

Determinants of functional independence and quality of life in children with spina bifida: a physical therapy perspective.

MAGC Schoenmakers¹
CSPM Uiterwaal²
VAM Gulmans¹
RHJM Gooskens³
PJM Helders¹

- ¹ Department of Pediatric Physical Therapy, Wilhelmina Children's Hospital, University Medical Center, Utrecht, the Netherlands
- ² Julius Center for Health Sciences and Primary Care, Wilhelmina Children's Hospital, University Medical Center, Utrecht, the Netherlands
- ³ Rudolf Magnus Institute for Neuroscience, Department of Neurology and Neurosurgery, Wilhelmina Children's Hospital, University Medical Center, Utrecht, the Netherlands

Submitted for publication



ABSTRACT

OBJECTIVES: To investigate determinants of functional independence and study which functional abilities were determinants for 'health related quality of life' (HRQL) in children with myelomeningocele.

DESIGN: Cross-sectional study by means of clinical assessment, 'disability' measurement, and questionnaires.

SETTING: Outpatient spina bifida clinic at a university hospital.

PARTICIPANTS: 122 children with myelomeningocele. Mean age 7.9; range 1-18 years.

MAIN OUTCOME MEASURES: Functional independence as measured by the 'Pediatric Evaluation of Disability Inventory' (PEDI), and quality of life as measured by the 'Spina Bifida HRQL Questionnaire'. Uni- and multivariate logistic regression models were used to investigate factors that were determinants for these outcomes. Results were expressed as odds ratios (OR) and 95% confidence intervals (CI).

RESULTS: Lesion level below L3 (OR: 0.4, CI: 0.1-1.0), mental status of IQ \geq 80 (OR: 4.2, CI: 1.2-14.9), having no contractures in lower extremities (OR: 3.4, CI: 1.3-8.8), and having normal strength of knee extensor muscles (OR: 4.1, CI: 1.4-11.5), were most strongly associated with independence in self-care. Mental status (OR: 16.1, CI: 2.8-93.9), having no contractures in lower extremities (OR 1.5, CI: 1.4-5.3), and normal strength in knee extensors (OR: 11.0, CI: 1.3-97.0), were the most important determinants for independence in mobility. Concerning functional abilities, being independent with regard to mobility was the most important determinant for HRQL (OR: 5.3, CI: 1.6-17.4).

CONCLUSIONS: In children with myelomeningocele, good muscle strength, mental ability and being independent in mobility, appeared to be much more important for daily life function and quality of life than other medical indicators of the disorder. This information is of clinical significance in planning a comprehensive and realistic rehabilitation program.



INTRODUCTION

Spina bifida is a complex congenital disorder that represents a broad spectrum of neural tube defects, including spina bifida aperta and spina bifida occulta. The incidence varies in different parts of the world, but is generally 0.4-1.0 per 1000 live births in the USA [1], and a higher incidence is found in Northern Europe. In myelomeningocele (MMC), the most common and more serious form of spina bifida aperta, both brain and spinal cord are often malformed, resulting in hydrocephalus, Chiari II malformation, muscle weakness and lack of sensation in lower extremities, bladder and bowel incontinence, as well as cognitive dysfunction [2-5].

The functional consequences of spina bifida can be displayed using the disablement framework as described by Verbrugge and Jette [6], consisting of four different domains: 'pathology', 'impairments', 'functional limitations' and 'disability'. Several factors might influence functional outcome, such as intra-individual factors (coping, life style), extra-individual factors (medical care, rehabilitation) and risk factors. This model has a lot in common with the most recent International Classification of Functioning, Disability and Health (ICF) designed by the World Health Organization (WHO) [7].

Most studies focus on the relation between 'pathology' and outcome. Hydrocephalus, lesion level and presence of neural tissue in the sac, are considered important factors that influence the functional prognosis of patients with spina bifida [2-5,8,9]. Hunt et al. and Bowman et al. [2,3,5] reviewed complete cohorts of patients with myelomeningocele from birth into adulthood. These important studies reflect the results of treatment that was carried out in the sixties and seventies. As treatment strategies change with time, ongoing research is therefore extremely important.

Physical therapy is a common intervention for children with spina bifida. Traditionally, therapy sessions focussed on improvement of muscle strength, muscle tone, and prevention of contractures, to opti-

mize childhood development and functioning [10]. However, it is not clear how these impairments are related to functional independence and 'health related quality of life'. In various other chronic diseases, 'impairment' parameters do not significantly correlate with functional abilities and 'disability' parameters [11,12].

In this study, we investigate which factors of the 'disability process' domains are the most important determinants for functional independence regarding self-care and mobility in children with myelomeningocele. Regarding HRQL, our main interest was to find out which functional abilities were most important determinants, in order to find sufficient and realistic goals for physical therapy treatment.



METHODS

Patients

The study group consisted of children born between 1981-2000 (aged 1 to 18 years), who have been followed-up in the outpatient spina bifida clinic of the Wilhelmina Children's Hospital. Participants included those with a diagnosis of spina bifida aperta (myelomeningocele). Patients with meningocele and those with cervical (myelo)meningocele, encephalocele, or cord traumas were excluded. Non-Dutch speaking patients were also excluded. One hundred and thirty patients met the inclusion criteria, and 122 were willing to participate in the study (response rate 94%) carried out from January 1999 to September 2001. The Medical Ethics Committee of our hospital approved the protocol of this descriptive, cross-sectional study. Informed consent was obtained from the parents and from the patients themselves if they were older than 12 years of age.

Procedures

From the domain of 'pathology', the investigated determinants for functional independence on self-care and mobility were presence of shunted hydrocephalus, number of shunt revisions, and lesion level. From the 'impairment' domain, intelligence, contractures in lower extremities, muscle-tone, and muscle strength from upper and lower extremities were included. In order to find out which functions from the domain of functional abilities might influence the quality of life in children with myelomeningocele, we included ambulation level and hand function as well as independence in self-care, mobility and social function (caregiver assistance scores of the 'Pediatric Evaluation of Disability Inventory' (PEDI) [1,13,14].

Data were obtained by physical examination, and during a face-to-face interview regarding functional abilities (PEDI-scores) and HRQL. All measurements were performed by the same experienced pediatric physical therapist (MS). Parents were questioned about their educational level.

Measurements with regard to 'pathology'

Information on the presence of shunted hydrocephalus and the number of shunt revisions was obtained from the medical record. Determination of the neurosegmental motor level was based on muscle strength of the lower extremities as described by McDonald et al. [15]. Many different systems are used to classify neurological level in spina bifida patients, while agreement in levels from L3 and downwards is lacking [16]. For the purposes of the analyses, we therefore divided the neurological level into two groups: below L3, and from L3 up to thoracic.

Measurements with regard to 'impairments'

Information on mental status, such as Intelligence Quotients (IQs) (WISC-R) in children older than 4 years, was obtained from the psychological records. IQs ≥ 80 were considered as normal [2,3,5]. Body weight and body height were measured, crown-to-heel for ambulators and upper-arm span for non-ambulators.

Range of motion in lower extremities was measured in a standardized way with a two-legged 360-degree goniometer, and compared to reference values for children [17]. The percentages of range of motion-loss were assessed on a 5-point scale according to Spiegel et al. [18]. Range of motion-loss of 5% or more in one or both extremities was defined as a contracture.



Muscle tone of upper extremities, trunk, and lower extremities was scored on a 5-point scale ranging from severe hypotonia (1), mild hypotonia (2), normal tone (3), mild hypertonia (4), to severe hypertonia (5) as described by Shurtleff [19]. Both mild or severe hypotonia and hypertonia were classified as an abnormal score.

Muscle strength of upper and lower extremities was graded 0-5, according to the standard Manual Muscle Testing (MMT) as described by Daniel's & Worthingham [20]. In children under the age of 5 years, muscle grading on a 6-point scale is not reliable. Therefore, in these patients strength was graded as absent, weak, or full strength as advocated by McDonald et al. [21]. In upper extremities, elbow flexors and hand muscles were tested. In lower extremities, flexors, extensors and abductors of the hip, as well as extensors of the knee and dorsal flexors of the ankle joint were tested, as these muscle groups seem to correlate strongly with ambulation level [15]. Children with full strength or grade 5 were classified as having normal muscle strength.

Measurements regarding functional abilities and quality of life

Ambulatory status was modified from Hoffer et al. [22]. We grouped patients into non-ambulators (completely wheelchair-dependent or walking in therapeutic situations) and functional-ambulators (household or community walkers).

In children older than 4 years, hand function was measured with the Movement Assessment Battery for Children (Movement-ABC) [23]. This instrument has been developed to evaluate gross and fine motor function in children aged 4-12⁺ years. Percentile scores of the child's motor abilities can be compared with a normal age-matched sample of children [23].

To evaluate the level of functional independence we used the caregiver assistance scores of the Dutch adapted version of the PEDI [24,25]. The PEDI evaluates function in terms of *capabilities* (functional skills) and *performance* of what the child actually does in response to the

environment (the amount of caregiver assistance required to accomplish daily tasks) in three domains: self-care, mobility and social function [24]. Reference values are provided for children between 0.5-7.5 years. Normal values are defined in the range of 2 SD (50 ± 20). In children > 7.5 years of age, results were calculated as a scaled score. In healthy children > 7.5 years of age, all functional skills should be mastered leading to a score of 100, which was considered to be normal [25]. We defined patients with normal caregiver-assistance scores as being functional independent. The intra-interviewer and inter-interviewer reliability of the adapted Dutch version of the PEDI are excellent. Intra-class Correlation (ICC) Coefficients were all but one above 0.90. The content validity and construct validity are acceptable [25]. Quality of life was measured with a Dutch-translation of the 'Spina Bifida HRQL Questionnaire' [26]. This instrument is a disease specific and age-related instrument for children (5-12 years) and adolescents (13-20 years) with spina bifida. For children, the questionnaire is used as a proxy report, whereas adolescents complete the questionnaire by themselves [26]. In a reference group of Canadian children with spina bifida, the mean score (SD) was 168 (24); for adolescents the mean score (SD) was 182 (30). Reproducibility was good (ICC = 0.78 for children; 0.96 for adolescents). Internal consistency was good (Cronbach's $\alpha = 0.93$ for children, and 0.94 for adolescents). Construct validity correlations were 0.63 (children) and 0.37 (adolescents) [26]. The 'Spina Bifida HRQL questionnaire' is not yet cross-culturally adapted; therefore the median of our study population was used as cut-off, to determine patients with higher or lower level of quality of life.

Statistical analysis

Descriptive analysis (means, standard deviations; medians, P25-P75) was used to characterize functional independence (PEDI caregiver



assistance scores) and 'health related quality of life' (Spina Bifida HRQL scores).

To identify the most important independent factors associated with the defined outcome parameters (functional independence and HRQL) we first conducted univariate logistic regression analysis. We assessed whether the associations were independent of known confounders. Factors concerning functional independence were adjusted for age and body mass index (BMI). Factors concerning HRQL were adjusted for age, mental status and educational level of parents. With regard to strength of upper and lower extremities we first looked for the muscle group that was most strongly associated with independence in self-care and mobility. This muscle group was used for further analysis as an indicator muscle for lower and upper extremity strength respectively.

Next, in order to find out which was the most important determinant, we used multivariate logistic regression models with all statistically significant factors from the univariate analysis as independent variables, for each outcome parameter. Analysis was performed with the Statistical Package for Social Sciences (SPSS 9.0).

RESULTS

Patient characteristics are listed in Table 1.

Table 1 General data and clinical characteristics of 122 children with myelomeningocele.		
	Number	
Age in years (SD)	7.9	(5.2)
Age distribution:		
- 1-3.9 years (%)	39	(32)
- 4-7.9 years (%)	25	(20)
- 8-12.9 years (%)	33	(27)
- 13-18.0 years (%)	25	(20)
males (%)	54	(44)
Body Mass Index in kg/m ² (SD)	18.4	(4.6)
Educational level of parents:		
- bachelor or university level (%)	47	(38.6)
- lower than bachelor level (%)	75	(61.4)
Shunted hydrocephalus (%)	97	(80)
Shunt revisions:		
- 0 (%)	40	(41)
- 1 (%)	24	(25)
- 2-4 (%)	25	(26)
- more than 4 (%)	8	(8)
Lesion level		
- thoracic (%)	25	(20)
- upper lumbar (L1-L3) (%)	26	(21)
- lower lumbar (L4 –L5) (%)	45	(37)
- sacral (%)	26	(21)

**Table 1** continued.

	Number	
Mental status in children > 4 years ^a : mean IQ (SD)	83.3	(19.8)
Ambulatory status in children > 2.5 years ^b		
- non-ambulant (%)	53	(52)
- functional ambulant (%)	50	(48)
PEDI		
- deviant self care CA-score (%)	85	(69.6)
- deviant mobility CA-score (%)	67	(54.9)
- deviant social function CA-score (%)	32	(26.2)
≤ 7.5 years: - self care mean CA-score (SD)	30.7	(18.0)
- mobility mean CA-score (SD)	27.1	(15.7)
- social function mean CA-score (SD)	43.9	(12.6)
> 7.5 years: - self care mean CA-score (SD)	66.4	(20.5)
- mobility mean CA-score (SD)	88.9	(19.2)
- social function mean CA-score (SD)	86.1	(22.02)
Spina Bifida HRQL questionnaire in children > 5 years ^c		
- ≤ 12 years median score (25-75 th centile)	166	(151-186)
- > 12 years median score (25-75 th centile)	185	(176-198)
Abbreviations: SD = standard deviation, IQ = intelligence quotient, PEDI = Pediatric Evaluation of disability Inventory, CA = caregiver assistance, HRQL = health related quality of life.		
a = number of valid values (n = 83); b: n =103, c: n = 73.		

With regard to the muscle group most strongly associated with independence in self-care, we found hand muscle to contribute statistically significantly (OR: 4.4, CI: 1.5-36.2), whereas elbow flexor muscles did not (OR: 4.6, CI: 0.6-37.8). Independence in mobility was significantly associated with all lower extremity muscles groups: hip flexor muscles (OR: 4.7, CI: 1.7-13.1), hip abductor muscles (OR: 6.3, CI: 1.9-29.2), hip extensor muscles (OR: 4.9, CI: 1.4-17.0), and knee extensor muscles (OR: 4.1, CI: 1.4-11.5), and foot dorsal flexor muscles (OR: 5.5, CI: 12.2-14.2). Multivariate logistic regression modeling indicated that the odds ratio of knee extensor muscle strength remained significant (OR: 4.2, CI: 1.4-15.1), whereas the others did not. For further analyses, hand muscles and knee extensor muscles were used as indicator muscles for upper and lower-extremity muscle strength respectively.

Results regarding functional independence in self-care and mobility are shown in Table 2. The factors from the domains of 'pathology' and 'impairments', that are univariately associated with functional independence in self-care and mobility, after being adjusted for confounders (age, BMI), are shown in the middle column (univariate analysis). The results of analyses, aimed to find out which component from the different domains was independently associated with each of the two outcome parameters for functional independence are presented in the right column (multivariate analysis). Independence in self-care was statistically significantly associated with lesion level, having no contractures, mental status and muscle strength of lower extremities. From the multivariate logistic regression model, none of the odds ratios remained significant. Independence in mobility was significantly associated with hydrocephalus, lesion level, mental status, having no contractures, muscle tone above the cele level, and muscle strength of lower extremities. From the multivariate logistic regression model, the odds ratios of mental status, having no contractures, and muscle strength of lower extremities remained significant.



Table 2 Determinants for functional independence (= normal PEDI scores on caregiver assistance) regarding self-care and mobility.

	Univariate analysis	Multivariate analysis
	odds ratio (95% CI)	odds ratio (95% CI)
SELF-CARE		
<i>Pathology</i>		
- non shunted hydrocephalus	3.59 (0.23 – 10.49)	-
- less than two shunt revisions	1.14 (0.80 – 4.77)	-
- lesion level below L3	0.37 (0.14 – 0.98)	1.31 (0.23 – 7.31)
<i>Impairments</i>		
- mental status (IQ ≥ 80)	4.23 (1.20 – 14.94)	2.73 (0.75 – 9.57)
- no contractures lower extremities	3.39 (1.31 – 8.75)	2.43 (0.85 – 6.99)
- normal muscle tone above cele level	2.77 (0.94 – 8.15)	-
- normal strength of knee extensors muscles	4.06 (1.44 – 11.46)	3.62 (0.63 – 20.83)
- normal strength of hand muscles	4.39 (0.53 – 36.23)	-
MOBILITY		
<i>Pathology</i>		
- non shunted hydrocephalus	2.76 (1.01 – 7.56)	0.99 (0.20 – 5.00)
- less than two shunt revisions	1.36 (0.61 – 3.03)	-
- lesion level below L3	0.12 (0.04 – 0.31)	0.34 (0.05 – 2.61)
<i>Impairments</i>		
- mental status (IQ ≥ 80)	16.89 (4.24 – 67.28)	16.09 (2.76 – 93.93)
- no contractures lower extremities	3.66 (1.48 – 7.66)	1.45 (1.40 – 5.32)
- normal muscle tone above cele level	8.27 (2.73 – 25.06)	0.32 (0.05 – 1.98)
- normal strength of knee extensors muscles	13.37 (4.63 – 38.67)	10.99 (1.25 – 96.97)
- normal strength of hand muscles	35.06 (0.00 – 59.72)	-

All values that are statistically significant are indicated in bold.

Abbreviations: CI = confidence interval, IQ = intelligence quotient, PEDI = Pediatric Evaluation of Disability Inventory

Results regarding quality of life are shown in Table 3. Factors that remained significant after being adjusted for age, mental status, and educational level of the parents are shown in the column of univariate analysis. HRQL was significantly associated with being functional ambulant (household or community walker), and being independent with regard to mobility. From the multivariate logistic regression model, the odds ratio of being independent in mobility remained significant.

Table 3 Functional abilities associated with 'health related quality of life' (HRQL).

	Univariate analysis	Multivariate analysis
	odds ratio (95% CI)	odds ratio (95% CI)
<i>Functional abilities</i>		
- functional ambulant	4.52 (1.01 – 20.15)	2.17 (0.76 – 6.22)
- normal hand function	7.29 (0.29 – 179.77)	-
- independent in self care (normal CA-scores)	0.25 (0.33 – 1.90)	-
- independent in mobility (normal CA-scores)	3.61 (1.33 – 40.13)	5.26 (1.59 – 17.41)
- independent in social function (normal CA-scores)	3.34 (0.47 – 23.79)	-
All values that are statistically significant are indicated in bold.		
Abbreviations: CI = confidence interval, CA = caregiver assistance.		



DISCUSSION

From our study it is suggested that lesion level, mental status, contractures, and muscle strength of lower extremities particularly determine independence in self-care. All these factors are mutually dependent. Mental status, having no contractures, and muscle strength of lower extremities were the most important determinants for functional independence in mobility. With regard to HRQL, being independent in mobility seemed to contribute more to HRQL, than other functional abilities such as being independent in self-care or being wheelchair-dependent.

Knowledge of determinants of independence is of utmost importance for treatment in order to “focus on realistic goals rather than wasting efforts on attempting the impossible” [2]. The relation between ‘pathology’ (hydrocephalus, Chiari II malformation, lesion level) and outcome in patients with myelomeningocele has been studied extensively [2-5,8], but few has been published on the impact of ‘impairments’ and ‘functional limitations’. Dahl et. al. [27] studied self-care skills in 35 young children with myelomeningocele. They found many of these children to be slow in the development of independence in self-care; 60% needed moderate or maximal caregiver assistance. Poor self-care skills were related to both the child’s function above and below the level of the cele. Low intelligence, hypotonia above the level of the cele, being non-ambulant and poor executive function seemed to be significant risk factors for poor self-care skills. These results are partially in agreement with our data. We found poor self-care skills in 69% of our patients. We also found mental status to be an important determinant for independence in self-care, but muscle tone above the level was not. Nevertheless, motor function below the cele level appeared to be also important for complete independence in self-care. With regard to mobility we found that variables in the ‘pathology’ domain such as the presence of shunted hydrocephalus, the number of shunt revisions and lesion level seemed to be of less importance.

Several researchers have published information on the relation between lesion level, muscle strength and ambulatory status [22,28,29]. The level of neurological lesion and associated strength of lower extremity muscles are the most important factors influencing ambulatory status. However, there is a large discrepancy in ambulatory outcomes across different lesion groups. McDonald et al. [29] studied the relationship between patterns of strength and ambulation in 291 children with myelomeningocele. They reported that m. iliopsoas strength was found to be the best predictor of ambulation with the m. quadriceps, m. tibialis anterior, and mm. glutei also contributing significantly. In our study the same muscle groups, particularly knee extensors, appeared to be significantly associated with independence in mobility, including transfers, indoor and outdoor ambulation, and climbing stairs. This information is of clinical relevance for rehabilitation interventions. We agree with McDonald et al. [29] when they recommend the use of specific patterns of lower-extremity muscle strength, rather than lesion level, to predict walking abilities. We found lesion level to be of less importance, than muscle strength of lower extremities with regard to independence in mobility.

Our finding of contractures being an important determinant for mobility is in agreement with findings of other authors [28,30]. It is questionable to what extent contractures can be prevented or treated by physical therapy. Often, surgical procedures are needed to restore the congenital muscle imbalance. Further research is needed to investigate whether physical therapy programs designed to increase power of unaffected muscles, lead to functional improvement [31]. In future research it is important to identify relevant strength thresholds for specific functional activities in order to create realistic rehabilitation programs.

Regarding HRQL, our main interest was to find out which functional abilities were most important determinants for HRQL, in order to find sufficient and realistic goals for physical therapy treatment. From current literature there is growing evidence that physical functioning of patients with spina bifida, is related to HRQL [1,32,33]. Pit-Ten Cate



et al. [32] showed that children with spina bifida had lower scores on quality of life with regard to self-care, incontinence and mobility, compared to children with hydrocephalus alone. Lesion level, type of spina bifida, and presence of hydrocephalus itself were not significantly related to quality of life. McCormick et al. [13] studied the impact of caring for a child with spina bifida on family function. The impact on the family was related less to the clinical diagnosis, than to the child's functioning at home. We found that being independent in mobility seemed to contribute more to HRQL, than other functional abilities such as being independent in self-care or being wheelchair-dependent.

Our study has some limitations. Regarding HRQL, we focussed on 'disability' aspects only, but from psychosocial literature it is suggested that personal and environmental factors might be of great importance [13,14,32]. Kirpalani et al. [1] studied the influence of parental hope on HRQL in children and adolescents with spina bifida. Although physical functioning explained a significant proportion of variance in HRQL, parental hope explained an additional 19 to 24% of the variance. The study of Pit-Ten Cate et al. [32] also confirmed that other factors than disability are associated with quality of life. They found that severity of the condition and family resources independently predicted quality of life. Therefore, it is extremely difficult for health professionals to predict future health related quality of life and furthermore it is always from the outsider's perspective. We agree with Kirpalani et al. [1] that patients and families are the best judges of their own quality of life. For future research, a close collaboration between behavioral and medical researchers is needed, as it will assist in the search for better ways to prevent disability [31].

When interpreting our findings it should be considered that in children with spina bifida, physical and intellectual disabilities often become more manifest when these children grow older. Realities in adulthood might be totally different from childhood [2]. Therefore, long-term follow-up remains important.

Our observations, that being independent in mobility appeared to contribute more to HRQL than being wheelchair-dependent, should be kept in mind when planning a realistic rehabilitation program regarding ambulation. Strategies to persist walking abilities with aids is a common goal in physical therapy, but the use of these aids should never disadvantage the patient in terms of independence.



CONCLUSIONS

Our findings showed that parameters on the level of ‘impairments’ were significantly associated with ‘disability’. We found that good muscle strength, having no contractures, and mental ability appeared to be much more important for daily life function of spina bifida patients than other medical indicators of disorder. Being independent in mobility appeared to contribute more to HRQL, than being independent in self-care or being wheelchair-dependent.

ACKNOWLEDGEMENTS

We would like to thank the families and children who participated in this study. We also acknowledge Carla Tims for her technical support in the data-management of this study.

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