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Quality of performance of everyday activities in children with spina bifida: a population-based study

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Abstract

Aim: The aim of this study was to evaluate the quality of the performance of everyday activities in children with spina bifida.

Methods: Fifty children with spina bifida (of 65 children in a geographic cohort), aged 6 to14 years, were evaluated with Assessment of Motor and Process Skills.

Results: Compared with age-normative values, 60% of the children with spina bifida were found to have motor ability measures below 2 SD and 48% process ability measures below 2 SD. Most of the children with spina bifida had difficulties performing well-known everyday activities in an effortless, efficient and independent way, relating to both motor and process skills. The motor skills hardest to accomplish involved motor planning and the process skills hardest to accomplish were adaptation of performance and initiations of new steps, thus actually getting the task done.

Conclusion: To reach autonomy in life, children with spina bifida may need particular guidance to learn not only how to do things but also how to get things done.

INTRODUCTION

'I suppose he can do it, it's somehow just never done'. This phrase is often heard within the clinic when parents whose children were born with spina bifida (SB) are asked how well their children perform everyday activities.

Spina bifida is a congenital condition that often leads to gross and fine motor dysfunctions (1) and to neurogenic bladder and bowel dysfunctions (2). It has also been recognized that attention and memory problems are often present in children with SB (3–5), as are executive dysfunctions, even among those with normal IQ and especially in the group that has developed hydrocephalus (HC) (4,6). Furthermore, delays and limitations in self care have been reported in children and adolescents with SB (7,8). Studies of long-term outcome have shown that problems seen in childhood often persist in adulthood (9,10). According to Oekeshott (10), only 37% of the individuals with SB are totally independent at 30 years of age, 30% need supervision and occasional help and 33% are dependent i.e. need daily care. Independence failure in self care has been found to be a hindering factor for employment in young adults with SB (11). For a person with bowler and bladder dysfunction, the consequences of an unsuccessful self care might even be fatal (9). It is therefore of vital interest to understand more about *why* children with SB have difficulties performing everyday activities independently.

Taking care of oneself requires the successful performance of everyday activities of varying complexity. Independence in everyday life activities in children and adolescents with SB has been studied from the perspective of to what extent different spina bifida-specific conditions (lesion level, motor functions, hydrocephalus or mental status) are related to level of independence (12,13). Most existing studies have been based on questionnaires filled in by the parents or the child. These studies have focused on what the child is considered capable of doing or usually do (7,8,12,13). However, no description of the specific performance skills that imply strength or weakness of performance, i.e. description of the quality of the performance, is available for this group. Evaluating the ability of the actual 'doing' with a direct observational method could give an idea of what obstacles to performance the child with SB faces (14). No study has, to the best of our knowledge, evaluated the difference in the quality of the performance of everyday activities between children with SB and typically developed children. The aim of this study was to apply a standardized observational assessment method to evaluate

Abbreviations
Lipo-MMC, Lipo-myelomenigocele; ADL, Activities of daily living; AMPS, Assessment of Motor and Process Skills; HC, Hydrocephalus; MMC, Myelomeningocele; SB, Spina bifida

the quality of the performance of everyday activities in children with SB compared with children without known disabilities.

METHODS

Participants

All children born in the period 1993–1999 diagnosed with myelomeningocele (MMC) or lipo-myelomeningocele (lipo-MMC), living in the Swedish region of Västra Götaland, including the counties of Halland and Värmland on December 31, 2006 (n = 65), were identified. This region represents 23% of the population of Sweden. All of the 65 identified children except one received annual routine check-ups at the Regional Rehabilitation Centre, Queen Silvia Children's Hospital in Gothenburg. These 64 children were regarded as potential participants for this study. Exclusion criteria were: (a) motor dysfunction of such severity that the child was unable to drive a manual wheelchair independently (n = 2) or (b) had autism as an additional diagnosis (n = 1). Following this, 61 children/families were invited to participate in connection with their annual routine check-ups during 2006–2007. Fifty of them accepted to participate, giving a consent rate of 82%. Those who did not accept are called non-participants in this study.

The Regional Ethical Review Board in Gothenburg, Sweden was consulted prior to the study (Dnr 574-05), and formal ethical committee approval was deemed unnecessary.

Instruments

The instrument used, Assessment of Motor and Process Skills (AMPS), is a standardized, criterion-based observational assessment, designed to evaluate the quality of the performance of well-known and self-chosen everyday activities (15). The person is observed and rated by a trained and AMPS-certified occupational therapist, during the actual performance of a task in a natural environment. The AMPS indicates whether a person has sufficient performance skills to manage well-known everyday activities in a competent manner i.e. in an effortless, efficient, safe and independent way (15). The performance skills evaluated are regarded as universal goal-directed actions that are comprised in and support all activities of daily living (ADL) tasks. In this instrument, they are expressed by 16 motor skill items and 20 process skill items. Motor skill items are the observed actions that the performer needs to accomplish to move him/her or the objects used in the task with sufficient posture, coordination, mobility, strength and effort to perform the task. Process skill items embrace the actions carried out to start the different steps of the activity and to proceed until the goal of the activity is reached. To do so, the performer needs to initiate, to seek information, to use knowledge and objects, to organize him/her and the steps of the task (both temporal and spatial), to adapt and compensate for difficulties that occur during the performance and keep heading towards the goal. The AMPS manual offers a choice of 83 standardized activities, nine of them are personal ADL tasks and 74 instrumental ADL tasks. Each activity is calibrated according to their defined level of difficulty. The child him/herself chooses two or three tasks that he/she is motivated to do and normally does. The activities serve as means of assessment, where the purpose is to evaluate the quality of the performance of familiar activities of a defined difficulty, but n*ot* to investigate if the child can or cannot perform specific tasks.

After the observations, the occupational therapist rates the quality of the child's performance of the motor skills and the process skills needed to complete the task on a four-point scale: 4 – adequate skill, 3 – questionable skill, 2 – ineffective skill and 1 – markedly deficient skill. The raw ordinal scores obtained are then converted into two linear person-ability-measures (logits), one for motor- and one for process ability, using the many-faceted Rasch analysis in the AMPS software program (http://www.ampsintl.com/software.htm) (15,16). Though the Rasch analysis, the person's ability measures are adjusted for item difficulty, task challenge and rater severity (15). The AMPS software program provides cut-off values that indicate the level below which the performer needs assistance. Internationally based age-normative values derived from 5651 children in the AMPS international database are also available in the AMPS software program (15). The AMPS has been found to have excellent intra- and inter-rater reliability (17) and validity for use from the age of 3 years to adults (18), in different cultures (19), and for different diagnoses, including developmental disabilities (20).

The Hoffer scale (21) was used to classify the ambulation level of the children with SB. This scale describes the ambulatory status of an individual with SB on a 4-point scale: 1 – community ambulator, 2 – household ambulator, 3 – non-functional ambulator (only for training) and 4 – non-ambulatory (Table 1).

Procedure

The families received a posted invitation to participate in this study and were asked to sign written information and consent form. The child also received written information.

Each child was interviewed prior to the assessment, to determine what activities he/she routinely performs in everyday life. With this information as a source, the occupational therapist asked the child to choose two tasks from a pre-selection of four to five familiar tasks from the AMPS manual. If the child seemed uncertain, the parents were also consulted. Once the tasks were chosen, the child and the occupational therapist agreed verbally how the task was going to be performed. All observations were carried out by the same rater (the first author).

Data analysis

Fisher's exact test was used to analyze differences in demographic data between the study sample and the non-participants, with the exception of differences in age, for which an independent sample *t*-test was used. Fisher's exact test was also used to study gender differences in the study sample.

Table 1 Demographic characteristics of the study group of children with spina bifida and non-participants

	Study group n = 50 (%)	Non participants n = 11 (%)	p-value
Gender			
Female	24 (48)	4 (36)	ns
Male	26 (52)	7 (63)	ns
Age			
Mean, years: months	10:5	10:8	ns
Range, years	6–14	6–13	
Hydrocephalus			
Shunted (HC+)	39 (78)	7 (64)	ns
Not shunted (HC–)	11 (22)	4 (36)	ns
Type of SB			
MMC	45 (90)	8 (73)	ns
Lipo-MMC	5 (10)	3 (27)	ns
Ambulation (Hoffer)			
1 – in community	23 (46)		
2 – in household	7 (14)		
3 – for training, non-functional	8 (16)		
4 – non-ambulator	12 (24)		
School form			
Mainstream school	41 (82)		
Special education	5 (10)		
Preschool	4 (8)		
Additory diagnosis			
Epilepsy	3 (6)		
Cerebral palsy	1 (2)		
ADHD	1 (2)		

The results from the study group of children with SB were analyzed in two ways. First their results were compared with the age-normative values available from the AMPS software program. As those age norms are based on the results of children without known disabilities from different regions of the world, an additional comparison with Nordic children was needed. Fifty Nordic children without known disabilities, matched for age and sex to the study group, were obtained randomly from the AMPS international database. The motor and process ability measures from the study group were compared with the measures from this control group using a non-parametric test, the Mann–Whitney *U*-test, as normal distribution could not be assumed.

The study group was also divided into subgroups using a two-step procedure. First the children with shunt-treated hydrocephalus (HC+) and those without (HC–) were separated, and then the HC+ group were divided into ambulators (those who were classified as level 1 or 2 on the Hoffer scale) and non-ambulators (level 3 or 4), resulting in three subgroups (all in the HC– group were ambulators). A non-parametric test, the Kruskall–Wallis test, was used to analyze differences between the subgroups. All statistical tests were completed with SPSS for Windows version 15.0 (SPSS, Inc., Chicago, IL,USA). The level of significance was set to $p < 0.05$ for all statistical tests.

Descriptive statistics were used to report the performance skills that were most often rated as markedly deficient in the SB group (score 1).

RESULTS

The mean age of the children in the study group was 10 years 5 months (SD 2y), range 6–14 years. The demographic data for the children with SB and the non-participants are shown in Table 1.

Comparing the outcome measures from the study group with age-normative data reported in the AMPS software program, it was found that regarding motor ability measures, 60% of the study group achieved scores below 2 SD from their age mean. Twenty-four percent attained results that were low but in the average range and 16% achieved results in the mean range (±1 SD). None achieved motor ability measures above 1 SD from the mean. Regarding the process ability measures, 48% fell below 2 SD from their age mean, 26% of the individuals were within the average range but low, and 26% of the individuals achieved measures in the mean range or higher (Table 2). There were no significant gender differences in children with SB for motor ability ($p = 0.57$) nor for process ability ($p = 0.41$) regarding the proportions of scores below 2 SD. Children from all three subgroups were found among those that fell below 2 SD both on motor and on process ability measures. However, The Kruskall–Wallis test showed significant differences between the groups ($p = 0.009$) indicating that a larger proportion was shunt-treated and non-ambulators (Table 2).

The comparison of the motor and process ability measures from the study group with those from the matched control group confirmed the results from the comparison with the international age norms, as the children with SB had significantly lower medians of both motor ($p < 0.001$) and process ability measures ($p < 0.001$) than their peers in the control group (Table 3).

Among the 16 motor skill items rated in AMPS, the five that were most often rated as marked deficiency (score 1) in children with SB were; 'positions' (severe deficits in positioning the arm or body appropriately in relation to task objects), 'bends' (failure to bend or twist the body appropriately to the task), 'reaches' (failure to secure task objects when reaching), 'calibrates' (severely deficient regulation of

Table 2 ADL motor and process ability measures of children with spina bifida compared with the internationally based age norm from the AMPS data base

	HC+ non ambulator n = 20	HC+ ambulator n = 19	HC– ambulator n = 11	Total n = 50 (%)
Motor ability measure				
Over age norm (+2 SD)	0	0	0	0 (0)
High (+1 SD)	0	0	0	0 (0)
Within mean range	1	2	5	8 (16)
Low (–1 SD)	0	8	4	12 (24)
Under age norm (–2 SD)	19	9	2	30 (60)
Process ability measure				
Over age norm (+2 SD)	0	0	0	0 (0)
High (+1 SD)	1	1	1	3 (6)
Within mean range	0	6	4	10 (20)
Low (–1 SD)	4	5	4	13 (26)
Under age norm (–2 SD)	15	7	2	24 (48)

Table 3 Measures (logits) for ADL motor and process ability of 50 children with spina bifda compared with a matched control group of Nordic children

	Spina bifida			Control			
	Min	Max	Median	Min	Max	Median	p-value
Motor ability measure	−1.34	2.05	0.87	0.66	3.31	1.99	p < 0.001
Process ability measure	−1.76	1.75	0.19	0.12	2.10	0.94	p < 0.001

force or speed of task related actions) and 'paces' (severely deficient rate of task performance). Of the 20 process skills, the five most often rated as markedly deficient were 'accommodates' (i.e. severe deficit to modify actions to overcome problems), 'initiates' (fail to initiate actions or steps of tasks), 'notices-responds' (failure to respond to task-relevant cues from the environment), 'inquires' (asks many questions related to information that had already been discussed/clarified prior to beginning the task) and 'adjusts' (fails to change workplaces or adjust switches and dials to overcome problems) (Fig. 1).

DISCUSSION

The result indicates that the children in this study, ambulators as well as non-ambulators, were to a large extent unable to perform self-chosen and well-known everyday activities in an effortless, efficient, safe and independent manner. A majority of the children not only had lower motor ability measures but also lower process ability measures compared both with international age norms and with a matched control group of Nordic children without known disabilities. As the AMPS evaluates the overall performance of well-known and familiar activities, the deficits seen in this study are likely to be present in most ordinary everyday life tasks, in many performance areas. Even though 82% of the children attended mainstream schools, only about one fourth of them had process ability measures that were within the mean range or higher compared with the age norms.

This study is the first, as far as we know, which used an observation-based assessment to compare the quality of the overall performance of well-known activities of children with SB and children without known disabilities. Through the direct observation of the child's actual performance of the task, this study provides new knowledge about the quality of the performance of activities that children with SB considered themselves capable of doing.

The motor ability measures were, as expected, low in the study group. It is noticeable that the motor skills hardest to accomplish were the ones that needed motor planning and adaptations, i.e. to position your body in an efficient way and to be able to reach and secure objects and to calibrate the right force in movements. The process skill hardest to accomplish for the children in the study group were: adaptation of performance and initiation of new steps. The children with SB often apply the strategy of asking even if it is not needed. In ordinary daily situations, the adult might often facilitates the child's performances instinctively and unconsciously, just by answering the child's questions. This can make it hard to notice if the child with SB actually performs the activities autonomously, or only does them when someone provides small prompts and cues concerning what and how to do, and thereby initiate the doing. The results from this study therefore confirm earlier studies describing dysexecutive syndrome in connection with SB (4,6) and

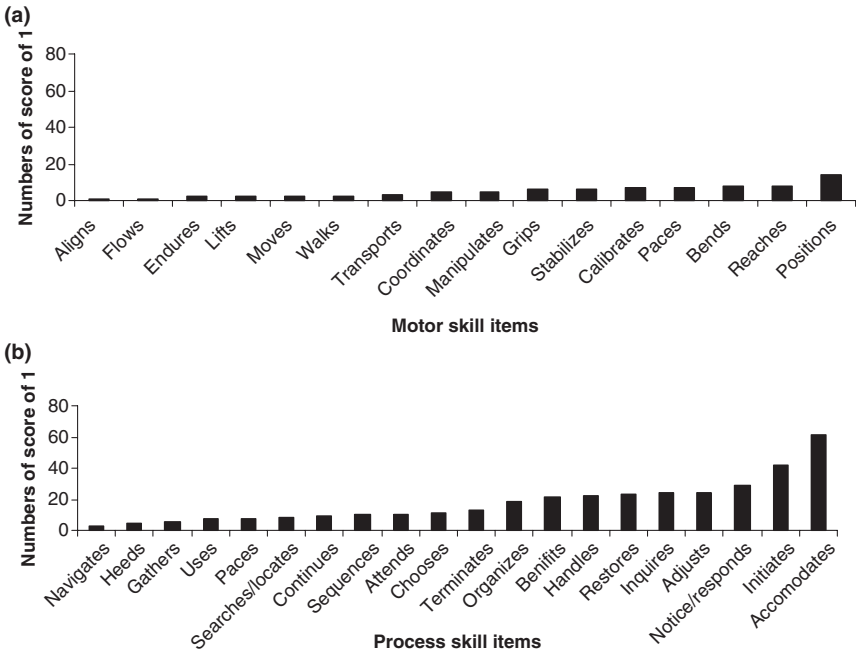

Figure 1 Numbers of score of 1 (markedly deficient) rated on the 100 ADL tasks performed by the spina bifida group distributed over the AMPS skill items. (a) motor skill items. (b) process skill items.

explain the findings of Holmbeck et al. (3) that the children with SB do need more parental guidance than their typically developed peers. In view of these findings, a low ability to perform everyday activities, should not be labelled as being 'delayed', as discussed by Davis et al. (7), as this leads to the idea that the child will catch up one day. This study found that even if they reached the milestones (i.e. being capable) (7) their performance was often less efficient and autonyms than the performance of typically developed children at the same age. It would perhaps be more appropriate to consider the problems in doing something autonomously as a common characteristic of SB. An urgent issue is therefore to start to explore intervention methods, which could give children with SB the right support from an early age to enable them to compensate for or overcome these deficits.

Even though the sample in this study is relatively small and heterogeneous, it gains strength and validity from the fact that the study is population-based in a geographic region that represents approximately one fourth of the population of Sweden, comprising both rural and urban communities. Therefore the results most probably can be generalized to a larger population of children with SB.

As expected, the children who were shunt-treated and non-ambulators were shown to have more deviant results than the ones less severely disabled. This confirms the previous studies (12,13) which have reported that children who have shunt-treated hydrocephalus and are non-ambulant are less independent in everyday life. Further, our study contributes new information by showing that all three subgroups of children with SB (Table 2) were represented among individuals falling below 2 SD on both motor and process ability measures compared with normally developed children.

Approximately one fourth of the children in the SB group did, however, have results that were within the mean range or higher regarding process skills. It would be interesting to study those children more carefully, especially the ones that that achieved results that were higher than 1 SD from the mean, to understand what could explain their successful performance, achieved despite different levels of ambulation and incidence of hydrocephalus (Table 2).

This study was cross-sectional, and as such, did not answer the question as to whether the performance problems observed will follow the individuals in our cohort throughout life. Nevertheless, it can be presumed from looking at long-term outcome studies (9,10) that the performance deficiencies most probably will remain. It has been stated earlier that adolescents with SB need guidance during their transition to adulthood (9,22). Our study supports this, as it suggests that it is not enough to learn how to do things (being capable). The results of this study imply that even if the child with SB seems to know how to do things, he/she still might have problems getting them done, efficiently, on his/her own.

It has been found that adolescents with SB perceive that they do not participate as much as their peers do in common activities during adolescence (23). It will not, given the findings from our study, be sufficient to just give opportunities for participation. Consequently, the child born with SB will *also* need support to be able to seize the opportunities given. Difficulties reaching independence are probably not primarily due to lack of experience or, as has been suggested, caused by the overprotection of parents (24). Rather, overprotection is probably the logical response from the parents when they perceive that their child 'never get things done'. Therefore we claim that the knowledge gained from this study highlights the importance of informing parents and caregivers of children with SB from an early stage. To back up their child's autonomy in everyday activities, the parents need knowledge concerning the underlying reasons why their child does not get things done.

CONCLUSIONS

Children with spina bifida are, to a large extent, unable to perform well-known and self-chosen activities in an effortless, efficient, safe and independent way. This study suggests that this it due to deficiencies in both motor and process skills. We claim that, to reach autonomy in daily life, children born with spina bifida need particular guidance to overcome process deficits, to learn not only how to do things but also how to get things done.

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