Cognitive status of young adults with spina bifida

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The cognitive status of 168 Dutch young adults (103 females, 65 males; mean age 20 years 9 months, age range 16 to 25 years) with spina bifida (SB) was examined. The main purpose was to establish the effect of the type of SB (occulta or aperta) and the effect of hydrocephalus (HC) within the group with SB aperta (AHC+). Results indicated, on average, a lower cognitive status of persons with AHC+ (n=111) than of persons with SB occulta (n=37) and of persons with SB aperta without HC (AHC– ; n=20). Almost half the young adults with AHC+ had cognitive impairments of some sort. These included more domain specific impairments (70%) as well as a more general cognitive deficit (30%). Cognitive status of persons with SB occulta and of those with AHC– was similar to that in the healthy population. The presence of associated pathology, rather than SB per se, has a negative effect on cognitive status.

The availability of new medical treatment has meant that the life expectancy of patients with spina bifida (SB) has increased and a significant population of young adults with SB has come into existence (Lorber 1971, Ouden et al. 1996, McDonnell and McCann 2000, Bowman et al. 2001). As a result, a growing number of young adults with SB are facing challenges with respect to education, vocation, housing, and relationships. In the terms of the *International Classification of Functioning, Disability and Health* (World Health Organization 2001), restrictions in participation in these domains are not explained by the severity of neurological impairments, rather, indirect effects have been suggested. Neurological impairments, such as the type of SB and hydrocephalus (HC; Loomis et al. 1994), limit neuropsychological (cognitive and affective) functions, and the neuropsychological functions in turn restrict participation (Hommeyer et al. 1999). This study examines the relation between neurological parameters and cognition.

In children with SB under 16 years of age the presence of HC has been identified as the major illness parameter that affects the level of cognitive functioning (Wills 1993). Illness parameters that are related to HC (i.e. the necessity of shunting and number of shunt revisions, encephalitis, epileptic seizures, and additional structural abnormalities of the central nervous system) also affect the level of cognitive functioning (Spain 1974, Tew and Laurence 1975, Dennis et al. 1981, Shaffer et al. 1985, Wills et al. 1990, Friedrich et al. 1991, Wills 1993, Kokkonen et al. 1994, Snow et al. 1994, Holler et al. 1995, Fletcher et al. 1996, Bier et al. 1997, Fletcher et al. 1997, Hunt et al. 1999). Some studies have found that the level of the lesion is related to cognitive functioning (Shaffer et al. 1985, Wills 1993, Bier et al. 1997). Among non-illness parameters, socioeconomic status contributes to differences in the cognitive and social outcome of patients with SB (Bier et al. 1997).

Few studies have addressed the cognitive outcome of persons with SB over the age of 16 years (Friedrich et al. 1993, Loomis et al. 1994, Snow et al. 1994, West et al. 1995, Bier et al. 1997, Dise and Lohr 1998, Mataro et al. 2000). The limitations of these studies are either a selection bias (e.g. participants having been included on the basis of intelligence or of HC), a rather wide age range, or an uncertainty about the presence of HC in the participants. As a result, the proportion of the population of young adults with HC with impairment(s) in intelligence, cognitive flexibility, problem-solving ability, and processing efficiency remains to be established. Insight into the cognitive status and factors affecting the cognitive status of this group when they reach young adulthood will provide paediatricians with a better understanding of the results of treatment that has been given to them.

The purpose of this study was to assess the cognitive status of young adults with SB, including the often-neglected group with SB occulta. In order to explore possible effects of primary neurological parameters, i.e. type of SB (occulta vs aperta) and presence or absence of HC, we divided the group into three subgroups and compared the cognitive data obtained from the three groups. Furthermore, we investigated and compared the effects of secondary parameters (complications of HC) and of associated pathology (epileptic seizures and additional brain malformations). Level of lesion, being associated with both types of SB and HC, was not entered as a separate variable.

See last page for list of abbreviations.

Method

This multicentre study was part of the Adolescents with Spina Bifida in the Netherlands (ASPINE) project, a cross-sectional study of physical and cognitive abilities, health care, participation in society, and life satisfaction of young people with SB. The ethics and research committees of the participating institutions have approved ASPINE.

PARTICIPANTS

To be included, participants had to have SB aperta or occulta (International Classification of Diseases, Ninth revision, Classification of Mental Disorders codes 741 and 756.17 respectively, World Health Organization 1979), be aged between 16 and 25 years, and have sufficient command of the Dutch language. Participants were recruited by 11 of the 12 Dutch Spina Bifida Teams, in coordination with the Dutch Spina Bifida Patients Association, organizations for sheltered homes, and rehabilitation centres. Written invitations to participate were signed by the patient's physicians or by the management of the sheltered homes. Written informed consent was obtained from the participants or their parents, where applicable.

Invitations were sent to 350 prospective participants which yielded 181 (52%) positive responses. Known causes of non-participation were unknown address (29%) and time limitations of the invitee (29%). Participants did not differ from non-participants in age, sex, type of SB, level of lesion, or being shunted for HC.

Ten of the 181 individuals who were willing to participate were only interviewed by telephone; they were not included in the present study. Two participants with comorbidity that could independently induce serious physical and/or cognitive impairments were excluded: one had serious heart disease and one had a chromosome disorder. Participants with pathology associated with SB were not excluded from the analysis, apart from one participant who could not be examined neuropsychologically because of blindness which was acquired after having been shunted for HC early in life. Data for 168 participants were included for the analysis.

MATERIALS

The headings under which the tests are listed below represent the measurement pretensions of the tests for the healthy population. Tests were administered according to their respective manuals.

General intelligence

The Standard Progressive Matrices (Raven 1996, Raven et al. 1998) is a multiple choice test requiring accuracy of discrimination and evaluation of logical relations in visual displays. It provides an index of fluid intelligence (Heaton et al. 1986). We used a 20-minute time limit for the results of which recent Dutch normative scores are available (Bouma et al. 1996). The dependent variable is the number of adequate solutions, corrected for age and sex, and converted to an IQ score. Mean IQ score of the healthy population is 100 (SD 15).

Memory and verbal learning

The Wechsler Memory Scale (Wechsler 1974) provides a global measure of memory. The subtests are: Personal and Current Information, Orientation to Time and Place, Mental Control, Logical Memory, Digit Span, Visual Reproduction, and Associative Learning. In this study only the age-corrected memory quotient (MQ; population mean 100; SD 15) was used as the dependent variable.

The Verbal Learning Test (VLT; Mulder et al. 1996) is a recent Dutch version of the California Verbal Learning Test (Delis et al. 1987). Participants are orally presented with a 'shopping list' (list A) consisting of 16 common nouns drawn from four semantic categories (fruits, clothing, tools, and spices). The items are presented five times (trials) in identical order. In each trial participants recall as many nouns as they can think of. After list A, a second (interfering) list is presented, of which the participant is again asked to recall as many nouns as possible. This is followed by a free and cued recall of list A. In the latter, the participant is provided with the semantic categories of list A. After 20 minutes filled with distracting tests, free and cued recall of list A is again determined. Finally, the participant is asked to identify the 16 nouns of list A among 28 distracting nouns. Thanks to elaborate psychometric studies in the Dutch population (Mulder et al. 1996), learning indices can be compared with population normative values. The indices are corrected for age and sex and normative scores are calculated. The normative scores (z scores multiplied by two), have a mean of 0 (SD 2). For the purpose of this study, we inspected the normative scores and selected the three most informative learning parameters: Total Recall of list A (the total number of recalled items across five trials), Learning slope (the rate of learning of additional items across five trials), and Consolidation (the ability to remember learnt items over a longer period of time).

Executive functioning

In the Wisconsin Modified Card Sorting Test (Nelson 1976), four stimulus cards are presented depicting one of four different shapes, which also differ in colour and number. The response cards have one of three categories – shape, colour, or number – in common with the standard stimuli but none of the cards are identical to the standard. The participant is asked to sort the cards and is informed that the examiner will tell them whether their choice was correct or not. After six correct responses the participant is asked to sort differently. The ability to maintain and to shift a cognitive set is inferred from the number of categories found and applied correctly; we scored the number of correct categories. The maximum is 6, which is also the expected number in individuals without cognitive impairment.

The Trail Making Test (TMT; Reitan and Wolfson 1996) is a paper and pencil test. Part A requires the participant to connect 25 circles, placed randomly on an A4 sheet of paper and numbered 1 to 25, in sequential order. In part B, 13 of the 25 circles are numbered from 1 to 13 and 12 contain one of the first 12 letters of the alphabet. The participant has to connect the numbers and letters as quickly as they can, alternating the sequences. The time to complete the task is recorded for both parts. We took the time (in seconds) for completing part B and the time difference between part B and part A (B–A) as measures of divided attention (Corrigan and Hinkeldey 1987).

Word production according to lexical rules (UNKA test; Jennekens-Schinkel et al. 1990) addresses the interface of memory, language, and behavioural regulation. The participant is asked to generate as many words as they can think of that begin with the prescribed letters (U, N, K, A). Production time is 60 seconds per letter. The dependent variable is the

total number of correct words. Data on the performance of the typically developing population is available (A Jennekens-Schinkel, personal communication 2001).

Reaction time

Reaction time was measured with the Time Tapper test (Fetrics, Holland VOF). Participants were asked to react as quickly as possible to stimuli appearing on a panel in front of them by lifting their finger from the rest button, moving to and pressing the reaction button, and then returning to the rest button. In the two simple types, single stimuli were presented (respectively a green light and a tone) and the participant had to react to every stimulus. The third and fourth types were of the 'go–no–go' type. Stimuli were presented singly or in combination and the participant had to react only to a target subset (either of an intra- or of an intermodal nature). The dependent variables were the median decision times (numbers of milliseconds between stimulus presentation and lifting the finger from the rest button) and the median motor times (numbers of milliseconds between lifting the finger from the rest button to touching the reaction button) averaged across the four conditions.

DATA COLLECTION

Medical records were examined according to a fixed protocol and participants underwent a physical examination by a clinician. They were interviewed in a semi-structured way and were assessed by a neuropsychologist. Data were gathered with respect to type of SB, level of lesion (defined as the lowest completely unimpaired level on both sides measured with sensitivity to pin prick and light touch), HC, number of shunt revisions, ventriculitis, epilepsy, corpus callosum malformation, cerebral bleeding or ischaemia, total number of surgical interventions other then shunting and shunt revisions, ambulation (Hoffer et al. 1973), incontinence (defined as wetting at least once a month), living arrangement, and educational/vocational status.

The total examination was conducted within 3½ hours. Participants were randomly picked for examination, so that half of the participants started with the physical examination and the other half started with the neuropsychological examination. The order of the neuropsychological tests was fixed and was ordered in such a way that there would be no interference between the different tests. The 1 hour 30 minute neuropsychological examination included a short break. For logistical reasons, participants were split randomly into two groups. Both groups were administered the tests of intelligence, memory, and executive function. Additionally, one group was administered the VLT and the other group the reaction speed tasks. Twenty-five participants did not complete the test battery, mostly because of lack of time. For these participants the missing values were substituted by the mean score of the total sample on that particular test.

DATA ANALYSIS

Statistical analysis was performed using SPSS for Windows (version 10.0.7). Illness characteristics and sex of the three clinical groups of participants were compared using the χ^2 test. Age was compared with analysis of variance (ANOVA).

Because group size was small for some variables, and because some variables violated the assumption of normality, cognition and speed of the three clinical groups were compared by means of ANOVA and, if appropriate, by the non-parametric Kruskal–Wallis test. Statistically significant group differences were analyzed post hoc using the independent samples *t*-test with a Bonferroni correction (alpha/number of tests) and, if appropriate, the Mann–Whitney *U* test. Alpha was set at 0.05. Parametric and non-parametric analyses did not yield any different results, therefore we only report the parametric data.

For case finding, the clinical significance of poor performances was defined by setting a cut-off point and considering scores from 2SD worse than the population mean of the variable as 'deviation scores' in the IQ, MQ, VLT, UNKA tests. In the absence of suitable population data, the reference value was assigned to the group with SB occulta for the TMT-B, TMT-(B-A), and reaction speed measures. For the Wisconsin Modified Card Sorting Test, the clinically significant cut-off point of five categories was used because failing to find all six categories is pathognomonic for impairment. Between the three clinical groups, group differences in the proportions of participants who obtained deviation scores were compared using the χ^2 test. Furthermore, we summed the deviation scores to a 'total cognitive deviation' score, in order to explore the range (0 to 6) across cognitive domains.

For analysis of the effect of number of drain revisions and of associated pathology on the cognitive status within the group of persons with SB aperta with HC (AHC+), we used the cognitive measure that correlated most strongly with all other cognitive measures, i.e. IQ. Statistical analyses were performed using the independent samples *t*-test.

Results

Demographic and neurological characteristics of the groups of persons with SB occulta, SB aperta without HC (AHC–), and SB aperta with HC (AHC+) are summarized in Table I. Sixty percent of the participants were female. The AHC– had a significantly smaller proportion of females than the two other groups ($p=0.043$). Mean age of all participants was 20 years 9 months; age was not significantly different between the groups. All AHC+ participants had been shunted for HC early in life. The majority (63%) among them having had two or more shunt revisions. Early in life, one of 37 participants in the SB occulta group and five of 20 participants with AHC– had had transient signs of raised intracranial pressure which had never required shunting. As they did not differ from those without HC, these six participants with presumably compensated HC were treated as non-hydrocephalic participants for the analyses of cognition and reaction speed. The distribution of levels of lesion was similar for persons with SB occulta and AHC–. Among persons with AHC+ the proportion of high level lesions was significantly larger ($p<0.001$) than among persons without HC. The measure of collinearity (Cramer's Φ) was 0.406 between type of SB and level of lesion, 0.742 between type of SB and HC, and 0.585 between level of lesion and HC. Pathology associated with SB, i.e. encephalitis, epilepsy, corpus callosum malformation, cerebral bleeding, and ischaemia of the brain, were uncommon and not restricted to persons with AHC+. Persons with AHC+ had undergone more surgical interventions than persons with SB occulta ($p=0.002$). Persons with HC were more often wheelchair dependent and incontinent than those without HC ($p<0.001$). Also, they lived more often in sheltered homes, were more often unemployed or had sheltered work, and had received more special education ($p<0.001$).

ANOVA revealed significant differences between the groups of persons with SB occulta, AHC, and AHC+ on all but one of the cognitive and speed tasks (Table II, Fig. 1). Post-hoc analysis showed no significant difference between the groups with SB occulta and AHC–. Mean scores of individuals with AHC+ on all but one test were significantly worse ($p<0.01$) than of both SB occulta and AHC–. Figure 1 clearly shows that cognitive functioning of persons with SB occulta and AHC– was similar and near to normal and that cognitive functioning of persons with AHC+ was below average in most domains. Specifically, for AHC+ individuals' IQ ranged from 55 to 117, whereas for persons with AHC– , IQ ranged from 69 to 117.

Table I: Demographic and neurological characteristics of study sample

Characteristics	*SB occulta n=37 n (%)*	*AHC– n=20 n (%)*	*AHC+ n=111 n (%)*
Sex			
Female	25 (68)	7 (35)	69 (62)
Male	12 (32)	13 (65)	42 (38)
Age, mean (SD)	20.6 (3.2)	21.0 (3.1)	20.7 (2.9)
Hydrocephalus			
Shunted	0	0	111 (100)
Compensated	1 (3)	5 (25)	0
No	36 (97)	15 (75)	0
Level of lesion			
≥L2	5 (14)	2 (10)	62 (56)
L3 – L5	14 (38)	7 (35)	44 (40)
≤S1	18 (49)	11 (55)	5 (5)
Shunt revisions			
No shunt or no revision	37 (100)	20 (100)	16 (14)
1	–	–	25 (23)
2–4	–	–	35 (32)
More than 5	–	–	35 (32)
Corpus callosum			
Malformation	1 (3)	1 (5)	7 (6)
Bleeding/ischaemia			
Yes	–	–	1 (1)
Encephalitis			
Yes	2 (5)	–	10 (9)
Epilepsy			
Yes	2 (5)	–	11 (10)
Total nr of surgical interventions			
Mean (SD)	4.9 (3.4)	8.3 (7.8)	8.7 (5.9)
Ambulation			
Wheelchair dependent	2 (5)	3 (15)	75 (68)
Continence			
Incontinence	14 (38)	11 (55)	90 (81)
Living arrangements			
With parents	21 (57)	11 (55)	54 (49)
Sheltered home	1 (3)	2 (10)	42 (38)
Independently alone	7 (19)	7 (35)	12 (11)
Independently together	8 (22)	–	3 (3)
Vocational status			
Unemployed	3 (8)	2 (10)	25 (23)
Student ordinary education	21 (57)	11 (55)	32 (29)
Student special education	2 (5)	2 (10)	23 (21)
Paid normal work	11 (30)	5 (25)	18 (16)
Sheltered work	–	–	13 (12)

Two participants with corpus callosum malformations had had encephalitis, two other participants with corpus callosum malformations had antiepileptic medication, and one participant with a corpus callosum malformation had had a cerebral bleed as a result of shunting for HC. AHC–, spina bifida aperta without hydrocephalus, AHC+, spina bifida aperta with hydrocephalus.

The majority of deviation scores (Table III, Fig. 1) were obtained by The AHC+ participants: about one-fifth had an IQ or MQ below 70. Between one-third and one-half of AHC+ participants had a deviation score on the VLT. Almost half failed on the Wisconsin Modified Card Sorting Test. A quarter deviated on TMT-B; when corrected for completion time of part A, this percentage dropped to 12%. Only 7% had a deviation score on the UNKA. As far as reaction speed was concerned, deviating decision times were found in 44% of The AHC+ participants and deviating motor times in one quarter of AHC+ participants. Participants of both other groups obtained considerably fewer deviation scores. Four participants with SB occulta had deviation scores in consolidation (VLT). Among those with AHC–, three of 20 participants had a deviation score on the Wisconsin Modified Card Sorting Test and three of 12 had a deviation score on both decision time and motor time of the reaction speed task. When exploring generality versus specificity of the deviations, six persons with AHC– had one deviation score and none had 2 or more deviation scores. Similar to persons with AHC–, of the persons with SB occulta, 24% had one or more deviation scores and one person had 2 deviation scores. In contrast, 79% of the persons with AHC+ had one or more deviation scores, 44% had 2 or more deviation scores, and 15% had a more general deviation with 4 or more deviation scores.

IQ significantly correlated with all but one of the other cognitive scores. Pearson's r ranged from 0.21 to 0.73. The majority of the correlations (7 of 10) were higher than 0.40. IQ was found to be the most robust measure of general cognitive status. Within the group with AHC+, mean IQ decreased with increasing number of shunt revisions: 0 or 1 revision (mean IQ=86), 2 to 4 revisions (mean IQ=86), and 5 or more revisions (mean IQ=77). Post-hoc analysis showed the difference between the groups with no or one shunt revision and with five or more shunt revisions to be significant ($p<0.0167$).

As far as associated pathology was concerned, when grouping together all AHC+ participants with associated pathology and comparing this group ($n=24$) with the group without associated pathology, a statistically significant IQ difference appeared: former group mean IQ 75 versus mean IQ 85 for the latter group ($p=0.003$). Further analyses showed that in particular, corpus callosum malformation (four participants of seven vs 17% of those without corpus callosum malformation) with epilepsy (six participants of 11 vs 16% of those without epilepsy) was related to IQs below 70. Of the five persons with associated pathology but without HC, the one who had had encephalitis had a low-average IQ of 84. IQs of the other four individuals were average.

Discussion

The purpose of the present study was to assess the cognitive status of young adults with SB aperta or occulta, a sparsely studied issue which is, however, relevant to establishing the effects of SB and HC on cognitive status later in life and for understanding psychosocial outcome (Hommeyer et al. 1999).

Three participant groups were distinguished: SB occulta, AHC+, and AHC–. Our results indicate that the cognitive status

of persons with AHC+ is in the below-average range when compared with the population mean. Moreover, the performances of persons with SB occulta and AHC– are similar to those in the typically developing population. The conclusion is that the presence of HC is the main culprit for a poor cognitive status and SB aperta, per se, has no negative effect.

As for the group of persons with SB occulta, the conclusion that the cognitive abilities of persons in this group are normal confirms the findings of Friedrich et al. (1993) in a previous study using a small number ($n=10$) of patients with lipomyelomeningocele. However, it should be noted that this study included three persons with HC. Also in our study, the group of participants with SB occulta did not consist only of persons with lipomyelomeningocele but also included other types of SB occulta. We now conclude that in the wider population of individuals with SB occulta, cognitive abilities are similar to those of the general population.

As mentioned earlier, the group with AHC– performed similarly to individuals with SB occulta on the cognitive tests. Few deviation scores were found and none of the persons with AHC– had 2 or more deviation scores. Together with Friedrich et al. (1991), we conclude that those with AHC– do not have serious cognitive problems. Considering the small number of participants in this subgroup, further research is warranted.

The group of individuals with AHC+ scored below average on most cognitive tasks. The findings are roughly similar to those described for children (Mapstone et al. 1984, Wills et al. 1990, Wills 1993, Yeates et al. 1995, Fletcher et al. 1997, Snow 1999) and older persons (West et al. 1995, Bier et al. 1997, Dise and Lohr 1998, Hommet et al. 1999). Differences in operational methods hamper specific comparisons. Although persons with AHC+ scored below average as a group, it should be noted that half of the AHC+ participants had a (near to) normal cognitive status. This finding adds to the available literature on the heterogeneity of those with SB (Wills 1993, Snow et al. 1994, West et al. 1995) supporting the notion that the prototypical SB patient is an untenable abstraction.

Cognitive status is not influenced solely by the presence of HC. Corpus callosum malformation and epilepsy appear to be negatively related to intelligence. Also, in line with previous findings (Holler et al. 1995) but contradicting recent findings (Ralph et al. 2000), within the group of persons with HC, having had five or more shunt revisions had a negative effect on IQ. It should be noted that in this study both a higher level of lesion and having associated pathology is positively related to the number of shunt revisions (data not shown). It may well be that the associated pathology is the major disruptive factor in cognition. Epilepsy was rare in the present study and its effect could not be assessed independently, but the finding of lower IQ in those with associated pathology (including epilepsy) may be understood on the same basis as the negative effect of seizures (Ralph et al. 2000).

Three features of the present study have to be mentioned. Every effort was made to approach all those with SB, including those with SB occulta, and to examine every individual we were able to trace. Non-response analysis showed that the participant group was similar to the non-participant group on important demographic and illness characteristics. Therefore, we feel that our findings can be generalized over

Table II: Mean score and 95% CI on cognitive tests for participants with spina bifida (SB) occulta, and SB aperta without (AHC–) and with (AHC+) hydrocephalus

Test	*Measure*	*SB occulta*	*AHC–*	*AHC+*	*n*
Raven SPM	IQ	97[a]	93[b]	83[a,b]	37/20/111
		93–101	88–98	80–86	
WMS	MQ	108[a]	100[b]	91[a,b]	37/20/111
		104–113	96–104	88–94	
VLT	List A total recall	-0.4[a]	-0.5[b]	-4.0[a,b]	23/10/54
		-1.3–0.5	-1.6–0.6	-4.6 – -3.4	
	Learning slope	-0.5[a]	-0.1[b]	-3.2[a,b]	
		-1.5–0.5	-1.3–1.1	-3.9 – -2.5	
	Consolidation	-1.0[a]	-0.4[b]	-2.5[a,b]	
		-1.9–0.0	-1.3–0.5	-3.2 – -1.8	
WMCST	Total categories	6.0[a]	5.8[b]	4.9[a,b]	37/20/111
		–	5.6–6.0	4.6–5.2	
TMT	Time part B (s)	61[a]	67[b]	91[a,b]	37/20/110
		53–69	57–77	83–99	
	Difference B–A (s)	35	41	47	
		28–42	33–49	40–54	
UNKA	Total words	48[a]	42	38[a]	37/20/111
		43–54	36–47	35–40	
RT	Mean decision time (ms)	373[a]	392[b]	445[a,b]	15/12/60
		353–393	359–425	429–460	
	Mean motor time (ms)	120[a]	127	142[a]	
		109–130	107–146	134–150	

[a]Significant difference between SB occulta and AHC+, $p<0.0167$; [b]significant difference between AHC– and AHC+, $p<0.0167$; SPM, Standard Progressive Matrices (Raven 1996, Raven et al. 1998); WMS, Wechsler Memory Scale (Wechsler 1974); MQ, memory quotient; VLT, Verbal Learning Test (Mulder et al.1996); WMCST, Wisconsin Modified Card Sorting Test (Nelson 1976); TMT, Trail Making Test (Reitan and Wolfson 1996); UNKA, word production according to lexical rules (Jennekens-Schinkel et al. 1990); RT, reaction times.

Table III: Numbers (%) of deviation scores found in groups of young adults with spina bifida (SB) occulta and SB aperta without (AHC–) and with hydrocephalus (AHC+)

Test	*Measure*	*SB occulta*	*AHC–*	*AHC+*
Raven SPM	IQ	1 (3)	– (0)	22 (20)[a]
WMS	MQ	– (0)	– (0)	16 (14)[a]
VLT	List A total recall	2 (9)	– (0)	22 (41)[a]
	Learning slope	1 (4)	– (0)	21 (39)[a]
	Consolidation	4 (17)	– (0)	12 (22)[a]
WMCST	Total categories	– (0)	3 (15)	50 (45)[b]
TMT	Time part B	2 (5)	– (0)	26 (23)[c]
	Difference B–A	3 (8)	– (0)	13 (12)[c]
UNKA	Total number of words	– (0)	– (0)	10 (9)[a]
RT	Mean decision time	– (0)	3 (25)	34 (57)[c]
	Mean motor time	– (0)	3 (25)	15 (25)[c]
Total cognition deviation score	Number of deviation scores			
	0	27 (73)	14 (70)	24 (22)
	1	9 (24)	6 (30)	38 (34)
	2	1 (3)	– (0)	21 (19)
	3	– (0)	– (0)	12 (11)
	4	– (0)	– (0)	10 (9)
	5	– (0)	– (0)	4 (4)
	6	– (0)	– (0)	2 (2)

[a]2SD below population mean; [b]below maximum score; [c]2SD below mean SB occulta. SPM, Standard Progressive Matrices (Raven 1996, Raven et al. 1998); WMS, Wechsler Memory Scale (Wechsler 1974); MQ, memory quotient; VLT, Verbal Learning Test (Mulder et al. 1996); WMCST, Wisconsin Modified Card Sorting Test (Nelson 1976); TMT, Trail Making Test (Reitan and Wolfson 1996); UNKA, word production according to lexical rules (Jennekens-Schinkel et al. 1990); RT, reaction times.

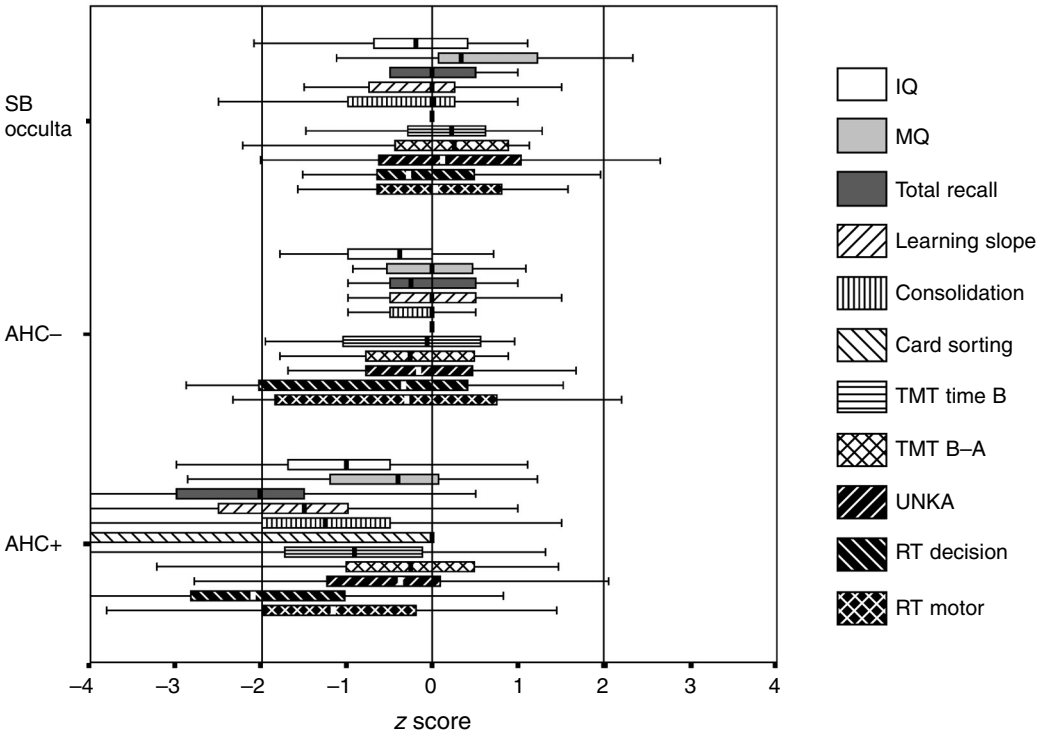

Figure 1: *Cognitive status in relation to type of spina bifida and hydrocephalus. For purpose of graphical display by means of boxplots, all scores were converted to z scores. Hinges mark 25th and 75th centiles and length of box corresponds with interquartile range. Whiskers show range of values falling within 1.5 of hinges. SB, spina bifida; AHC–, SB aperta without hydrocephalus; AHC+, SB aperta with hydrocephalus; MQ, memory quotient; TMT, Trail Making Test (Reitan and Wolfson 1996); UNKA, word production according to lexical rules (Jennekens-Schinkel et al. 1990); RT, reaction times.*

the population of young adults with SB in the Netherlands. Of course, only those patients were included who were diagnosed with SB occulta at birth or later on in life when the defect caused physical complaints. There certainly are many persons with undiscovered SB occulta. This selection bias will probably make little difference to cognitive outcome, in which we found the full breadth of normality in this group.

The second feature to be mentioned is that we did not exclude patients with an IQ below 70. We insisted on including these participants for the reason that learning disability* is a possible outcome of the neural tube defect. Excluding participants with learning disability would possibly lead to a too optimistic view of cognitive status.

The third feature is the application of an arbitrary but very strict rule for determining deviation scores. We wanted to minimize the chance of overestimating the true state of affairs regarding cognitive deficits. Our results may, therefore, even underestimate the number of persons with a cognitive deficit.

One problem with our study was that logistic reasons necessitated the population to be randomly split into two groups which were administered a core battery of tests (assessing intelligence, memory, and executive functioning) and part of the remaining tests (verbal learning or reaction speed). The missing values restrained the analysis of the group data and prohibited statistical analysis of the deviation sum score. On some occasions participants could not complete all of the assessment. By substituting a missing value on a particular test by the mean score of the total sample we tried to avoid biasing the results in favour of detecting deviating scores in the clinical subgroups.

A final shortcoming of this study was that reference scores for a healthy, age-appropriate population were not available for all tests. Based on the clinical intuition that patients with SB occulta are not different from the typically developing population in major aspects of cognitive functioning, we took the mean score and standard deviation of the occulta group as a reference. Our choice was supported by the data on tests for which normative scores are available.

Conclusion

Our results suggest that the cognitive status of young adults with SB occulta and AHC– is normal. Almost 50% of young adults with AHC+ have a cognitive impairment of some sort. Cognitive status appears to be negatively influenced by multiple shunt revisions and, particularly, by pathology associated with HC.

Future research should concentrate on a more detailed examination of cognition and should include a more qualitative analysis of the performance, for instance by a comprehensive error analysis. Further research should also concentrate on the consequences of cognitive impairments for education, vocation, and social participation.

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*US usage: mental retardation.

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List of abbreviations

AHC+	Spina bifida aperta with hydrocephalus
AHC–	Spina bifida aperta without hydrocephalus
HC	Hydrocephalus
MQ	Memory quotient
SB	Spina bifida
TMT-A	Trail Making Test part A
TMT-B	Trail Making Test part B
UNKA	Test of word production according to lexical rules
VLT	Verbal Learning Test