Functional independence among young adults with spina bifida, in relation to hydrocephalus and level of lesion

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Knowledge about the level of functional independence that can be expected in adulthood might support decisions on the treatment of newborn infants with spina bifida. This study determined functional independence among young adults with spina bifida and its relationships with pathological characteristics known from birth (hydrocephalus and level of lesion). Data were collected from medical records and by physical examination. Functional independence was assessed on six domains (self-care, sphincter control, transfers, locomotion, communication, and social cognition) using the Functional Independence Measure (FIM). Participants were 165 patients with spina bifida (69 males, 96 females; age range 16 to 25y, mean 20y 9mo [SD 2.9]; 117 with hydrocephalus). Patients without hydrocephalus were independent for all FIM domains except sphincter control, as were patients with hydrocephalus with a lesion level below L2. Most patients with hydrocephalus and a lesion at L2 or above were dependent as regards sphincter control (98%), locomotion (79%), and self-care (54%), and quite a few needed support in transfers (38%), social cognition (29%), and communication (15%).

Spina bifida is a complex congenital disorder, the consequences of which are related to the level of lesion and the presence of hydrocephalus (Verhoef et al. 2004). Because of medical improvements in recent times, more patients born with spina bifida survive childhood and grow up into adulthood (Hunt 1999, Bowman et al. 2001). For this group, being independent in activities of daily living is an important prerequisite for independent living and social participation. It is important for patients and their families, as well as for caregivers, to have reasonable expectations about future functional independence because such knowledge can form the basis of the treatment decision of newborn infants with spina bifida.

Independence of young adults with spina bifida in activities of daily living has been described in few studies. Börjeson and Lagergren (1990) conducted interviews among 26 adolescents with myelomeningocoele and concluded that two-thirds of them were independent in dressing, bathing, and hygiene. Buran et al. (2004) found that most of their 66 adolescents with spina bifida were independent according to the Functional Independence Measure for children (WeeFIM; Msall et al. 1994) in the areas of eating, grooming, upper and lower body dressing, locomotion, and most transfers. The lowest scores on the WeeFIM were found for the items, bladder and bowel management, toilet transfer, and stair mobility. Andren and Grimby (2000) found that most of their 20 adult patients with spina bifida were completely independent as regards eating and the social and cognitive items of the Functional Independence Measure (FIM), and showed modified independence for the other items. However, these studies did not describe relations between functional independence and disease characteristics.

Several studies found a relation between level of lesion and independence in activities of daily living of children and young adults with spina bifida (Msall et al. 1994, Staal et al. 1996, Padua et al. 2002, Schoenmakers 2003). Higher levels of independence in activities of daily living, have been found in children and adults with meningocoele than in patients with myelomeningocoele (Kalucy et al. 1996).

However, these studies suffer from several limitations, such as using small numbers of patients, including broad age ranges, not using standardized measures of independence, or not describing subgroups at all. For this reason, they do not provide sufficient evidence of the prognosis of functional independence in newborn patients with spina bifida to support decision making.

The aim of the present study was to describe the degree of independence of young adults with spina bifida in terms of activities of daily living for the total group as well as for subgroups of patients, based on differences in the presence of hydrocephalus and the level of lesion.

Methods

PARTICIPANTS

Young adults with different types of spina bifida (aperta and occulta; International Classification of Diseases [ICD; World Health Organization 2003] codes 741 and 756.17) aged between 16 and 25 years and living in the Netherlands were included. Excluded were patients with spina bifida occulta without any neurological loss, non-Dutch-speaking patients, or patients with comorbidity independently causing more physical and/or cognitive problems than the spina bifida itself.

Patients were recruited from 11 of the 12 multidisciplinary spina bifida teams in the Netherlands. The Dutch Association for Patients with Spina Bifida also invited members to participate. In addition, rehabilitation centres, special housing facilities, and special schools were approached to find potential participants.

Table I: Scoring levels of Functional Independence Measure

Level		*Description*
No helper		
7	Complete independence	All tasks described as making up the activity are typically performed safely, without modification, assistive devices or aids, and within a reasonable amount of time
6	Modified independence	One of the following may be true: activity requires an assistive device; the activity takes more than reasonable time, or there are safety (risk) considerations
Helper		
5	Supervision or set-up	Patient requires no more help than standby, cueing or coaxing, without physical contact, or, helper sets up needed items or applies orthoses
4	Minimal contact assistance	Patient requires no more help than touching, and expends 75% or more of effort
3	Moderate assistance	Patient requires more help than touching, or expends half (50%) or more (up to 75%) of effort
2	Maximal assistance	Patient expends less than 50% of effort, but at least 25%
1	Total assistance	Patient expends less than 25% of effort, patient does not perform activity, or assistance of two helpers is needed

Table II: Hoffer classification (with an extra category of normal ambulators; Hoffer et al. 1973)

Classification	*Description*
Non-ambulators	Patients who are wheelchair bound
Non-functional ambulators	Patients who are able to walk in a therapy session, but use their wheelchair afterwards
Household ambulators	Patients who walk only indoors and with apparatus. They may use wheelchair for some indoor activities at home and school and for all activities in community
Community ambulators	Patients who walk indoors and outdoors for most of their activities may need crutches, braces or both. They use a wheelchair only for long trips out of community
Normal ambulators	Patients without any mobility problems, not using any devices for mobility at all

INSTRUMENTS

Data were collected by means of an interview and a standardized physical examination (peformed by MV).

Hydrocephalus was defined as either having a shunt at the time of the physical examination or having had one previously.

In accordance with the International Standards for Neurological and Functional Classification of Spinal Cord Injury (Maynard et al. 1997), the level of lesion was defined as the lowest completely unimpaired dermatome level on both sides measured with sensitivity to pin prick and light touch. Sensory level was used instead of motor level because it could be performed in a more standardized way, took little time, and was feasible in all patients (despite contractures, arthrodeses, or other medical problems). Patients were categorized into three subgroups based on the level of lesion: high (L2 and above), middle (L3 to L5), and low (S1 and below; Shurtleff et al. 1975, Swank and Dias 1992, Staal et al. 1996).

The FIM was used to describe the degree of independence in activities of daily living. The FIM consists of 18 items in six domains of physical and cognitive functioning: self-care, sphincter control, transfers, locomotion (FIM motor score), communication, and social cognition (FIM cognition score). Each item is scored on a 7-point scale (Maynard et al. 1997) varying from 'complete independence' (level 7) to 'total assistance' (level 1; Table I). The FIM can be used as an observational instrument or as a questionnaire with a very strong correlation between both scores (Karamehmetoglu et al. 1997). In the present study, the FIM items were rated based on observations during the preceding physical examination, supplemented by information from the participants, if necessary.

Although several studies have used the FIM to measure functional independence in this group of patients, a few of these studies have revealed shortcomings of this instrument in terms of the locomotion and sphincter control scales (Linacre et al. 1994, Grimby et al. 1996, Andren and Grimby 2000). We, therefore, also used the Hoffer classification (Hoffer et al. 1973) to describe ambulation (Table II). In our study a further category of normal ambulators was added to distinguish between persons without any mobility problems (normal ambulators) and those with minor mobility problems (community ambulators). Additional questions were also asked about the frequency of accidents involving soiling with either urine or faeces and requiring the need to change clothes or napkins (with or without the use of condom, urethral, or suprapubic catheters).

STATISTICS

Data were analyzed with descriptive statistics using SPSS (version 10). We decided to present frequencies of FIM score levels for items and domains instead of mean item and domain scores, because the aim of the present study was to describe percentages of patients in various subgroups who are independent for certain activities of daily living. For this goal, mean item or domain scores were less useful.

First, the score distribution was described for all 18 FIM items separately. For the purpose of this description, the item scores (range 1–7) were recorded into three categories: 1 to 5, 6, and 7 (Table I). FIM scores of 1 to 5 were merged because these scores all indicate the need of help from others in varying degrees, and because the lower categories applied to few patients.

Second, a description of independence for each of the six

FIM domains was prepared for six subgroups of patients. These subgroups were based on two disease characteristics: the presence or absence of hydrocephalus, and a high, middle, or low level of lesion. For each subgroup of patients we then determined for every domain of the FIM the percentage (and binomial 95% confidence interval) of patients who were independent or completely independent. Independence for a domain was defined as having a minimum score of 6 on all items of that domain of the FIM; complete independence was defined as having a minumum score of 7 on all items of a domain (http://www.swogstat.org/stat/public/binomial_conf.htm).

Table III: Independence in terms of activities of daily living according to 18 items of Functional Independence Measure (*n*=165)

Activity	*No helper*		*Helper*
	Level 7 (%)	*Level 6 (%)*	*Level 1–5 (%)*
Self-care			
Eating	95.2	1.2	3.6
Grooming	97.0	2.4	0.6
Bathing	70.9	6.7	22.4
Dressing upper body	90.9	6.7	2.4
Dressing lower body	59.4	24.2	16.4
Toileting	67.9	12.7	19.4
Sphincter control			
Bladder	12.7	9.1	78.2
Bowel	24.8	9.1	66.1
Transfers			
Bed, chair, wheelchair	88.5	3.6	7.9
Toilet	70.9	15.8	13.3
Bath or shower	42.4	44.2	13.3
Locomotion			
Walk/wheelchair	36.4	57.0	6.7
Stairs	30.9	29.7	39.4
Communication			
Comprehension	55.8	35.8	8.5
Expression	83.0	15.8	1.2
Social cognition			
Social interaction	90.3	9.7	0
Problem solving	58.2	30.3	11.5
Memory	57.6	33.3	9.1

Levels are defined as follows: 1–5, supervision, set-up, and assistance; 6, modified independence; 7, complete independence.

ETHICAL APPROVAL

The medical ethics committee approved the Adolescents with Spina Bifida in the Netherlands (ASPINE) study. Informed consent was obtained from all participants.

Results

PATIENTS

A total of 350 patients were invited by mail to participate in the ASPINE study, of whom 181 agreed to do so. Participants and non-participants were similar in age (20.4y [SD 3.0] versus 20.3y [SD 3.1]), sex (41 vs 49% male), type of spina bifida (79 vs 86% aperta), level of lesion (19 vs 23% L2 and above, 66 vs 64% L3 to L5, and 15 vs 13% S1 and below) and being shunted for hydrocephalus (67 vs 64%). Eleven invited patients with spina bifida occulta were excluded because no neurological loss was determined; two were excluded because of comorbidity independently inducing serious physical and/or cognitive disorders (one had a serious heart disease and one a chromosome disorder). In all, 168 patients participated in this study.

Data for the present study on functional independence were complete for 165 patients. The mean age of the study population was 20 years 9 months (range 16–25y [SD 2.9]). Forty-two per cent of patients were male, 84% had spina bifida aperta, and 71% of patients had hydrocephalus. Forty-four per cent of patients had a level of lesion of L2 and above, 40% L3 to L5, and 16% S1 and below.

OUTCOME ON THE 18 FUNCTIONAL INDEPENDENCE MEASURE ITEMS FOR THE TOTAL GROUP

In the self-care domain, patients were least likely to be completely independent for bathing, dressing the lower body, and toileting (Table III). As regards sphincter control, 12.7% of the patients were completely independent in terms of bladder management, and fewer than one-quarter for bowel

Table IV: Percentage of patients with spina bifida who were independent for domains of Functional Independence Measure, in subgroups based on hydrocephalus and level of lesion

	No hydrocephalus			
Level of lesion:	*L2 and above (n=7)*	*L3 to L5 (n=21)*	*S1 and below (n=20)*	*Total (n=48)*
Percentage independent; minimum score 6 on all items in domains listed				
Personal care	85.7 (42.1–99.6)	100 (83.9–100)	100 (82.2–100)	97.9 (88.9–100)
Sphincter control	14.3 (0.4–57.9)	28.6 (11.3–52.2)	50.0 (27.2–72.8)	35.4 (22.2–50.5)
Transfers	100 (59.0–100)	100 (83.9–100)	100 (82.2–100)	100 (92.6–100)
Locomotion	57.1 (18.4–90.1)	100 (83.9–100)	100 (82.2–100)	93.8 (82.8–98.7)
Communication	100 (59.0–100)	100 (83.9–100)	100 (82.2–100)	100 (92.6–100)
Social cognition	100 (59.0–100)	100 (83.9–100)	100 (82.2–100)	100 (92.6–100)
Percentage completely independent; minimum score 7 on all items in domains listed				
Personal care	71.4 (29.0–96.3)	95.2 (76.2–99.9)	100 (82.2–100)	93.8 (82.8–98.7)
Sphincter control	14.3 (0.4–57.9)	28.6 (11.3–52.2)	50.0 (27.2–72.8)	35.4 (22.2–50.5)
Transfers	57.1 (18.4–90.1)	76.2 (52.8–91.8)	100 (82.2–100)	83.3 (69.8–92.5)
Locomotion	28.6 (3.7–71.0)	61.9 (38.4–81.9)	100 (82.2–100)	72.9 (58.2–84.7)
Communication	71.4 (29.0–96.3)	81.0 (58.1–94.6)	95.0 (75.1–99.9)	85.4 (72.2–93.9)
Social cognition	57.1 (18.4–90.1)	90.5 (69.6–98.8)	90.0 (68.3–98.8)	85.4 (72.2–93.9)

Numbers in parentheses are 95% confidence intervals.

management. Regarding locomotion, most problems related to negotiating stairs, for which more than one-third needed help. More than half of the patients used a device for locomotion, including walking aids as well as wheelchairs.

The mean total FIM motor score for the whole group was 76.6 (SD 14.0; range 21 to 91) and the mean total FIM cognitive score was 33.1 (SD 2.4; range 25 to 35). Mean total FIM score was 109.6 (SD 15.5; range 53 to 126; minimum 18, maximum 126). Healthy persons will obtain the maximum FIM score of 126.

FUNCTIONAL INDEPENDENCE FOR PATIENTS WITH SPINA BIFIDA IN RELATION TO HYDROCEPHALUS AND LEVEL OF LESION

Almost all patients without hydrocephalus were independent for all domains of the FIM except for sphincter control, whereas patients with hydrocephalus were much more likely to be dependent on the assistance of others (Table IV). The percentages of patients with hydrocephalus who were independent varied from 5% for sphincter control to 88% for communication.

Patients without hydrocephalus showed no difference related to level of lesion in terms of transfers, communication, or social cognition. Patients without hydrocephalus with a level of lesion of L2 and above were less likely to be independent for locomotion, personal care, and sphincter control than patients with lower levels of lesion, but this group was very small. Within the group of patients with hydrocephalus, clear relationships were found between level of lesion and independence. The greatest differences were seen for locomotion, personal care, and transfers. Patients with a higher level of lesion (especially those with level of lesion L2 and above) were less independent for all FIM domains than patients with lower levels of lesion.

SPHINCTER CONTROL AND INCONTINENCE

The FIM scores for bladder and bowel management were low (Table III). Nearly all patients with hydrocephalus were dependent (score 1 to 5; Table IV). Table V shows that 52% of the patients with hydrocephalus and 20% of those without hydrocephalus had accidents at least weekly. In both subgroups, with and without hydrocephalus, patients with higher lesions were more likely to be incontinent.

LOCOMOTION AND MOBILITY

Almost all patients without hydrocephalus were independent for locomotion, whereas only 46.2% of patients with hydrocephalus were independent for locomotion according to the FIM score (Table IV). According to the Hoffer classification, 47.9% of all patients with hydrocephalus were non-ambulators. Three-quarters of patients with hydrocephalus and a high level of lesion were non-ambulators (Table VI). Of the patients without hydrocephalus, almost all were normal or community ambulators, in contrast to one-third of the patients with hydrocephalus.

Discussion

Our study shows that patients without hydrocephalus were, with a few exceptions, independent for all FIM categories except sphincter control. Within the group of patients with hydrocephalus, independence was closely related to level of lesion. Patients with hydrocephalus and a middle or low level of lesion were largely independent for all FIM categories except for sphincter control. Of all patients with hydrocephalus and a high level of lesion, fewer than half were independent in terms of self-care, whereas about one-fifth were independent for locomotion and only one patient was independent (or continent) as regards sphincter control.

Our study confirms earlier results showing that most patients with spina bifida are independent in terms of activities of daily living (Börjeson and Lagergren 1990; Kalucy et al. 1996; Staal et al. 1996; Andren and Grimby 2000, 2004; Padua et al. 2002; Buran et al. 2004). However, our larger study adds to the literature by providing figures for different subgroups of persons with spina bifida including estimation of the precision of these figures.

One limitation of our study is that only half of the patients invited for this study participated. However, no statistically significant difference found between participants and non-

Table IV: continued

Hydrocephalus			
L2 and above (n=66)	*L3 to L5 (n=45)*	*S1 and below (n=6)*	*Total HC+ (n=117)*
45.5 (33.1–58.2)	82.2 (68.0–92.0)	83.3 (35.9–99.6)	61.5 (52.1–70.4)
1.5 (0.0–8.2)	8.9 (2.5–21.2)	16.7 (0.4–64.1)	5.1 (1.9–10.8)
62.1 (49.3–73.8)	93.3 (81.7–98.6)	100 (54.1–100)	76.1 (67.3–83.5)
21.2 (12.1–33.0)	77.8 (62.9–88.8)	83.3 (35.9–99.6)	46.2 (36.9–55.6)
84.8 (73.9–92.5)	91.1 (78.8–97.5)	100 (54.1–100)	88.0 (80.7–93.3)
71.2 (58.8–81.7)	93.3 (81.7–98.6)	100 (54.1–100)	81.2 (72.9–87.8)
16.7 (8.6–27.9)	57.8 (42.2–72.3)	83.3 (35.9–99.6)	35.9 (24.2–45.3)
0 (0.0–5.4)	4.4 (0.5–15.2)	16.7 (0.4–64.1)	2.6 (0.5–7.3)
6.1 (1.7–14.8)	44.4 (29.6–60.0)	100 (54.1–100)	25.6 (18.0–34.5)
0 (0.0–5.4)	22.2 (11.2–37.1)	83.3 (35.9–99.6)	12.8 (7.4–20.3)
36.4 (24.9–49.1)	46.7 (31.7–62.1)	50.0 (11.8–88.2)	41.0 (32.0–50.5)
27.3 (17.0–39.6)	44.4 (29.6–60.0)	50.0 (11.8–88.2)	35.0 (26.5–44.4)

participants. We therefore assume that we can generalize our results to the population.

Even with the large number of patients involved in our study, two of the subgroups were very small (patients without hydrocephalus with a high level of lesion, and patients with hydrocephalus and a low level of lesion), which was reflected in the broad confidence intervals found for those subgroups. However, we feel that the results are in line with what could be expected and we preferred to present the figures for each subgroup separately rather than merging them with those for other groups.

Another comment must be made about the measurement of level of lesion. We found the information in the medical records about level of lesion at birth to be insufficient; methods of measurements varied (motor level vs sensitivity level) or were not recorded, or no level was described at all. We decided, therefore, to use a standardized measurement of level of lesion performed during the physical examination. Because of this cross-sectional design, the level of lesion might in some cases deviate from the level of lesion at birth (e.g., due to tethered cord). The use of a standardized assessment of level of lesion at an early age with a regular follow-up is recommended, to enable prognostic research in the future. Furthermore, improvements in medical treatment in previous decades might lead to better functional outcomes, especially in the subgroups with the most severe spina bifida. However, such a development has not yet been demonstrated and should be the subject of further studies.

In our study, not having hydrocephalus was found to be a strong predictor of independence in terms of activities of daily living. Level of lesion provided little additional information on expectations of independence within this group. Within the group of patients with hydrocephalus, by contrast, the level of lesion offered important additional information on functional outcome. Patients with a high level of lesion were less likely to be independent as regards most FIM motor and cognitive domains. This is in agreement with the sparse literature (Msall et al. 1994), and also confirms the relationships between level of lesion, cognitive impairments, and dependence in terms of activities of daily living as described earlier (Staal et al. 1996, Schoenmakers 2003).

Our study was largely based on one standardized outcome instrument, the FIM. Although this instrument is well validated and often used (Davidoff et al. 1990, Heinemann et al. 1994, Linacre et al. 1994, Grimby et al. 1996, Hajek et al. 1997, Karamehmetoglu et al. 1997, Andren and Grimby 2000, Voll et al. 2001), its pros and cons have to be kept in mind. Few patients were rated as independent for sphincter control. This may be due to the nature of the sphincter control items in the FIM (Linacre et al. 1994, Grimby et al. 1996). Our additional description of the frequencies of incontinence showed that the percentages of patients having bladder or bowel accidents were comparable to the percentages of patients having a score below 6 on the FIM sphincter control items. It might be inferred that the FIM score for this domain reflects continence rather than independence for bladder and bowel control in this patient group. Future studies using the FIM might benefit from separate recording and reporting of frequency of incontinence and dependence in bladder and bowel management. Although both of these sub-items are included in the FIM, its scoring instructions unfortunately state that only the lower of the two scores has to be recorded.

Another potential disadvantage of using the FIM for this group of patients is the lack of distinction between mobility with a wheelchair and walking with other aids (Grimby et al. 1996, Andren and Grimby 2000). This is why we added the Hoffer classification (Hoffer et al. 1973). This additional information about walking or using a wheelchair in the future can

Table V: Frequency of incontinence for urine and/or faeces for six subgroups of patients with spina bifida, based on hydrocephalus and level of lesion

Frequency of incontinence	*No hydrocephalus*				*Hydrocephalus*			
	L2 and above (n=7)	*L3 to L5 (n=21)*	*S1 and below (n=20)*	*Total (n=48)*	*L2 and above (n=66)*	*L3 to L5 (n=45)*	*S1 and below (n=6)*	*Total (n=117)*
Never any accidents	14.3	28.6	45.0	33.3	3	8.9	16.7	6.0
Less than once a month	28.6	9.5	10.0	12.5	15.2	11.1	16.7	13.7
Once a month but less than once a week	28.6	28.6	25.0	27.1	24.2	37.8	0	28.2
Once a week but less than daily	14.3	23.8	5.0	14.6	19.7	8.9	50	17.1
Daily accidents	14.3	9.5	15.0	12.5	37.9	33.3	16.7	35.0

Table VI: Mobility for six subgroups of patients with spina bifida, based on hydrocephalus and level of lesion according to adapted Hoffer classification

Ambulation	*No hydrocephalus*				*Hydrocephalus*			
	L2 and above (n=7)	*L3 to L5 (n=21)*	*S1 and below (n=20)*	*Total (n=48)*	*L2 and above (n=66)*	*L3 to L5 (n=45)*	*S1 and below (n=6)*	*Total (n=117)*
Normal ambulator (no devices)	28.6	61.9	100	72.9	1.5	28.9	50	14.5
Community ambulator	28.6	28.6	0	16.7	9.1	26.7	33.3	17.1
Household ambulator	0	4.8	0	2.1	9.1	20.0	16.7	13.7
Non-functional ambulator	0	4.8	0	2.1	6.1	8.9	0	6.8
Non-ambulator (manual or electric wheelchair)	42.9	0	0	6.3	74.3	15.5	0	47.9

be very valuable for parents and patients in the future. In fact, the frequencies we found for independence in the domain of locomotion using the FIM closely resembled the frequencies of ambulation derived from the Hoffer classification, which is due to the FIM locomotion domain being a combination of two items: walking/wheelchair and stairs.

A final comment about the FIM must be made regarding a possible ceiling effect in the communication and social cognition domains. Our results for cognition were rather favourable, showing that even in the group of persons with hydrocephalus and a higher level of lesion, most were independent. However, previous research on other patients with other diagnoses, such as spinal cord injury, stroke, and other neurological impairments, has shown that the FIM might be rather insensitive to mild and moderate neurocognitive impairment and cannot replace neuropsychological testing (Davidoff et al. 1990, Heinemann et al. 1994, Hajek et al. 1997, Voll et al. 2001). Results of the ASPINE study published previously have shown that cognitive impairments are present in about half of our respondents with spina bifida aperta and hydrocephalus, and not in respondents without hydrocephalus. These were mainly mild cognitive impairments; about one-fifth of the patients with spina bifida aperta and hydrocephalus had an IQ of less than 70 (Barf et al. 2003).

Conclusion

From the results of our study, a more detailed and precise prognosis can be given to parents about the functional independence of their child at adult age. If a child were born with spina bifida without hydrocephalus, we would expect this child to become independent in terms of activities of daily living, except for sphincter control (incontinence).

If a child is born with spina bifida and is also affected with hydrocephalus, it is important to determine the level of lesion. If the child has a level of lesion of L3 or below, this child is likely to be independent for almost all activities of daily living, with the smallest percentage (78%) being independent for locomotion. However, incontinence will be a major problem for this child.

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