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Activities and functional assessment 1 year after spinal fusion for paralytic scoliosis

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Abstract The results of spinal fusion in patients with paralytic deformities are usually presented as the correction of the deformity. When evaluating the surgical results in such patients, it is advantageous to classify the patients into subgroups because of the varying dysfunction and disabilities. The aim of this study was to evaluate the effect of spinal fusion in patients with paralytic scoliosis in relation to function in terms of Impairments, activities in terms of Disabilities, and dependence in terms of Handicaps 1 year postoperatively, with emphasis on subgroups. A total of 94 patients with paralytic scoliosis and 18 different diagnoses were evaluated 1 year after surgery. The patients were classified according to whether or not they could understand verbal instructions. The patients were also grouped according to the Scoliosis Research Society classification of diagnoses. A set of instruments was used with the variables classified according to WHO's International Classification of Impairments, Disabilities, and Handicaps (ICIDH). This consists of the patient's/relative's motives for surgery, persistent skin discoloration, ambulating or use of wheelchair, use of a brace, sitting balance, weight distribution on a sitting surface, angle of scoliosis, reaching, pain estimation, activities of daily living (ADL) Klein and Bell, care given, time spent resting, and seating supports. The set of instrument also included a follow-up questionnaire comprising 12 different areas, where the patients/relatives assessed the results of surgery. The study showed that spinal fusion in paralytic scoliosis led to showed improvements in

the whole group of patients at the Impairment level in the angle of scoliosis, sitting balance, weight distribution, and reduced number of patients with persistent skin discoloration, and at the Handicap level in reduced time for resting during the day, reduced number of seating supports in the wheelchair, and in the use of a brace. Most of the parameters were unchanged. The results in the subgroups were almost the same as in the whole group, although pain and reaching at the Impairment level, and ADL at the Disability level, could not be measured in the patients who were unable to understand verbal instructions. The subjectively assessed results showed that seating posture was ranked positively irrespective of the motive for surgery. The study showed that the patients with paralytic scoliosis maintained or improved their function and level of independence in terms of Impairment and Handicap 1 year postoperatively. The subjective results assessed by the patients/relatives also showed a positive outcome of surgery. Weight distribution on a seating surface was improved, but still uneven, and with respect to better sitting balance and increasing time sitting in a wheelchair, this can involve a risk for pressure sores and needs further investigation. When introducing outcomes including the Disability level, one must take the importance of homogeneity in the groups into consideration.

Key words Paralytic scoliosis · Spinal fusion · Activity · Function · The International Classification of Impairments, Disabilities, and Handicaps (ICIDH)

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Introduction

“Treatment of neuromuscular scoliosis has undergone an evolution over the past 50-year period. There was initially some enthusiasm for casting and bracing” [15], but “probably no other group of patients with spinal deformities benefits from corrective surgery as much as the paralytics” [20].

Paralytic scoliosis is a condition that is frequently progressive. Untreated curvatures will have adverse effects on the patient’s function and daily living [4]. Patients with paralytic scoliosis constitute a heterogeneous group with different diagnoses and varying dysfunction and disabilities, but the results of surgery are usually presented as the correction of various angles of the deformity. Such an approach considers the individual at an organ level according to Impairment from the taxonomy of The International Classification of Impairments, Disabilities, and Handicaps (ICIDH) [23]. The use of outcomes at all levels of Impairments, Disabilities, and Handicaps focuses on the perspective of illness, instead of a narrow disease perspective, when considering outcome of surgery [12]. In the traditional field of surgery, there are few prospective studies with large numbers of patients, and the outcome is often focused only on the spinal deformity. Considering past work, in 1924 Hibbs [10] reported on 59 patients with infantile paralysis and scoliosis who were treated with spinal fusion. He stated that the patients seemed to experience relief from fatigue and an improvement in general physical well being. In a study of 22 patients with spinal muscular atrophy fused with Harrington instrumentation, Aprin et al. [1] found no change in the ability to perform tasks involving daily hygiene, dressing and eating, but sitting balance was improved for most of them and the self-rated results were mostly positive. Brown et al. [5] presented a study of gross motor activities pre- and postoperatively in patients with spinal muscular atrophy. Thirty-four patients were spinal corrected with Harrington instrumentation and six patients with Luque segmental spinal instrumentation. The study compared the two instrumentations, and found no difference in postoperative function. An analysis of the total population showed a slightly improved sitting balance.

Askin et al. [2] reported poor results 1 year after corrective surgery in a prospective study on functional assessment including physical ability and independence in daily activities for 20 patients with neuromuscular disorders and spinal deformities. The patients or the caregivers stated that the cosmetic appearance had improved. The authors suggest that despite the failure to show an improvement in function, surgery should be performed to improve the cosmetic appearance and maintain the level of function.

The results from previous studies are inconsistent and show a need for more systematic investigations. Since evaluation is dependent on the patient’s ability to cooper-

ate, classifying the patients into subgroups, those who understand and those who do not understand verbal instructions [12], is advantageous. That means we can evaluate Impairment, Disabilities, and Handicap in the group of patients who understand verbal instructions. For the other group, who are not able to understand verbal instructions, it is more important to evaluate Impairments and Handicap. In that respect, the choice of assessment can be guided by the patient’s ability to understand verbal instructions, irrespective of his/her disorders.

Aim

The aim of this study was to evaluate the effect of surgical correction in relation to function in terms of Impairment, activities in terms of Disabilities, and dependence in terms of Handicap 1 year postoperatively, in patients with paralytic scoliosis, with emphasis on subgroups.

Materials and methods

Patients

This prospective study with 1-year postoperative evaluation consists of 94 consecutive patients with paralytic scoliosis, who were evaluated preoperatively from March 1992 to September 1996. Of the 100 preoperatively evaluated patients, 6 dropped out of the study: 2 patients had died, 2 patients refused to participate in the postoperative investigation, and 2 patients did not participate due to long travel distance. In this 1-year postoperative material of 94 patients, there were 55 females and 39 males with a mean age of 15.7 years (range 5–44 years) and a median of 15 years, with 18 years in the third quartile and 12 in the first quartile. There were 18 different diagnoses represented (Table 1).

Posterior segmental instrumentation with sublaminar wires, hooks and screws was used for all patients. In all wheelchair bound patients with pelvic obliquity, the instrumentation and fusion extended to the pelvis. In 37 patients, those with myelomeningocele or severe spasticity, an anterior instrumentation with Zielke apparatus was combined with the posterior segmental instrumentation. In the remaining 63 patients, only the posterior design was used.

The patients were classified, as had been done preoperatively, according to whether or not they could understand verbal instructions. One year postoperatively, there were 64 patients (of 67 evaluated preoperatively) in the group who were able to understand verbal instructions and 30 patients (of 33 evaluated preoperatively) who were not able to understand verbal instructions.

The patients were also grouped according to the Scoliosis Research Society classification of diagnoses (Table 1). Most of the mentally retarded patients were found in the group with upper motor neuron lesions ($n = 41$ of 45 evaluated preoperatively). These patients with upper motor neuron lesions were also grouped according to whether they could understand verbal instructions ($n = 15$) or could not understand verbal instructions ($n = 26$). All patients in the group with lower motor neuron lesions ($n = 37$ of 39 evaluated preoperatively) could understand verbal instructions.

There were ten patients 10 (of ten evaluated preoperatively) with myopathies, and with one exception all of them could understand verbal instructions. There were six patients (of six evaluated preoperatively) with congenital abnormalities, and half of this group could understand verbal instructions.

Table 1 Number of patients and their diagnoses

Diagnosis	<i>n</i>	Set of instruments
<i>Upper motor neuron lesion</i>	41	A set of instruments (Table 2), consisting of a combination of existing instruments and new ones [19], was used. This set of instruments consists of three parts: general information, activities and function, and a follow-up questionnaire. In the first, general information, the patients/relatives were asked about the motive for surgery and were given the following answer alternatives: respiration, pain, seating position, cosmetics with yes/no alternatives. An open question was also included to permit other answers. Other general information variables that were evaluated postoperatively were: persistent skin discoloration, ambulating, or use of a wheelchair, and use of a brace. In the second part, concerning activities and function, sitting balance, weight distribution on seating surface, angle of scoliosis, reaching, pain estimation, ADL Klein and Bell index, care given in physical effort and time consumption, time used for resting, and seating supports in a wheelchair were evaluated. The third part, a follow-up questionnaire, consisted of a subjective assessment where the patients and relatives, or only the relatives when the patient could not take part, evaluated the results of surgery. The assessment included questions about how surgery affected 12 different areas: seating posture, reaching, pain, respiration, gastrointestinal function, daily activities, ambulating, school, work, care given, need for rest, and cosmetic appearance (Table 2). The response alternatives were: very positive, positive, unchanged, negative, very negative. The results are presented as positive (very positive and positive), unchanged, or negative (very negative and negative).
Cerebral palsy	24	
Rett's syndrome	9	
Injury of spinal cord	4	
Aicardis syndrome	1	
Charcot-Marie-Tooth	1	
Encephalopathy	1	
Microcephaly	1	
<i>Lower motor neuron lesion</i>	37	
Myelomeningocele	23	
Spinal muscular atrophy	9	
Poliomyelitis	3	
Central core disease	1	
Guillian-Barré syndrome	1	
<i>Myopathies</i>	10	
Myopathies	5	
Muscle dystrophy	4	
Duchenne and Becker		
Arthrogryposis	1	
<i>Congenital abnormalities</i>	6	
Malformation, multiple	3	
Chromosomal abnormalities	2	
Prader Willi's syndrome	1	

Table 2 The set of instruments developed by Samuelsson et al. [19] for use pre- and postoperatively with paralytic scoliosis patients, including general information, activities and function, and follow-up questionnaire

Variables	Pre-operative	Post-operative
General information		
Personal data	X	
Diagnoses	X	
Patient's/relative's motive for surgery	X	
Mental status	X	
Persistent skin discoloration	X	X
Ambulating/wheelchair use	X	X
Occupation	X	
Understands/does not understand verbal instructions	X	
Brace	X	X
Activities and function		
Sitting balance 1–7 points [16]	X	X
Weight distribution EMED system: force (N), pressure (N/cm ²), area (cm ²) [19]	X	X
Angle of scoliosis (Cobb)	X	X
Reaching to defined body parts in sitting position 0–60 points [19]	X	X
Pain estimation [19]	X	X
ADL Barthel 0–100 points [13]	X	
ADL Klein & Bell 0–303 points (was done in those with ADL Barthel score > 30) [11]	X	X
Care given to the patient in terms of physical effort and time consumption [19]	X	X
Time used for resting in h/week [14]	X	X
Seating supports in wheelchair [19]	X	X
Follow-up questionnaire [19]		
Sitting posture		X
Reaching		X
Pain		X
Respiration		X
Gastrointestinal function		X
Daily activities		X
Ambulating		X
School		X
Work		X
Care given		X
Need for rest		X
Cosmetic appearance		X

Statistical methods

To detect any significant differences between the pre- and postoperative results, Wilcoxon's rank sum test was used. For the bivariate analysis of correlation we used Spearman's rank correlation test.

Results

General information activities and function, and dependence

The results for the whole group ($n = 94$) 1 year postoperatively showed reduced numbers of patients with persistent skin discoloration ($P < 0.001$) and fewer patients who used a brace ($P < 0.001$). There were no differences in the use of a wheelchair – all who were ambulating continued to ambulate postoperatively. There were some improvements in the whole group postoperatively according to all levels in the International Classification of Impairments, Disabilities, and Handicaps (ICIDH). At the level of Impairments, there were changes in sitting balance ($P < 0.01$), the angle of scoliosis ($P < 0.001$), and weight distribution ($P < 0.001$), and at the Disability level an improvement was found only in ADL ($P < 0.05$). Improvements at

the Handicap level included a reduction in resting time ($P < 0.001$), and less need for seating supports in the wheelchair ($P < 0.001$) (Table 3).

When the patients were classified into two subgroups, those who understood and those who did not understand verbal instructions, the results were almost the same as for the whole group. The difference was that sitting balance was not significantly improved in the group who understood verbal instructions ($n = 64$). This group had also, as in the whole group, shown improvement at the Disability level for bathing/hygiene. For the group who did not understand verbal instructions ($n = 30$), there was no difference compared with the whole group, and ADL, pain, and reaching were not measured in this group (Table 4). Both groups showed improvements in terms of a reduced number of persistent skin discoloration ($P < 0.05$ and $P < 0.001$) and in the use of a brace ($P < 0.05$ and $P < 0.001$).

When the patients were classified according to the Scoliosis Research Society Classification of diagnoses, the results for the group with upper motor neuron lesions ($n = 41$) showed improvements at the Impairment level for sitting balance, weight distribution on a seating surface, angle of scoliosis, and reaching. There were no changes at the Disability level. There were improvements at the Handicap level in reduced need for rest and seating

Table 3A Comparison of the 1-year postoperative results with the preoperative results ($n = 94$), and their classification according to the International Classification of Impairments (I), Disabilities (D), and Handicaps (H) [23] (p . points, q quartile)

Variables	Preoperative results		Postoperative results		P-value
	Median	(range: q3-q1)	Median	(range: q3-q1)	
Sitting balance 1–7 p. (I)	4	(6–2)	5	(6–3)	0.0035**
Weight distribution when sitting on one seating surface in % (I)	94	(100–75)	71	(90–59)	0.0001***
Angle of scoliosis in ° (Cobb) (I)	80	(98–57)	28	(55–16)	0.0001***
Reaching 0–60 p. (I)	55	(60–42)	56	(60–43)	NS
Pain estimation 1–15 p. (I)	1	(7–1)	1	(2–1)	NS
ADL Klein & Bell 0–303 p. (D)	243	(274–212)	240	(277–211)	NS
Care given (physical effort) 0–30 p. (H)	15	(19–7)	16	(20–8)	NS
Care given (time consumption) in min (H)	150	(320–60)	180	(281–41)	NS
Time used for resting in h/week (H)	2	(7–0)	0	(5–0)	0.0007***
Seating supports in wheelchair 0–7 supports (H)	2	(4–0)	1	(3–0)	0.0001***

* $P < 0.05$; ** $P < 0.01$;
*** $P < 0.001$

Table 3B One-year postoperative evaluation of ADL (activities of daily living) measured by the Klein & Bell index ($n = 39$)

Variables	Preoperative results		Postoperative results		P-value
	Median	(range: q3-q1)	Median	(range: q3-q1)	
Dressing 0–103 p. (D)	95	(103–73)	84	(103–66)	NS
Elimination 0–46 p. (D)	34	(46–27)	38	(43–32)	NS
Mobility 0–68 p. (D)	46	(60–29)	48	(62–29)	NS
Bathing/hygiene 0–56 p. (D)	48	(56–40)	53	(56–43)	0.0199*
Eating 0–30 p. (D)	30	(30–30)	30	(30–30)	NS
Total 0–303 p. (D)	243	(274–212)	240	(277–211)	NS

* $P < 0.05$

Table 5 Comparison of the 1-year postoperative results with the preoperative results in patients with upper motor neuron lesions ($n = 41$)

Variables	Preoperative results		Postoperative results		P-value
	Median	(range: q3-q1)	Median	(range: q3-q1)	
Sitting balance 1–7 p. (I)	3	(6–1)	5	(6–1)	0.016*
Weight distribution when sitting on one seating surface in % (I)	90	(99–74)	72	(90–61)	0.0179*
Angle of scoliosis in ° (Cobb) (I)	70	(87–48)	24	(46–15)	0.0001***
Reaching 0–60 p. (I)	48	(60–24)	56	(60–26)	0.0423*
Pain estimation 1–15 p. (I)	1	(8–1)	1	(3.5–1)	NS
ADL Klein & Bell 0–303 p. (D)	243	(275–197)	228	(272–211)	NS
Care given (physical effort) 0–30 p. (H)	18	(20–14)	18	(23–12)	0.0321*
Care given (time consumption) in min (H)	253	(368–113)	260	(325–135)	NS
Time used for resting in h/week (H)	7	(14–3)	5	(7–1.5)	0.002**
Seating supports in wheelchair 0–7 supports (H)	3	(5–1)	2	(3–1)	0.0021**

* $P < 0.05$; ** $P < 0.01$;*** $P < 0.001$

In the analysis of the groups with upper and lower motor neuron lesions, myopathies, and congenital abnormalities, the results showed the same pattern as in the whole group, except that the groups with upper motor neuron lesions, including both those who understood and those who did not understand verbal instructions, and the patients with congenital abnormalities assessed daily activities as positive and the myopathies assessed reaching as negative.

An analysis was done of the correlation between the patients'/relatives' subjective assessments in the follow-up questionnaire and the differences between pre- and postoperative results of function, activities, and dependence. In the whole group ($n = 94$) there were few correlations between the objective results and the subjective assessments.

The subjective assessments of pain were related to and corresponded with the objective results for the same area ($r = 0.50$; $P < 0.001$), and the same pattern was found in need for rest ($r = 0.46$; $P < 0.001$). The objectively evaluated time used for resting was correlated to work ($r = 0.79$; $P < 0.05$), but this included only nine patients.

Patients' motives for surgery in relation to results of the follow-up questionnaire

Most of the patients/relatives had only one motive, although some of them had two or three motives for surgery. The results for patients'/relatives' motives for surgery showed that patients/relatives most often wanted to obtain improvements in respiration, pain, and seating posture (Table 8).

An analysis of the relationship between the patients'/relatives' motives for surgery and the postoperative subjective evaluation in the follow-up questionnaire of the effect on similar areas was done. Seating posture was

ranked as positive in between 81 and 94% of cases, irrespective of the preoperatively stated motive for surgery. In general, the ratings in the other areas on the questionnaire were higher if the area corresponded to the motive for surgery indicated preoperatively.

Discussion

Several authors have reported the importance of surgical correction in paralytic scoliosis in order to maintain an upright position and to preserve the patient's level of function [2, 8, 9, 17].

In a previous prospective study of surgical correction in patients with paralytic scoliosis, Osebold et al. [18] reported unchanged daily activities and ambulating, but improved sitting balance in 40 patients with myelomeningocele, surgically corrected with Harrington instrumentation, where almost half of this group also had an anterior fusion. Berned-Müller et al. [3] reported poor results in motor skills and unchanged daily activities in 14 patients with myelomeningocele, corrected using Harrington instrumentation. They also reported a decrease in ambulating ability due to contractures in hips, worsened by straightening of the lumbar lordosis.

The present study has demonstrated some improvements postoperatively at the Impairment and Handicap levels, while the functional level was mostly maintained. Reaching, pain estimation, care given, and activities of daily living (ADL) did not change. These results are in line with previous studies, but our study included a larger sample.

Askin et al. [2] reported unchanged daily activities, locomotion, and sitting balance postoperatively in a group of 20 patients surgically corrected with either Harrington instrumentation or the Galveston technique. The study included patients with different diagnoses and different in-

Table 6 Comparison of the 1-year postoperative results with the preoperative results in patients with upper motor neuron lesions who were able to understand verbal instructions ($n = 15$) and in patients with upper motor neuron lesion who were *not* able to understand verbal instructions ($n = 26$)

Variables	Understand verbal instructions			Not understand verbal instructions		
	Preoperative results		Postoperative results	Preoperative results		Postoperative results
	Median	(range: q3-q1)	Median (range: q3-q1)	Median	(range: q3-q1)	Median (range: q3-q1)
Sitting balance 1-7 p. (I)	4	(7-1)	6 (7-3)	2	(6-1)	3 (6-1)
Weight distribution when sitting on one seating surface in % (I)	82	(97-68)	69 (74-61)	91	(100-75)	83 (91-61)
Angle of scoliosis in ° (Cobb) (I)	62	(85-45)	30 (45-15)	71	(102-57)	22 (46-11)
Reaching 0-60 p. (I)	48	(60-24)	56 (60-26)	-	-	-
Pain estimation 1-15 p. (I)	1	(8-1)	1 (4-1)	-	-	-
ADL Klein & Bell 0-303 p. (D)	243	(275-197)	228 (272-211)	-	-	-
Care given (physical effort) 0-30 p. (H)	11	(17-0)	11 (17-6)	19	(20-18)	21 (24-18)
Care given (time consumption) in min (H)	87	(203-3)	120 (190-30)	340	(410-259)	288 (360-265)
Time used for resting in h/week (H)	4	(5-0)	1 (4-0)	10	(20-7)	7 (11-3)
Seating supports in wheelchair 0-7 supports (H)	1	(4-0)	1 (3-0)	4	(5-1)	3 (4-1)

$$**P < 0.05; **P < 0.01; ***P < 0.001$$

tellectual function, and showed that the mentally retarded patients had the same pattern, but with lower scores than the others. The authors do not explain how ADL was assessed in the mentally retarded group. It is obvious that patients with paralytic scoliosis always represent different diagnoses and different intellectual function, which limits the possibility of analysing them as a group.

When assessing at the Disability level, the patient's co-operation is needed. It is not possible to ask patients who do not understand verbal instructions to perform activities or to estimate the subjective values of pain [12]. In this study there were a lot of missing values when trying to describe these patients as a group. Disability cannot be measured properly in subgroups who do not understand verbal instructions. For those who are able to understand verbal instructions, the preoperative levels are already high and no improvements can be expected in these subareas. The results concerning change at the Impairment and Handicap levels were similar in the whole group and the two subgroups. Time used for resting and seating supports in the wheelchair were significantly reduced in both groups. Sitting balance was significantly improved for those who were not able to understand verbal instructions, and the same tendency was seen in the other group, although it was not significant. The postoperative level for both groups was the same, and weight distribution was changed in both groups. In a study dealing with idiopathic and neuromuscular scoliosis, Smith and Emans [21] reported that for normal weight distribution in a sitting position in individuals without deformities, 60% of the weight was on one ischial tuberosity. In the group who are not able to understand verbal instructions, the postoperative results are still poor according to Smith and Emans, and there may be a risk for pressure sores/skin discoloration. This risk may also be influenced by better sitting balance and the patient's increased time sitting in a wheelchair during the day.

In the analysis of upper motor neuron lesions, where the patients were divided into subgroups, we found that the group who understood verbal instructions showed improvements at the Impairment level, and that the group who did not understand verbal instructions showed improvements at both the Impairment and Handicap levels.

The group with lower motor neuron lesions was homogeneous with respect to understanding verbal instructions. As previously described, improvements can be seen at the Impairment level and also one small improvement at the Handicap level. The patients already had a good functional level to start with at the Impairment, Disability and Handicap levels. Nevertheless, no change constitutes a good result, indicating that the patients are on the same functional level postoperatively, since without surgical correction the patient's deformity would probably lead to progression.

Since the group is not homogeneous at all the defined levels and the distribution is wide, it is important to

Table 7 Comparison of the 1-year postoperative results with the preoperative results in patients with lower motor neuron lesions ($n = 37$)

Variables	Preoperative results		Postoperative results		
	Median	(range: q3-q1)	Median	(range: q3-q1)	<i>P</i> -value
Sitting balance 1–7 p. (I)	4	(6–3)	5	(6–3)	NS
Weight distribution when sitting on one seating surface in % (I)	99	(100–76)	71	(90–58)	0.0124*
Angle of scoliosis in ° (Cobb) (I)	92	(111–68)	36	(63–19)	0.0001***
Reaching 0–60 p. (I)	57	(60–49)	57	(59–48)	NS
Pain estimation 1–15 p. (I)	1	(6–1)	1	(1–1)	NS
ADL Klein & Bell 0–303 p. (D)	245	(272–209)	252	(277–205)	NS
Care given (physical effort) 0–30 p. (H)	14	(18–5)	13	(19–5)	NS
Care given (time consumption) in min (H)	125	(243–46)	95	(245–20)	NS
Time used for resting in h/week (H)	0	(1–0)	0	(0–0)	NS
Seating supports in wheelchair 0–7 supports (H)	1	(3–0)	1	(2–0)	0.004**

* $P < 0.05$; ** $P < 0.01$;

*** $P < 0.001$

Fig. 1 Assessments of patients/relatives, reported as percentages, concerning how surgery affected 12 areas, each classified according to the WHO International Classification of Impairments (I), Disabilities (D) and Handicaps (H) [23], 1 year after surgery ($n = 94$)

Table 8 Patients'/relatives' motives for surgery, for the whole group, for those who understand versus those who do not understand verbal instructions, and by diagnosis (1 upper motor neuron lesions, 1 : 1 upper motor neuron lesions who understand verbal in-

structions, 1 : 2 upper motor neuron lesions who do not understand verbal instructions, 2 lower motor neuron lesions, 3 myopathies, 4 congenital abnormalities)

Motive	Whole group (<i>n</i> = 100)	Under-stands (<i>n</i> = 69)	Does not understand (<i>n</i> = 31)	Diagnosis					
				1 (<i>n</i> = 45)	1 : 1 (<i>n</i> = 16)	1 : 2 (<i>n</i> = 29)	2 (<i>n</i> = 39)	3 (<i>n</i> = 10)	4 (<i>n</i> = 6)
Predetermined areas									
Respiration	29	22	7	9	4	5	12	5	5
Pain	21	17	4	8	4	4	9	4	4
Sitting posture	49	35	14	6	12	14	16	7	7
Cosmetic appearance	11	11	0	4	4	0	6	0	0
Open questions									
Communication	5	3	2	5	3	2	0	0	0
Care given	4	1	3	4	1	3	0	0	0
Get rid of brace	9	7	2	4	2	2	4	1	1
Strength in spine	7	4	3	3	0	3	2	1	1
Better balance	7	3	4	4	1	3	1	0	0
Gastrointestinal	6	4	2	1	0	1	3	1	1
Progression	13	7	6	7	1	6	5	1	1
Persistent skin discoloration	1	1	0	0	0	0	1	0	0

analyse whether certain patients gain or lose function. For example, one patient with spinal muscular atrophy reported that after the surgical correction he no longer needed to support himself with his arms and hands, and was now able to drive the tractor on the family farm.

Patients with myelomeningocele have poor sensibility in their lower extremities, and they constitute a risk group for development of pressure sores. The results showed that 5 of 37 patients still had persistent skin discoloration and that, despite better weight distribution, the distribution was not even, and this is a risk factor according to Smith and Emans [21].

The myopathies with progressive Duchenne muscular dystrophy (DMD) represented an exception compared to other groups. They became worse at reaching the mouth, because the spinal fusion made the collapsing spine straighter and longer with an erect position, this in the context of a weak upper extremity. This is important information for the patient and relatives when considering early instrumentation and fusion, which has been advocated in patients with DMD [17] in an attempt to prevent progression of deformity and prolong sitting independence.

However, the groups with myopathies and congenital abnormalities are small and are not homogeneous within groups, so it is therefore difficult to draw any general conclusions from the results.

Improved seating posture was an overall motive for surgery on the part of the patients/relatives, and the results from the questionnaire demonstrated that sitting posture and cosmetic considerations were the most positively assessed factors and that the expectations of the patients/relatives concerning the effect of surgery were partly fulfilled. Assessed improvement in ability to sit was also confirmed by Dias et al. [7] in a telephone interview with 22 caregivers of children who had undergone surgical correction. Askin et al. [2] reported that 75% of patients/caregivers were delighted with the positive postoperative cosmetic appearance. Despite the objective overall improvement in the present study in sitting balance and the pa-

tients'/relatives' positive subjective assessment of sitting posture, we found no correlation between those variables. The reason for this may be that sitting balance is not the only variable that explains subjectively assessed sitting posture and, especially for the group who did not understand verbal instructions, it was difficult to get objectively assessed variables. The analysis is limited, since only areas in which change occurs were included, and the questions related to effect of surgery. In this study it was shown that the instruments at the Disability level were not applicable to all subgroups, which highlights the difficulty of assessing activities, particularly in the group of patients who do not understand verbal instructions. There might have been an improvement in carrying out measurements if a scale with more alternatives had been used. Another approach would be to individualise the different activities and use a goal attainment scale as described by Streiner and Norman [22].

Conclusion

The patients with paralytic scoliosis maintained or improved their function and level of independence in terms of Impairments and Handicaps 1 year after surgery. The subjectively assessed results obtained from the patients/relatives also showed a positive outcome of surgery.

Weight distribution on a seating surface was improved, but still uneven, and with respect to better sitting balance and increased time sitting in the wheelchair, this can involve a risk for pressure sores and needs further investigation.

When introducing outcomes including the Disability level, one must take the importance of homogeneity in the groups into consideration.

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References

1. Aprin H, Bowen R, MacEwen D, Hall JE (1982) Spine fusion in patients with spinal muscular atrophy. *J Bone Joint Surg [Am]* 64: 1179–1187
2. Askin GN, Hallet R, Hare N, Webb JK (1997) The outcome of scoliosis surgery in the severely physically handicapped child. An objective and subjective assessment. *Spine* 22: 44–50
3. Berned-Müller E, Nordwall A, von Wendt L (1992) Influence of surgical treatment of scoliosis in children with spina bifida on ambulation and motoric skills. *Acta Paediatr* 18: 173–176
4. Bradford DS (1987) Neuromuscular spinal deformity. In: Bradford DS, Lonstein JE, Moe JH, Ogilvie JW, Winter RB (eds) *Moe's textbook of scoliosis and other spinal deformities*, 2nd edn. Saunders, Philadelphia, pp 271–305
5. Brown JC, Zeller JL, Swank SM, Furumasa J, Warath SL (1989) Surgical and functional results of spine fusion in spinal muscular atrophy. *Spine* 14: 763–770
6. Cassidy C, Craig CL, Perry A, Karlin LI, Goldberg MJ (1994) A reassessment of spinal stabilization in severe cerebral palsy. *J Pediatr Orthop* 14: 731–739
7. Dias RC, Miller F, Dabney K, Lipton G, Temple T (1996) Surgical correction of spinal deformity using a unit rod in children with cerebral palsy. *J Pediatr Orthop* 16: 734–740
8. Dubousset J, Herring JA, Shufflebarger HJ (1989) The crankshaft phenomenon. *J Pediatr Orthop* 9: 541–550
9. Hensinger RN, MacEwen GD (1976) Spinal deformity associated with heritable neurological condition spinal muscular atrophy, Friedreich's ataxia, familial dysautonomia, and Charcot-Marie-Tooth disease. *J Bone Joint Surg [Am]* 58: 13–24

10. Hibbs RA (1924) A report of fifty-nine cases of scoliosis treated by the fusion operation. *Bone Joint Surg* 6: 15
11. Klein RM, Bell B (1982) Self-care skills: behavioural measurement with the Klein & Bell ADL scale. *Arch Phys Med Rehabil* 63: 335–338
12. Larsson E-L, Aaro S, Ahlinder P, Öberg B (1998) Evaluation of activities and function in patients with paralytic scoliosis. *Eur Spine J* 7: 294–301
13. Mahoney FI, Barthel DW (1965) Functional evaluation: the Barthel index. *M State Med J* 14: 61–65
14. Meguro K, Ueda M, Yamaguchi T, et al (1990) Disturbance in daily sleep/wake pattern in patients with cognitive impairment and decreased daily activity *J Am Geriatr Soc* 38: 1176–1182
15. Miller A, Temple T, Miller F (1996) Impact of orthoses on the rate of scoliosis progression in children with cerebral palsy. *J Pediatr Orthop* 16: 332–335
16. Mulcahy CM, Pountney TE, Nelham RL, Green EM, Billinton GD (1988) Adaptive seating for motor handicap: problems, a solution, assessment and prescription. *Br J Occup Ther* 51: 347–352
17. Murbarak SJ (1993) Spinal fusion in Duchenne muscular dystrophy – fixation and fusion to the sacropelvis? *J Pediatr Orthop* 13: 752–757
18. Osebold WR, Mayfield JK, Winter RB, Moe JH (1982) Surgical treatment of paralytic scoliosis associated with myelomeningocele. *J Bone Joint Surg [Am]* 64: 841–856
19. Samuelsson K, Larsson E-L, Normelli H, Öberg B, Aaro S, Tropp H (1996) Development of an instrument for clinical evaluation after surgery for neuromuscular scoliosis. *Eur Spine J* 5: 400–406
20. Shook JE, Lubicky J (1991) Paralytic spinal deformity. Scoliosis. In: Bridwell KH, DeWald RL (eds) *The textbook of spinal surgery*. Lippincott, Philadelphia, pp 279–322
21. Smith RM, Emans JB (1992) Sitting balance in spinal deformity. *Spine* 17: 1103–1109
22. Streiner DL, Norman GR (1995) *Health measurement scales. A practical guide to their development and use*. Oxford Medical Publications, Oxford, pp 45–53
23. WHO (1993) *The International Classification of Impairments, Disabilities and Handicaps (ICIDH)*. New edition of the manual of classification relating to the consequences of disease. WHO, Geneva