



The Etiological Understanding of Spine Deformities in Patients with Neuromuscular Disorders

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Abstract

Purpose: The etiology understanding is the corner stone for proper management of the spine deformities in patients with neuromuscular disorders.

Methods: Progressive spine deformities have been encountered in 71 patients. All showed different forms of neuromuscular disorders. We proceeded with applying prompt evaluation criteria which was mainly based on the etiological diagnosis of each type of NMS (clinical phenotype and genotype). Assessing the Functional Independence Measure (FIM), we observed deterioration in some aspects. The Cobb's angle was assessed in two occasions i.e. prior to corrective surgeries plus the post-surgery results. Followed by one year postoperative re-assessment. Surgical corrections have been performed via posterior pedicle screw fixation.

Results: Surgical correction to re-align the C-shaped thoracolumbar scoliosis. Lumbar hyperlordosis were common in cerebral palsy patients, though decreased thoracic kyphosis or even thoracic lordosis occurs more frequently in Duchenne Muscular atrophy patients. There was noticeable improvement in functional status according to Functional Independence Measure (FIM).

Conclusion: Etiology understanding is the paramount element in re-aligning corrective surgeries of the spine in patients with progressive spine deformities. We concluded that rigid

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hyper lordosis has been and still the key problem encountered in patients with neuromuscular disorders.

Keywords

Neuromuscular Scoliosis; Cerebral Palsy; Muscular Dystrophies; Spinal Muscular Atrophy

Abbreviations

MD: Muscular Dystrophy; SMA: Spinal Muscular Atrophy; DMD: Duchenne Muscular Dystrophy; CP: Cerebral Palsy; NMS: Neuromuscular Scoliosis; FIM: Functional Independence Measure

Introduction

Neuromuscular Scoliosis (NMS) can be defined as a clinical manifestation of mal-development of the spine causing effectively progressive scoliosis.

Scoliosis can occurs in a remarkable number of patients with cerebral palsy and in patients with Duchenne Muscular atrophy [1-3]. Progressive spinal malalignment can cause a constellation of unpleasant complications.

The first and foremost procedure for proper management of patients with NMS have been carried out via detailed clinical assessment in correlation with the clinical and the radiological phenotype in all patients. Each category of these disorders have been thoroughly studied.

Material and Methods

Ethical approval: The protocol was reviewed and approved by the Institutional Review Board of Ilizarov Center (IRB No. 2 (62) 05.03.2019). The study was carried out prospectively between the years 2015-2019.

Table 1 showed the distribution of patients of with neuromuscular scoliosis via the etiological diagnosis. A constellation of pelvic and spinal mal-alignments, assessment performed and studied prior to corrective surgeries.

	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.	15.4±1.	14.5±3.	14.0±3.
		37	55	17	61

Table 1: Distribution of selected patients of with neuromuscular scoliosis via the etiological diagnosis.

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)

Table 2: Showed the characteristics of radiographic phenotypic mal-alignment in four categories of patients with neuromuscular scoliosis before surgery with a total of 71 patients.

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Inclusion criteria were confirmed through the definite diagnosis of MD, CP, DMD or SMA (were mainly based on clinical phenotypic characterization and genotype). The average age was ≤ 18 years old. Signed informed consents have been obtained from guardians.

Indications for surgical correction of deformity based on precise evaluation of the progressivity of scoliosis with curve $\geq 40^\circ$ Cobb, and for lumbar hyper-lordosis, patients were assessed in accordance with magnitude of spinal mal-alignment.

The radiological phenotype was the baseline evaluative tool, which included Antero-Posterior (AP) and lateral spine radiographs in supine and standing positions and AP pelvis radiographs in standing positions have been organized (Fig 1,2).

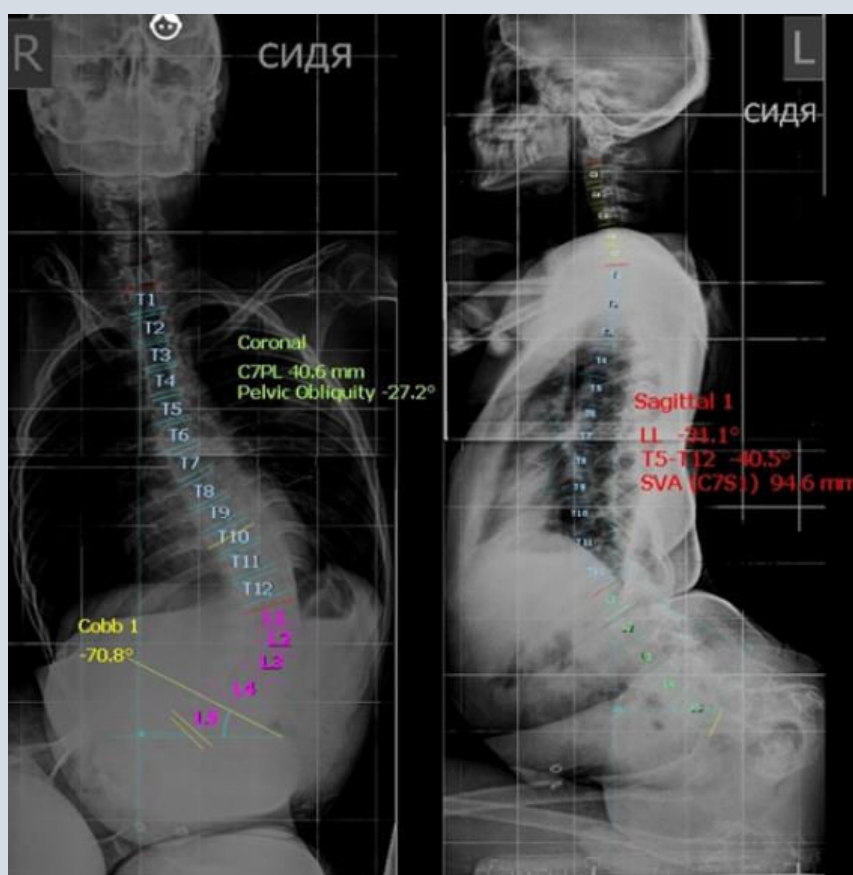


Figure 1: AP and lateral spine radiographs of an 18-year-old patient with severe spinal mal-alignment secondary to DMD (pre-operative radiographs).

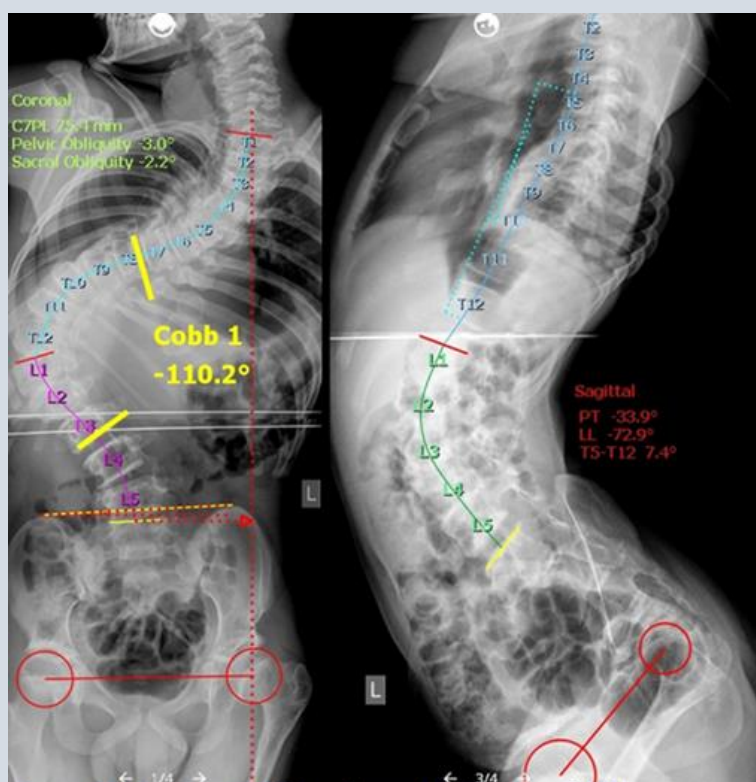


Figure 2: AP and lateral spine radiographs of a 14-year-old patient with lordoscoliosis secondary to CP, GMFCS level V (pre-operative radiographs).

Computed tomography of the spine and MRI for spine and chest. Prior to hospitalization, additional investigations to rule out any associated cardiothoracic abnormalities have been carried out accordingly.

Surgical corrections have been performed via posterior pedicle screw fixation.

Tomographic assessment to rule out associated cardio-thoracic abnormality has been performed.

Statistics

The importance of variations between the different clinical entities described in this paper have been evaluated via studying the diverse pathological features of each diagnostic category. We performed our statistics through the application of Microsoft Excel software and SPSS Statistics.

Results

Mean functional independence measure score in all patients prior to corrective re-alignment was 49.7 ± 32.02 , with a predominance of non-ambulatory patients (89% vs 11%). Among patients with CP were 6 ambulatory and 26 non-ambulatory cases (GMFCS IV-V).

In group DMD there were slightly more mild scoliosis than other groups (DMD $53.41 \pm 24.66^\circ$ Cobb vs MD $69.67 \pm 24.29^\circ$ Cobb, SMA $78.20 \pm 26.47^\circ$ Cobb, CP $76.20 \pm 29.02^\circ$ Cobb, $p=0.076$).

AP and lateral spine radiographs of an 18-year-old patient showed post-operative correction using a multi-support transpedicular system at the level of T4-S2 (Fig 3). In Fig. 4 AP and lateral spine radiographs of a 14-year-old showed operative correction of spinal deformity using a multi-support transpedicular system at the T2-S2-allar level (Fig. 4) (Table 3).

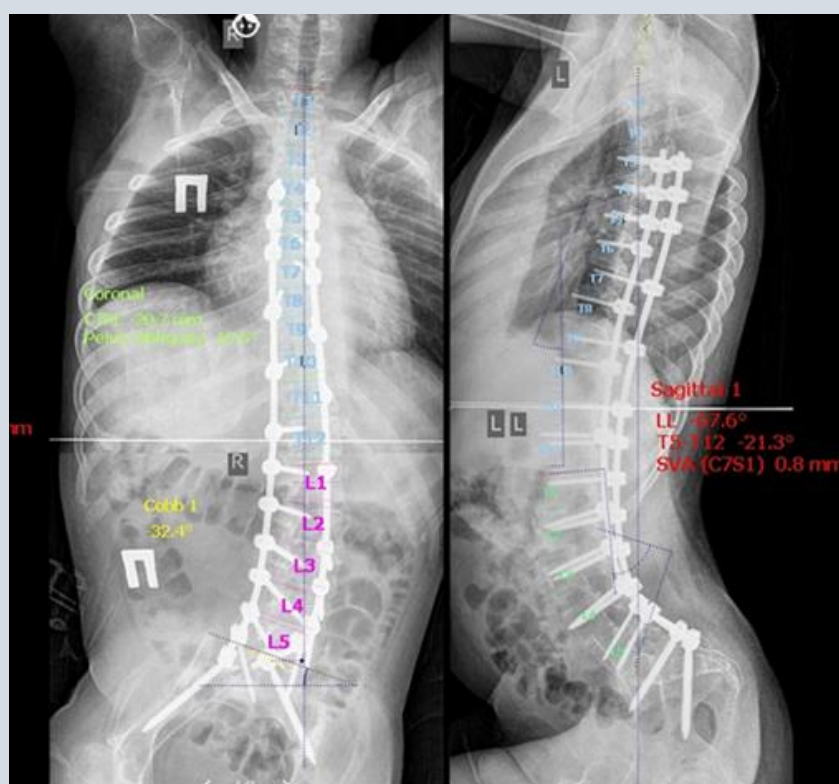


Figure 3: AP and lateral spine radiographs of an 18-year-old patient showed post-operative correction using a multi-support transpedicular system at the level of T4-S2.

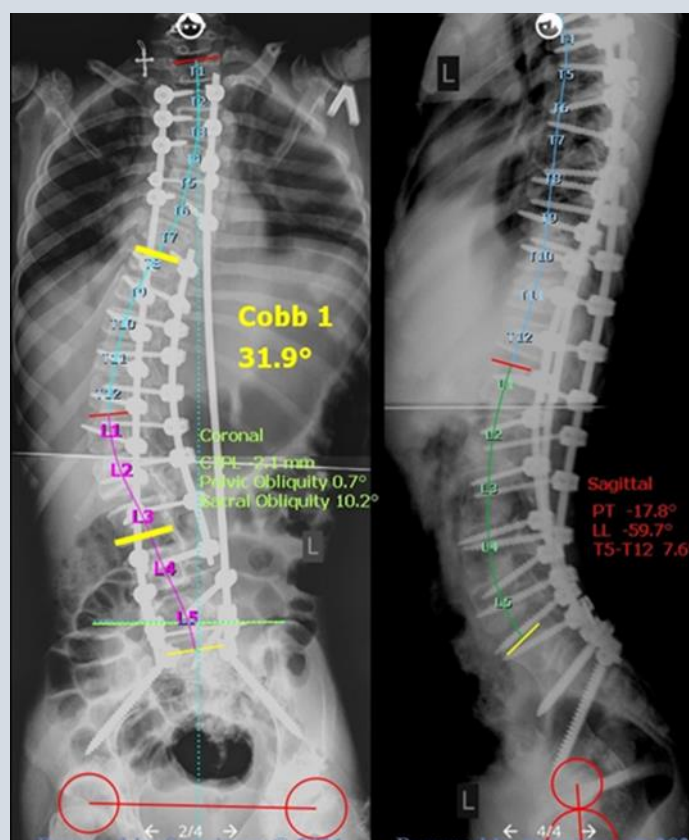


Figure 4: AP and lateral spine radiographs of a 14-year-old showed operative correction of spinal deformity using a multi-support transpedicular system at the T2-S2-allar level.

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±250.5	438.17±262.9	340.00±165.3	342.81±197.3	364.90±212.7
	3	6	4	4	2
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

Table 3: Showed the constellation of criteria such as blood loss, duration of intervention, and the period of hospitalization.

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Intraoperative Neurophysiological Neuro-Monitoring (IONM) was performed in 21 patients.

38 patients (53.5%) manifested anemia secondary to profuse bleeding. Blood transfusion was warranted (Table 4,5).

Unfortunately, because of logistical reasons, we were unable to perform radiographic evaluation on all patients in regard to one year postoperative results. COVID-19 trip restrictions has been an additional obstacle. Only 45 patients visited our Institute for re-evaluation. Evaluation of the results one year after surgery showed the average improvement of scoliosis correction, please refer to (Table 4 Vs Table 5). FIM score was re-assessed accordingly and promising results evolved. The percentage of ameliorating FIM score 1 year after surgery was 47.30 ± 30.43 (vs preoperative 49.7 ± 32.02).

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

Table 4: Showed the post-operative (7-21 days) results of the radiographic phenotype in all patients (N=71 patients).

NMS group	MD (N=7)	SMA (N=9)	DMD (N=7)	CP (N=22)	Total (N=45)
Measurements	Mean $\pm$ SD	Mean $\pm$ SD	Mean $\pm$ SD	Mean $\pm$ SD	Mean $\pm$ SD
Major scoliotic curve, Cobb	36.64 $\pm$ 24.54 (N=7)	45.22 $\pm$ 23.30 (N=9)	20.54 $\pm$ 16.12 (N=7)	30.72 $\pm$ 12.51 (N=22)	32.96 $\pm$ 18.73 (N=45)
Secondary scoliotic curve, Cobb	40.23 $\pm$ 34.74 (N=3)	17.27 $\pm$ 4.05 (N=3)	21.20 (N=1)	27.65 $\pm$ 23.26 (N=2)	27.67 $\pm$ 21.87 (N=9)
Pelvic obliquity, Cobb	3.41 $\pm$ 5.4 (N=7)	12.61 $\pm$ 6.7 (N=9)	6.20 $\pm$ 4.76 (N=7)	5.45 $\pm$ 5.26 (N=22)	6.68 $\pm$ 6.17 (N=45)
Thoracic kyphosis Th5-12, Cobb	28.14 $\pm$ 7.99 (N=7)	25.57 $\pm$ 23.03 (N=9)	10.62 $\pm$ 9.10 (N=6)	23.70 $\pm$ 13.91 (N=21)	22.99 $\pm$ 15.49 (N=43)
Lumbar lordosis L1-S1, Cobb	56.60 $\pm$ 6.01 (N=7)	45.27 $\pm$ 13.89 (N=9)	38.63 $\pm$ 12.77 (N=6)	56.58 $\pm$ 15.29 (N=21)	51.71 $\pm$ 14.89 (N=43)

Table 5: Represent the 1 year postoperative assessment via the radiographic criteria in 45 patients.

Discussion

The main tasks for comprehensive corrections of spine mal-alignment in patients with NMS are; etiology understanding, natural history, risk factors, conservative treatment and eventually surgical interventions.

The hazards of scoliosis is strongly correlated to the etiological background, the severity (whether progressive or non-progressive), and the general condition of the patient. For instance, the severity of scoliosis can be evaluated and to certain extent can be anticipated particularly in patients with cerebral palsy and in spinal muscular atrophy [4].

The phenotype of spine mal-alignment is diverse and correlated to the etiology and the pathological background. In other spine pathologies as in open spina bifida and in patients with deterioration of subacute ascending myelopathy. Ryabykh, et al., observed apparent differences in the grade of spinal deformities. Lumbar kyphosis has been described as the most common form of spinal mal-alignments [5].

In our study, we encountered that CP patients are most commonly presented with hyper rotational lordo-scoliosis. The latter was accompanied with hip contractures and/or dislocation [6,7]. Our findings were to certain extent compatible with the results showed by Yazici and Senaran [8].

There were some queries regarding the commencement of the scoliosis, was it stemmed from hip dislocation? Is the role of hip dislocation causing effectively to pelvic rotation and scoliosis? Or, progressive spine mal-alignment may cause pelvic deformities such as obliquity and dislocation. Such queries are still unclear [9]. Patients with DMD have been categorized in four types [10].

The corrective surgeries applied NMS patients are diverse and are mainly connected to the underlying etiology. It is mandatory to consider conservative programs. Corticosteroids have been recommended in DMD patient, though the results were unconfirmed. Special exercises are recommended to lessen the spine deformities especially in patients with CP. In addition to bracing, other postural and stretching exercises in conjunction with the administration of botulinum toxin injection might be helpful [9-14]. Panagopoulos, et al., applied intrathecal baclofen pumps associated with selective dorsal rhizotomy, though reports showed progression of scoliosis can occurs [14,15].

Sato, et al., described the management of postural deformity in patients with cerebral palsy. Messina, et al., and Brunner, et al., and others discussed the role of bracing in patients with NMS [16-18].

There are pros and cons regarding the advantages and disadvantages regarding the application of pedicle screw system [19-21].

Spinopelvic fixation is a treatment of choice applied to most of the patients with NMS 20-21.

In patients with rigid lordo-scoliosis with Cobb's angle of $>70^\circ$, we might refer to anterior release and fusion procedures, aiming to provide reasonable correction.

In rigid large scoliotic curve, different types of osteotomies can be applied with promising results [20].

The implemented procedures of osteotomies in patients with neuromuscular diseases is under dispute and has been discussed thoroughly [20-22].

In our patients, Intraoperative Spinal Cord Monitoring (IONM) turned out to be disadvantageous [23].

Conclusion

There is almost always a dilemma arises in connection with corrective spine surgeries among the spine surgeons. Panic of late onset mechanical/pathological complications is the main burden confronting spine surgeons. Despite the definite and solid diagnosis of each group of patients with neuromuscular disorders, yet every patient can develop unexpected set of unpleasant outcome. Furthermore, despite the general rules for diagnosis and management, but

nevertheless every single patient requires precise assessment in accordance with his individualistic criteria, even in patients with typical diagnostic criteria. Finally, we wish to stress that etiology understanding of every single patient is the fundamental element in approaching persuasive surgical results.

Limitations

We believe that, a small group of patients has insufficient impact and can impose certain limitations on the conclusions made. Recognizing the specific etiological spectrum with the need for accurate verification of the underlying pathology are mandatory. Thence fore, extensive studies are of utmost importance in connection with correlation of a comprehensive databases can be of tremendous help to further understand the wide spectrum of spine deformities in NMS patients.

Authors Contributions

EF, OS, DS, PO, SK, AAK and SR all made substantial contribution, analysis, and interpretations. AAK and SGK modified the paper contents. EF, OS, DS, PO and SR performed the surgical part. All read and approved the final version of the paper.

Conflict of Interest and Source of Funding

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