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ARTICLE

Adults with spina bifida: ambulatory performance and cognitive capacity in relation to muscle function

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STUDY DESIGN: Cross-sectional study.
OBJECTIVE: Describe and compare ambulatory performance and cognitive capacity in relation to muscle function in an adult cohort with spina bifida. Also, explore factors associated with ambulation in participants with muscle function level 3.
SETTING: Specialist clinic for adults with spinal cord disorders in Stockholm, Sweden.
METHODS: A total regional cohort of adults ($n = 219$) with spina bifida was invited, 196 (104 women, mean age 35 years, SD 13 years) participated. Mode of mobility, cognitive capacity and muscle function were investigated. For participants with muscle function level 3, factors associated with ambulation were investigated using multivariate logistic regression analysis.
RESULTS: In all, 84 participants (42%) were community ambulators, 22 (12%) household ambulators and 90 (46%) wheelchair users. There was a linear association between the lower degree of muscle function and scoliosis ($P < 0.001$). Mode of mobility varied despite similar muscle prerequisites in participants with muscle function level 3 ($n = 58$). Factors associated with ambulation in participants with muscle function level 3 were the absence of scoliosis, lower BMI and higher cognitive capacity.
CONCLUSIONS: Cognitive capacity and mode of mobility varied widely across the cohort. However, in participants with muscle function level 3, despite similar muscular prerequisites, a large variation in the mode of mobility was found, suggesting that other factors were involved. It is important to prevent scoliosis, support a healthy lifestyle, as well as offer cognitive screening and support to promote ambulatory function and optimise independence in the everyday lives of adults with spina bifida.

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INTRODUCTION

Spina bifida (SB) is a complex congenital neural tube malformation involving multiple body systems [1, 2] leading to a multifaceted disability. Due to better medical treatment, 75% of people with SB reach early adulthood [3]. In most cases, SB includes impaired motor and sensory function, leading to partial or complete paralysis and/or sensory loss below the malformation. Mode of mobility ranges from ambulation in the community to the use of a powered wheelchair. Musculoskeletal conditions, pain, bladder and bowel-related problems, and pressure ulcers are common [1]. Further, tethered cord syndrome and orthopaedic conditions such as contractures, hip dislocations, scoliosis, and kyphosis have been frequently reported [2, 4].

Most people with open SB have shunted hydrocephalus, migration abnormalities of the central nervous system and Chiari II malformation [2, 4]. A higher level of spinal lesion is a marker of more severe anomalous brain development which, in turn, is associated with reduced independence [5]. The cognitive impairments, often affecting executive function, prospective memory, timing and time management [6], are highly variable between persons [2]. Timing and attention impairments are associated with hydrocephalus [7] and brain dysmorphologies, such as the Chiari II malformation [8], while movement impairments are caused by spinal cord dysfunction and cerebellar dysmorphologies that influence sensory-motor timing and motor regulation [5].

Impaired gait function [9–12] is common and associated with the level and degree of spinal malformation. Gait is a complex activity requiring interactions between supraspinal locomotor and cognitive networks [13]. Long-term functional outcomes such as ambulation and sitting balance are closely related to the neurological level of malformation [12, 14, 15], but the mode of mobility is difficult to predict since it is influenced by factors such as age, body mass index (BMI), orthopaedic deformities and cognitive status [15]. Ambulatory persons with sacral or low-lumbar SB usually retain their walking ability into adulthood, whereas those with a high-lumbar or thoracic level of malformation often become wheelchair users [4, 14, 16]. A key component for ambulatory performance is an adequate function in knee extensors [16], corresponding to muscle function (MF) level 3, consistent with the classification of muscle function groups [9]. The aim of this paper was to describe and compare ambulatory performance and cognitive capacity in relation to MF in an adult cohort with SB. The paper also aimed to explore factors associated with ambulation in adults with MF level 3.

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MATERIALS AND METHODS

Study design

A cross-sectional study conducted by a multidisciplinary team at a regional clinic for spinal cord disorders, investigating medical, physical and cognitive functioning associated with the level of muscle function.

Participants

A near-total regional cohort of adults (≥18 years) with SB ($n = 219$) were consecutively invited to participate in conjunction with regular clinical follow-up.

Data collection procedure

The study is part of a larger research project aiming to describe the living and health issues of a regional cohort of adults with SB enrolled at a specialised clinic for adults with spinal cord disorders [17]. In short, the data collection was conducted through regular clinical assessments and structured interviews by three persons, a physiotherapist (MB), an occupational therapist (supervised by GH) and a nurse who were part of the clinic's SB multi-professional team with over 10 years experience of adults with SB. Medical records were used to validate the provided information.

Muscle function was assessed via manual muscle testing of the lower extremities using a 0–5 graded scale [18], and participants were classified according to the categories developed by Bartonek [9], with an additional category for those who had no loss of muscle strength, Table 1. For participants with an asymmetrical motor function, the most severely impaired side was used for classification [15] in order to avoid overrating their function. As knee extensor function is a key component of gait, participants in MF level 3 were further explored. Hip, knee and ankle joint contractures of more than 20 degrees were registered [9]. A clinical examination including visual inspection (by the physiotherapist) for scoliosis in a sitting position was used in combination with participant statement and medical records to register scoliosis and/or previous spinal surgery resulting from scoliosis. Both conditions are hereinafter referred to as scoliosis. The clinical examination also included height (m) in a standing position or, for participants who were unable to stand, in lying position (from joint to joint in case of contractures) and measurement of weight (kg). BMI was calculated (kg/m^2). The American Spinal Injury Association (ASIA) Impairment Scale (AIS) was used according to the International Standards for Neurological Classification of Spinal Cord Injury [19].

Each interview included sociodemographic, medical and physical factors, presence, and location of pain on examination day (yes/no), use of assistive devices and orthoses, and self-reported maximal walking distance (verified by control questions to ensure accuracy and categorised as: >1000 m, ≤1000 m, ≤100 m, ≤10 m, 0 m, Table 3). Further, the ambulatory function was assessed according to Hoffer [14, 20]. The participants were dichotomised as ambulatory (community and household ambulators) or wheelchair users (non-functional ambulators and wheelchair users).

Physical exercise was self-reported and categorised as no physical exercise, moderate exercise (minimum 30 min, 1–2 times per week) and vigorous physical exercise (minimum 30 minutes at least three times weekly). This was also verified by the assessors via control questions about their exercise regimes.

Cognitive capacity was assessed using three tests. The coding test for psychomotor speed and executive function and the block design test for spatial/psychomotor ability and executive function, both from the Wechsler Abbreviated Scale of Intelligence, WASI [21]. Further, the FAS test was used, a phonemic word fluency test for verbal executive ability and mental speed [22]. Results for the subtests were scaled according to age, with a mean of 10 (SD 3) and a range of 1–19. To calculate the scores on the FAS test, the results were first converted to Z values.

Table 1. Muscle function (MF) levels.

Level	Muscle function
MF 0	No loss of muscle strength
MF 1	Weakness of foot intrinsic muscles Good-to-normal plantar flexors, grades 4–5
MF 2	Fair or less plantar flexion, grade ≤3 Fair or better knee flexion grade ≥3 Poor to fair or better hip extension and/or hip abduction, grade ≤2–3
MF 3	Good to normal hip flexion and knee extension, grades 4–5 Fair or less knee flexion grade ≤3 Traces of hip extension, hip abduction and below knee muscles
MF 4	No knee extension activity Poor or less hip flexion grade ≤2 Fair or good pelvic elevation, grades 2–4
MF 5	No muscle activity in the lower limbs No pelvic elevation

Data analysis

The Shapiro–Wilk test ($P < 0.05$) was used to analyse normal distribution. Descriptive data were presented as numbers and proportions. Mean and standard deviation (SD) were used for normally distributed variables, while median (Md) and interquartile ranges (IQR) were used for non-normal distributions. The differences were analysed using the Chi-square test for dichotomous variables, one-way ANOVA for normally distributed variables with more than two groups, Student's t test for normally distributed variables comparing two groups, and the Mann–Whitney U test for variables with non-normal distribution.

The Cochran–Armitage test of trend was used to determine whether there was a linear association between the muscle function and dichotomic variable. Statistically significant differences are presented.

For persons with MF level 3, factors potentially associated with ambulation [15, 23] age [15], sex, height, weight, BMI [9], scoliosis [9, 23], daily bladder and/or bowel incontinence, sensory function in the feet and cognitive capacity [15] (the coding test [21], FAS test [22] and block design test [21]) were investigated using bivariate logistic regression analysis. According to the "rule of thumb" of ten persons per variable, five variables were included as there were 50 participants [24]. Age was considered a possible confounder and was therefore included, together with the variables with the lowest P values. A multivariate model was performed using backwards enter mode. The collinearity between the variables in the model was investigated using Spearman's rank correlations and were preset at less than 0.6. The results from the regression analysis were presented with odds ratios (OR) including 95% confidence intervals (CI) and Hosmer–Lemeshow goodness-of-fit statistics were used for model fit of the final multivariate model.

The analyses were performed using SPSS version 24 (IBM Corp., Armonk, NY, USA). The statistical significance was determined at $P \leq 0.05$.

RESULTS

There were 196 persons (89%) (104 women, 53%) with a mean age of 35 years (SD 13) invited to participate, Table 2, Fig. 1. Nineteen persons declined follow-up and four did not respond. Seventy-eight percent, 153 out of 196 (80 women), participated in the cognitive assessment, Fig. 1. Of those who declined, some had previously completed another neuropsychiatric assessment (not included in the present investigation), but in most cases, no reason was given. No significant differences were seen in age, sex, prevalence of hydrocephalus, muscle or ambulatory function between those persons who participated in the cognitive assessment and those who declined.

The cohort comprised 84 (42%) community ambulators, 22 household ambulators (12%) and 90 (46%) wheelchair users (including non-functional walkers). Out of 196 participants, 179 (91%) had myelomeningocele, 13 (7%) lipomeningocele and four (2%) spina bifida occulta. The most common level of neurological impairment according to the American Spinal Injury Association Impairment Scale (AIS) [19] was L3 in 82 participants (41%). Mean height was 158 cm (SD 14), mean weight 69 kg (SD 19) and median BMI 27 (IQR 23–31). For demographic and structural characteristics see Table 2, for mobility and the use of orthoses and assistive devices see Table 3.

There was a significant difference in mean height between participants in MF level 0, 1 and 2 compared to participants in MF level 3, 4 and 5 ($P < 0.000$) with the successively lower height of participants in MF level 1 (170 cm) to MF level 5 (145 cm).

Table 2. Demographic and structural characteristics, for the total cohort and divided into muscle function (MF) levels for total group.

Participants, *n* (%)	Total 196 (100)	MF level 0, 16 (8)	MF level 1, 19 (10)	MF level 2, 36 (18)	MF level 3, 58 (30)	MF level 4, 33 (17)	MF level 5, 34 (17)
Sex, women, *n* (%)	104 (53)	9 (56)	9 (47)	19 (53)	29 (50)	18 (55)	20 (59)
Men, *n* (%)	92 (47)	7 (44)	10 (53)	17 (47)	29 (50)	15 (45)	14 (41)
Age, mean (SD)	35 (13)	32 (11)	36 (15)	38 (16)	35 (12)	36 (13)	33 (11)
Md (IQR)	33 (23–46)	27 (22–45)	37 (23–50)	37 (22–50)	33 (24–45)	34 (22–47)	32 (23–47)
Length (cm), mean (SD)	158 (14)	169 (11)	170 (14)	162 (12)	159 (11)	150 (10)	145 (13)
Md (IQR)	157 (150–167)	173 (161–179)	170 (158–178)	164 (155–170)	160 (153–167)	150 (141–157)	145 (140–151)
Weight (kg), mean (SD)	69 (19)	72 (21)	74 (20)	68 (16)	72 (18)	69 (22)	59 (17)
Md (IQR)	67 (56–81)	72 (58–79)	67 (63–87)	69 (54–79)	68.5 (61–83)	65 (57–81)	56.5 (47–71)
BMI, mean (SD)	28 (13)	25 (5)	26 (5)	26 (5)	29 (7)	31 (9)	29 (8)
Md (IQR)	27 (23–31)	24 (22–28)	25 (21–28)	27 (22–28)	28 (24–32)	28 (24–36)	28 (22–35)
Weekly exercise							
Vigorous >30 min ≥3 times/week	23 (12)	2 (12)	3 (16)	4 (11)	8 (14)	4 (12)	2 (6)
Moderate >30 min 1–2 times/week	77 (39)	6 (38)	8 (42)	14 (39)	17 (29)	17 (52)	15 (44)
No exercise	96 (49)	8 (50)	8 (42)	18 (50)	33 (57)	12 (36)	17 (50)
Neurological level, *n* (%)							
T3-T12 AIS A, B, C	49 (25)	–	1 (5)	1 (3)	3 (5)	12 (36)	32 (94)
T3-T12 AIS D	1 (1)	–	–	1 (3)	–	–	–
L1-L2 AIS A, B, C	35 (17)	–	–	1 (3)	14 (24)	19 (58)	1 (3)
L1-L2 AIS D	5 (3)	–	4 (20)	1 (3)	–	–	–
L3 AIS A, B, C	68 (34)	6 (37)	2 (11)	24 (66)	33 (57)	2 (6)	1 (3)
L3 AIS D	14 (7)	1 (6)	7 (37)	5 (14)	1 (2)	–	–
L4-S1 AIS A, B, C	10 (5)	0	2 (11)	1 (3)	7 (12)	–	–
L4-S1 AIS D	7 (4)	2 (13)	3 (16)	2 (5)	–	–	–
AIS E	7 (4)	7 (44)	–	–	–	–	–
Hydrocephalus, *n* (%)	123 (63)	5 (31)	4 (21)	16 (44)	39 (67)	28 (85)	31 (91)
Tethered cord symp, *n* (%)	58 (30)	4 (20)	7 (37)	19 (53)	23 (39)	15 (46)	12 (35)
Pain, *n* (%)	78 (40)	2 (13)	7 (37)	19 (53)	23 (40)	15 (46)	12 (35)
Scoliosis[a]	90 (46)	3 (19)	7 (37)	13 (36)	15 (26)	20 (61)	32 (94)

SD standard deviation, *IQR* interquartile range, *BMI* body mass index, *AIS* American spinal injury association impairment scale.
[a]Surgery $n = 32$, scoliosis registration in the medical record (incl radiographs) $n = 18$, clinical examination $n = 40$.

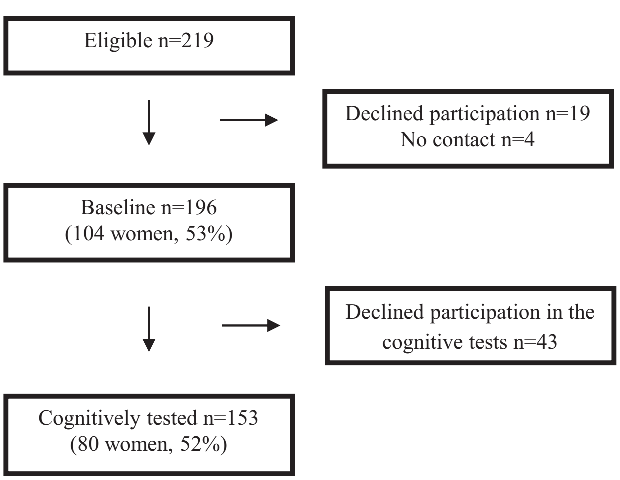

Fig. 1 Flowchart of participants. Displaying number of eligible, declining and participating persons as well as number of cognitively tested participants.

Further, there was a significant linear association between a lower degree of muscular function and a higher proportion of participants with hydrocephalus ($P < 0.001$), a higher proportion of participants with contractures in the lower extremities ($P < 0.001$) and scoliosis ($P < 0.001$). There was only a significant difference in BMI between MF level 2 and 4 ($P = 0.033$). Other variables were not associated ($P > 0.05$) with MF level.

In the subtests for cognitive capacity, participants with MF level 4 and 5 had significantly lower scores. On the coding tests, participants with MF level 5 showed significantly lower results compared to all other MF levels. Further, in the block design test, participants in MF level 5 had significantly lower results compared to all other MF levels, except MF level 4. On the FAS test, participants with MF level 4 and 5 showed significantly lower results compared to participants with MF levels 2 and 3, Table 3.

For participants with MF level 3, the most common level of malformation, (29 women), 16 (28%) were community ambulators, 19 (32%) household ambulators and 23 (40%) wheelchair users (including the non-functional ambulators). The community ambulators (mean age 39 years, SD 14) and the wheelchair

Table 3. Mobility, use of orthoses and assistive devices, and results from the Coding, Block design and FAS tests for the total cohort and divided into muscle function (MF) levels for total group.

Participants, *n* (%)	**Total 196 (100)**	**MF level 0, 16 (8)**	**MF level 1, 19 (10)**	**MF level 2, 36 (18)**	**MF level 3, 58 (30)**	**MF level 4, 33 (17)**	**MF level 5, 34 (17)**
Ambulatory function							
Community ambulation	71 (36)	16 (100)	19 (100)	26 (72)	9 (16)	1 (3)	–
Community ambulation[a]	13 (6)	–	–	6 (17)	7 (12)	–	–
Household ambulation[b]	17 (9)	–	–	1 (3)	14 (24)	2 (6)	–
Household ambulation[c]	5 (3)	–	–	–	5 (8)	–	–
Non-functional ambulation[d]	17 (9)	–	–	2 (5)	12 (21)	3 (9)	–
Do not walk[e]	73 (37)	–	–	1 (3)	11 (19)	27 (82)	34 (100)
Maximal walk distance							
> 1000 m	54 (27)	16 (100)	14 (74)	20 (56)	4 (7)	–	–
≤1000 m	29 (15)	–	5 (26)	11 (31)	12 (20)	1 (3)	–
≤100 m	28 (14)	–	–	3 (8)	23 (40)	2 (6)	–
≤10 m	10 (5)	–	–	1 (3)	7 (12)	2 (6)	–
0 m	75 (38)	–	–	1 (3)	12 (21)	28 (85)	34 (100)
Orthoses							
Insole	25 (13)	4 (25)	6 (32)	11 (31)	4 (7)	–	–
SMO[f]	2 (1)	–	–	2 (6)	–	–	–
AFO[g]	41 (21)	–	–	6 (17)	21 (36)	9 (27)	5 (15)
KAFO (open or closed)[h]	12 (6)	–	–	–	8 (14)	3 (9)	–
Prosthesis	2 (1)	–	1 (5)	1 (3)	–	–	–
Walking aids							
Cane/crutches	16 (8)	–	1 (5)	1 (3)	13 (22)	1 (3)	–
Walker	9 (5)	–	1 (5)	4 (11)	4 (7)	–	–
Support from walls	3 (3)	–	1 (5)	–	1 (2)	1 (3)	–
Wheelchair							
Manual	52 (27)	–	–	6 (17)	24 (41)	12 (36)	10 (29)
Powered	4 (4)	–	–	–	3 (5)	1 (3)	–
Both manual and powered	67 (34)	–	–	4 (11)	19 (33)	20 (61)	24 (71)
Cognitive assessment, *n*	153	10	13	30	50	25	25
HC, *n*	98	3	4	14	34	20	23
Coding[i], mean (SD)	6.8 (3.1)	7.9 (2.8)	8.2 (3.4)	8.0 (2.5)	7.4 (3.1)	6.0 (2.5)	3.7 (2.1)
Block design[i], mean (SD)	6.9 (2.7)	8.1 (3.4)	8.0 (3.4)	7.6 (2.5)	7.4 (2.9)	5.9 (1.1)	5.8 (2.0)
FAS[i,j], mean (SD)	7.3 (3.8)	6.3 (2.4)	7.2 (4.3)	7.9 (3.9)	8.9 (4.1)	5.8 (3.0)	5.3 (2.5)

[a]Wheelchair use only for long distances outdoor.
[b]wheelchair outdoor.
[c]Wheelchair both in and outdoor.
[d]ambulation during therapy.
[e]wheelchair use for mobility.
[f]Supramalleolar orthoses.
[g]Ankle foot orthoses.
[h]Knee ankle foot orthoses.
[i]Reference value for the general population is 10 (SD = 3). The range of scores on the scale is 1–19 (mean = 10; SD = 3).
[j]To calculate the score on the FAS scale, results were first converted to *Z* values.

users (mean age 37 years, SD 11) were significantly older than the household ambulators (mean age 29 years, SD 11) ($P =$ 0.012 and $P = 0.017$). The ambulatory participants had a lower mean BMI (26, SD 3.7 and 27, SD 5.6) than the wheelchair users (mean of 32 with two outliers with a BMI of 47 and 49, SD 7.4) ($P = 0.001$). Further, there were no significant differences regarding prevalence of pain, tethered cord symptoms or results from the cognitive tests.

For participants with MF level 3, the bivariate logistic regression analysis showed that sex, height, BMI and scoliosis were

Table 4. Bivariate and final multivariate logistic regression analysis of factors associated with ambulation in persons with muscle function (MF) level 3.

Variable, $n = 50$	Bivariate				Multivariate			
	β	*P* value	OR	95% CI	β	*P* value	OR	95% CI
Age	−0.038	0.127	0.962	0.916–1.011	−0.034	0.368	0.967	0.898–1.041
Sex (ref woman)	1.371	**0.027**	3.939	1.168–13.281	2.064	0.052	7.874	0.981–63.235
BMI[a]	−0.208	**0.003**	0.812	0.708–0.933	−0.271	**0.015**	0.763	0.613–0.948
Scoliosis (ref yes)[b]	−1.804	**0.011**	0.165	0.041–0.656	−3.497	**0.009**	0.030	0.002–0.415
Coding test	0.034	0.088	1.034	0.995–1.075	0.070	**0.030**	1.073	1.007–1.143
Height	0.104	**0.005**	1.109	1.032–1.193				
Weight	−0.028	0.150	0.973	0.937–1.010				
Incontinence bladder (ref yes)	−0.618	0.411	0.593	0.124–2.350				
Incontinence bowel (ref yes)	−0.888	0.177	0.411	0.113–1.494				
Hydrocephalus (ref yes)	−0.862	0.200	0.422	0.113–1.578				
Block design test[c]	0.024	0.290	1.024	0.980–1.070				
Sensory function in feet[d]	−0.254	0.666	0.776	0.245–2.453				
FAS test	0.036	0.551	1.037	0.920–1.169				

[a]Body Mass Index (kg/m^2), [b]presence of scoliosis and/or spine surgery, [c]raw scores, [d]according to the International Standards for Neurological Classification of Spinal Cord Injury. *P* values < 0.05 in bold, Overall model fit (Hosmer and Lemeshow test): $\chi^2 = 1.495$, df = 8, $P = 0.993$. Cox & Snell $R^2 = 0.468$, Nagelkerke $R^2 = 0.637$.
Analysis of ambulators (community and household ambulators) versus wheelchair users (non-functional ambulators and wheelchair).

significantly associated with mode of mobility, Table 4. These variables were therefore included in the multivariate model, together with age (possible confounder) and the coding test (*P* value <0.1). Height was not included in the multivariate analysis as it is closely associated with BMI, which was considered more relevant for ambulation [15]. In the final multivariate model, BMI, scoliosis and the coding test were independently associated with ambulation, Table 4. For every increasing unit of BMI, the OR of being ambulatory decreased by ~24% (95% CI 0.613–0.948, $P = 0.015$). The participants with scoliosis had an ~97% lower OR (95% CI 0.002–0.415, $P = 0.009$) of being ambulatory. For every increasing unit of the coding test, the OR of being ambulatory increased by approximately 7% (95% CI 1.007–1.143, $P = 0.030$).

DISCUSSION

In this cohort of nearly 200 adults with SB, ambulatory function and cognitive capacity were investigated in relation to different muscle function. Interestingly, in participants with MF levels 2–4 (corresponding to a lumbar malformation), ambulatory function was highly variable. Mode of mobility, use of assistive devices and orthoses, as well as maximal walking distance, varied between participants with similar muscular prerequisites for walking, indicating that ambulatory function is complex and depends on multiple factors. In the most common MF level (level 3), the ambulatory participants had a lower BMI, lower presence of scoliosis and higher cognitive capacity.

Walking aids were used by over half (51%) of the ambulatory persons with MF 3 and by 15% in MF2 (Table 3), indicating that ambulation is highly challenging for persons with MF level 3 category. Thus, optimising assistive devices is a prerequisite for promoting functional independence in persons with SB. Combining ambulation and wheelchair at different circumstances could be beneficial and enable flexibility in everyday life. Wheelchair use could be regarded as a limiting factor, associated with barriers [25], although it is also the most essential mobility device [25]. Thus, a retained walking function (when possible) could enhance the quality of life as it may improve accessibility and social availability. However, clinical experience and the experience of adults with SB [26] indicate that if walking is too challenging using a wheelchair could be a viable decision.

Adults with SB seem positive about exploring their cognitive capacity, potentially reflecting a perceived gap of knowledge as most of the participants (78%) approved the testing. For children with SB, cognitive screening and support are currently part of SB follow-up in many well-resourced countries. The guidelines for the care of adults with SB [27] recommend a full neuropsychological assessment for persons with cognitive problems. However, our study showed that cognitive problems are widespread and common. To enable optimal individual support in coping with aspects of daily life, we suggest that global cognitive screening such as Montreal Cognitive assessment [28] is offered when persons with SB are enrolled in adult care and at follow-up. For persons, whose screening results indicate the need for further investigation, we suggest that they are offered a full neuropsychological assessment, including assessment of psychomotor speed and executive function.

In this study, almost two thirds of the participants (63%) had hydrocephalus which can be a major determinant for cognitive function [2, 4, 7] depending on severity. However, no information could be found in the medical records about the severity of the hydrocephalus, only information about whether hydrocephalus was present or not. Hydrocephalus is a rough measure that offers minimal information about the impact on cognitive or physical function i.e., the consequences for everyday life. This means that a person with well-functioning hydrocephalus can be very different from a person with multiple complications and who has received multiple operations. Thus, we have chosen not to focus on hydrocephalus in the manuscript.

Almost half of the cohort (46%) had scoliosis. This is in line with a review by Heyns et al. [23] in which just over 50% of the study cohort had scoliosis. More than nine out of ten participants with MF level 5, all wheelchair users, had scoliosis. According to clinical experience, scoliosis may negatively affect the sitting posture in a wheelchair making it difficult to achieve an ergonomically sound position.

We explored factors associated with ambulation in participants with MF level 3. In this cohort, the ambulatory function was highly variable despite similar muscular prerequisites suggesting that other factors are important in predicting the level of ambulation. The ambulators had better prerequisites for walking with a lower BMI, lower presence of scoliosis and better results from the coding test for psychomotor speed and executive function. Among these factors, scoliosis and BMI had the highest impact. The association between scoliosis and ambulatory status has been previously indicated [29]. A high BMI is associated with a greater risk of cardiovascular disease, pressure ulcers, decreased participation in physical activity, psychosocial consequences, and can negatively affect mobility and transfers [30]. As this was a cross-sectional study, the causality dilemma of "which came first"—a higher BMI resulting from increased wheelchair use or a higher BMI being the reason for using a wheelchair more—could not be answered. In this study, ambulatory persons demonstrated significantly higher coding test results, an easily administered and sensitive test of psychomotor capacity, that is relatively unaffected by intellectual capacity, education or learning [31]. Further, the household ambulators, combining ambulation with wheelchair use for longer distances, were younger than both the community ambulators and wheelchair users, potentially indicating a transition of mobility over time. The factors that potentially affect a changed mode of mobility would be interesting for future studies.

Strengths and limitations

This study comprises a large regional cohort of adults with SB, estimated to represent around 25% of the national cohort. The entire cohort was invited to participate and more than 90% of the 219 adults who met the eligibility criteria agreed to participate. The cohort is assumed to represent more than 90% of adults with SB in the Greater Stockholm area. This is due to the extensive system of follow-up and detection of new cases. We consider it a strength that the cohort represents adult persons with SB with a wide age range (18–73 years), as well as mode of mobility and cognitive capacity.

Patients did not receive standard radiographs as part of the data collection process, therefore there is a level of uncertainty concerning the prevalence of scoliosis. However, as our numbers are in line with previous literature, we believe they are largely accurate.

Of the participants, 21% declined cognitive assessment, of which a few had previously performed similar tests. However, no obvious differences in characteristics (age, sex, prevalence of hydrocephalus, muscle, or ambulatory function) could be detected between the participants who declined and those who performed the tests. Although the MF level 3 comprised a near-total regional cohort ($n = 58$) it was a relatively small group, hence the current results should be interpreted with caution. Further, the participants were classified according to the MF levels of the most severely impaired side, potentially resulting in some of the participants having better prerequisites for walking compared to those participants with a symmetric level of MF.

Clinical implications

First, it is important to focus on modifiable factors such as BMI, starting already in childhood. In a review in 2017 [32], Polfuss et al. concluded that it is "critical to initiate prevention efforts early with a multifactorial approach for this at-risk population". They might be offered nutrition and health-related lifestyle coaching and be provided with an easy way to measure and follow-up their weight (this also applies to wheelchair users). Moreover, as almost half of this cohort reported no weekly exercise, it is important to locate gyms that are also suited to physically impaired persons and/or provide a programme for home exercises. Even non-exercise physical activity has been reported to increase energy expenditure in persons with complete paraplegia [33] highlighting the importance of an active lifestyle. Second, to include spinal assessment in standardised clinical care is of importance to be able to prevent and address scoliosis. Third, adults with SB should be offered the opportunity to try out and modify orthoses and assistive devices as this could significantly improve gait pattern, and thereby help maintain ambulatory function and a higher level of physical activity.

The association between ambulation and cognitive function might suggest that people with SB have difficulties performing dual tasking, i.e., a cognitive and a motor task simultaneously. Difficulties in performing dual tasks or prioritising the most important tasks may have serious consequences in everyday life, with an increased risk of falls and consequential injuries. Falls are common in persons with Spinal Cord Injuries [34], who have similar prerequisites for ambulation as persons with SB. However, thus far, dual-task interference in adults with SB is an unexplored field. Further research is essential to identify the best intervention for this group.

CONCLUSION

Cognitive capacity and mode of mobility varied widely across the cohort and the latter differed between persons with similar muscle function. In participants with MF level 3, factors independently associated with ambulation included the absence of scoliosis, lower BMI, and better results on the coding test for psychomotor speed and executive function. It is important to prevent scoliosis when possible, support a healthy lifestyle, as well as offer cognitive screening and support to promote ambulatory function and optimise independence in the everyday lives of adults with SB.

DATA ARCHIVING

Due to Swedish and EU personal data legislation, the dataset is not publicly available but is available from the corresponding author upon appropriate request. Any sharing of data will be regulated via a data transfer and user agreement with the recipient.

REFERENCES

1. Wagner R, Linroth R, Gangl C, Mitchell N, Hall M, Cady R, et al. Perception of secondary conditions in adults with spina bifida and impact on daily life. Disabil Health J. 2015;8:492–8.
2. Copp AJ, Adzick NS, Chitty LS, Fletcher JM, Holmbeck GN, Shaw GM. Spina bifida. Nat Rev Dis Prim. 2015;1:1–18.
3. Bowman RM, McLone DG, Grant JA, Tomita T, Ito JA. Spina bifida outcome: a 25-year prospective. Pediatr Neurosurg. 2001;34:114–20.
4. Liptak GS, Garver K, Dosa NP. Spina bifida grown up. J Dev Behav Pediatr. 2013;34:206–15.
5. Fletcher JM, Copeland K, Frederick JA, Blaser SE, Kramer LA, Northrup H, et al. Spinal lesion level in spina bifida: a source of neural and cognitive heterogeneity. J Neurosurg. 2005;102:268–79.
6. Dennis M, Barnes MA. The cognitive phenotype of spina bifida meningomyelocele. Dev Disabilities Res Rev. 2010;16:31–9.
7. Del Bigio MR. Neuropathology and structural changes in hydrocephalus. Dev Disabilities Res Rev. 2010;16:16–22.
8. Miller E, Widjaja E, Blaser S, Dennis M, Raybaud C. The old and the new: supratentorial MR findings in Chiari II malformation. Child's Nervous Syst. 2008;24:563–75.
9. Bartonek A, Saraste H. Factors influencing ambulation in myelomeningocele: a cross-sectional study. Dev Med Child Neurol. 2001;43:253–60.
10. Gutierrez EM, Bartonek A, Haglund-Akerlind Y, Saraste H. Kinetics of compensatory gait in persons with myelomeningocele. Gait Posture. 2005;21:12–23.
11. Bendt M, Bartonek Å. Walking in adults with spina bifida with respect to muscle function. Eur J Physiother. 2016;18:154–60.
12. Thompson RM, Foley J, Dias L, Swaroop VT. Hip status and long-term functional outcomes in spina bifida. J Pediatr Orthopaedics. 2019;39:e168–e72.

13. Takakusaki K. Neurophysiology of gait: from the spinal cord to the frontal lobe. Mov Disord. 2013;28:1483–91.
14. Tita AC, Frampton JR, Roehmer C, Izzo SE, Houtrow AJ, Dicianno BE. Correlation between neurologic impairment grade and ambulation status in the adult spina bifida population. Am J Phys Med Rehabilitation. 2019;98:1045–50.
15. Bartonek A, Saraste H, Knutson LM. Comparison of different systems to classify the neurological level of lesion in patients with myelomeningocele. Dev Med Child Neurol. 1999;41:796–805.
16. Seitzberg A, Lind M, Biering-Sorensen F. Ambulation in adults with myelomeningocele. Is it possible to predict the level of ambulation in early life? Child's Nervous Syst. 2008;24:231–7.
17. Bendt M, Gabrielsson H, Riedel D, Hagman G, Hultling C, Franzen E, et al. Adults with spina bifida: a cross-sectional study of health issues and living conditions. Brain Behav. 2020;10:e01736.
18. Hislop HJ. Daniel´s and Worthingham´s muscle testing. Techniques of manual examination. Philadelphia, PA: Saunders Company; 1995.
19. Kirshblum SC, Burns SP, Biering-Sorensen F, Donovan W, Graves DE, Jha A, et al. International standards for neurological classification of spinal cord injury (revised 2011). J Spinal Cord Med. 2011;34:535–46.
20. Hoffer MM, Feiwell E, Perry R, Perry J, Bonnett C. Functional ambulation in patients with myelomeningocele. J bone Jt Surg Am Vol. 1973;55:137–48.
21. Wechsler D. WAIS-III/WMSIII technical manual: Updated. San Antonio: Psychological Corporation; 2002.
22. Tallberg IM, Ivachova E, Jones Tinghag K, Ostberg P. Swedish norms for word fluency tests: FAS, animals and verbs. Scand J Psychol. 2008;49:479–85. https://doi.org/10.1111/j.1467-9450.2008.00653.x.
23. Heyns A, Negrini S, Jansen K, Moens P, Schelfaut S, Peers K, et al. The prevalence of scoliosis in spina bifida subpopulations: a systematic review. Am J Phys Med Rehabilitation. 2018;97:848–54.
24. Vittinghoff E, McCulloch CE. Relaxing the rule of ten events per variable in logistic and Cox regression. Am J Epidemiol. 2007;165:710–8.
25. Chaves ES, Boninger ML, Cooper R, Fitzgerald SG, Gray DB, Cooper RA. Assessing the influence of wheelchair technology on perception of participation in spinal cord injury. Arch Phys Med Rehabilitation. 2004;85:1854–8.
26. Gabrielsson H, Hultling C, Cronqvist A, Asaba E. Views on everyday life among adults with spina bifida: an exploration through photovoice. Int J Qual Stud Health Well-being. 2020;15:1830702.
27. Guidelines for the care of people with Spina Bifida. available from https://www.spinabifidaassociation.org/: Spina Bifida Association; 2020 [cited 2019 0407].
28. Nasreddine ZS, Phillips NA, Bedirian V, Charbonneau S, Whitehead V, Collin I, et al. The Montreal Cognitive Assessment, MoCA: a brief screening tool for mild cognitive impairment. J Am Geriatrics Soc. 2005;53:695–9.
29. Müller EB, Nordwall A, Odén A. Progression of scoliosis in children with myelomeningocele. Spine. 1994;19:147–50.
30. Agha M, Agha R. The rising prevalence of obesity: part A: impact on public health. Int J Surg Oncol. 2017;2:e17.
31. Hoyer WJ, Stawski RS, Wasylyshyn C, Verhaeghen P. Adult age and digit symbol substitution performance: a meta-analysis. Psychol Aging. 2004;19:211–4.
32. Polfuss M, Bandini LG, Sawin KJ. Obesity prevention for individuals with spina bifida. Curr Obes Rep. 2017;6:116–26.
33. Holmlund T, Ekblom-Bak E, Franzén E, Hultling C, Wikmar LN, Wahman K. Energy expenditure in people with motor-complete paraplegia. Spinal Cord. 2017;55:774–81.
34. Jørgensen V, Butler Forslund E, Opheim A, Franzén E, Wahman K, Hultling C, et al. Falls and fear of falling predict future falls and related injuries in ambulatory individuals with spinal cord injury: a longitudinal observational study. J Physiother. 2017;63:108–13.

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AUTHOR CONTRIBUTIONS

MB, EBF, EF and ÅS designed the study. Authors MB, EBF and GH conducted the data analysis, while all authors (ÅS, EF, CH) contributed to the interpretation of the data. MB drafted the manuscript. All authors revised the manuscript for important intellectual content and approved the final version.

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ETHICS

We certify that all applicable institutional and governmental regulations concerning the ethical use of human volunteers were followed during the course of this research. Verbal and written information was provided before the participants gave their written informed consent. The study was approved by the Regional Ethical Review Board in Stockholm (Dnr: 2014/1111-31).

COMPETING INTERESTS

The authors declare no competing interests.

ADDITIONAL INFORMATION

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