery.[19]

Depending on the patient's condition, a preoperative review by a pediatric gastroenterologist and/or nutritionist may be appropriate to assess nutritional status and advise on any required regimens leading up to surgery. Postoperative review may be helpful in cases with resistant gastroparetic ileus. Prokinetic medications should ideally be prescribed with the guidance of a gastroenterologist or nutritionist.[17] In certain cases, parenteral feeding or a gastrojejunal feeding tube may be required. Preoperative evaluation of a hiatus hernia and gastroesophageal reflux disease (GERD) is useful to reduce the risk of postoperative aspiration pneumonia warranting prolonged intubation or/and artificial ventilation.[4]

Appropriate hydration, nasogastric aspiration/feeds and careful attention to serum electrolytes (especially serum K^+ levels) and supplementary nutrition is important to optimize outcomes in perioperative period.[17] In addition, low body mass index (BMI) in many of these patients makes them vulnerable to developing superior mesenteric artery syndrome with subsequent duodenal obstruction. Persistent and intractable vomiting around 5 to 7 days after the surgery with high index of suspicion should warrant the treating clinician in requesting a barium swallow study to aid in early detection and prompt management.[4] Patients with rigid curves on traction or bending films should alert the surgical team to consider the above measures perioperatively.[20]

Nutritional Needs and Complications

Patients with NMD are often malnourished due to low caloric intake, GERD, and increased metabolic requirements. Retrospective case series in cerebral palsy (CP) and spastic quadriplegia have established an association between preoperative serum albumin and lymphocyte count and immediate postoperative adverse events.[21] A preoperative serum albumin of <3.5 g/dL and a total body lymphocyte count less than 1.5/mm^3 were both associated with a higher rates of infection, prolonged intubation and longer inpatient stay.[21,22]

Vitamin D has been implicated in several studies to be a key player in postoperative outcomes in patients with NMS. Low vitamin D levels have been observed in patients with scoliosis.[23] Vitamin D has been recognized to have, in addition to bone metabolism, a central role in muscle function, inflammatory-mediated reactions, immunity, and fibrous tissue formation.[24-26] Afro-Caribbean patients are at a higher risk of low vitamin D levels compared with Caucasians. Patients are particularly prone to develop low vitamin D levels during the winter period compared with other seasons.[23] Preoperative vitamin D assessment and prescription of supplements should be a standard practice to improve outcomes.

Preoperative evaluation of the patient's overall nutritional status and gut optimization with supplementary nutrition has also shown to reduce the incidence of wound dehiscence and infection.[27] Observing the caloric intake and optimizing feeding both orally and via a gastro- or jejunostomy tubes as a part of workup prior to surgery is hence justified. Oral feeds should be resumed as soon as possible to avoid malnutrition with its consequences.[28]

Obesity as a form of malnutrition in patients with NMD should not be overlooked. It is common in the presence of CP, DMD, and spina bifida. Obesity in the presence of NMD is associated with increased rates of complications, including wound infection after spinal instrumentation, urinary tract infections and readmission following surgery.[29] Ramo et al[30] observed a greater risk of infection in patients with BMI >25 kg/m^2 undergoing posterior spinal fusion of NMS. Basques et al[31] reported a greater risk of infection with a BMI equal to or greater than the 95th percentile. Strategies such as early dietary and physical activity modification should be considered early to avoid postoperative complications.

Cardiac Complications

Clinically significant cardiac abnormalities in NMDs result in either cardiomyopathy or conduction abnormalities resulting in arrhythmias.[32] The severity and onset of these vary according to the type of NMD. Muscular dystrophies are an inherited group of disorders associated with progressive wasting of skeletal muscle. Among these group of disorders, DMD is one of the most common forms. The absence of Dystrophin in cardiac myocytes leads to loss of membrane integrity and excessive influx of calcium intracellularly. As a result, intracellular proteases are activated and degrade the contractile proteins.

Extensive fibrosis and loss of function leads to cardiomyopathy, which is the leading cause of death in cases with DMD.[33] Other patients may present with arrhythmias. Cardiac involvement is of particular concern in all muscular dystrophies and is thought to be present in up to 20% of cases.[34] The absence of skeletal symptoms does not preclude cardiac abnormalities. Other NMDs such as Friedreich's ataxia can be associated with left ventricular hypertrophy.[35] Chest cage deformity, for example, Parasol' deformity of spinal muscle atrophy can cause restrictive lung disease, hypoventilation with subsequent pulmonary hypertension (HTN) and Cor pulmonale.[36]

Progressive weakness may restrict physical activity and therefore mask the presence of cardiac symptoms. Preoperative screening for cardiac abnormalities is therefore imperative prior to surgery, especially in those with known risk factors.

Preoperative evaluation should be performed in close conjunction with a pediatric cardiologist familiar with NMDs. The American Heart Association recommends evaluation should occur in close proximity to the planned surgery, ideally within 3 to 6 months.[32] Baseline electrocardiography (ECG) and cardiac imaging in the form of transthoracic echocardiography are recommended to assess cardiac function.[37] Cardiac magnetic resonance imaging is gaining favour and may be indicated in patients where transthoracic echocardiography does not allow adequate evaluation of cardiac function, because of poor acoustic windows. It offers a number of advantages, including visualization of myocardial fibrosis. In some cases, dobutamine stress echocardiogram may provide additional information for cardiac evaluation. Optimization of cardiac function may include beta blockade, antiarrhythmic treatments, implantable defibrillation, and/or pacemakers.

Intraoperative blood loss may cause significant stress to already impaired cardiac function. Numerous strategies may be employed to reduce the blood loss and its effect cardiac functions. These are discussed in more detail in the following section but include maintenance of normothermia, administration of antifibrinolytics, normovolemic hemodilution, blood salvage, and controlled hypotension. Postoperative hemodynamic monitoring may be required until cardiorespiratory function returns to baseline. In some instances, ionotropic agents may be required to support cardiac function.

Blood Loss

Neuromuscular patients have an almost 7 times higher risk of extensive blood loss (defined as greater than 50% of estimated total blood volume [EBV]) during surgery in comparison with patients with idiopathic scoliosis.[38] This is primarily related to the extensive approach, and long operative time. Other contributing factors are reduced coagulation factor reserve and increased fibrinolytic activity.[39] Patients with DMD are at a particular risk of excessive blood loss during corrective surgery due to lack of dystropin in all muscle layers, including arterial smooth muscles coupled with subsequent poor vasoconstrictive response. Noordeen et al[40] showed a significantly higher blood loss in patients with DMD than in patients with spinal muscular atrophy undergoing the same surgical procedure. The amount of blood loss was directly proportional to operative time and was independent of patient's age, curve magnitude and the number of levels fused.[40]

Proper intraoperative positioning to relieve intra-abdominal pressure (in posterior fixation surgeries), judicious use of hypotensive anesthesia, strict subperiosteal dissection as well as proper hemostatic techniques are the key steps in minimizing intraoperative blood loss. Recycling and transfusing lost blood back into the patient using cell salvage can reduce the need to transfuse blood. For several years, preoperative autologous blood transfusion has been used, by some centers, and is an excellent alternative to cross-matched blood as it eliminates the majority of transfusion-related adverse events.[41]

Theroux et al[42] investigated the effect of use of desmopressin acetate in reducing the blood loss during NMS surgery and observed no difference in blood loss from the placebo group despite the increase in the von Willebrand factor and factor VIII levels. ε-Aminocaproic acid was tested by Florentino-Pineda et al[43] during surgery for idiopathic scoliosis which reduced blood loss significantly and may also be used in NMS.

Genitourinary Complications

Urinary tract infections (UTIs) are the commonest postoperative genitourinary complication. The incidence is as high as 20% to 22% in most of the reported series. Most case are due to prolonged catheterization. Patients with myelomeningocele are particularly at higher risk of developing UTIs due to the presence of neurogenic bladder. Stagnant urine and bacterial multiplication in these patients also predispose them to an increased risk of pyelonephritis. Other minor genitourinary complications include hematuria and chronic retention.[44] Prophylactic antibiotics and regular bladder wash/catheter care is the cornerstone in preventing life-threatening genitourinary infections.

Wound Complications

The incidence of postoperative wound infection is highest in NMS among all spectrum of disorders undergoing spinal deformity surgery. The incidence of infection is reported as 4% to 14% versus 1% to 5% in idiopathic scoliosis.[3,44-49] The incidence of superficial infection has been reported to be 7% to 14%, and deep infection 5% to 8%.[49] Wound complications are the single most important factor associated with prolonged hospital stay and utilization of scarce hospital resources.

Sponseller et al[50] observed deep wound infection to be closely related to the degree of cognitive impairment and use of allograft. Other factors like serum albumin levels, total white blood cell count, prior spinal surgery, estimated intraoperative blood loss, staged anterior and posterior surgery were found to be statistically insignificant.[50] In a retrospective case control study of 151 cases of NMS, Master et al[9] found the presence of a ventriculoperitoneal shunt before surgery to be the only factor

statistically associated with an increased risk for wound infection. These 2 studies also reported a high incidence of pseudoarthrosis and prolonged hospital stay with coexistent postoperative wound infection.

Cahill et al[51] reported that patients with myelomeningocele are at highest risk of postoperative wound infection (infection rate of 19.2%). This is almost a 40-fold increased risk in comparison to patients with idiopathic scoliosis (infection rate of 0.5%). Many authors observed infection rates of >20% in their series of corrective deformity surgery in spinal dysraphism.[52,53] High incidence of deep wound infection in myelomeningocele was attributed to a variety of reasons. Key factors reported were defective midline/paramedian musculature, which made the closure difficult, proximity of dura to the skin with high incidence of dural leak/cerebrospinal fluid cutaneous fistula and previous multiple surgeries with poor soft tissue quality.[9,51]

Neurological Injury

The incidence of neurological injury in surgery for neuromuscular scoliosis is higher when compared with idiopathic scoliosis.[54] Spinal cord monitoring (SCM) during spinal procedures is utilized to allow early detection of neurological insult that results from instrumentation and correction of spinal deformities. It is now considered a standard of care during pediatric surgery for spinal deformity.[55] The recovery of postoperative neurologic deficits shows a directly proportional relationship to the speed of removal of instrumentation.[56] Theoretically, earlier detection of spinal cord dysfunction allows immediate intervention and the possible prevention of neurologic deficit. In most spinal injury units, spinal cord monitoring has replaced the Stagnara wake-up test. The latter, however, is still of clinical importance should there be a significant change in intraoperative monitoring. However, the wake-up test is not always practical in patients with neuromuscular scoliosis, owing to muscle weakness and attenuated cognitive function.[57] There is conflicting evidence in the literature regarding the reliability of SCM for patients with NMS. Some argue that baseline measurements are absent in a large proportion of patients with NMS, and when present are not of sufficient or reliable amplitude to be allow reliable monitoring.[58] Others propose that careful control of anesthesia allows for reliable cortical and subcortical responses in a sufficient proportion of the patient population thereby justifying the regular use of SCM in scoliosis surgery for these patients.[59]

DiCindio et al[60] were able to reliably monitor both transcranial motor evoked potentials (TcMEP) and somatosensory spinal evoked potentials (SSEP) from the posterior tibial nerve[60]. This was caveated with the fact that patients with severe CP presented the most challenging patients for successful SCM. The authors were able to reliably monitor SSEPs in 82% of patients with CP and 86% of patients with other neuromuscular disorders. On the other hand, TcMEPs were monitorable in 63% of patients with mild or moderate CP and only 39% of those with severe CP. Of those with other neuromuscular disorders, 86% had recordable MEPs at baseline. In their recent series of patients, Hammett et al[61] were unable to yield reliable baseline SCM readings in 13% of patients and intraoperative monitoring was therefore abandoned. They were only able to obtain useful readings in 75% of patients with severe CP. They observed 2 patients who had clinical neurological deficits with no change in intraoperative SCM. Other studies have attempted to improve the reliability of SCM with the use of an epidural electrode.[55,62] SCM can reliably be obtained in approximately 98% of patients with NMS,

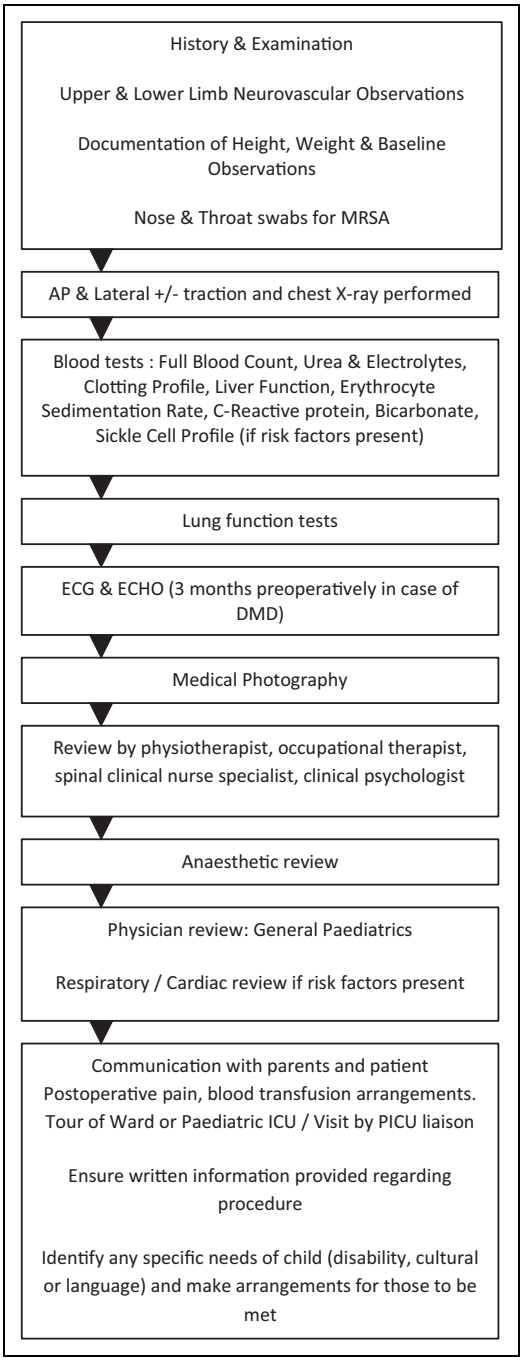

Figure 1. Preoperative pathway for management of patients undergoing surgery for neuromuscular scoliosis.

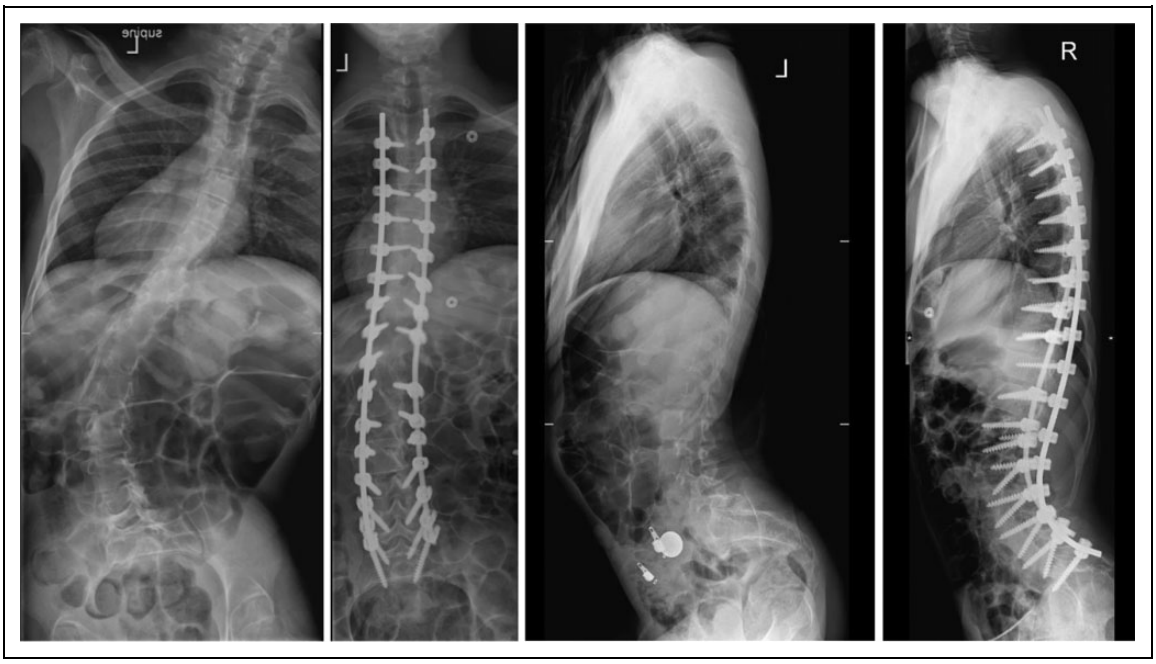

Figure 2. Preoperative and postoperative anteroposterior and lateral radiographs.

using SSEPs with epidural placed electrodes rather than somatosensory cortical evoked potentials. A decline in amplitude of 50% of the initial baseline reading was considered as significant.[55]

Careful consideration must be made as to the clinical utility of SCM in this patient group and balanced against the cost to healthcare providers. Diefenbach et al[63] estimated the cost per case for intraoperative monitoring to amount to \$790 $\pm$ \$681 or 1.4% of the overall spend. Hammett et al[61] commented in their study that on no occasion did changes in intraoperative SCM lead to an alteration in the surgical plan. While some patients had anesthetic interventions or adjustment of monitoring leads, no patients had instrumentation removed or altered. For SCM to be a worthwhile cost each case should be considered on an individual basis. The loss of residual motor function, protective sensation, or continence may have a significant impact on patients and their caregivers. Any savings achieved through lack of SCM may be incurred at a greater degree through inpatient or social care costs if complications do occur. In any case, the literature suggests that the use of SCM is associated with lower rates of complications and length of inpatient stays in this patient group.[2]

Proposed Pathway

The cost of a multidisciplinary pathway is inherently expensive and incurred through clinician time, increased manpower requirements and investigations. We can justify this increased cost through the evidence, which suggests an overall reduction in complications and improved patient outcomes. In order for the multidisciplinary team to be both clinically effective and financially viable, such procedures must be considered in volume and as such surgery of this kind should be performed in select specialized units. This ties in with recommendations from the UK Spinal Services: Getting It Right First Time Programme National Specialty Report.[64]

The ultimate goal of preoperative optimization is to provide a safe and effective management pathway for these vulnerable patients. Below is our institutional pathway for the optimization of patients undergoing surgery for neuromuscular scoliosis (Figure 1). This pathway has been developed through a review of the evidence and literature described in this article and multidisciplinary consensus. We believe this to be an effective strategy in reducing perioperative morbidity and mortality and would recommend this approach in other specialized institutions.

Clinical Vignette

A 15-year-old patient with CP (Gross Motor Function Classification System grade V) was diagnosed with NMS at the age of 11 years. Curve progression was observed during a course of conservative treatment. A progressive increase in his curve was noted over a 4-year period. He experienced increasing pain in his back and from costopelvic impingement. A loss of ability to sit in his wheelchair was also observed and he was admitted to his local hospital twice for lower respiratory tract infections requiring intravenous antibiotics. Following counseling and extensive discussion with the parents regarding the risks and benefits of corrective deformity surgery a decision was made to undergo surgical management of his scoliosis. Four weeks preoperatively, he was admitted for assessment. He was reviewed by the physiotherapists, occupational therapists, speech and language team, pediatricians, dietitians, pediatric intensivists,

and the surgical team. A full set of blood tests were done, including blood count, liver and kidney function tests, coagulation profile, and serum albumin levels. The pediatric intensive care team performed an overnight sleep study. To conclude the inpatient assessment, a report was formulated outlining detailed recommendations for presurgical optimization. Subsequently, the patient was discussed in the multidisciplinary team meeting. The night before his planned surgery, he was admitted overnight, whereupon he was noted to be febrile and coughing. On clinical review by the pediatric team, he was diagnosed and treated for a lower respiratory tract infection and his surgery was rescheduled.

Three weeks later, he was readmitted and underwent instrumented fusion between T4 and S1 (Figure 2). Lumbo-pelvic fixation is associated with extensive soft tissue dissection, increased operative time, blood loss, and wound infection. In this case, good control of pelvic obliquity was achieved without extension to the pelvis and the sacrum was chosen as the lower instrumented level. The procedure was uneventful, and he was successfully extubated without requirement for supplemental respiratory support. He spent a total of 3 days in the pediatric intensive care unit where he was reviewed by various teams and his progress assessed. Percutaneous endoscopic gastrostomy tube feeding resumed immediately after surgery as planned in the preoperative multidisciplinary team assessment. The surgical wound was reviewed daily and healed without complication. Thirteen days following surgery, having made an excellent recovery, he was discharged back to his usual place of residence.

Conclusions

Spinal deformities commonly develop in patients with neuromuscular disorders causing significant pain and functional impairment. Surgical correction carries a high risk of postoperative complications. A multidisciplinary team approach may reduce the risk of complications and subsequently improves patient outcomes.

Declaration of Conflicting Interests

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