ORIGINAL ARTICLE
DEFORMITY
http://dx.doi.org/10.1590/S1808-185120222104262160

# FRONTAL AND SAGITTAL PATTERNS OF NEUROMUSCULAR SPINAL DEFORMITIES BASED ON NOSOLOGICAL PROFILE

## *PADRÕES FRONTAIS E SAGITAIS DAS DEFORMIDADES NEUROMUSCULARES DA COLUNA VERTEBRAL, SEGUNDO O PERFIL NOSOLÓGICO*

## *PATRONES FRONTAL Y SAGITAL DE LAS DEFORMIDADES NEUROMUSCULARES DE LA COLUMNA VERTEBRAL SEGÚN EL PERFIL NOSOLÓGICO*

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## ABSTRACT

Objective: Reveal frontal and sagittal patterns of spinal deformity depending on neuromuscular nosology for surgery and outcome planning. The characteristics of spinal deformity vary according to the pathology. In cerebral palsy, muscular dystrophies, and spinal muscular atrophy, specific features of deformities are poorly written, especially in the sagittal profile. Methods: The evaluation criteria were age, gender of the patients, the volume of blood loss, duration of hospitalization, measurement of the deformity curve, thoracic and lumbar kyphosis (Cobb angle), pelvic obliquity concerning the horizontal line, the percentage of curve correction. Cobb angle was measured preoperatively before hospital discharge (up to 21 days postoperatively) and one year after surgery. Results: The cohort of 71 patients with spinal deformities due to neuromuscular diseases included four groups: muscular dystrophy (MD), spinal muscular atrophy (SMA), Duchenne muscular dystrophy (DMD), and cerebral palsy (CP). The most characteristic deformity in the frontal plane was C-shaped thoracolumbar scoliosis with rotation of the pelvis; rotation of the vertebrae increased according to the magnitude of scoliosis. Lumbar hyperlordosis was common in patients with PD, whereas decreased thoracic kyphosis or even thoracic lordosis occurs more frequently in patients with DMD. Moderate correction of scoliosis was observed in all groups. There was no significant improvement in functional status, according to the FIM. Conclusion: The findings showed that rigid hyperlordosis is the main problem of spinal deformities in neuromuscular patients. Scoliosis and pelvic obliquity can be well corrected in NMS by pedicle screw construction with standard maneuvers and pelvic screw fixation. ***Level of Evidence IV; Lesser quality prospective study.***

**Keywords:** Scoliosis; Cerebral Palsy; Muscular Dystrophies; Spinal Muscular Atrophy.

## *RESUMO*

*Objetivo: Revelar padrões frontais e sagitais de deformidade espinhal depende da nosologia neuromuscular para cirurgia e planejamento de resultados. As características da deformidade espinhal variam de acordo com a patologia. Na paralisia cerebral, nas distrofias musculares e na atrofia muscular espinhal, as características específicas das deformidades estão mal escritas, especialmente no perfil sagital. Métodos: Os critérios de avaliação foram: idade, sexo dos pacientes, volume de perda de sangue, duração da internação hospitalar, medida da curva de deformidade, cifose torácica e lombar (ângulo Cobb), obliquidade pélvica em relação à linha horizontal, a porcentagem da correção da curva. O ângulo Cobb foi medido no pré-operatório antes da alta hospitalar (até 21 dias de pós-operatório) e um ano após a cirurgia. Resultados: A coorte de 71 pacientes com deformidades espinhais devido a doenças neuromusculares incluiu quatro grupos: distrofia muscular (DM), atrofia muscular espinhal (AME), distrofia muscular de Duchenne (DMD) e paralisia cerebral (PC). A deformidade mais característica no plano frontal era a escoliose toracolombar em forma de C com a rotação da pélvis; a rotação das vértebras aumentou de acordo com a magnitude da escoliose. A hiperlordose lombar era comum em pacientes com DP, enquanto que a diminuição da cifose torácica ou mesmo a lordose torácica ocorre com maior frequência em pacientes com DMD. A correção moderada da escoliose foi observada em todos os grupos. Não houve melhora significativa no status funcional, de acordo com a FIM. Conclusão: Os achados mostraram que a hiperlordose rígida é o principal problema das deformidades espinhais em pacientes neuromusculares. A escoliose e a obliquidade pélvica podem ser bem corrigidas no NMS através da construção de parafusos pediculares com manobras padrão e fixação de parafusos pélvicos.* ***Nível de Evidência IV; Estudo prospectivo de menor qualidade.***

***Descritores:*** *Escoliose; Paralisia Cerebral; Distrofias Musculares; Atrofia Muscular Espinhal.*

## *RESUMEN*

*Objetivo: La revelación de los patrones frontal y sagital de la deformidad de la columna vertebral depende de la nosología neuromuscular para la planificación de la cirugía y los resultados. Las características de la deformación de la columna vertebral varían según la patología. En la parálisis cerebral, las distrofias musculares y la atrofia muscular espinal, las características específicas de las deformidades están mal escritas, especialmente en el perfil sagital. Métodos: Los criterios de evaluación fueron la edad, el sexo de los pacientes, el volumen*

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Received on 03/17/2022 accepted on 09/15/2022
Reviewed by: Helton Delfino

*de pérdida de sangre, la duración de la hospitalización, la medición de la curva de deformación, la cifosis torácica y lumbar (ángulo de Cobb), la oblicuidad pélvica en relación con la línea horizontal, el porcentaje de corrección de la curva. El ángulo de Cobb se midió antes del alta hospitalaria (hasta 21 días después de la operación) y un año después de la misma. Resultados: La cohorte de 71 pacientes con deformidades espinales debidas a enfermedades neuromusculares incluía cuatro grupos: distrofia muscular (DM), atrofia muscular espinal (AME), distrofia muscular de Duchenne (DMD) y parálisis cerebral (PC). La deformación más característica en el plano frontal era la escoliosis toracolumbar en forma de C con rotación de la pelvis; la rotación de las vértebras aumentaba según la magnitud de la escoliosis. La hiperlordosis lumbar fue común en los pacientes con EP, mientras que la disminución de la cifosis torácica o incluso la lordosis torácica ocurre más frecuentemente en los pacientes con DMD. Se observó una corrección moderada de la escoliosis en todos los grupos. No hubo una mejora significativa del estado funcional según el FIM. Conclusión: Los resultados mostraron que la hiperlordosis rígida es el principal problema de las deformidades de la columna vertebral en los pacientes neuromusculares. La escoliosis y la oblicuidad pélvica pueden corregirse bien en el SMN mediante la construcción de tornillos pediculares con maniobras estándar y la fijación de tornillos pélvicos.* ***Nivel de evidencia IV; Estudio prospectivo de menor calidad.***

***Descriptores:*** *Escoliosis; Parálisis Cerebral; Distrofias Musculares; Atrofia Muscular Espinal.*

## INTRODUCTION

Neuromuscular scoliosis (NMS) can be defined as a secondary deformity of the spine with a leading scoliotic component caused by muscle imbalance due to neuropathic or myopathic conditions.

Scoliosis develops in about 20-25% of patients with cerebral palsy (CP) and nearly all children with DMD (Duchenne muscular dystrophy) and spinal muscular atrophy (SMA).[1-3] Loss of trunk balance, pain, discomfort in the back, and problems of verticalization are indications for orthopedic correction of spinal deformity in NMD patients.

The authors aimed to analyze a mixed NMS cohort consisting of patients with various types of muscular dystrophy (MD), CP, DMD, and SMA, focusing on radiological characteristics of spinal deformities in each of these diseases before and after surgery.

## METHODS

The study was carried out prospectively in 2018-2019 years, in patients with NMS who were surgically treated in our center due to MD, CP, DMD, and SMA. Inclusion criteria were: confirmed MD, CP, DMD, or SMA diagnosis (neurological and/or genetic examination), age ≤18 years old, the presence of spinal deformity, and signed informed consent to participate in the study. Only patients indicated for surgery and who eventually underwent surgery were included.

Indications for surgical correction of deformity were progressive scoliosis with curve ≥40° Cobb and lumbar hyperlordosis with pronounced sagittal imbalance.

A standard examination for all patients included anterior-posterior and lateral X-rays of the spine, the anterior-posterior X-ray of the hip joint, computed tomography (CT) scans of the spine and the chest, and magnetic resonance imaging (MRI) of the spine. X-rays of the spine were taken while standing or sitting with or without support, lying in some patients. Before a hospitalization, a standard examination was performed by a pulmonologist and a cardiologist, including, among other things, spirometry, blood and urine tests, electrocardiography and echocardiogram, and densitometry in some patients.

All patients underwent surgical correction of spinal deformity with posterior pedicle screw fixation. The evaluation criteria included: age, gender of patients, the volume of blood loss (mL), hospitalization length, and measurement of the deformity curve, thoracic kyphosis and lumbar lordosis (Cobb angle), pelvis obliquity relative to the horizontal line (Cobb angle), the percentage of deformity correction. The Cobb angle was measured preoperatively before discharge from the hospital (up to 21 postoperative days) and one year after surgery.

### Statistics

The significance of differences between data groups was evaluated using the criteria depending on the number of observations and distribution type of parameter values within groups. Statistical calculation was performed using Microsoft Excel software and SPSS Statistics.

We conducted this study in compliance with the principles of the Declaration of Helsinki. The Institutional Review Board of Ilizarov Center (IRB No. 2 (62) 05.03.2019) reviewed and approved the study's protocol.

## RESULTS

The nosological profile of 71 patients with NMS was as follows: 13 patients with MD, 12 with SMA, 14 with DMD, and 32 with CP (Table 1). Males predominated (63% vs. 37%). The average age was 14.0±3.61 years old. The angle of the main scoliotic curve ranged from 13.8° Cobb to 152.6° Cobb (mean 70.87±27.99° Cobb) (Table 2).

The mean FIM score in the whole NMS group before surgery was 49.7±32.02, predominating non-ambulatory patients (89% vs. 11%). Among patients with CP were six ambulatory and 26 non-ambulatory cases (GMFCS IV-V).

CT of the chest demonstrated significant deviation in four patients: atelectasis, hypostasis, and lung fibrosis. MRI of the spine did not reveal any significant pathology. Spirometry was done in 45 patients and showed variable results from the absence of lung function violations (13 patients) to extremely severe dysfunction (five patients). The rest of the patients (17) had a mild decrease in respiratory function. Spirometry could not be done in 26 patients due to peculiarities of mental and physical development (the patients could not execute instructions, or the procedure was not effective).

In group DMD, there was slightly more mild scoliosis than in other groups (DMD 53.41±24.66° Cobb vs. MD 69.67±24.29° Cobb, SMA 78.20±26.47° Cobb, CP 76.20±29.02° Cobb, p=0.076).

S-shaped scoliosis with a secondary scoliotic curve was relatively common in patients with MD (38%), SMA (42%), and DMD (29%), and less often in patients with cerebral palsy (2 patients, 6%), everyone else had C-shaped scoliosis (Table 2).

Pelvic obliquity was significantly less in MD patients (10.28±10.50° Cobb, p=0.047), with a relatively uniform distribution among the other nosological groups (Table 2, Figure 1a).

The tendency to thoracic hypokyphosis, and sometimes even thoracic lordosis, was more pronounced in the DMD group (p=0.004) (Figure 1b and 2).

More pronounced lumbar hyperlordosis with extremely pronounced rotation occurred in CP patients (p=0.003) (Figure 1c and Figure 3).

Growth-friendly instrumentation without pelvic screws was implanted for 14 patients (growing rod system), and the final correction with pelvic fixation was produced for 57 patients. The main types

**Table 1**. Distribution of selected patients with neuromuscular scoliosis by nosological profiles, gender and age (N=71 patients).

| | MD (n = 13) | SMA (n = 12) | DMD (n = 14) | CP (n = 32) | Total (n = 71) |
|---|---|---|---|---|---|
| Males | 9 | 7 | 9 | 20 | 45 |
| Females | 4 | 5 | 5 | 12 | 26 |
| Age (years old) | 14.0±4.40 | 11.3±4.37 | 15.4±1.55 | 14.5±3.17 | 14.0±3.61 |

**Table 2.** Major measured radiographic parameters in four groups of patients with neuromuscular scoliosis before surgery (N=71 patients).

| NMS group | MD (N = 13) | SMA (N = 12) | DMD (N = 14) | CP (N = 32) | Total (N = 71) |
|---|---|---|---|---|---|
| **Measurements** | **Mean ± SD** | **Mean ± SD** | **Mean ± SD** | **Mean ± SD** | **Mean ± SD** |
| Major scoliotic curve, Cobb | 69.67±24.29 (N=12) | 78.20±26.47 (N=12) | 53.41±24.66 (N=14) | 76.20±29.02 (N=32) | 70.87±27.99 (N=70) |
| Secondary scoliotic curve, Cobb | 52.34±25.81 (N=5) | 39.94±28.49 (N=5) | 32.30±10.59 (N=4) | 62.40±11.74 (N=2) | 44.71±23.14 (N=16) |
| Pelvic obliquity, Cobb | 10.28±10.50 (N=13) | 18.46±8.25 (N=12) | 17.99±7.76 (N=14) | 13.87±10.98 (N=32) | 14.79±10.13 (N=71) |
| Thoracic kyphosis Th5-12, Cobb | 28.39±24.50 (N=13) | 41.44±27.46 (N=12) | 5.99±22.42 (N=14) | 30.97±21.12 (N=32) | 27.34±25.37 (N=71) |
| Lumbar lordosis L1-S1, Cobb | 58.97±19.97 (N=13) | 46.58±22.64 (N=12) | 30.69±39.82 (N=14) | 65.81±39.10 (N=32) | 54.38±36.15 (N=71) |

CP - cerebral palsy, DMD - Duchenne muscular dystrophy, MD – muscular dystrophy, N - number of patients, NMS – neuromuscular scoliosis, SD - standard deviation, SMA - spinal muscular atrophy.

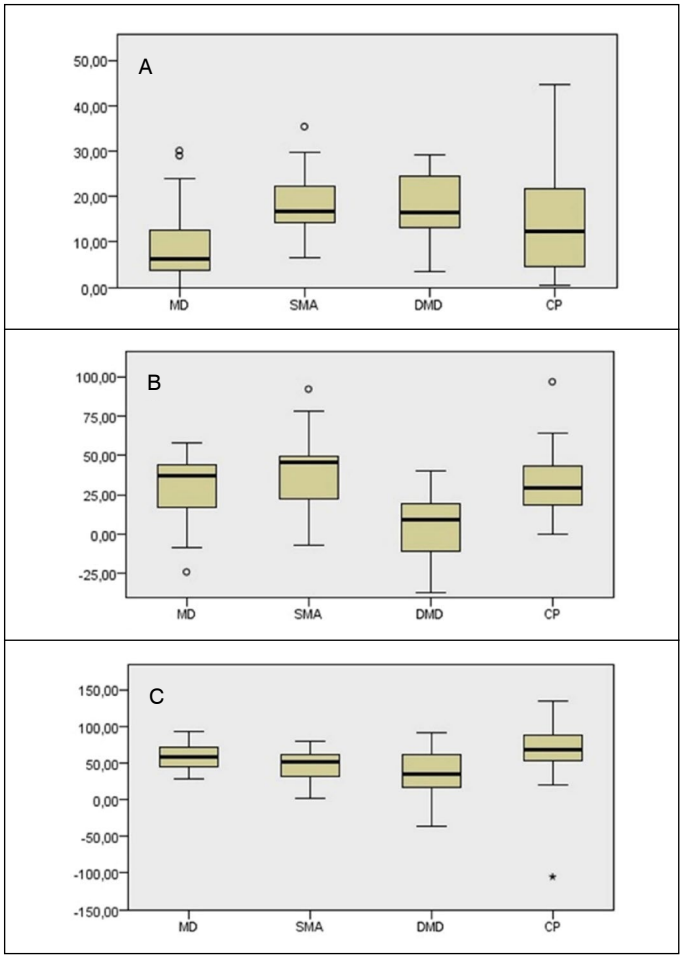


**Figure 1.** Pelvic obliquity (A), thoracic kyphosis (B), lumbar lordosis (C) measured prior to surgery (Kruskal-Wallis criterion for independent samples).

of pelvic fixation were sacral (S1, S2 bilateral pedicle screws) and iliac screws (iliac or S2 alar iliac screws). Osteotomies, excluding facetectomy, were not performed in any patient. All bone grafts were usually used for additional fusion. Operating times were the longest in patients with CP (217.69±68.05 min, p =0.035), with relatively little blood loss (342.81±197.34 ml) (Table 3). The greatest blood loss was observed in patients with SMA (438.17±262.96 ml, p=0.177) (Table 3). Intraoperative neurophysiological neuromonitoring (IONM) was done in 21 patients (spontaneous electromyography and motor evoked potentials recording). In most cases, it was unstable basally or not called up, most likely due to muscle dystrophy, pathology of the spinal cord path, or cortical atrophy.

Severe post-hemorrhagic anemia and the need for blood transfusion occurred in 38 patients (53.5%). For all patients, after surgery performed CT of the spine, significant screw malposition was not found.

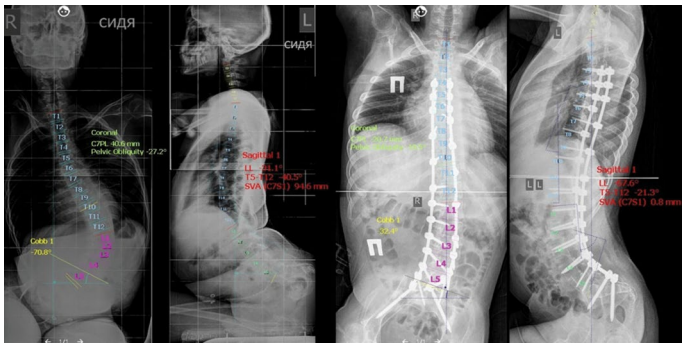


**Figure 2.** Spine X-rays of an 18-year-old patient with lordoscoliosis secondary to DMD before (a) and after (b) operative correction using a multi-support transpedicular system at the T4-S2-allar levels.

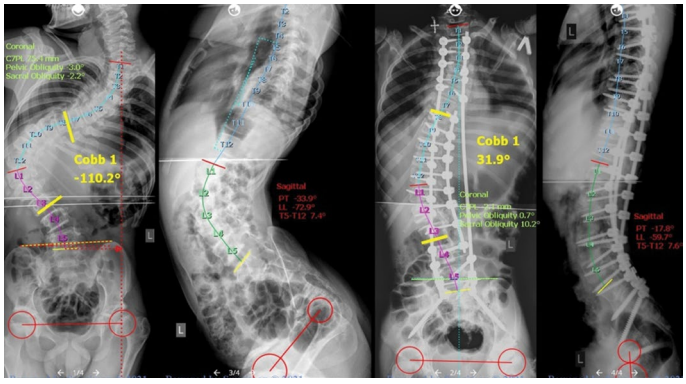


**Figure 3.** Spine X-rays of an14-year-old patient with lordoscoliosis secondary to CP, GMFCS level V, before (a) and after (b) operative correction of spinal deformity using a multi-support transpedicular system at the T2-S2-allar level.

The average time of in-patient treatment was 22.97±8.1 days (from 9 to 55 days), which is 2-2.5 times more than most patients after spinal deformity correction in our hospital. The longest hospital stay was in patients with MD (29.00±10.85 days, p=0.189), then in SMA (25.08±4.44 days) and DMD (23.31±7.60 days), and the shortest in patients with CP (19.59±6.59 days) (Table 3).

Moderate scoliosis correction was seen in all groups measuring 45% in MD, 47% in SMA, 61% in DMD, and 63% in CP (Table 2 vs. Table 4). Correction of the pelvic obliquity was 62%, 50%, 65%, and 67% in the groups, respectively (Table 2 vs. Table 4). DMD thoracic hypokyphosis (p=0.001) and CP lumbar hyperlordosis (p=0.001) were more rigid to surgical correction.

Unfortunately, X-rays evaluation one year after surgery could not be performed in all patients, which is due to the large territory of the country, the difficulties of transporting such patients, which often takes 2-3 days, COVID-19 trip restrictions, as well as the poor quality of X-ray at the living place (impossibility to perform vertical X-ray images of the entire spine). In addition, some of these patients

(1 with DMD and 1 with CP) did not take lateral X-rays in the outpatient clinic at the living place for unclear reasons. Only 45 cases came up for analysis. Evaluation of the results one year after surgery showed an average loss of scoliosis correction of 0.57±8.96° Cobb, 0.56±17.09°Cobb, 1.72±6.48°Cobb and 6.92±9.24°Cobb during a year, respectively for MD, SMA, DMD, and CP (Table 4 vs. Table 5). FIM score was also measured only in 45 patients. Among them, 33 patients (46%) improved, and patients (27%) deteriorated in functional independents. The average postoperative FIM scores one year after surgery was 47.30±30.43 (vs. preoperative 49.7±32.02). In addition, one patient with CP died six months after surgery because of acute cardiorespiratory insufficiency of an unknown cause.

## DISCUSSION

*Etiology, risk factors, conservative treatment, and natural history of NMS*

A large number of neuromuscular diseases could provoke the occurrence of spinal deformity.[4] The most common pathologies leading to NMS are CP, DMD and another MD, SMA, myelomeningocele, traumatic myelopathy, and spinal cord tumors.[4] The occurrence of spinal deformity depends on the nosology and level of nervous system damage, so the prevalence of spinal deformities in neuromuscular disease is extremely variable.[4] There are a large number of works, which are concentrated on prediction models for the NMS development in different pathology, conservative and preoperative treatment, reporting the results of surgical deformity correction, and vantages and disadvantages of surgery.[4]

The risks of scoliosis development and the natural course of the disease depend on pathology, severity, functional status of the patient and timely treatment. For example, scoliosis development predictable in non-ambulatory patients with CP and SMA.[4] In our study, most patients were non-ambulatory (89%).

Features of spinal deformity also vary depending on pathology. If in cases of myelomeningocele and posttraumatic myelopathy difference is obvious and depends on the level of pathology, and is often accompanied by lumbar kyphosis,[5] in other diseases difference is poorly written.

In our study group CP patients were characterized by hyper rotational lordoscoliosis associated with hip contractures and/or dislocation. Our data are confirmed by the findings reported by Yazici and Senaran who demonstrated a high incidence of such sagittal disorders in CP. Another question, what were the first: hip dislocation and pelvic obliquity or scoliosis? Hip dislocation may lead to pelvis rotation and scoliosis; on the contrary deformity may cause pelvic obliquity and initiate hip dislocation. It remains unclear.[6]

SMA patients showed moderately mobile kyphoscoliosis in our group. Merlini et al one of the first to notice that when children with SMA begin to sit, their backs become kyphotic and then scoliotic a curve develops, pelvic obliquity as a rule commensurate to scoliosis. Most studies indicate that thoracolumbar C-shaped scoliosis develops in most SMA patients, less often - S-shaped or local lumbar.[7] Sagital (kyphosis and lordosis) and axial (vertebrae rotation) features are rarely considered for analysis. It is noted that the greater the degree of scoliosis, the greater the amount of vertebrae rotation.

For patients with DMD, some authors described following types of scoliotic deformity: progressive collapsing kyphoscoliosis with significant rotation, progressive hyperlordotic scoliosis and straight

**Table 3**. Blood loss, surgery time and length of hospital stay (N=71 patients).

| NMS group | MD (N = 13) | SMA (N = 12) | DMD (N = 14) | CP (N = 32) | Total (N = 71) |
|---|---|---|---|---|---|
| **Measurements** | **Mean ± SD** | **Mean ± SD** | **Mean ± SD** | **Mean ± SD** | **Mean ± SD** |
| Blood loss (mL) | 378.46±250.53 | 438.17±262.96 | 340.00±165.34 | 342.81±197.34 | 364.90±212.72 |
| Surgery duration (min) | 177.31±43.10 | 181.67±49.14 | 190.71±51.06 | 217.69±68.05 | 198.89±59.54 |
| Length of hospital stay (days) | 29.00±10.85 | 25.08±4.44 | 23.31±7.60 | 19.59±6.59 | 22.97±8.13 |

CP - cerebral palsy, DMD - Duchenne muscular dystrophy, MD – muscular dystrophy, N - number of patients, NMS – neuromuscular scoliosis, SD - standard deviation, SMA - spinal muscular atrophy.

**Table 4**. Major measured radiographic parameters in following patients with neuromuscular scoliosis 7-21 days after surgery (N=71 patients).

| NMS group | MD (N = 13) | SMA (N = 12) | DMD (N = 14) | CP (N = 32) | Total (N = 71) |
|---|---|---|---|---|---|
| **Measurements** | **Mean ± SD** | **Mean ± SD** | **Mean ± SD** | **Mean ± SD** | **Mean ± SD** |
| Major scoliotic curve, Cobb | 38.78±14.53 (N=12) | 42.07±21.88 (N=12) | 21.16±16.89 (N=14) | 28.21±17.57 (N=32) | 30.99±18.91 (N=70) |
| Secondary scoliotic curve, Cobb | 34.62±18.35 (N=5) | 33.90±17.45 (N=5) | 18.90±7.71 (N=4) | 19.80±11.17 (N=2) | 28.26±15.55 (N=16) |
| Pelvic obliquity, Cobb | 3.88±3.41 (N=13) | 9.24±5.75 (N=12) | 6.30±6.57 (N=14) | 6.18±6.12 (N=32) | 6.30±5.86 (N=71) |
| Thoracic kyphosis Th5-12, Cobb | 25.95±12.70 (N=13) | 16.24±8.86 (N=12) | 9.72±12.29 (N=14) | 27.00±14.00 (N=32) | 21.58±14.31 (N=71) |
| Lumbar lordosis L1-S1, Cobb | 48.48±13.07 (N=13) | 39.02±7.67 (N=12) | 47.21±16.10 (N=14) | 57.03±20.80 (N=32) | 50.48±17.98 (N=71) |

CP - cerebral palsy, DMD - Duchenne muscular dystrophy, MD – muscular dystrophy, N - number of patients, NMS – neuromuscular scoliosis, SD - standard deviation, SMA - spinal muscular atrophy.

**Table 5**. Major measured radiographic parameters in following patients with neuromuscular scoliosis 1 year after surgery (N=45 patients).

| NMS group | MD (N = 7) | SMA (N = 9) | DMD (N = 7) | CP (N = 22) | Total (N = 45) |
|---|---|---|---|---|---|
| **Measurements** | **Mean ± SD** | **Mean ± SD** | **Mean ± SD** | **Mean ± SD** | **Mean ± SD** |
| Major scoliotic curve, Cobb | 36.64±24.54 (N=7) | 45.22±23.30 (N=9) | 20.54±16.12 (N=7) | 30.72±12.51 (N=22) | 32.96±18.73 (N=45) |
| Secondary scoliotic curve, Cobb | 40.23±34.74 (N=3) | 17.27±4.05 (N=3) | 21.20 (N=1) | 27.65±23.26 (N=2) | 27.67±21.87 (N=9) |
| Pelvic obliquity, Cobb | 3,41±5.4 (N=7) | 12.61±6.7 (N=9) | 6.20±4.76 (N=7) | 5.45±5.26 (N=22) | 6.68±6.17 (N=45) |
| Thoracic kyphosis Th5-12, Cobb | 28.14±7.99 (N=7) | 25.57±23.03 (N=9) | 10.62±9.10 (N=6) | 23.70±13.91 (N=21) | 22.99±15.49 (N=43) |
| Lumbar lordosis L1-S1, Cobb | 56.60±6.01 (N=7) | 45.27±13.89 (N=9) | 38.63±12.77 (N=6) | 56.58±15.29 (N=21) | 51.71±14.89 (N=43) |

CP - cerebral palsy, DMD - Duchenne muscular dystrophy, MD – muscular dystrophy, N - number of patients, NMS – neuromuscular scoliosis, SD - standard deviation, SMA - spinal muscular atrophy.

sagittal profile and non-progressive curves up to 30°Cobb. At the same time, it was noted that the kyphotic and scoliotic components increase in sitting position. The general trend for DMD scoliosis is the occurrence of thoracolumbar C-scoliosis with pelvic tilt, as with other types of NMS. In our study, we noted the predominance of thoracic hypokyphosis and back straightening, also found in some other studies.

It is necessary to start with conservative treatment.[8] Steroids in DMD patient results in a substantially decreased need for spinal deformity surgery. Development of NMS in CP can prevent or slow down timely by special exercises, bracing, a custom sitting system, passive stretching, postural management, and botulinum toxin injection.[9-11] The use of intrathecal baclofen pumps and selective dorsal rhizotomy, on contrary, can cause the progression of scoliosis.[12,13] In SMA cases, early medical treatment by gene therapy with an adeno-associated viral vector or intrathecal injections of an antisense oligonucleotide medication can prevent disease progression and deformity development.[14,15]

In general, studies of the natural course of scoliosis in patients with SMA and DMD (without specific medicament treatment) have shown substantial progression of spinopelvic deformities.[7]

Bracing can be useful when deformity is mobile.[10,16,17] The role of bracing in some patients with NMS (for example, in non-ambulatory CP, in all SMA and DMD cases) is limited because of the effect on respiratory movements of the chest, risk of bedsores and even more movement restriction. Custom-sitting system with multiple support can slow down deformity progression in non-ambulatory patients.[4,10]

*Indications for surgery, types of surgery, and complications*

Surgery can be considered in patients with scoliosis angle > 40-50°Cobb, which causes difficulties in daily care and positioning, leading to significant pain, alterations in skin integrity and bed sores, and worsening pulmonary and cardiac function.[4] The aims of deformity correction: achieving a balanced spine, prevention of curve progression, decreased pain, restoration of skin integrity, reduction of the chest and abdominal cavity deformity, improving urine passage, daily care facilitation, improvement in functionality and quality of life. The timing of surgery should be considered on an individual case basis.[5,18] Inpatients with NMS, skeletal immaturity, and rigid curves >30°Cobb early surgical treatment can be considered, and they have the risk of deformity progression almost 100%.[18] They usually performed the growth-friendly surgical treatment: growing rods or anterior growth modulating procedures.

It's clear now that pedicle screw system gives an advantage in deformity correction; their main disadvantage is that there are restrictions on spine movement.[18] On the one hand, scoliosis straightens better, but patents have more limited movement freedom, which can lead to a decrease in functional independence and quality of life. There are still heated debates now.[19] Most patients will have a spinopelvic fixation.[1] The exception is ambulatory patients with NMS with adequate head control, without hip subluxation or dislocation, and small degree of pelvic obliquity (<15°); they may not need pelvis fixation.[20]

Anterior release and fusion procedure also improve, especially in cases with rigid lordoscoliosis and large thoracic curves (>70°Cobb).

Different types of osteotomies (facetectomy, Ponte vertebrectomy, Smith-Peterson osteotomy, pedicle subtraction osteotomy, and vertebral column resection) in rigid large scoliotic curves can help provide better intraoperative curve correction.[18] An increased number of complications are described in more of these techniques. In addition they increase amount of blood loss and surgery time. Therefore, the use of osteotomies in patients with neuromuscular diseases is controversial and needs to choose carefully. As a rule, in patients with SMA, CP, and MD, especially in non-ambulatory cases, we generally avoid osteotomy or perform only multilevel facetectomy.

Correction and posterior instrumentation of the thoracic and lumbar spine and pelvic fixation can be recommended for cases with initial pelvic obliquity exceeding 15°. Scoliosis with thoracic hypokyphosis and even a fully lordotic spine were common for DMD cases and were surgically addressed for realignment and sagittal balance of the spine.

In our group, IONM was useless in most patients, and recent studies have explored the frequent unreliability of IONM in NMS patients.[21] In our opinion, in this direction, it would be useful to conduct additional studies and find the optimal way of IONM.[22] The reported overall complication rate in NMS is variate from 6% to 40%.[12] In CP, complication after surgery developed in 17-68%.[5]

The general tendency for complication in NMS is as follows: the lower functional status is accompanied by a higher risk of complication,[16] pulmonary complications are most common, implant-related complications and infection meet a little bit less often.[16] Younger age of surgery associated with a higher risk of pseudoarthrosis and neurological deterioration.

Studies are showing that the respiratory function of NMS patients can be improved by proper periodical management, such as training program in which they used noninvasive positive pressure ventilation and mechanical insufflation-exsufflation before and after surgery.[21]

*Long-term results of surgery*

It should be remembered that the functional status of patients with NMS varies greatly, so in some cases, researchers cannot assess directly quality of life or functional status of the patient, in such cases, evaluation of the quality of life and satisfaction of their caregivers will help.[4]

Most studies show improvement in the quality of patients' and caregivers' life and caregivers' satisfaction.[23-26] They found that most of the patient caregivers felt that those patients treated surgically were more comfortable than those who were not. Not all research results demonstrated significant changes in the quality of life of the patients or improvement in vital signs.[27]

## CONCLUSION

Spinal deformity in NMS patients is characterized by specific manifestations with prevailing deformity components depending on the nosology. The findings showed that lordosis was the key problem of spinal deformities in neuromuscular patients and was often rigid to surgical correction. Lumbar hyperlordosis with significant rotation were common for CP patients, whereas the thoracic hypokyphosis often occurred in DMD patients. Scoliosis and pelvic obliquity can be well corrected in NMS by pedicle screw construction with standard maneuvers and pelvic screw fixation. The data obtained can be practical for the planning of surgical corrections of spinal deformities and identifying correction maneuvers.

### Limitations

A small group of patients has weak strength and sampling power and imposes limitations on the conclusions. Recognizing the specific nosological spectrum with the need for accurate verification of the underlying pathology multicenter prospective studies with databases of high-volume spinal centers, including neuromuscular scoliosis patients, can be used to prove or disprove our assumptions.

All authors declare no potential conflict of interest related to this article.

**CONTRIBUTIONS OF THE AUTHORS:** Each author contributed significantly to this manuscript. EUF, OMS, DMS, PVO, and SOR: substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data, or the creation of new software used in work. EUF, OMS, DMS, PVO, and SOR: drafted the work or revised it critically for important intellectual content. EUF, OMS, DMS, PVO, and SOR: approved the version to be published. EUF, OMS, DMS, PVO, and SOR: agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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