Functionality and quality of life after neuromuscular scoliosis surgery

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Abstract

Introduction: The objective of this study was to evaluate surgical complications in patients with scoliosis operated on in our Center, as well as their functionality and quality of life after surgical treatment.

Methods: A retrospective case series study was carried out. Medical history was reviewed and the patient and primary caregiver were interviewed. Results after surgery were assessed for complications, function, quality of life and caregivers´ degree of satisfaction.

Results: Complication rate was 26.7%, due to infection and hemodynamic alterations. Positive results were obtained regarding function and quality of life of these patients, being more significant in terms of the ability to be seated and activities of daily living.

Conclusions: Surgery of patients with neuromuscular scoliosis improves the capacity and balance of sitting, favoring digestive, respiratory and social life capacity. Although rate of complications is high, satisfaction rates on the part of the caregivers are elevated.

Key words: Spine; scoliosis; neuromuscular; cerebral palsy; quality of life; functionality

Level of Evidence: IV

Funcionalidad y calidad de vida luego de la cirugía de escoliosis neuromuscular

Resumen

Introducción: El objetivo de este trabajo fue estudiar las complicaciones de la cirugía en pacientes con escoliosis neuromusculares operados en nuestro Centro, así como la funcionalidad y la calidad de vida luego del tratamiento quirúrgico.

Materiales y Métodos: Se realizó un estudio retrospectivo, de tipo serie de casos. Se revisó la historia clínica, y se entrevistó al paciente y al cuidador principal. Luego de la cirugía, se evaluaron las complicaciones, la funcionalidad, la calidad de vida y el grado de satisfacción de los cuidadores.

Resultados: La tasa de complicaciones fue del 26,7%, por infección y alteraciones hemodinámicas. Se obtuvieron resultados positivos en la funcionalidad y la calidad de vida de los pacientes, y fueron más significativos en cuanto a la capacidad de estar sentado y las actividades de la vida diaria.

Conclusiones: La cirugía de los pacientes con escoliosis neuromuscular mejora la capacidad y el equilibrio de estar sentado, lo que favorece la capacidad digestiva, respiratoria y de vida social del paciente. Si bien se trata de cirugías con altas tasas de complicaciones, las tasas de satisfacción por parte de los cuidadores son altas.

Palabras clave: Columna; escoliosis; neuromuscular; parálisis cerebral; calidad de vida; funcionalidad.

Nivel de Evidencia: IV

Conflict of interests: The authors have reported none.

Introduction

Although neuromuscular scoliosis can result from diverse neurologic or muscular disorders (cerebral palsy, myelodysplasia, muscle dystrophy), its main underlying etiology is the dysfunction of muscle forces that act upon the vertebral column, what leads to trunk misbalance.[1] The different kinds of neuromuscular scoliosis share some characteristics, such as large curvatures at an early age, curvature rigidity, progression independent from growth, and concomitant kyphosis.

Oftentimes it is associated with pelvic obliquity, which causes unequal weight distribution while sitting, more pressure on the skin, and pain, thus generates frequent ischiatic and trochanteric sores. All this jeopardizes comfortable vertical sitting posture on his or her wheelchair in patients with general body disorders, what is key to these patients' mobility and social interaction, and requires from their parents for making a difficult decision: to either choose surgical correction or accept progressive and disabling deformity in their children. Patients with neuromuscular scoliosis come as a challenge due to the complexity of their deformities and their general health fragility.[2,3]

At the time of formulating a therapeutic plan, not only the curvature degree but also patients' needs and quality of life, the large percentages of perioperative complications and the underlying condition's natural history should be taken into account.

The aims of this study are to evaluate the surgical complications in the cases of neuromuscular scoliosis operated on at our Centre, to evaluate patients' function and quality of life after surgical treatment, and to assess caregivers' satisfaction level with the treatment.

Materials and Methods

We carried out a retrospective study of the case-series type in which we revised the patients operated on due to scoliosis of neuromuscular origin at our Centre between 2010 and 2015. All the procedures were undertaken by the same surgical team. Data were collected from records in the patients' surgical block and medical history. We interviewed patients and their caregivers to have questionnaires about surgical impact on different areas replied to.

Studied variables were: preoperative and postoperative Cobb angles, days of ICU admittance, total number of days of hospital admittance, and number of RBC transfusions. We also studied postoperative complications: superficial infection; deep infection (defined as the need for surgical toilet); implant-related complications; pulmonary complications: pneumonia, atelectasis, pleural effusion, pneumothorax, and long-term need for ventilator; neurologic complications; and non-union. The same author was in charge of assessing the angles of the curvatures using the same instrument.

We assessed patients' quality of life after the surgery by a questionnaire about the impact that surgery had had on different areas: 1, very negative; 2, negative; 3, no changes; 4, positive; and 5, very positive. The variables questioned about were: posture while sitting, pain, respiratory function, gastrointestinal function, day-to-day activities (eating, bathing, hair combing), mobility, schooling, rest, need for cares and cosmetic appearances. We assessed as "positive" those answers that were "very positive" or "positive", and as "without changes" or "negative" those ones that were "negative" or "very negative". This scale was used by Larsson in three of their studies.[4-6] On the other hand we evaluated the degree of caregivers' satisfaction by asking them—How satisfied are you with postoperative results? We posed five possible options to answer: 1, very unsatisfied; 2, unsatisfied; 3; indifferent; 4; satisfied; and 5, very satisfied. Data were analyzed by the chi square statistical method.

The authors have stuck to the protocols about publication of patients' data in the institution that the study was conducted at. All the patients included in the study and their guardians received sufficient information and granted their Informed Consent to participate in this study.

Results

We included 15 patients (7 females and 8 males) (Table). Their average age at the time of the surgery was 12.1 years old (ranging from 4 to 19). The average follow-up was 2.6 years (ranging from 1 to 4.4). The patients' conditions were: cerebral palsy (8 cases, 53%), myelitis (2 cases, 13%), myelomeningocele (1 case, 6.7%), muscle dystrophy (1 case, 6.7%), and minicore myopathy (1 case, 6.7%).

According to the Gross Motor Function Classification System (GMFCS) for cerebral palsy, five patients belonged to level V (62.5%); one, to level IV (12.5%) and two patients belonged to level III (25%). The types of curvatures were: thoracolumbar curvature (10, 67%); lumbar curvature (2, 13%); thoracic curvature (1, 6.7%); and two patients (13%) had double-curvature—thoracic and lumbar curvatures, with average 94º- Cobb angle. Eighty-seven percent of the patients (13) had pelvic obliquity. All of them were subject to posterior approach. The procedures we used were: instrumented arthrodesis with pedicle screws and bars (80%), Luque-Galveston instrumentation (13%) and growth bars (6.7%). In patients with pelvic obliquity we included the iliac bones in the fixation. The Cobb angle decreased 41.3% on average (ranging from 6 to 81.3%). Patients needed 15 days (ranging from 8 to 31) of hospital admittance, with average ICU admittance of three days (ranging from 1 to 7).

ISSN 1852-7434 (online) • ISSN 1515-1786 (printed)

Table. Patients' data

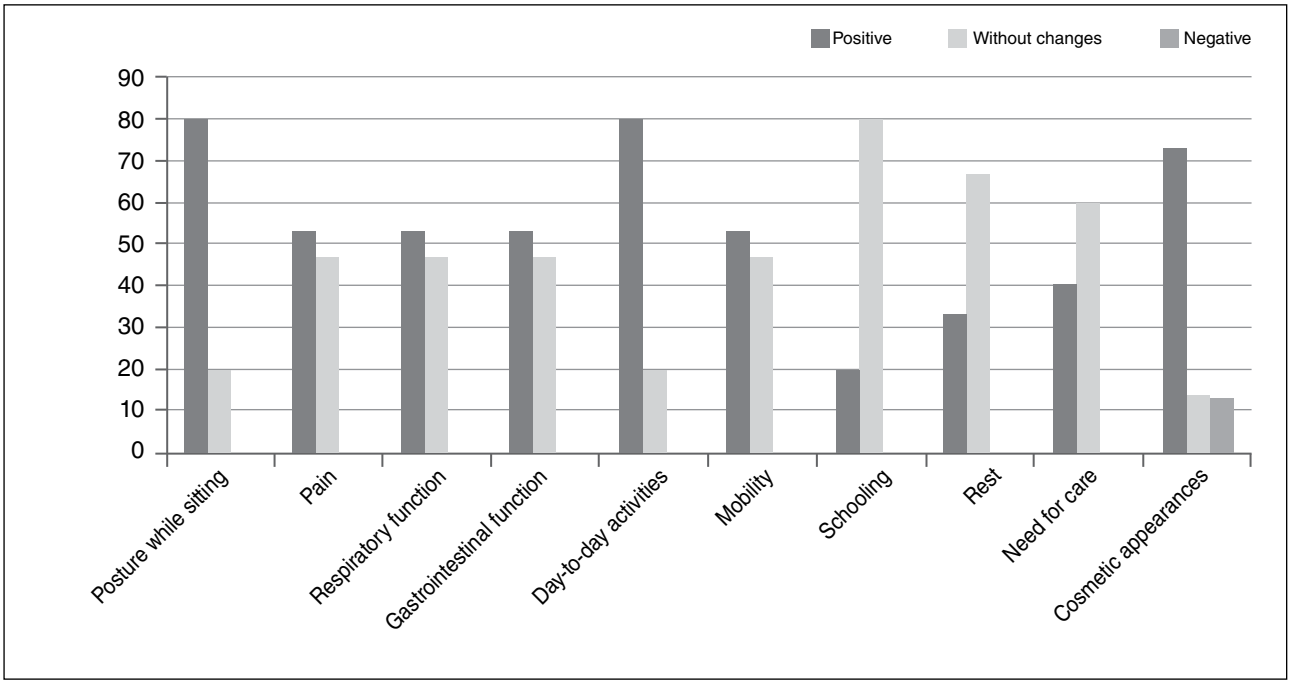
n	Age	Diagnosis	Type of curvature	Preoperative Cobb (º)	Postoperative Cobb (º)	Follow-up	Treatment	Admittance days	ICU admittance	Weight (kg)	RBC transfusion	Complications
1	15	CP GMFCS V	Lumbar	57	10	4.4	Luque-Galveston	8	3	31	1800	Deep infection
2	14	CP GMFCS V	Lumbar	54	22	3.6	Pedicle screws	9	1	23	900	No
3	18	CP GMFCS III	Thoracolumbar	117	40	1.25	Pedicle screws	13	2	41	900	No
4	14	CP GMFCS V	Thoracic	92	35	2.58	Pedicle screws	12	3	24	1800	No
5	14	CP GMFCS III	Thoracolumbar	73	37	2.33	Pedicle screws	11	2	31	1200	No
6	11	CP GMFCS V	Thoracolumbar	87	44	4.33	Luque-Galveston	31	6	32	900	Hemothorax Hypovolemic shock, Heart failure
7	12	CP GMFCS IV	Thoracolumbar	135	80	3.25	Pedicle screws	11	2	24	1200	No
8	10	Myelomeningocele	Thoracolumbar	160	83	2.58	Pedicle screws	21	2	21	900	Deep infection
9	11	Muscular dystrophy	Thoracolumbar	128	24	1.8	Pedicle screws	12	4	21	900	No
10	11	Rett Syndrome	Thoracolumbar	53	15	1.8	Pedicle screws	15	3	22	600	No
11	12	CP GMFCS V	Thoracolumbar	58	55	3.8	Pedicle screws	25	7	38	1750	Hypovolemic shock, Lobe atelectasis
12	9	Post-infectious myelitis	Thoracolumbar	73	12	1.0	Pedicle screws	19	2	30	1200	No
13	4	Post-vaccination myelitis	Double thoracic and lumbar	130-80	40-30	1.0	Growth bars	9	2	12	300	No
14	8	Minicore myopathy	Double thoracic and lumbar	113-35	15-30	3.4	Pedicle screws	13	3	21	450	No
15	19	Spinal atrophy	Thoracolumbar	85	35	2.75	Pedicle screws	15	3	46	1050	No

CP GMFCS = Cerebral Palsy *Gross Motor Function Classification System*

Four patients (26.7%) suffered surgical complications: deep infection that required surgical toilet and long-term antibiotic treatment (2 cases, 13%), immediately postoperative hypovolemic shock (2 cases, 13%), and respiratory complications—one patient suffered lobar atelectasis in left lung. The need for RBC transfusion was 1050 ml (ranging from 300 to 1800).

In Figure 1, there is detail about patients' quality of life. Regarding posture while sitting, 80% answered positively, whereas 20% showed no changes (p=0.01). As regards pain, respiratory function, gastrointestinal function and mobility, 53% of the patients underwent positive

changes, whereas 47% did not undergo any change (p=0.35). With respect to day-to-day activities (hair combing, teeth brushing, bathing), 80% of the patients underwent positive changes, while 20% of them did not show changes (p=0.01). Regarding schooling, 80% did not show changes after the surgery, whereas 20% showed



▲ **Figure 1.** Results by patients' functional variables after surgery.

positive changes (p=0.01). Sixty-seven percent did not undergo any change in rest, and 33 % showed positive changes (p=0.01). With respect to the patients' need for care after the surgery, 60% did not experience differences while 40% of them showed positive changes (p=0.15). When it comes to cosmetic appearances, 73% showed positive changes after the surgery, 14% did not show changes whatsoever, and 13% showed negative changes (p=.25). It is worth highlighting that caregivers who answered negatively made a point of surgical scars.

Caregivers' answers to questions about satisfaction levels with the surgery were as follows: satisfied (9.60%) and very satisfied (6.40%). Figure 2 shows the X-rays of one of the patients included in the study.

Discussion

There is controversy over the benefits spinal surgery brings about to patients with neuromuscular scoliosis. In fact, the studies that assess function and quality of life after the surgery include patients with very heterogeneous conditions. Most of these studies are retrospective and use scores which are not the most adequate ones for these types of patients.

Larsson et al. published several studies which analyze function and quality of life after the surgery.[4-6] In 2005, they reported a prospective study which included 82 patients with average follow-up of 9.5 years. The patients' average age at the time of surgery was 22 years old. Conditions were varied and heterogeneous—most patients suffered cerebral palsy and myelomeningocele.

To assess the patients' quality of life, they used the Klein-Bell test and evaluated function with the same parameters we used in our study. Although most variables showed positive changes after the surgery, such as posture while sitting, cosmetic appearance and day-to-day activities, these changes were not statistically significant. Another one of their studies[5] reports positive changes in balance and weight distribution on bearing surfaces while sitting (p=0.01, p=0.001).

Ersberg and Gerdhem[7] conducted a study which assessed preoperative and postoperative quality of life in 211 patients operated on due to scoliosis, among who 32 suffered scoliosis of neuromuscular origin. The patients' average age at the time of the surgery was 14.7 years old and average hospital admittance was 12 days. These authors evaluated quality of life by the EQ-5D and SRS-22r scores. In the patients operated on due to neuromuscular scoliosis, there were no significant changes at postoperative years one and two (p=0.4 and p=0.2) as compared with preoperative standards, although the article criticizes that these scores have not been designed for these types of patients.

Thacker et al.[8] published a retrospective study which evaluated 30 patients who, at the time of the surgery, averaged 10.6 years of age. To assess the patients' skills after the surgery, they used the Hospital Rancho Los Amigos' score; two thirds of the patients experienced functional improvement during follow-up, and most benefits involved capability to keep sitting posture.

ISSN 1852-7434 (online) • ISSN 1515-1786 (printed)

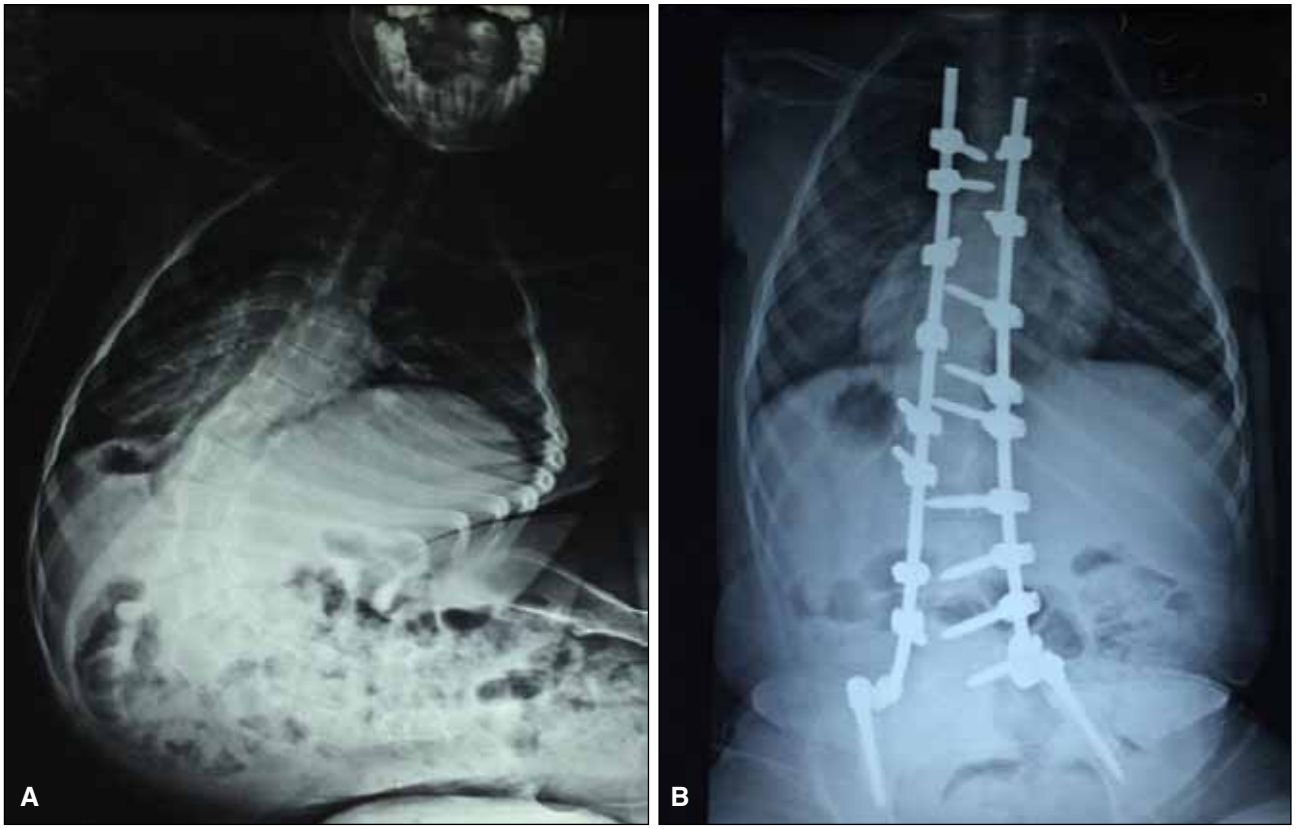


Figura 2. Eleven year-old patients with diagnosis of muscular dystrophy. **A.** Preoperative X-ray with thoracolumbar curvature of 128°. **B.** X-ray one year after the surgery with Cobb angle=24°.

Although ours is a retrospective study which includes a limited number of patients, our results were similar to those in the other studies. It is worth mentioning that most benefits after the neuromuscular scoliosis surgery were: capability to keep sitting posture and to carry out day-to-day activities. Moreover, caregivers' high rates of satisfaction with the surgical procedure are also worth highlighting.

Conclusions

Decision-making to carry out spinal surgery in these patients is difficult and it should be undertaken by a multidisciplinary team. In those patients who do not walk, capability to keep sitting posture decreases due to spinal deformity and pelvic obliquity.

Similarly to the other authors, we conclude that surgery for neuromuscular scoliosis improves capability to keep sitting posture and balance; this way, it favors patients' digestive and respiratory function as well as patients' social life. Although these surgeries are associated with high complication rates, caregivers' satisfaction levels are high.

All in all, new high-quality studies are warranted to assess the benefits that spinal surgery brings about to these patients.

Bibliography

1. Sarwark J, Sarwahi V. New strategies and decisión making in the management of neuromuscular scoliosis. *Orthop Clin North Am* 2007;38:485-96.
2. Herkowitz HN, Garfin SR, Elismont FJ, Bell GR, Balderston RA. *Rothman-Simeone The Spine*, 6ª ed. Philadelphia: Saunders; 2011.

3. Burgos J, Izquierdo E, Sarramea L. Patología de la columna vertebral, Sociedad Latinoamericana de Columna, Buenos Aires: Editorial Médica Panamericana; 2016, cap. 14.

4. Larsson EL, Aaro S, Ahlinder P, Oberg B. Preoperative evaluation of activities and function in patients with paralytic scoliosis. *Eur Spine J* 1998;7;294-301.

5. Larsson EL, Aaro S, Oberg B. Activities and functional assessment 1 year after spinal fusion for paralytic scoliosis. *Eur Spine J* 1999;8:100-9.

6. Larsson EL, Aaro SI, Normelli HC, Oberg BE. Long-term follow-up of functioning after spinal surgery in patients with neuromuscular scoliosis. *Spine* 2005;30(19):2145-52.

7. Ersberg A, Gerdhem P. Pre- and postoperative quality of life in patients treated for scoliosis. Initial experiences with the SweSpine Registry. *Acta Orthopaedica* 2013;84(6):537-43.

8. Thacker M, Hui JHP, Wong HK, Chatterjee A, Lee EH. Spinal fusion and instrumentation for paediatric neuromuscular scoliosis: retrospective review. *J Orthop Surg (Hong Kong)* 2002;10(2):144-51.

ISSN 1852-7434 (online) • ISSN 1515-1786 (printed)