

## Associations between orthopaedic findings, ambulation and health-related quality of life in children with myelomeningocele

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### Abstract

**Purpose** Modern principles for treatment of patients with myelomeningocele include early closure of the neural tube defect, neurosurgical treatment of hydrocephalus and treatment aimed at minimizing contractures and joint dislocations. The aim is to achieve a better survival rate and a better quality of life (QOL). Better ambulatory function is thought to improve the management of activities of daily living. This study focused on evaluating which factors might affect ambulation, function and health-related QOL in children with myelomeningocele.

**Methods** Thirty-eight patients with neurological deficit from myelomeningocele were examined in an unbiased follow-up. This included a physical examination using validated methods for ambulatory function and neuromuscular

status, chart reviews and evaluation of radiographs in terms of hip dislocation and spine deformity. The Pediatric Evaluation of Disability Inventory (PEDI) was used to measure mobility, self-care and social function, and the Child Health Questionnaire (CHQ-PF50) was used to measure QOL.

**Results** Muscle function class, quadriceps strength, spasticity in hip and/or knee joint muscles and hip flexion contracture as well as the ambulatory level all affected functional mobility as well as self-care/PEDI. Patients with hip dislocation, spinal deformity or those who were mentally retarded also had significantly worse functional mobility. Besides being affected by the severity of the neurological lesion, self-care/PEDI was significantly impaired by hip flexion contracture and absence of functional ambulation. General health-related QOL was significantly lower in this patient group than for US norms. Nonambulatory and mentally retarded patients had a significantly lower physical function of their QOL (CHQ).

**Conclusions** The severity of the disease, i.e. reduced muscle strength and occurrence of spasticity around hip/knee, affected ambulation, functional mobility and self-care. Acquired deformities (hip dislocation and spine deformity) affected functional ambulation only. Patients with reduced functional mobility and self-care experienced lower physical QOL. Children with myelomeningocele had significantly reduced QOL compared to healthy individuals.

**Keywords** Spina bifida · Ambulation · Physical factors · Functional ability · Quality of life

### Introduction

With modern principles for treatment of patients with myelomeningocele; i.e. early closure of the neural tube defect and neurosurgical treatment of hydrocephalus, which

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have been used since the beginning of the 1970s, the early mortality rate has dropped. Also, morbidity due to kidney disease has been substantially reduced with the introduction of clean intermittent catheterization for management of neurogenic bladder. Individuals with myelomeningocele should now be expected to survive into adulthood. The emphasis of medical care is shifting to enhancing quality of life (QOL) and promoting independence [1].

The ability of an individual to care for oneself is highly dependent on the functional mobility of that individual. Achieving independent ambulation has traditionally been the goal of orthopaedic treatment and is the aspect of mobility that receives the most attention. It is assumed that maximizing ambulatory function will promote future independent living and higher QOL; however, the relationship of ambulation to other functional skills and QOL has not been fully explored.

The purpose of this study was to investigate which factors might negatively influence ambulatory function and explore the connection between ambulation, function and health-related QOL. Increased knowledge might help to predict future ambulation, thereby improving treatment planning. It might also answer the question of whether better ambulatory function renders a better QOL in children with myelomeningocele.

## Materials and methods

### Patients

Patients were recruited from two centres with well-established treatment for children with myelomeningocele: the Walter Reed Army Medical Center, Washington DC, and the Kennedy Krieger Institute, Baltimore, MD, USA. All patients fulfilled the inclusion criteria: (1) there was a myelomeningocele that had been surgically treated in the neonatal period; (2) there was the presence of some motor deficit regardless of level of spina bifida; (3) age was between 3 and 16 years at the time of inclusion in the study; (4) treatment occurred at one of the two centres at the time of the study; (5) acceptance and ability to participate in the study. Eleven children at the Walter Reed Army Medical Center and 27 patients at Kennedy Krieger Institute completed the study, so the total number of patients included was 38.

### Physical examination

Two physiotherapists who had not taken part in the treatment of the patients, either one of the authors (Å.B) or a physiotherapist especially trained by her for this study,

performed the physical examinations. All methods used for physical examination, Table 1, have been validated, shown to be reliable and previously used in published studies.

The children were divided into five classes of muscle function according to their lower limb muscle strength [2, 3]. The development of muscle function classes was based on a comparison of six different classification systems with the aim to facilitate comparison of treatment results with special consideration of ambulation [4]. This classification was found to be reliable and work well both for clinical judgments and for outcome documentation in the Swedish studies of these authors.

Presence of spasticity in the lower limb muscles was documented. Spasticity was considered present if the patient showed one or more of the following signs: static abnormal posturing with increased muscle tension despite full passive motion of a joint, velocity-dependent catch response, resistance throughout passive range of motion or clonus. For the purpose of statistical analyses, patients were grouped into two groups; no spasticity of lower limbs or around ankle joint, or occurrence of spasticity around the knee and/or hip joints.

The range of motion of the lower limb joints was assessed according to the recommendations of the American Academy of Orthopaedic Surgeons [8]. Joint contractures of the lower limb were defined for the ankle if  $\geq 15$  degrees of fixed equinus foot, for knee if  $\geq 20$  degrees of extension contracture or for hip if  $\geq 20$  degrees of extension contracture were observed. Hip flexion contracture was measured using Thomas' test [9]. All contractures were measured in the supine position.

The ambulatory function was assessed using the criteria of Hoffer et al. [5], with some modifications [3]. For the purpose of statistical analyses, patients were grouped together as either "ambulatory", i.e. ambulating but using wheelchair only for long distances outdoors (Grade 1–3) or "nonambulatory", i.e. using wheelchair both in- and outdoors (Grade 4–6).

Key functional capabilities and performance were measured by the Pediatric Evaluation of Disability Inventory (PEDI, Table 1) [6]. In a parent-report questionnaire, administered in an interview session, the parents were asked questions that reflect the three content domains: self-care (73 questions), mobility (59 questions) and social function (65 questions). All children had scaled scores that provide an estimate of the level of capability regardless of age and can be used for the evaluation of children over the age of 7.6 years if their functional abilities fall below those expected of 7.5-year-old children with no disabilities. The scaled scores range from 0 to 100, where 100 reflects the best function. The PEDI can be used for the comparison of children of all age groups; however, no norm scales exist for children above the age

**Table 1** Definitions and descriptions of used gradings of muscle function classes, functional ambulation, PEDI and CHQ-PF50

Questionnaire	Purpose	Grading/ description	Definition/description
Muscle function class [2–4]	Muscle strength of the lower limbs	I	Weakness of intrinsic foot muscles; Good to normal foot plantarflexion (grade 4–5)
		II	Fair or less foot plantarflexion (grade 3 or less); fair or better knee flexion (grade 3 or more) Poor to fair or better hip extension and/or hip abduction (grade 2–3 or more)
		III	Good to normal hip flexion and knee extension (grade 4–5); fair or less knee flexion (grade 3 or less) Trace of hip extension, hip abduction and below-knee muscles
		IV	No knee extension activity; poor or less hip flexion (grade 2 or less); fair or good pelvic elevation
		V	No muscle activity in the lower limbs; no pelvic elevation
Functional ambulation, modification [3, 5]	Ambulatory function	Ambulatory patients	1 Community ambulation, no orthoses (sometimes insoles) Ability to keep up with peers when walking outdoors according to the criteria of the PEDI
			2 Community ambulation with need of orthoses, no need for walking aid. Wheelchair used only for long distances outdoors
			3 Household ambulation with need for orthoses and need for walking aid. Wheelchair used only outdoors (and for long distances indoors)
		Nonambulatory patients	4 Household ambulation with orthoses and walking aid. Wheelchair used both in- and outdoors
			5 Nonfunctional ambulation. Ambulation during therapy, in school and for limited time at home. Wheelchair use for mobility
			6 Nonambulators
PEDI [6]	Measures capability and performance of functional activities	Mobility	Score calculated from 59 questions concerning the individual's mobility
		Self-care	Score calculated from 73 questions concerning the individual's self-care
		Social function	Score calculated from 65 questions concerning the individual's social function
CHQ-PF50 [7]	General HRQL measures physical, social and mental components.	PF	Physical Functioning: presence and extent of physical limitations
		RP	Role/Social-Physical: presence and extent of limitations in school work or activities with friends as result of physical health
		GH	General Health: overall health and illness
		BP	Bodily Pain/Discomfort: indicator of physical health
		FA	Family-limitations in Activities: frequency of disruption to "usual" family activities
		REB	Role/Social-Emotional/Behavioural: capturing limitations in psycho-social well-being, due to emotional or behavioural difficulties
		PT	Parental Impact/Time: amount of <i>limitations in personal time</i> experienced by the parent due to child's health, emotional well-being, attention of learning disabilities, child's ability to get along with others, and general behaviour
		PE	Parental Impact/Emotional: amount of <i>distress</i> experienced by the parent due to child's health, emotional well-being, attention of learning disabilities, child's ability to get along with others, and general behaviour
		SE	Self Esteem: social confidence, school abilities, self-regard
		MH	Mental Health: frequency of both negative and positive states
		BE	General Behaviour: overt behaviour
		FC	Family-Cohesion: family relationships in general
		Summary scores	Physical (PhS) and Psychosocial (PsS) summary score, mean norm value is 50, SD 10 (US)

of 7.5 years. The questionnaire has previously been used in populations with myelomeningocele.

### Review of charts and radiographs

Chart reviews for all examined patients were performed to collect information on previous medical and surgical treatments as well as complication(s) of the disease and other concomitant diseases.

Radiographs of the hips and spine were located, and if older than one year a new examination was undertaken. The prevalence of hip dislocation was noted. Four patients did not have new radiography performed; however, a judgment of the occurrence of hip dislocation was possible through clinical evaluation in combination with older radiographs.

For the spine, the existence of scoliosis or kyphosis, including Cobb angle [10] and apex of the curve, was noted. Radiographs of the spine older than one year were used in five patients, who did not show any clinical signs of scoliosis at physical examination.

### Questionnaires

Parents underwent a nurse interview session. Information on mental retardation was collected from the Individual Education Program (IEP) of the specific patient. The IEP is a written document that is developed for each public school child who is eligible for special education and is a mandated requirement of the “Individuals with Disabilities Education Act” of the United States.

A validated questionnaire on general health-related QOL, the Child Health Questionnaire-50 Parent Form (CHQ-PF50) [7], was answered by the parent (Table 1). It measures the physical, social and mental components of health-related QOL. The questionnaire has 12 subscales and each is scored from 0 to 100, where 100 reflects the best function. It also offers an opportunity to calculate physical and psychosocial summary scores (PhS and PsS respectively), each with a mean norm value of 50. The CHQ has been validated in many countries and used for a variety of different diseases, including myelomeningocele [11]. The national norms for a population sample of US children have been used for comparison with the results from our study group.

### Ethics

The Institutional Review Boards at the Johns Hopkins Hospital, Baltimore, MD and the Walter Reed Army

Medical Center, Washington, DC, United States, approved the study.

### Statistical methods

Distributions of variables are given as means, standard deviations (SD) and ranges.

When comparing two groups, Fisher’s exact test for dichotomous variables, chi-square test for nonordered categorical variables and Mantel–Haenszel chi-square test for ordered categorical variables were used. Mann–Whitney’s *U*-test was used for continuous variables.

The Student’s *t*-test was used for comparison with the US norm values.

The Spearman rank correlation coefficient ( $r_s$ ) was used for correlation analyses.

All tests were two-tailed and conducted at the 5% significance level.

### Results

Age distribution, gender, racial origin and neurological findings of the patients are found in Table 2. Thirty-five (92%) patients had a shunt, of which 69% had been revised twice or more. Nine patients had mild or moderate mental retardation (none had severe or profound mental retardation).

### Neuromuscular status and orthopaedic manifestations

Information on the musculoskeletal status of the patients is shown in Table 3, divided up by functional ambulation. Nineteen children were ambulatory and nineteen non-ambulatory.

The only child in age group 3–4 years was ambulatory (Grade 2 acc. to Hoffer). Among the eight children aged between five and eight years at examination, six were ambulatory and two were nonambulatory. One of these two nonambulatory children was 5.4 years at examination, had muscle function class III, hip contractures of more than 20°, bilateral hip dislocations and a scoliotic curve of 35°. The other nonambulatory child, 7.6 years of age, had muscle function class V, spasticity around hip and knee joints with hip and knee contractures of more than 20° as well as equinus, unilateral hip dislocation and a kyphoscoliotic spinal curvature (95° thoracic kyphosis and 26° scoliosis).

Patients with weak quadriceps muscles (strength of 1–3), less lower limb muscle strength or spasticity around the hip and/or knee joints were significantly more often

**Table 2** Age, gender, racial origin and neurological findings in 38 patients with myelomeningocele [Mean (SD)/Median (Min–Max) or *n*(%)]

	All patients ( <i>n</i> = 38)
Age at examination	
Mean (SD)/median (Min–Max)	11.1 (3.4) 11.3 (3.8–16.8)
Age group	
3–4	1
5–7	8
8–10	9
11–12	6
13–15	10
16–18	4
Gender	
Male	20 (52.6%)
Female	18 (47.4%)
Racial origin of parents	
Caucasian	26 (68.4%)
Non-Caucasian	11 (31.6%)
Neurology	
Arnold Chiari malformation	35 (92.1%)
Type 1	8 (22.9%)
Type 2	23 (65.7%)
Type unknown	4 (11.4%)
Shunt	35 (92.1%)
Shunt revisions	
Revised 0–1 times	11 (31.4%)
Revised ≥2 times	24 (68.6%)
Tethering	16 (42.1%)
Syrinx	8 (21.1%)
Mental retardation <sup>a</sup>	
No	25 (73.5%)
Yes	9 (26.5%)

<sup>a</sup> Information not available on all patients

nonambulators. All ambulatory patients had quadriceps muscle strength of 4–5.

Eleven percent of ambulatory patients were mentally retarded compared to 44% of nonambulators, *ns* (*p* = 0.076). Occurrence of shunt, shunt revisions twice or more, tethering or syrinx was not more frequently seen among the nonambulators.

Patients with hip or knee flexion contracture of 20° or more or equinus of at least 15°, approximately one-third of the patients, were significantly more often nonambulators. Thirteen patients (34%) had uni- or bilateral hip dislocations. 17/19 (89%) of the ambulatory patients had located hips, while 11/13 (84%) patients with hip dislocation(s) were nonambulatory, *p* = 0.0052.

Nine patients had a scoliotic spine with a curvature of at least 30° (Cobb) at the present examination, and another two patients were previously surgically treated for a scoliotic deformity, and now had a straight spine. Twelve patients had a major sagittal deformity (either lordosis or kyphosis of 70° or more), of which two also had a scoliotic deformity. Out of the total 21 patients with spine deformity, only five patients, all with muscle function class I–III and without spasticity or joint contracture, were ambulators. Out of the 16 nonambulators with spinal deformity, four patients had muscle function class I–III and 12 had class IV–V, but all 16 had either spasticity around hip/knee, joint contractures or hip dislocation.

PEDI/functional skills

Table 4 depicts functional skills as measured by PEDI. All three domains; mobility, self-care and social ability were tested against factors found to affect the ability to ambulate (Table 3). Testing was also performed for ambulatory versus nonambulatory patients and for normally intelligent versus mentally retarded children.

Functional mobility and functional self-care both were significantly better in patients with better lower limb muscle strength, quadriceps muscle strength of grade 4–5, no spasticity around the hip or knee joint, less than 20° hip flexion contracture or in ambulatory patients.

Function due to mobility also was significantly deteriorated by occurrence of hip dislocation, spine deformity and by being mentally retarded. None of the factors studied affected the social ability domain significantly.

Quality of life

Quality of life as measured with the CHQ was significantly reduced in children with myelomeningocele as compared to the US normative sample in all subscales but self esteem (SE), general behaviour (BE) and family cohesion (FC), Fig. 1. The summary scores were also significantly lower than the US norms, 31.5 vs. 53.0 (*p* < 0.0001) for the physical component summary score (PhS) and 47.9 vs. 51.2 (*p* = 0.036) for the psychosocial component summary score (PsS), Fig. 2.

Comparisons of QOL for subgroups of the study population in terms of ambulatory function or mental retardation are presented in Fig. 3. Physical functioning was significantly affected by ambulatory function (PF for ambulatory patients 59.1 vs. 40.6 for nonambulators, *p* = 0.020) as well as by mental retardation (PF for normally intelligent children 58.7 vs. 27.8 for mentally retarded children, *p* = 0.010).

**Table 3** Characteristics of 38 patients with myelomeningocele divided up by ambulatory function [n(%)]

	All patients n = 38	Ambulatory function <sup>a</sup>		p-Value
		Ambulatory patients n = 19	Non-ambulatory patients n = 19	
<b>Neuromuscular status</b>				
Neurological level of deficiency = muscle function class				
I	5 (13.1%)	5 (26.3%)	0	<0.0001
II	12 (31.6%)	11 (57.9%)	1 (5.3%)	
III	7 (18.5%)	3 (15.8%)	4 (21.1%)	
IV	8 (21.0%)	0 (0%)	8 (42.1%)	
V	6 (15.8%)	0	6 (31.6%)	
Muscle strength knee extensors, grade				
0–3	14 (36.8%)	0	14 (73.7%)	<0.0001
4–5	24 (63.2%)	19 (100%)	5 (26.3%)	
Spasticity				
None or in ankle joint muscles only	28 (73.7%)	19 (100%)	9 (47.4%)	0.0004
Spasticity in hip and/or knee joint muscles	10 (26.3%)	0	10 (52.6%)	
<b>Neurological problems</b>				
Mental retardation				
No	25 (73.5%)	16 (88.9%)	9 (56.2%)	ns (0.076)
Yes	9 (26.5%)	2 (11.1%)	7 (43.8%)	
<b>Orthopaedic findings</b>				
Hip flexion contracture $\geq 20^\circ$	12 (32.4%)	1 (5.3%)	11 (61.1%)	0.0007
Knee flexion contracture $\geq 20^\circ$	12 (31.6%)	0	12 (63.1%)	<0.0001
Equinus $\geq 15^\circ$	10 (26.3%)	2 (10.5%)	8 (42.1%)	0.042
Status of the hip				
Located	25 (65.8%)	17 (89.5%)	8 (42.1%)	0.0052
Dislocated, uni- or bilateral	13 (34.2%)	2 (10.5%)	11 (57.9%)	
Spine deformity <sup>b</sup>	21 (55.3%)	5 (26.3%)	16 (84.2%)	0.0008

<sup>a</sup> Comparison between ambulatory (wheelchair use only for long distances outdoors, Grade 1–3 of functional ambulation acc. to Hoffer) and nonambulatory patients (wheelchair use both in-and outdoors, Grade 4–6 of functional ambulation acc. to Hoffer)

<sup>b</sup> Defined as scoliosis with Cobb angle  $\geq 30^\circ$  and/or major sagittal deformity (either lordosis or kyphosis of  $70^\circ$  or more) OR a previously surgically treated spine deformity with no curvature at present examination

Correlations were found between the physical performance (PF) of the QOL and all three domains of PEDI, self-care ( $r = 0.51$ ,  $p = 0.0012$ ), mobility ( $r = 0.60$ ,  $p < 0.0001$ ) and social ability ( $r = 0.32$ ,  $p = 0.049$ ). These results were also reflected in the physical summary score (PhS score), which was correlated with self-care ( $r = 0.34$ ,  $p = 0.035$ ) and mobility ( $r = 0.38$ ,  $p = 0.020$ ).

## Discussion

A considerable number of orthopaedic procedures as well as physiotherapeutic efforts and orthotic device production are performed with the aim to keep the lower extremities straight and hip joints located. Whether these efforts really have a positive impact from the patient's perspective and whether a higher level of ambulation really is correlated to

a better QOL provide the focus for this paper. We also wanted to delineate between internal or external factors that might affect the ability of growing individuals with myelomeningocele to have independent mobility.

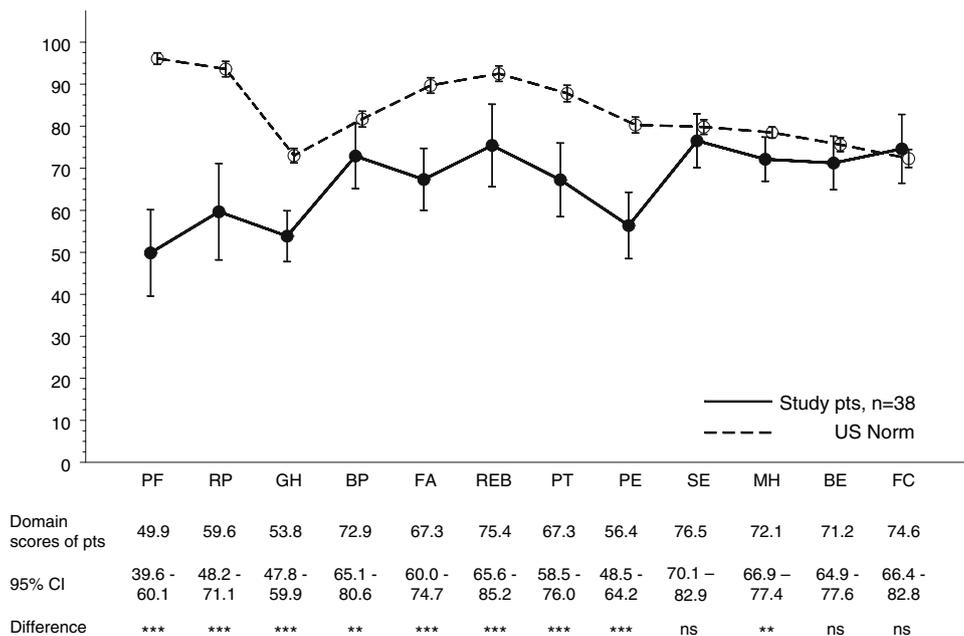
It might be questionable as to whether the younger children in the study had acquired their final ambulatory function. In the eight children less than eight years of age, six were already ambulators. The other two (nonambulatory) children had low muscle strength in combination with contractures and deformities to an extent that will make future ambulation doubtful. Therefore, we considered the grading of the ambulatory function reliable enough for analysis.

The neurological lesion itself did affect whether the patient was an ambulator or not. Patients with stronger lower limb muscles were significantly more often ambulating and all ambulatory patients had quadriceps muscle

**Table 4** PEDI/functional skill domains in 38 patients with myelomeningocele, divided up according to subgroups of clinical status [Mean (SD)/ (Min–Max)]

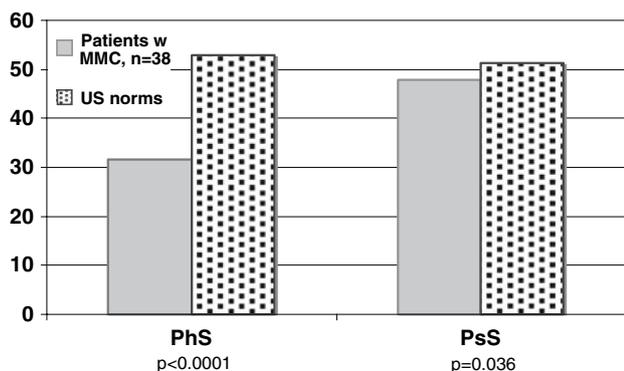
	<i>n</i>	Mobility	<i>p</i> -Value	Self-care	<i>p</i> -Value	Social ability	<i>p</i> -Value
All patients	38	44.3 (13.5) (11.0–59.0)		61.1 (12.3) (15.0–73.0)		61.4 (4.8) (41.0–65.0)	
Patient subgroups according to							
Neurological level of deficiency = muscle function class							
I–III	24	52.1 (7.4) (28.0–59.0)	<0.0001	65.1 (7.3) (48.0–73.0)	0.033	62.3 (3.2) (55.0–65.0)	ns (0.21)
IV–V	14	31.1 (11.0) (11.0–49.0)		54.2 (15.9) (15.0–73.0)		59.9 (6.5) (41.0–65.0)	
Muscle strength knee extensors, grade							
0–3	14	31.1 (11.0) (11.0–49.0)	<0.0001	54.2 (15.9) (15.0–73.0)	0.033	59.9 (6.5) (41.0–65.0)	ns (0.21)
4–5	24	52.1 (7.4) (28.0–59.0)		65.1 (7.3) (48.0–73.0)		62.3 (3.2) (55.0–65.0)	
Spasticity in hip and/or knee joint muscles							
No	28	49.7 (9.8) (16.0–59.0)	0.0001	65.0 (8.7) (40.0–73.0)	0.0013	61.5 (5.3) (41.0–65.0)	ns (0.25)
Yes	10	29.3 (10.9) (11.0–50.0)		50.2 (14.6) (15.0–68.0)		61.1 (3.2) (56.0–65.0)	
Hip flexion contracture $\geq 20^\circ$							
No	25	49.6 (11.6) (16.0–59.0)	0.0004	65.3 (9.0) (40.0–73.0)	0.0026	62.2 (5.0) (41.0–65.0)	ns (0.070)
Yes	12	34.2 (11.6) (11.0–50.0)		53.1 (14.5) (15.0–70.0)		59.9 (4.3) (52.0–65.0)	
Status of the hip							
Located	25	47.7 (13.7) (11.0–59.0)	0.0061	62.3 (12.8) (15.0–73.0)	ns (0.28)	62.2 (3.3) (52.0–65.0)	ns (0.34)
Dislocated	13	37.8 (10.8) (16.0–50.0)		58.8 (11.3) (40.0–73.0)		59.8 (6.6) (41.0–65.0)	
Spine deformity <sup>a</sup>							
No	24	51.7 (9.8) 56.0 (28.0–59.0)	0.0007	65.1 (9.2) 71.0 (46.0–73.0)	ns (0.056)	62.9 (2.8) 64.0 (55.0–65.0)	ns (0.055)
Yes	14	38.4 (13.3) 40.0 (11.0–58.0)		57.8 (13.7) 62.0 (15.0–73.0)		60.2 (5.7) 63.0 (41.0–65.0)	
Functional ambulation							
Ambulatory patients	19	54.6 (4.4) (46.0–59.0)	<0.0001	66.6 (5.8) (52.0–73.0)	0.012	62.7 (2.8) (55.0–65.0)	ns (0.11)
Nonambulatory patients	19	34.1 (11.5) (11.0–50.0)		55.5 (14.5) (15.0–73.0)		60.1 (5.9) (41.0–65.0)	
Mental retardation							
No	25	47.9 (11.9) 52.0 (20.0–59.0)	0.032	65.0 (7.8) 66.0 (47.0–73.0)	ns (0.063)	62.8 (2.4) 63.0 (56.0–65.0)	ns (0.092)
Yes	9	39.1 (11.8) 39.0 (16.0–51.0)		56.9 (11.1) 54.0 (40.0–73.0)		58.0 (7.9) 62.0 (41.0–65.0)	

<sup>a</sup> Defined as scoliosis with Cobb angle  $\geq 30^\circ$  and/or major sagittal deformity (either lordosis or kyphosis of  $70^\circ$  or more) OR a previously surgically treated spine deformity with no curvature at present examination



**Fig. 1** CHQ-PF50: results for subscales for patients with myelomeningocele versus national US norms. Mean with 95% CI. QOL as measured with the CHQ was significantly reduced in children with myelomeningocele as compared to the US normative sample in all subscales but self esteem (SE), general behaviour (BE) and family cohesion (FC). Subscales of CHQ: PF = Physical Functioning;

RP = Role/Social-Physical; GH = General Health; BP = Bodily Pain; FA = Family-Activities; REB = Role/Social-Emotional/Behavioural; PT = Parental Impact-Time; PE = Parental Impact-Emotional; SE = Self Esteem; MH = Mental Health; BE = General Behavior; FC = Family-Cohesion



**Fig. 2** CHQ-PF50: summary scores for patients with myelomeningocele vs. national US norms. Both physical and psychosocial summary scores were significantly lower among patients with myelomeningocele than US norms. PhS = physical summary score, PsS = psychosocial summary score

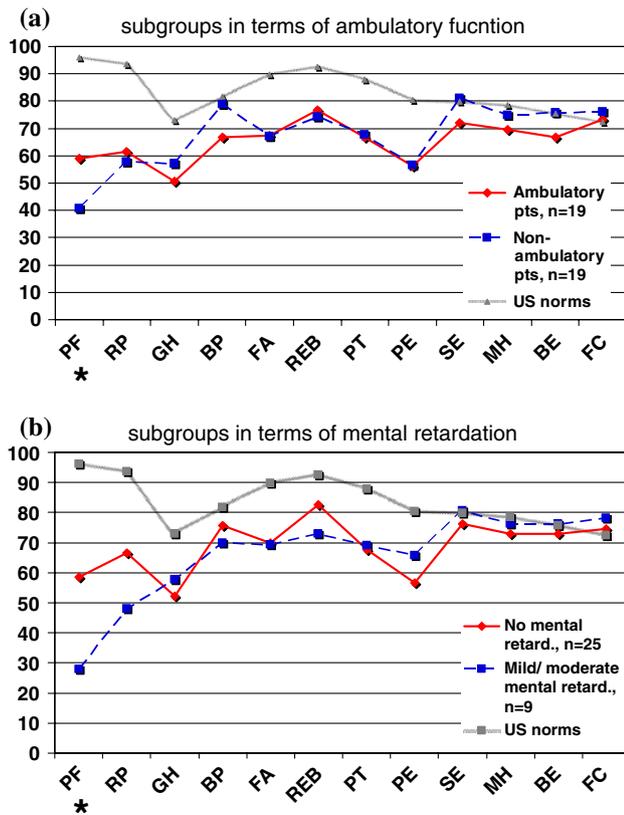
strengths of 4–5. The role of the knee extensor muscle strength in ambulation was in accordance with earlier studies by McDonald et al. [12], Mazur and Menelaus [13] and Huff and Ramsey [14], who all highlighted the importance of a quadriceps muscle strength grade of 4–5 for walking with ankle-foot orthoses.

Existence of spasticity around the hip and knee joints as well as spine deformity were deleterious for ambulation.

The spasticity itself might also be a contributing factor to contractures of hips and knees, which were strongly associated with the loss of walking ability. These findings are in accordance with earlier publications [3, 5, 15–18]. Patients with hip dislocation were also significantly more often nonambulators than those with contained hips.

Even though only two out of nine (22%) mentally retarded patients were ambulators compared to 16 out of 25 (89%) normally intelligent children, this difference did not reach statistical difference. This might be due to the small number of patients in the study. However, in the measurements on physical function, patients with mental retardation showed significantly worse results both in terms of functional mobility domain/PEDI and physical function (PF) of the CHQ.

Functional mobility as well as self-care was significantly affected by the severity of the neurological symptoms; muscle function class, quadriceps strength and spasticity in hip and/or knee joint muscles. Furthermore, functional mobility was also significantly affected by secondary orthopaedic deformities/conditions. This discloses possible relationships between secondary orthopaedic deformities and mobility, which need further studies to verification. Larger patient groups with less heterogeneity in terms of age, physical function, neurological lesion and associated orthopaedic deformities as well as regression analysis will



**Fig. 3a–b** CHQ-PF50: Comparisons of QOL for subgroups of the study population. Subscales of CHQ: PF = Physical Functioning; RP = Role/Social-Physical; GH = General Health; BP = Bodily Pain; FA = Family-Activities; REB = Role/Social-Emotional/Behavioural; PT = Parental Impact-Time; PE = Parental Impact-Emotional; SE = Self Esteem; MH = Mental Health; BE = General Behaviour; FC = Family-Cohesion. **a** Subgroups in terms of ambulatory function. Physical functioning was significantly lower in nonambulatory patients (ambulatory patients 59.1 vs. 40.6 for nonambulators,  $p = 0.020$ ). No other significant differences of subscales or summary scores were found between the two groups. US norm scale added for reference. **b** Subgroups in terms of mental retardation. Physical functioning was significantly lower in patients with mental retardation (patients without mental retardation 58.7 vs. 27.8 for patients with mental retardation,  $p = 0.010$ ). No other significant differences of subscales or summary scores were found between the two groups. US norm scale added for reference

be needed to obtain scientific proof; these prerequisites could not be obtained for this study group.

Correlations between PEDI scores and walking ability, intelligence or independence in activities of daily living agree with earlier publications [19]. However, none of these abovementioned factors affected the social ability. As one may expect, social abilities can develop despite impaired functional mobility or a reduced level of self-care.

General (health-related) QOL in this patient group was significantly lower than for the US norms. QOL in terms of PF was associated with the ambulatory function and mental retardation, i.e. ambulatory and nonretarded patients had

less limitation. PF was also correlated to all three PEDI domains, namely self-care, mobility and social ability. This fact strongly shows the importance of functional abilities in achieving a better (physical) QOL. This is in line with earlier publications: Rendeli et al. [11] found that disability was inversely correlated to the physical aspect of QOL, and Schoenmakers et al. [20] found that being independent with regard to mobility was the most important determinant of health-related QOL. They also found that mental status, having no contractures in lower extremities and normal strength in knee extensors were the most important factors in mobility independence. The present findings, as well as the earlier literature [1, 20, 21], support surgical correction of musculoskeletal deformities, treatment of tethered cord syndrome, and they point out the importance of maximizing functional independence, good muscle strength and mental ability to achieve a better QOL.

The goal for treatment of children with spina bifida has always been to give these children a life that is as good as it can be. However, the means of doing this has been under debate for many years. One area of debate is that of ambulatory function and is the question of whether to prevent hip dislocation. In this study, we found that physical QOL was significantly reduced and the mental well-being reduced in terms of effect on family and parents, but not for the child itself. These negative effects on QOL can thus already be seen during childhood. Therefore, it is important to prevent deterioration and to enhance improvement in physical abilities during childhood.

If improvement is achieved, the physical abilities and thereby the physical QOL may reach a higher level until maturity, which will also decrease the risk of “negative over-spill” into mental well-being. The patient will then be given the best opportunities to handle his or her own life when entering adulthood.

### Conclusion

Ambulatory ability as well as functional mobility was significantly affected by the severity of the neurological symptoms; muscle function class, quadriceps strength and spasticity in hip and/or knee joint muscles. They were also significantly affected by musculoskeletal complications such as hip flexion contracture, hip dislocation and spine deformity, all of which are consequences that orthopaedic treatment aims to reduce.

The general (health-related) QOL for children with myelomeningocele was significantly lower than for the US norms. Functional physical abilities were shown to be of importance for achieving a better QOL. Having a higher level of functional mobility, self-care and social ability improved the physical QOL.

In order to clarify the existence of relationships between secondary orthopaedic deformities and mobility, further studies are needed. Until this has been done, a judgment cannot be made as to whether preventive treatments for musculoskeletal complications affect the QOL of the patient.

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## References

- Bier JA, Prince A, Tremont M, Msall M (2005) Medical, functional, and social determinants of health-related quality of life in individuals with myelomeningocele. *Dev Med Child Neurol* 47:609–612
- Hislop H (1995) Daniel's and Worthingham's muscle testing: techniques of manual examination. Saunders, Philadelphia, PA
- Bartonek A, Saraste H (2001) Factors influencing ambulation in myelomeningocele: a cross-sectional study. *Dev Med Child Neurol* 43:253–260
- Bartonek A, Saraste H, Knutson LM (1999) Comparison of different systems to classify the neurological level of lesion in patients with myelomeningocele. *Dev Med Child Neurol* 41:796–805
- Hoffer MM, Feiwell E, Perry R, Perry J, Bonnett C (1973) Functional ambulation in patients with myelomeningocele. *J Bone Joint Surg Am* 55:137–148
- Haley SM, Coster WJ, Ludlow LH, Haltiwanger JT, Andrellos PJ (1992) Pediatric evaluation of disability inventory (PEDI). PEDI Research Group, New England Medical Center Hospitals, Boston, MA
- Landgraf JM, Abetz L, Ware JE (1999) Child's health questionnaire. In: The CHQ user's manual. HealthAct, Boston, MA
- American Academy of Orthopaedic Surgeons (1988) Joint motion. Method of measuring and recording. Churchill Livingstone, Edinburgh, UK
- Bartlett MD, Wolf LS, Shurtleff DB, Stahell LT (1985) Hip flexion contractures: a comparison of measurement methods. *Arch Phys Med Rehabil* 66:620–625
- Cobb J (1948) Technique for study of scoliosis. In: Blount WP, AAOS (ed) AAOS instructional course lectures. J. Edwards, Inc., Ann Arbor, MI, pp 261–275
- Rendeli C, Ausili E, Tabacco F, Caliendo P, Aprile I, Tonali P, Salvaggio E, Padua L (2005) Assessment of health status in children with spina bifida. *Spinal Cord* 43:230–235
- McDonald CM, Jaffe KM, Mosca VS, Shurtleff DB (1991) Ambulatory outcome of children with myelomeningocele: effect of lower-extremity muscle strength. *Dev Med Child Neurol* 33:482–490
- Mazur JM, Menelaus MB (1991) Neurologic status of spina bifida patients and the orthopedic surgeon. *Clin Orthop Relat Res March* (264):54–64
- Huff CW, Ramsey PL (1978) Myelodysplasia. The influence of the quadriceps and hip abductor muscles on ambulatory function and stability of the hip. *J Bone Joint Surg Am* 60:432–443
- Bartonek A, Saraste H, Samuelsson L, Skoog M (1999) Ambulation in patients with myelomeningocele: a 12-year follow-up. *J Pediatr Orthop* 19:202–206
- Bartonek A, Gutierrez EM, Haglund-Akerlind Y, Saraste H (2005) The influence of spasticity in the lower limb muscles on gait pattern in children with sacral to mid-lumbar myelomeningocele: a gait analysis study. *Gait Posture* 22:10–25
- Asher M, Olson J (1983) Factors affecting the ambulatory status of patients with spina bifida cystica. *J Bone Joint Surg Am* 65:350–356
- De Souza LJ, Carroll N (1976) Ambulation of the braced myelomeningocele patient. *J Bone Joint Surg Am* 58:1112–1118
- Tsai PY, Yang TF, Chan RC, Huang PH, Wong TT (2002) Functional investigation in children with spina bifida—measured by the Pediatric Evaluation of Disability Inventory (PEDI). *Childs Nerv Syst* 18:48–53
- Schoenmakers MA, Uiterwaal CS, Gulmans VA, Gooskens RH, Helders PJ (2005) Determinants of functional independence and quality of life in children with spina bifida. *Clin Rehabil* 19:677–685
- Selber P, Dias L (1998) Sacral-level myelomeningocele: long term outcome in adults. *J Pediatr Orthop* 18:423–427