SURGICAL MANAGEMENT OF NEUROMUSCULAR SCOLIOSIS: APPROACHES, PITFALLS AND OUTCOMES

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Background. Scoliosis is a common problem among neuromuscular disorders with increased incidence of morbidity and mortality. The management of neuromuscular scoliosis (NMS) is complex and requires a cooperative multidisciplinary team to provide meticulous perioperative care. Conservative treatment, such as bracing, can delay but not replace surgical correction. However, surgery has the risk of higher perioperative complication rates in this patient population.

Aim. It is a retrospective study to evaluate clinical outcome, approaches, pitfalls in management of NMS patients undergoing surgical correction.

Materials and methods. We retrospectively evaluated patients with NMS who underwent surgery for scoliosis from 2004 to 2018. Sixty-eight patients were included in the study after meticulous preoperative checkup. In most cases a single-pedicle screw construct was used, but in a few others a hybrid construct was used. A follow up was performed to make a clinical and radiological assessment and recorded all measurements and any complications. A telephone questionnaire was used for functional evaluation and patient/parent satisfaction with surgery. Only 52 patients could participate in the questionnaire.

Results. The mean follow up period was 43.28 months, and the mean age at the time of surgery was 14.29 years. The mean preoperative Cobb’s angle was 71.7°, while that of final follow-up was 34.6°. The mean Cobb’s correction percentage was 53.25%. For correction of fixed pelvic obliquity ≥15°, a sacropelvic extension was done in 60.29% of cases. Complications occurred in 39.71% of operated cases; chest related in 36.11% (of all complications), hardware-related 16.67%, visceral complications (as paralytic ileus) in 13.89%, decubitus ulcer and delayed wound healing in 13.89%, deep wound infection in 8.33%, CNS complications (as status epilepticus) in 8.33%, and death in 2.78% (one case). The results of the questionnaire indicated favorable functional outcomes and patient/family satisfaction with surgery. Only 52 patients could participate in the questionnaire.

Conclusion. Despite the perioperative difficulties seen in patients with NMS, patients who had relatively higher postoperative morbidity and mortality, most patients/parents were satisfied with the results of the spinal deformity surgery. The patients/parents would recommend surgery to other patients with similar disorders.

Keywords: neuromuscular scoliosis; outcome; pitfalls.

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ХИРУРГИЧЕСКОЕ ЛЕЧЕНИЕ НЕРВНО-МЫШЕЧНОГО СКОЛИОЗА: ДОСТУПЫ, ТРУДНОСТИ И РЕЗУЛЬТАТЫ

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Обоснование. Сколиоз часто сопутствует нервно-мышечным заболеваниям и характеризуется повышенной заболеваемостью и смертностью. Лечение нервно-мышечного сколиоза комплексное, оно предполагает скоординаированную работу мультидисциплинарной команды узких специалистов и тщательный периоперационный уход. Такие консервативные методы, как использование корсета, могут отсрочить, но не заменить хирургиче-
скую коррекцию. При этом операция у этой группы пациентов связана с повышенным риском периоперационных осложнений.

Цель — изучить клинические исходы, доступы и сложности в хирургическом лечении пациентов с нервно-мышечным сколиозом.

Материалы и методы. Ретроспективно изучены данные пациентов с нервно-мышечным сколиозом, которым в период с 2004 по 2018 г. была выполнена хирургическая коррекция сколиоза. После комплексного предоперационного обследования в исследование было включено 69 пациентов. В большинстве случаев использовали только транспедикулярную фиксацию, но в некоторых случаях применяли гибридную конструкцию. Наблюдение включало клинические исследования и радиологическую оценку с записью всех измерений и любых осложнений. Для оценки функционального состояния, удовлетворенности пациента/родителей пациента операцией проводили опрос по телефону, в этом опросе участвовали только 52 пациента.

Результаты. Средняя продолжительность наблюдения составила 43,28 мес., средний возраст на момент операции — 14,29 года. Среднее значение угла Кобба было 71,7°, при этом на момент окончательного осмотра он составлял 34,6°. Среднее значение коррекции угла Кобба — 53,25 %. Для коррекции фиксированного перекоса таза ≥15° в 60,29 % случаев использована крестцово-тазовая фиксация. Осложнения развивались у 39,71 % оперированных пациентов. Осложнения, связанные с органами грудной клетки, составили 36,11 % всех осложнений, связанные с металлоконструкцией — 16,67 %, висцеральные осложнения (в виде паралической кишечной непроходимости) — 13,89 %, пролежни и заживление раны вторичным натяжением — 13,89 %, раневая инфекция — 8,33 %, осложнения со стороны центральной нервной системы (в виде эпилептического статуса) — 8,33 % и смерть — 2,78 % (у одного пациента). По результатам опроса наблюдали благоприятный функциональный исход и удовлетворение пациента/семьи проведенным лечением.

Заключение. Несмотря на периоперационные сложности, встречаемые у пациентов с нервно-мышечным сколиозом с относительно высоким риском послеоперационных заболеваемости и смертности, большинство пациентов/родителей были удовлетворены результатом хирургического лечения искривления позвоночника. Кроме того пациенты/родители рекомендовали бы операцию другим пациентам с подобным заболеванием.

Ключевые слова: нервно-мышечный сколиоз; исход; сложности.

Neuromuscular disorders are a variety of lesions affecting neurological (Upper motor neuron or lower motor neuron), or muscular disorders (myopathies) [1–3]. The incidence of development of scoliosis in such disorders is high with reported values of 25–100% according to the causative pathology [4].

NMS is different from adolescent idiopathic scoliosis in that it usually progresses even after skeletal maturity, usually has large thoracolumbar curve collapsing into kyphosis, and associated with large pelvic obliquity with involvement of the sacrum as a part of the spinal curvature [2, 5].

Spinal deformity in the patients with neuromuscular disease exists concurrently with disorders of multiple systems making management difficult requiring a cooperative team. Orthopedic comorbidities like contractures and deformity of the hips, knees, and feet are commonly present. Urologic dysfunction, insensate skin, and pressure sores are complex consequences of neuromuscular disorders that require evaluation and care by surgical subspecialists. Cognitive delay, seizures, hydrocephalus, and subtle changes in mental function may complicate the evaluation of many neuromuscular conditions. Feeding difficulties, nutritional insufficiency and cardio-pulmonary insufficiency are especially important considerations in the preoperative evaluation of patients with spinal deformity because of their association with postoperative complications in these patients. Therefore, a multidisciplinary team is important for the complete evaluation of a patient with neuromuscular spinal deformity [4].

Conservative treatment, as bracing, can delay but not eliminate the need for corrective surgery. Bracing, wheelchair modifications, physical therapies, use of assistive devices, and systemic corticosteroids in Duchenne are the commonly used conservative therapies, but with no proven effect on NMS progression [6].

Surgical stabilization of NMS constitutes the mainstay of treatment in most cases with evident curve progression, and failure of conservative methods. The operative treatment of NMS is more complex than that of AIS as: patients with NM disorders are commonly health-debilitated, poor nutritional status, poor bone quality, and compromised cardiorespiratory functions. Moreover, prolonged surgery duration that may involve instrumenting to the
pelvis, with larger blood loss are adding complexity in NMS surgeries [6–8]. The main goal of surgery is to produce a stable balanced spine in sagittal and coronal planes over a level pelvis.

Fixation methods of thoracolumbar spine have developed over years, but recently there have been a rising preferable of using pedicle screw construct, whenever possible. Hybrid constructs entitles using thoracic hooks, or sublaminar wires / bands, together with pedicle screws in thoracic and lumbar vertebrae with adequate pedicle diameter, are also done frequently [6, 9, 10].

Some studies investigating the patient / family satisfaction from surgery and surgery outcome found difficulty in determining the functional benefits in patients, especially in those with the most severe involvement [11–13].

This study aimed to retrospectively evaluate a cohort group of patients undergoing surgical correction of Neuromuscular Scoliosis, and record approaches of management, preoperative & postoperative correction degree, pitfalls during management, outcome of surgery, and patient-parent satisfaction from surgery using a telephone questionnaire.

Materials and methods

A retrospective analysis of Neuromuscular scoliosis patients underwent surgical correction of scoliosis deformity at Pediatric orthopedic department of the medical university of Graz-Austria was done from data base of the hospital registry from 2004 till 2018.

We reviewed the medical records, and radiographs of 95 patients with NMS who had surgery with segmental spinal instrumentation. After excluding patients with incomplete records (n = 15), patients who had follow up less than one year (n = 12), the remaining 68 patients formed the study population. Out of the 68 patients, only 52 patients (76.47%) were available to answer the questionnaire. All surgeries were operated by a single senior author (VS). The radiographs were measured, the questionnaires were evaluated, and biomedical statistics were done by independent observers (EN, MS). All participants provided informed consent for our study.

Inclusion criteria:

- NMS patients with curves ≥50°.
- DMD patients with curves ≥15°.
- NMS patients with progressive curves ≥30° with cardio-pulmonary compromise due to spinal deformity with good fitness for surgery.
- All patients should be aware of surroundings, and communicative with others.

Exclusion criteria:

- All patients unfit for surgery.
- Families, or patients refusing surgical correction.

In DMD, scoliosis is rapidly progressive after patient loses ability to ambulate, which is associated with rapid decline in cardiopulmonary functions, that is why surgical correction of scoliosis deformity is advised earlier when cardiopulmonary functions are optimal [14].

Preoperative evaluation necessitated patient admission in hospital for 4-5 days to complete the checkup. Cardiological consultation (including Echocardiography), Pulmonological consultation including pulmonary function tests with chest x-ray, arterial blood gas analysis, routine preoperative laboratory investigations including complete blood count, liver and renal functions, prothrombin time & activity, screening for specific coagulation factors deficiency if suspected, urine analysis with bacteriological culture & antibiotic sensitivity, tests for gastroesophageal reflux & swallowing tests if suspected disorders, nutritional status assessment, and polysomnography were done for children with suspected respiratory distress at night time especially DMD and SMA.

Recent plain x-ray of the whole spine & pelvis in PA standing, or sitting AP in non-ambulant patients, and lateral views of spine were done to measure curve magnitude, thoracic kyphosis, lumbar lordosis, and degree of pelvic obliquity. Moreover, supine bending or traction radiographs obtained determining flexibility of the curve to determine the levels of instrumentation & fusion, and hand x-ray to estimate skeletal age & the remaining growth.

The patient, parents and/or legal guardians, physiotherapists, and caretakers were informed about indications of operation, goals of surgery, preoperative & postoperative procedures, and risks of complications at each stage of treatment.

Neuromonitoring (SSEP & MEP) were used when the patient was ambulant, active lower limb movement or sensation were preserved, or when the patient was continent for urine and stool.

Posterior approach for exposure of the thoracicolumbar spine was used in most cases when pos-
terior-only fusion is enough. Combined anterior-posterior approach was indicated in severe rigid deformities with Cobb’s angle above 90°, 2-staged procedure was done starting with anterior approach, either open or thoracoscopic, with anterior release and disectomies without instrumentation done with the help of thoracic surgeon, followed by Halo gravity traction. Ten to fourteen days later after recovery from 1st surgery, posterior spinal fusion & instrumentation was done, and this was needed in 3 cases (4.41%).

Only pedicle screw-construct was tried first for all patients. When there were too small pedicles for screw insertion, hybrid construct was done using hooks (transverse process, or laminar) for proximal thoracic vertebrae & pedicle screws for other vertebrae, and this was done in 5 cases (7.35%).

Extending the fixation to the pelvis was needed if fixed pelvic obliquity was ≥15° with various methods; sacroiliac screws, or only extending of fusion to 1st sacral segment that was usually done in most cases (60.29 % of 68 operated cases).

Postoperative care started immediately following surgery with patient admission to ICU. Weaning from mechanical ventilation was tried in the 2nd–4th postoperative day, when all parameters were stable. Parenteral broad spectrum antibiotics (3rd generation cephalosporin + ampicillin-sulbactam with dosing of 50 mg/kg/day) were used for 5–7 days postoperatively. Seating of the child and assisted walking was done with the help of physiotherapists. Training for daily activities of feeding, self-care, nursing and care whether independent or with parents and care givers were regularly made with physiotherapy.

Follow up and evaluation: the patients came to outpatient clinics every 3 months in the first year, then every 6 months in the second year, and every year for the next 2 years. Evaluation was made clinically and radiologically for measuring the Cobb’s angle, pelvic obliquity, thoracic kyphosis, lumbar lordosis, and thoracolumbar kyphosis if present. Checking of fusion mass, hardware breakage or loosening were also noted. Figures 1 & 2 show a case example both preoperative and postoperative radiologically and clinically.

A modified questionnaire [11, 12] has been developed, based on the difficulties faced by patients with functional daily activities, and satisfaction from surgery. The responses to all the fields were on a graduated scale from 1 (=good) to 5 (=bad) in functional activities (just like the educational scoring system in Austria for better understanding and estimating patient answers) and a scale from 1 (=yes), 2 (=maybe), or 3 (=no) in patient satisfaction part. Questionnaire was a telephone call held by pediatric orthopedic department secretaries, and questions were explained to the patients or their families. The modifications of previous questionnaires were; 1st: reversal of grading score (1=bad and 5=good),

![Fig. 1](image1.png)

**Fig. 1.** Preoperative (a–b), immediate postoperative (c–d), and final postoperative (e–f) PA and lateral x-rays of an example case
and 2nd in that it was held by a telephone call to let more patients to participate.

The mean follow-up period was 43.28 (12–136) months. Cerebral Palsy (CP) constituted the main causative pathology of NMS (53%), followed by Duchenne Muscle dystrophy (15%), spinal muscle atrophy (7%) and myelomeningocele (6%) as shown in Figure 3.

Results

Only 11 patients (16%) were able to ambulate, while the others were wheelchair bound. Table 1 summarizes the demographic data, preoperative and postoperative parameters of the whole patient population.

Regarding postoperative ICU stay, we found that more than 5 days ICU stay was encountered in 19/67 operated patients (28.4%), 6 patients (31.6%) had early postoperative complications making ICU care necessary, while 12 patients (63.2%) had no complications.

Surgery duration was calculated from the start of surgical incision till skin closure, and there was positive correlation between surgery duration and number of fused levels, and preoperative curve Cobb’s angle as correlation values were 0.29 and 0.30 respectively.

Many of the operated cases (58.82%) had one or more associated medical co-morbidities. Gastroesophageal reflux disease (GERD) was the most common comorbidity (36.76%), followed by epilepsy (30.4%). Other comorbidities like cardiopulmonary diseases, hydrocephalus, endocrinologic disorders, and visual disorders were less frequent.

Most (67.65%) NMS patients had one or more orthopedic co-morbidities. Feet deformities (paralytic club feet, fixed equinus, plano-valgus feet and cavo-varus deformities) constituted 47.7% of orthopedic comorbidities. Knee and hip flexion deformities were less often. Many of operated NM scoliosis cases (39.71%) had a hip instability either unilateral or bilateral. Hip surgeries for symptomatic instability of the hip were made, before and after scoliosis surgery, like soft tissue release or lengthening surgeries, pelvic osteotomies, femoral varus derotational osteotomies, or femoral head resection as salvage procedures.
Complications occurred in 39.71% of operated cases (27/68 cases) as follows (Figure 4).

1. **Chest-related**: (pneumonia, bronchopneumonia, atelectasis, pleural effusion, and haemothorax) in 13 cases (36.11% of all complications).

All chest related complications occurred in the early postoperative period, 54% of which have been managed conservatively with observation, antibiotics (bacterial pneumonia or bronchopneumonia) and antiedematous drugs (for pleural effusion), and 46% needed more invasive maneuvers either with: drainage chest tube in 4 cases (one haemothorax and 3 tense exudative pleural effusion not responding to conservative management), or bronchoscopy and lavage in 2 cases (one case of aspiration pneumonia and another of lung atelectasis). Pleural effusion developed in 6 cases: 3 of them

### Table 1

<table>
<thead>
<tr>
<th>Item</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean age at time of surgery</td>
<td>14.29 (8–20) years</td>
</tr>
<tr>
<td>Sex: Female %</td>
<td>52.94%</td>
</tr>
<tr>
<td>Percentage of patients able to ambulate</td>
<td>16.18%</td>
</tr>
<tr>
<td>The mean postoperative ICU stay (days)</td>
<td>5.39 (1–22)</td>
</tr>
<tr>
<td>The mean surgery duration (hours)</td>
<td>7.62 (4–10.33)</td>
</tr>
<tr>
<td>Percent of associated Medical comorbidities</td>
<td>58.82%</td>
</tr>
<tr>
<td>Percent of associated orthopedic comorbidities</td>
<td>67.65%</td>
</tr>
<tr>
<td>Percent of associated hip instability</td>
<td>39.71%</td>
</tr>
<tr>
<td>The mean preoperative Cobb's angle</td>
<td>71.7 (16–137°)</td>
</tr>
<tr>
<td>The mean immediate postoperative Cobb's angle</td>
<td>29 (4–72°)</td>
</tr>
<tr>
<td>The mean final postoperative Cobb's angle</td>
<td>34.6 (6–95°)</td>
</tr>
<tr>
<td>The mean absolute value of Cobb's coronal correction</td>
<td>37.5 (–15 to 112°)</td>
</tr>
<tr>
<td>The mean Cobb's correction percentage</td>
<td>53.25% (–18.75 to 100%)</td>
</tr>
<tr>
<td>The mean final loss of coronal correction</td>
<td>5 (–4 to 34°)</td>
</tr>
<tr>
<td>The mean Preoperative pelvic obliquity</td>
<td>12.37 (0–38°)</td>
</tr>
<tr>
<td>The mean immediate postoperative pelvic obliquity</td>
<td>6.67 (0–34°)</td>
</tr>
<tr>
<td>The mean final postoperative pelvic obliquity</td>
<td>8.25 (0–28°)</td>
</tr>
<tr>
<td>The mean absolute value of pelvic obliquity correction</td>
<td>4.24 (–23 to 31°)</td>
</tr>
<tr>
<td>The mean Pelvic obliquity correction percentage</td>
<td>21.7% (–460 to 100%)</td>
</tr>
<tr>
<td>The mean final loss of pelvic obliquity correction</td>
<td>1.58 (–6 to 16°)</td>
</tr>
<tr>
<td>The mean Preoperative thoracic kyphosis</td>
<td>36.15 (0–106°)</td>
</tr>
<tr>
<td>The mean immediate postoperative thoracic kyphosis</td>
<td>31.88 (15–55°)</td>
</tr>
<tr>
<td>The mean final postoperative thoracic kyphosis</td>
<td>23.48 (16–56°)</td>
</tr>
<tr>
<td>The mean Preoperative lumbar lordosis</td>
<td>40.29 (–30 to 104°)</td>
</tr>
<tr>
<td>The mean immediate postoperative lumbar lordosis</td>
<td>46 (15–80°)</td>
</tr>
<tr>
<td>The mean final postoperative lumbar lordosis</td>
<td>46.9 (15–84°)</td>
</tr>
<tr>
<td>Percentage of associated preoperative thoracolumbar kyphosis</td>
<td>33.82%</td>
</tr>
<tr>
<td>The mean preoperative thoracolumbar kyphosis</td>
<td>48.22 (25–114°)</td>
</tr>
<tr>
<td>The mean final postoperative thoracolumbar angle</td>
<td>2.74 (0–15°)</td>
</tr>
<tr>
<td>The mean number of fused segments/ levels</td>
<td>14.87 (8–18)</td>
</tr>
<tr>
<td>Percent of patients in which sacropelvic extension was done</td>
<td>60.29%</td>
</tr>
</tbody>
</table>
were managed conservatively, and the other 3 were drained with chest tube. Two out of the 3 cases, who developed pleural effusion that required drainage with chest tube, underwent combined approach of ventral release-dorsal fusion.

2. **Hardware-related** complications (crank shaft phenomenon, pseudoarthrosis, screw loosening, and scoliosis progression or decompensation) constituted 16.67% (6 cases); 4 cases needed reoperation. Two cases needed complete revision of the dorsal fusion with instrumentation (one case of pseudoarthrosis and another with crank shaft phenomenon) and both cases were from the earliest cases operated in the center of Medical University of Graz and both of them had hybrid instrumentation with hooks & pedicle screws. One case had distal decompensation of the lumbar curve that necessitated distal extension of the instrumentation to include the lumbar curve, and another case had aseptic loosening of the distal screws with solid fusion mass needed partial removal of the loss screws & part of the connecting rod.

3. **Visceral complications:** occurred in 5 cases (13.89%); 3 cases had gastrointestinal complications (2 cases had postoperative paralytic ileus, and one case developed postoperative pyloric stenosis with feeding difficulty) and all were managed conservatively. Two cases had urinary complications (one had permanent urinary diversion, and the other developed urinary tract infection and was managed conservatively with antibiotics).

4. **Decubitus Ulcer & wound healing complications:** in 5 cases (13.89%); 4 cases had decubitus ulcer over sacrum and all of them healed conservatively with daily dressing and all occurred in late postoperative period, and one case had a delayed wound healing and also managed conservatively over a period of 6 weeks postoperatively.

5. **Deep wound infection:** was encountered in 3 cases (8.33%), all of them occurred in the early postoperative period (within the first postoperative month) that needed one or more debridement operations with parenteral antibiotics according to bacteriological culture and antimicrobial sensitivity tests till complete clearing up of the infections with clinical and laboratory parameters. The causative microorganisms were different in the 3 cases; *Propionobacterium acne*, *Staphilococcus epidermidis* and *Staphilococcus aureus*.

6. **CNS complications:** 3 complications (8.33%) occurred in 2 patients; one case had postoperative status epilepticus, and another had viral encephalitis and status epilepticus. All complications occurred in the early postoperative period, and were managed conservatively.

7. **Death:** occurred in one case (2.78% of all complications and 1.47% of the whole case series). The patient died on table just after final closure of the wound without evident cause and was attributed to hemodynamic instability and all intraoperative laboratory and radiological investigations showed no abnormalities. Pelvi-abdominal ultrasonography, echocardiography, chest x-ray and intraoperative laboratory check-up were all without remarkable abnormalities. Cardiopulmonary resuscitation was tried several times without response.

Large curve magnitude was considered as indirect measure of curve rigidity. We divided patients into 2 groups; one included patients that had curves $<90^\circ$, and the other had curves $\geq 90^\circ$, and recorded the incidence of complications in both groups (Table 2). Using chi-square test to check whether curve rigidity increased incidence of complications, there was no statistical significance between both groups ($p$ value = 0.0688).
Functional assessment questionnaire (Fig. 5): The average answers for walking ability was 3.27, hand function 2.42, while average values for other functions ranged between 1.12-2.08. Walking ability did not improve from that of the preoperative state for non-ambulant patients and slightly improved for the ambulant patients. Regarding to hand functions; 12 patients (23% from those shared in questionnaire) reported worst scores (5), most of them (11/12) had tetraplegic CP and hand function did not change from preoperative state. Most of other functions had optimal score and improved compared with preoperative state.

Patient/family satisfaction from surgery (Fig. 6): The goal of surgery was achieved in almost all patients, also most of them felt better after surgery. Ten patients (19.23% from those shared in questionnaire) said that daily care didn't improve after surgery. Most of the patients would decide to do surgery again if they were offered surgery, and would recommend surgery for patients with similar condition.

We have made a comparison between the largest 2 populations within operated patients; Cerebral Palsy (CP), and Duchenne Muscle Dystrophy according to some comparable preoperative, postoperative parameters and regarding proportion

Table 2

<table>
<thead>
<tr>
<th>The presence of complications</th>
<th>Curve ≥90°</th>
<th>Curve &lt;90°</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yes</td>
<td>9 (60%)</td>
<td>18 (33.96%)</td>
<td>27</td>
</tr>
<tr>
<td>No</td>
<td>6 (40%)</td>
<td>35 (66.04%)</td>
<td>41</td>
</tr>
<tr>
<td>Total</td>
<td>15</td>
<td>53</td>
<td>68</td>
</tr>
</tbody>
</table>

Fig. 5. Summary of the responses of 1st part of the questionnaire regarding patients' functional status, and daily activities

Fig. 6. Summary of the responses of the 2nd part of the questionnaire concerned with patient/family satisfaction from surgery
of associated medical, orthopedic comorbidities and also regarding incidence of postoperative complications. The results of comparison were summarized in Tables 3 & 4.

### Discussion

Conservative treatment of NMS as bracing, can delay but not eliminate the need for corrective surgery. Bracing, wheelchair modifications, physical therapies, and systemic corticosteroids in Duchenne are the commonly used conservative therapies. However, nonoperative interventions have not been proven to affect neuromuscular scoliosis. It is considered to decrease morbidity and mortality associated with spinal deformity, and can be delayed with appropriate conservative methods [15, 16]. However, prolonged delay of surgery in a progressive NMS may increase the risk of complications [3, 4, 17].

Only 3 operated cases needed combined approach of ventral release (one open & 2 thoracoscopic), followed by posterior fusion and instrumentation in another setting. These cases had a large, rigid curve that exceeded 90 degrees with good preoperative lung functions. There is a debate in literature regarding whether one setting, or staged combined approach could be better for the patient [18, 19]. Tsirikos, et al. [18] concluded that two-stage anteroposterior spinal fusion provides safer and better results with less complications than single-stage procedure in

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**Table 3**

Comparison between CP and DMD groups regarding perioperative outcome parameters

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Cerebral Palsy Group ($n = 36$)</th>
<th>Duchenne Muscle Dystrophy Group ($n = 10$)</th>
<th>$p$ value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean age at surgery (years)</td>
<td>15 ± 4.4</td>
<td>13 ± 1.1</td>
<td>0.187</td>
</tr>
<tr>
<td>Mean surgery duration (hours)</td>
<td>7.66 ± 1.47</td>
<td>6.84 ± 0.58</td>
<td>0.093</td>
</tr>
<tr>
<td>Mean postoperative ICU stay (days)</td>
<td>6.03 ± 4.32</td>
<td>4 ± 1.94</td>
<td>0.158</td>
</tr>
<tr>
<td>Mean Number of fused segments</td>
<td>14.83 ± 1.28</td>
<td>15.4 ± 1.35</td>
<td>0.226</td>
</tr>
<tr>
<td>Mean preoperative cobb's angle</td>
<td>78.5 ± 18.97</td>
<td>44.3 ± 19.33</td>
<td>&lt;0.0001*</td>
</tr>
<tr>
<td>Mean final postoperative cobb's angle</td>
<td>39.22 ± 17.92</td>
<td>15.6 ± 9.36</td>
<td>0.0002*</td>
</tr>
<tr>
<td>Mean cobb's correction %</td>
<td>50.52 ± 17.51</td>
<td>63.47 ± 16.72</td>
<td>0.0425*</td>
</tr>
<tr>
<td>Mean loss of correction</td>
<td>6.22 ± 6.53</td>
<td>4.3 ± 4.24</td>
<td>0.3855</td>
</tr>
<tr>
<td>Mean preoperative pelvic obliquity</td>
<td>14.44 ± 8.89</td>
<td>6.3 ± 4.72</td>
<td>0.0081*</td>
</tr>
<tr>
<td>Mean postoperative pelvic obliquity</td>
<td>10.31 ± 6.82</td>
<td>4.5 ± 4.62</td>
<td>0.0153*</td>
</tr>
<tr>
<td>Mean pelvic obliquity correction %</td>
<td>14.27 ± 10.89</td>
<td>20.32 ± 9.66</td>
<td>0.841</td>
</tr>
<tr>
<td>Mean Thoracolumbar kyphosis</td>
<td>46.54 ± 24.87</td>
<td>44 ± 11.44</td>
<td>0.8162</td>
</tr>
</tbody>
</table>

*Note. Comparison was done using 2 tailed independent $t$ test and significance was considered when $p$ value was ≤0.05*.

**Table 4**

Comparison between CP and DMD groups according to proportion of associated medical, orthopedic comorbidities and postoperative complications

<table>
<thead>
<tr>
<th>Parameter</th>
<th>CP Group ($n = 36$)</th>
<th>DMD Group ($n = 10$)</th>
<th>$p$ value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Proportion of associated medical comorbidities</td>
<td>Mean: 0.722</td>
<td>Mean: 0.3</td>
<td>0.01326*</td>
</tr>
<tr>
<td></td>
<td>Variance: 0.2063</td>
<td>Variance: 0.2333</td>
<td></td>
</tr>
<tr>
<td>Proportion of associated orthopedic comorbidities</td>
<td>Mean: 0.66667</td>
<td>Mean: 0.8</td>
<td>0.39067</td>
</tr>
<tr>
<td></td>
<td>Variance: 0.2286</td>
<td>Variance: 0.17778</td>
<td></td>
</tr>
<tr>
<td>Proportion of postoperative complications</td>
<td>Mean: 0.38889</td>
<td>Mean: 0.1</td>
<td>0.04518*</td>
</tr>
<tr>
<td></td>
<td>Variance: 0.2444</td>
<td>Variance: 0.1</td>
<td></td>
</tr>
</tbody>
</table>

*Note. Comparison was done using 2 sample $z$ test for means and significance was considered when $p$ value was ≤0.05*.
the management of patients with NMS. We used the staged approach with a spacing of 10–14 days between the two operations time until the patients recovered adequately from the first operation. Halo gravity traction was done after ventral release which decreased the curve magnitude.

Regarding ICU stay, 28.4% of the patients needed more than 5 days stay in ICU, and about one third of them had early postoperative complication. Similar conclusions were made by Thacker et al. [20] as they reported better results and good perioperative care when postoperative ICU stay was short.

Many of the cases (48.65%) in whom surgery duration was ≥7.3 hour developed postoperative complication, and 72.22% of the complications were during the early postoperative period. In all cases of deep wound infection, the surgery duration was ≥8 hours. As it is logically expected and previously reported by previous studies [21, 22] that increased surgery duration was associated with adverse outcome specially postoperative wound infection, and this was also concluded by our study.

As previously stated, NMS is commonly associated with medical co-morbidities. In the current study, 58.82% of the patients had one or more co-morbidity. Gastroesophageal reflux was the commonest co-morbidity, followed by epilepsy, and cardiopulmonary disorders.

Epilepsy and antiepileptic drugs were associated with higher postoperative pulmonary complications as noticed by previous studies [23, 24]. In this study, 43% of the patients who had epilepsy as a medial comorbidity developed postoperative pulmonary adverse effect like pneumonia, aspiration pneumonia, bronchopneumonia, pleural effusion, or haemothorax. This difference was not of a statistical significance as compared with other patients with chest complications not suffering from epilepsy (p value = 0.18511).

Chidambaran et al. [25] have concluded that NMS in cerebral palsy with gastroesophageal reflux (GERD) as an associated co-morbidity undergoing spinal fusion were associated with increased rate of postoperative infections, either surgical site or organ space infection (like pneumonia or urinary tract infections). Possible explanations for this are; 1st: GERD predispose to malnutrition and aspiration pneumonia, 2nd: patients with GERD treated with acid inhibitors lose the protective acid barrier of the stomach increasing the risk for bacterial overgrowth, and 3rd: gastric acid inhibitors directly inhibit several leukocyte functions and modify gastrointestinal microflora quality and quantity [25]. In this study, GERD occurred in 36.76% of the operative group population, and postoperative infection developed in 16% of the patients that had GERD as a co-morbidity. Three cases of organ space infection, and only one case of deep surgical site infection. This difference was also not statistically significant as compared with those with postoperative infections not suffering from GERD (p value = 0.680699).

Regarding Cobbs angle correction, the previous studies showed variable good results of correction, either in spastic like CP, or muscular weakness like DMD. Bohtz et al. [25] made a study on NMS in CP, and their preoperative Cobb angle was 78.6 (50–120) degrees, and the mean correction of 64.3% (mean final Cobb angle: 28 degrees) was obtained. Similar results were recorded in a study made by Suk et al. [26] regarding NMS in a group of patients with DMD, SMA, and variable muscle dystrophies with a mean preoperative Cobb angle of 61.5 degrees, and a mean postoperative angle of 39 degrees. Another recent study with less degree of correction from Nordon et al. [11] included different neuromuscular diseases, primarily CP (55% of patients). The mean preoperative Cobb angle was 78.8 degrees, and the mean postoperative angle was 44.6 degrees, with a mean correction of 34.2 degrees.

In this present study, the mean preoperative Cobb angle was 71.7 degrees, while the mean immediate postoperative Cobb angle was 29°, and the mean final postoperative Cobb angle was 34.6°, with a mean correction percentage of 53.25%, and the mean final loss of correction was 5 degrees. In prospective of pelvic obliquity, our results showed insignificant differences. The mean preoperative pelvic obliquity was 12.37°, the mean immediate postoperative pelvic obliquity was 6.67°, and the mean final postoperative pelvic obliquity was 8.25°, with a mean final loss of correction of 1.58 degrees. Similar insignificant results were noted regarding thoracic kyphosis, and lumbar lordosis.

Sagittal plane deformities especially thoracolumbar kyphosis is a known risk factor for pseudoarthrosis, with reported relative risk variable between 2.7 to 3.6 when there is thoracolumbar kyphosis of 20 degrees [27].
The incidence of TL kyphosis in the operated patients was 34%. The mean preoperative angle was 48.22 degrees, and nearly all patients were corrected to the physiological range, with a mean final angle of 2.74 degrees. In all cases with TL kyphosis, there were no associated pseudarthrosis, and only one case developed a septic loosening of 1st sacral screw. Treatment of the NMS with only pedicle screw instrumentation is a rising concept, which eliminate the need for additional anterior release, and thus surgery risk [28]. Pedicle screws offer better grip with 3-column purchase, and a longer moment arm due to anatomic location of pedicle screw compared to other forms of implant as hooks, or sublaminar wires [29].

In the current study, two types of instrumentation were used; the main instrumentation was the only-pedicle construct, and the minority hybrid (thoracic vertebrae hooks + pedicle screws). Hybrid instrumentation was used in 7.35% of cases, while in the rest of patients only-pedicle screw construct was done. In two cases with hybrid instrumentation (40%); pseudarthrosis, and crankshaft phenomenon occurred that made complete revision with only-pedicle screw construct necessary. Another two cases of the hybrid instrumentation patients developed haemothorax, and pleural effusion that needed chest tube drainage.

Regarding complications; NMS is associated with longer hospital stay, and a five-fold higher frequency of mortality. NMS patients have a seven-fold higher risk of losing about half of their blood volume during surgery compared to those AIS [30–32]. The reported literature prevalence of postoperative complications following NMS correction surgeries is variable, and relatively high with reported range of 18–75%. In a meta-analysis of 15218 NMS patients, Sharma et al. [20] found prevalence rates of complications of 22.7% (pulmonary), 12.5% (implant, primarily removal, revision, or extension of instrumentation), 10.9% (infections), 3.0% (neurological), and 1.9% (pseudoarthrosis). Good knowledge of the postoperative complications will allow the surgeon to anticipate common problems, trying to prevent, and early detect and manage properly [20].

There are several ways to describe and classify postoperative complications. One common type is the time when complication occurred; intraoperative, early, and late postoperative. Another one is major, and minor complications. Another one is regarding the category of the complication; infections (surgical site or organ space), hardware related, chest, wound healing, visceral, etc. Most authors only report the complication which affect the outcome of the patient, and overlook those which did not compromise the patient, require further surgery, and/or affect the outcome [21].

In the current study, we have recorded any adverse effect, and categorized them into; major (that necessitated second operation for its management), moderate (that needed minimal intervention like bronchoscope, or chest tube), or minor (that could be controlled with conservative measures). The total number of complications were 36, and occurred in 27 patients. Major complications occurred in 8 patients (11.76%); 3 deep wound infection (4.4%) needed surgical debridement, and one death (1.47%). Two cases had hardware complications (2.94%); one distal decompensation needed distal extension of the dorsal fusion, and one distal loosening of the screws needed partial removal. One case of pseudarthrosis (1.47%), and one case of crankshaft phenomenon (1.47%), and both needed complete revision of the dorsal fusion with instrumentation. All the major complications, except for death, were adequately controlled with the surgical intervention, and the outcome of the patients was not adversely affected.

The moderate complications occurred in 7 patients (10.29%); 6 cases (8.82%) developed chest complications of pleural effusion & haemothorax needed chest tube drainage, and lung atelectasis & aspiration pneumonia needed bronchoscopic lavage, and one case (1.47%) had urine retention needed permanent urinary diversion. All cases were adequately controlled with the intervention, and the outcome was not affected. Regarding the minor complications 10 patients (14.71%) developed 19 complications; 5 cases of organ space infection (pneumonia and encephalitis), 3 cases of visceral complications (paralytic ileus and pyloric stenosis), 3 cases of decubitus ulcer, 2 cases of pleural effusion, 2 cases of minor aseptic loosening of distal screws, 2 cases of status epilepticus, and one case of delayed wound healing. All the cases were adequately controlled with conservative methods without any effect on the outcome.

Several studies investigated the patient-parent satisfaction from scoliosis correction surgery
in NMS, and the health-related quality of life in such patients, and most of these studies reported significant improvement of the health-related quality of life following surgery with patient-parent satisfaction [25, 33, 34]. Nordon et al. [11] investigated the correlation between the degree of correction of NMS, and patient quality of life and concluded that patients’ satisfaction following operation didn’t depend directly on the degree of deformity correction. They concluded also that, the disparity between satisfaction, and fulfillment of expectations might be due to unrealistic initial expectations, or misunderstanding of the main goal of the surgery.

In this study, our results of the patient satisfaction following surgery, and health-related quality of life following NMS correction surgery was similar to the previously reported literature results. Nearly all patients think that the goal of surgery was achieved. Most of them also believe that: they are doing better after the operation, they would decide to do surgery again if they can choose again, and they recommended surgery for other patients with similar conditions. Even those who gave relatively negative feedback regarding some daily care activities have reported that: there was no worsening following the operation, and these activities couldn’t be done before the NMS correction surgery.

Comparison between CP and DMD groups (both formed 67.65% of surgically operated groups) had showed some statistical differences regarding preoperative and postoperative cobb angle & pelvic obliquity. This could be attributed to the differences between preoperative cobb angle of both groups as we included NMS due to CP when Cobb angle was above 50°, while above 15° in DMD group. There were also statistical differences regarding proportion of associated medical comorbidities and postoperative complications. The differences regarding incidence of complications could be explained by; significant differences between both groups regarding preoperative curve magnitude, pelvic obliquity degree, and associated medical comorbidities.

One of the limitations of this study was that it included heterogeneous group of patients with various pathologies; spastic and paralytic lesions, which behave, and react to somewhat differently regarding rate of curve progression, and associated comorbidities. However, this was alleviated by standardizing the management protocol, indications of scoliosis correction, unifying the primary goal of surgery (limiting curve progression & establishment of balanced fused spine over a level pelvis), and also all cases were operated only by a single senior surgeon (V.S) by the same technique, and approach.

**Conclusion**

Scoliosis is a serious problem among neuromuscular disorders, and its management is usually complex due to associated co morbidities that needs a multidisciplinary cooperative team and it is adviceable to be done in a specialized equipped center.

NMS surgery is complex, and associated with a lot of perioperative and postoperative complications (developed in 40% of this study group). Despite increased incidence of increased morbidity, and mortality risks, functional outcome was optimal, and patient/family satisfaction from surgery was reached, and they recommended surgery to other patients with similar disorders.

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**Author contributions**

**Elsayed E. Negm:** corresponding author, writing the manuscript, measuring and evaluating the radiological data of patient list, evaluation of questionnaire, and making statistical analysis of data.

**Vinay Saraph:** all surgeries were done by him, revision of the manuscript.

**Mohamed S. Said:** shared in radiological measurements, writing and revision of manuscript, evaluation of the questionnaire.
All authors made a significant contribution to the research and preparation of the article, read and approved the final version before publication.

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