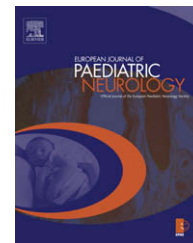




Official Journal of the European Paediatric Neurology Society



Original article

Motor profile and cognitive functioning in children with spina bifida

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ARTICLE INFO

Article history:

Received 14 May 2008

Received in revised form

17 January 2009

Accepted 19 January 2009

Keywords:

Spina bifida

Motor development

Cognition

Neuropsychology

ABSTRACT

Background: Spina bifida is a complex neuroembryological disorder resulting from incomplete closure of the posterior neural tube. Morbidity in the different fields of motor and cognitive neurodevelopment is variable in nature and severity, and often hard to predict. **Aims:** The current study investigates the relationship between cognitive functioning, fine motor performance and motor quality in children with spina bifida myelomeningocele (SBM) and SB-only, taking into consideration the cerebral malformations.

Material and methods: Forty-one children were included (22 girls and 19 boys aged between 6 and 14 years, mean age 10;0 years) in the study. A comprehensive assessment was conducted of cognitive functioning and motor profile, including fine motor and visual-motor functioning, and motor quality. The performance outcomes were analyzed for the total group of children and separately for the nonretarded children (FSIQ ≥ 70 , $N = 30$) to eliminate the influence of global intellectual impairment.

Results: Although the children with spina bifida showed increased incidence of cognitive and fine motor impairment, and impaired motor quality, after exclusion of the overall retarded children no associations were found between cognitive functioning and motor profile. In the comparison of SBM to SB-only specific differences were found for performance IQ, visual-motor functioning and motor quality, but not fine motor functioning.

Conclusion: Our findings underscore the role of cerebral malformation in spina bifida and its consequences for neuropsychological functioning. The complicated developmental interactions found strengthen the need for an individualized management of children with SB.

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Abbreviations: SB, spina bifida; HC, hydrocephalus; SBM, spina bifida myelomeningocele; ACM, Arnold–Chiari II malformation; CCD, MRI, magnetic resonance imaging; WISC, Wechsler Intelligence Scale for Children; VMI, visuospatial integration; VIQ, verbal intelligence quotient; PIQ, performance intelligence quotient; FSIQ, full-scale intelligence quotient; MD, manual dexterity.

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doi:10.1016/j.ejpn.2009.01.003

Please cite this article in press as: Vinck A et al., Motor profile and cognitive functioning in children with spina bifida, European Journal of Paediatric Neurology (2009), doi:10.1016/j.ejpn.2009.01.003

1. Introduction

Spina bifida (SB) is a neuroembryological disorder with complex physical and neuropsychological morbidity. The majority of children with myelomeningocele (SBM), the severe form of SB, develop hydrocephalus and associated cerebral malformations of the posterior cortex and white matter, midbrain, cerebellum, and corpus callosum.^{1–3} Although children with SBM tend to score within the low average to average range of general intelligence measures, they are at risk of specific cognitive difficulties, learning disabilities and motor deficits.^{4–6} Gross motor deficits are prominent and fine motor deficits are common.

According to the neuropsychological literature on SBM, motor abilities can be decomposed into visual-spatial and perceptual-motor abilities, gross and fine motor skills, and quality of motor movements.⁷ Apart from spinal lesions affecting limb functioning, physiological studies have revealed a diversity of relations between central nervous system (CNS) dysfunction or damage and motor limitations. Without trying to be exhaustive, we mention the following relations relevant for SBM. Hydrocephalus and dysfunction of the posterior parietal cortex, as well as corpus callosum anomaly can result in poor visual-spatial and perceptual-motor abilities. Damage to the cerebral motor cortex, pyramidal tract abnormalities, and cerebellar dysfunctions mainly influence gross and fine motor skills. Motor quality is particularly related to the functioning of the basal ganglia and the cerebellum.⁷

In children with SBM any of the CNS dysfunctions mentioned above can play a role to a certain degree. Therefore, a major issue that has both theoretical and clinical relevance is the involvement of cognitive and motor impairment, and the question to what extent these are interrelated. Erickson et al.⁷ distinguished between infancy (up to 5 years) and school-aged children, and found different types of relationships for the younger as compared to the older groups. Some authors attribute the children's poor fine motor skills to impaired hand coordination, a decrease in muscle force,^{8,9} impaired fingertip force^{9,10} and impaired motor quality,¹¹ which includes a kinetic tremor and dysmetria. These deficient fine motor skills result in poor handwriting and drawing and low legibility.^{11–14} Deficits in visual-motor integration and perceptual-motor coordination^{5,6,15} which require visual-spatial insight on the one hand, and adequate motor skills and motor quality on the other, are also consistently reported in children with SBM.

Even though more in-depth knowledge of motor functioning and quality in relation to cognitive and visual-motor impairment is indispensable for prognostic counselling and can have far-reaching implications for treatment strategies, in the literature information about the exact nature of the relationship between the various function domains in children with SB is as yet lacking. In the present study the motor functions of school-aged children with SB are measured extensively to explore to what extent motor performance and quality are related to deficiencies in the visual-motor domain and to the overall cognitive impairment. Secondly, to assess the role of the cerebral malformations in the observed deficits,

we compared the performance of the children with SBM to those with SB-only.

2. Material and methods

2.1. Patients

Eligible were 78 children diagnosed with SB born between 1988 and 1997 who had been referred for surgery to the Radboud University Nijmegen Medical Centre. Thirty-seven candidates were not included due to refusal to participate, insufficient command of the Dutch language, and loss of follow-up data. The study group thus comprised 41 children (22 girls and 19 boys aged between 6 and 14 years, mean age 10;0 years), of whom 22 had cerebral malformation. Three children could not be assessed neuropsychologically due to profound retardation and multiple disabilities.

All children had undergone spinal surgery and, if necessary, were shunted within the first months of life (Table 1).

The main diagnostic criterion was the presence of a defect in the closure of one or more vertebral arches in combination with a median skin defect and/or a cystic or lipomatous lump on the back, and/or a developmental anomaly of the spinal cord confirmed by magnetic resonance imaging (MRI). SB was specified according to the following characteristics: (1) type of spinal anomaly: open or closed; (2) cerebral malformation: hydrocephalus (HC), and/or Arnold–Chiari II malformation (ACM), and/or corpus callosum dysgenesis; and (3) neurological impairment of the lower part of the body. Hydrocephalus was diagnosed based on specified features on MRI or cranial computerized tomography (CCT). For the diagnosis of Arnold–Chiari II malformation MRI was taken as the only diagnostic instrument as CCT was considered insufficient. For seven (Table 1) of the 41 children a classification of corpus callosum dysgenesis was missing. Neurological impairment of the lower part of the body was scored as the uppermost affected spinal segment with decreased sensibility and/or decreased intentional movement.¹⁶ Sensibility was defined as observable reactions to pin prick and light touch. Intentional movement was defined as non-stereotypical, non-reflex movement.

Our study was approved by the Regional Committee on Research Involving Human Subjects and written consent was obtained from all parents.

2.2. Procedures

The same neuropsychologist assessed the cognitive and visual-motor functions of all children. In a separate visit two experienced paediatric physiotherapists assessed the children's motor functions. All tests comprised standardized measures with published age-based normative data.

2.3. Cognitive and visual-motor measures

Cognitive functioning was assessed using the Wechsler Intelligence Scale for Children (WISC-III), a general intelligence assessment battery for children aged between 6 and 16 years.

Table 1 – Patient characteristics specified for the complete and the nonretarded (FSIQ \geq 70) group.

	Complete group (N = 41)	Nonretarded group (FSIQ \geq 70, N = 30)	
		Cerebral malformations (N = 12)	No cerebral malformations (N = 18)
Age at spinal surgery (weeks)	Median: 1.0 25th percentile: 0.0 75th percentile: 56.0	Median: 0.0 25th percentile: 0.0 75th percentile: 1.0	Median: 65.5 25th percentile: 32.0 75th percentile: 121.25
Type of defect	MMC = 25 LMS = 13 Atypical SB = 3	Open = 12 Closed = 0 Atypical SB = 0	Open = 3 Closed = 12 Atypical SB = 3
Hydrocephalus	22/41	12/12	0/18
Arnold–Chiari II malformation confirmed by MRI	22/41	12/12	0/18 (1 unknown ^a)
Callosal dysgenesis confirmed by MRI	6/41 (7 unknown ^a)	4/12 (4 unknown ^a)	0/18 (1 unknown ^a)
Mean level of paresis	L2 (range: no impairment – T12)	L2 (range: no impairment – S2)	L2 (range: no impairment – S3)
Impaired cognitive functioning (FSIQ < 85)	18/38 ^b	6/12	5/18
Impaired fine motor functioning (MD-total > 5)	22/38 ^b	5/12	7/18
Impaired visuomotor integration (VMI < 85)	10/38 ^b	2/12	2/18
Motor quality: kinetic tremor	25/37 ^b	8/12	9/18
Motor quality: dysmetria	21/37 ^b	6/12	7/18
Sex	22 female, 19 male	4 female, 8 male	11 female, 7 male
Mean age (age range) (years; months)	10;0 (6;4–14;11)	9;9 (6;4–12;1)	9;10 (6;4–14;11)

MMC = myelomeningocele; LMS = lipomyeloschizis; SB = spina bifida; FSIQ = full-scale intelligence quotient; MD-total = manual dexterity total score; VMI = visual-motor integration; L = lumbar; T = thoracic.

a Unknown because callosal dysgenesis was not classified (six cases) or no MRI was performed (one case).

b The three non-testable children are not included, but were severely impaired in all domains.

The test comprises six verbal and seven performance subtests generating three different measures: verbal intelligence (VIQ), performance intelligence (PIQ), and full-scale intelligence quotient (FSIQ).¹⁷

Visual-motor integration measures were obtained from Beery's VMI¹⁸ and the 'disks' subtest of the Revised Amsterdam Children's Intelligence Test (RAKIT).¹⁹ In the VMI geometric patterns (e.g. a square, cube or circle) have to be copied by drawing and task difficulty is manipulated by increasing pattern complexity and by combining patterns. Correct reproductions were scored according to age norms. In the 'disks' test a playboard comprising nine fields is presented with each field containing two, three, or four differently arranged vertical pegs. The subject is given nine perforated disks with corresponding holes with the request to slip the disks over the corresponding peg arrangements on the playboard. The time it takes to complete the task is compared to age norms.

2.4. Fine motor functioning and motor quality measures

Fine motor functioning was measured with the Movement Assessment Battery for Children (MABC),^{20,21} an instrument evaluating gross and fine motor functioning in children aged between 4 and 12+ years. Because not all our children were able to perform the gross motor scale, only fine motor functions were evaluated using the manual dexterity (MD) scale,

which comprises three subtests: a movement speed task for both hands separately, a bi-manual coordination task, and an eye–hand coordination (drawing) task with the preferred hand. Scores per subtest range from 0 (good; 75% of age-related healthy peers) to 5 (very poor; 2% of age-related peers).

The quality of fine motor performance (kinetic tremor and dysmetria) was measured using the output on the MD scale. If oscillatory movements were present in the tracklayer of the scale's third (i.e. drawing) task, the child was classified as exhibiting kinetic tremor. Dysmetria was defined as an impaired ability to accurately control the range of movement during task execution.²² The presence of dysmetria was scored based on the standardized scoring system of the MABC evaluating the quality of motor performance during task execution. Criteria for dysmetria were: dysfluency, lack of adaptation to directional changes and inadequate accuracy, force and time regulation in two of the three MD tasks. The presence or absence of kinetic tremor and dysmetria during drawing was scored independently by the two physiotherapists. Disparate outcomes were discussed until a consensus was reached. As to the kinetic tremor scores, there was interrater agreement for 32 of the 37 children tested. The final decision in the five conflicting cases was based on the regularity in the oscillation movements. In the scoring of dysmetria proper adherence to the MABC scoring system with its clearly defined decision criteria had resulted in a 100% agreement.

Motor speed was assessed with the baseline speed subtest of the Amsterdam Neuropsychological Test Battery (ANT),²³ in which the children needed to press a button in response to a visual stimulus as fast as possible with the index finger of either the left or the right hand. Reaction times of the two hands are recorded and scored separately.

2.5. Statistical analysis

To investigate the effects of cerebral malformation children with SBM were compared to children with SB-only. To establish whether the proportion of children with impaired function, defined as one standard deviation below the mean, in this patient population would differ from the percentages expected on the basis of the normal distribution of scores, χ^2 tests were performed for FSIQ, VIQ, PIQ, MD-total, and VMI. To address the possibly confounding effect of cognitive impairment, we conducted separate analyses after having excluded the data of the children with a FSIQ below 70. Because the Shapiro–Wilk's test of normality showed half of the variables to be not normally distributed, nonparametric descriptive statistics and tests were applied. Table 1 presents the patient characteristics of the complete ($N = 41$) and the nonretarded group ($N = 30$).

To examine the relationship between the continuous variables of cognitive functioning (VIQ, PIQ, FSIQ), VMI, and MD, nonparametric Spearman's (ρ) correlation coefficients were calculated.

Mann–Whitney U tests were used to test the differences in cognitive functioning, visual-motor integration, fine motor functioning and speed between SBM and SB-only within the nonretarded group.

The abovementioned classification of scores as impaired or normal was used to examine associations between the different variables. For the three MD tasks of the MABC (MD-total) the sum score varied between 0 and 15, with higher scores indicating poorer performance. Children with a total score above 5 (corresponding to a percentile-score lower than 15 compared to the normal distribution of scores) were categorized as suffering from fine motor impairment.

Finally, odds ratios (ORs) were calculated for the non-retarded group to investigate associations between poor

cognitive functioning, impaired visual-motor integration and impaired fine motor output. Also, associations between these variables and cerebral malformation, kinetic tremor and dysmetria were analyzed. As the study population was relatively small for these types of analyses, we used the mid-p exact method to compute the 95% confidence intervals for the ORs using Episheet.²⁴

All other statistical analyses were performed with SPSS for Windows version 12.0.1 (SPSS Inc., Chicago, IL, USA) applying a significance level of $\alpha < 0.05$ (two-tailed).

3. Results

Table 1 provides an overview of the impaired cognitive and fine motor functions, impaired visual-motor integration, and poor fine motor quality for the complete group ($N = 41$) and for the nonretarded group ($N = 30$), specified for the SBM and SB-only patients. Sample sizes for the different variables vary since not all children were able to complete all tests.

For the complete group more than half of the patients were classified as impaired on fine motor functioning and motor quality; according to chi-square test, this deviates significantly from the expected test statistic. Although less outspoken, the results for the nonretarded group still showed a statistically significantly higher than expected incidence of impairment on fine motor functioning and motor quality.

3.1. Correlations between cognitive, motor and other performance outcomes

For the complete group strong and statistically significant correlations were found between the IQ measures, the VMI and MD-total (Table 2). The scatterplot, as depicted in Fig. 1, shows that in the complete group the strong correlation between FSIQ and MD-total was caused by six children with both severe global cognitive impairment (FSIQ < 70) and fine motor deficits (MD-score ≥ 12). Therefore, subsequent analyses were all performed on the nonretarded group only. After exclusion of the severely retarded children, correlations with MD-total were no longer statistically significant (Table 2).

Table 2 – Correlation coefficients for the complete and the nonretarded (FSIQ ≥ 70) group.

		VIQ	PIQ	FSIQ	VMI	MD-total
Cognitive functioning	VIQ	1	0.45 ^b	0.84 ^a	0.37 ^b	-0.05
	PIQ	0.69 ^a	1	0.84 ^a	0.63 ^a	-0.35
	FSIQ	0.92 ^a	0.91 ^a	1	0.58 ^a	-0.22
Visuomotor integration	VMI	0.66 ^a	0.79 ^a	0.77 ^a	1	-0.28
Fine motor functioning	MD-total	-0.52 ^a	-0.64 ^a	-0.60 ^a	-0.60 ^a	1

Spearman's (ρ) correlation coefficients of the complete group are presented below the diagonal and those of the nonretarded group above the diagonal. Correlations between the IQ measures and manual dexterity (MD) scores were negative, i.e. lower scores on the intelligence tests indicated a higher score on the MD tests reflecting a poorer performance.

VIQ = verbal intelligence quotient; PIQ = performance intelligence quotient; FSIQ = full-scale intelligence quotient; VMI = visuomotor integration; MD-total = manual dexterity total score.

a $p < 0.01$.

b $p < 0.05$.

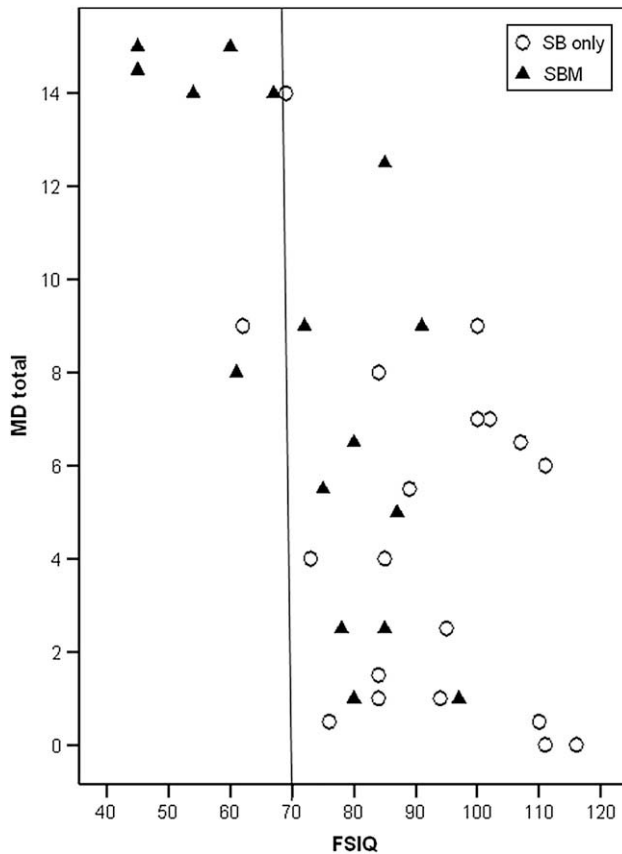


Fig. 1 – Scatterplot of the manual dexterity total score (MD-total) and the full-scale intelligence quotient (FSIQ) for the spina bifida children with and those without complex neuropathology.

3.2. Performance differences between nonretarded children with and without cerebral malformation

Within the nonretarded group, the children with SBM were statistically significantly more impaired than the children with SB-only on the PIQ and FSIQ measures, but not on VIQ. Similar subgroup differences were found for visual-motor

integration as measured by both the VMI and the disks task. The fine motor and simple speed tasks did not yield significant between-group differences: most nonretarded children showed performances falling within the low average range relative to standardized norm scores. The medians and interquartile ranges of the different subtests are presented in Table 3.

3.3. Associations between performance outcomes and cerebral malformation

Table 4 depicts the ORs for the associations between impaired cognitive functioning, visual-motor integration, fine motor performance, motor quality (kinetic tremor, dysmetria), and cerebral malformation (SBM) for the nonretarded group. As expected, increased ORs were mainly found among the IQ measures (VIQ, PIQ), but only the OR for the association between low PIQ and SBM was statistically significant. $PIQ < 85$ also showed a tendency to be associated with visual-motor integration (VMI), whereas fine motor functioning showed no association with any of the intelligence measures or VMI.

Kinetic tremor and dysmetria, the two aspects of motor quality, showed no association with fine motor performance (MD-total) or VMI. Thus, VMI was more related to the IQ measures than to motor functioning, and motor functioning and quality varied independently among the nonretarded children. Only weak associations were found between kinetic tremor and IQ measures. Cerebral malformation appeared to be associated with the IQ measures, especially PIQ, and with motor quality, but not with VMI or MD-total.

4. Discussion

The present study aimed to investigate the relationship between motor deficits and cognitive functioning in children with spina bifida.

Consistent with earlier reports we found our cohort of SB children to differ from normally developing peers in both cognitive and fine motor functioning.^{5,25} Even in the subgroup of nonretarded ($FSIQ \geq 70$) children both cognitive (FSIQ, PIQ)

Table 3 – Median (interquartile) of performance outcomes for the nonretarded ($FSIQ \geq 70$) children with (SBM) and without (SB-only) cerebral malformations.

Function	Subtest	SBM	N	SB-only	N	p^a
Cognitive function	VIQ	94 (20)	13	98 (23)	17	0.18
	PIQ	77 (12)	13	98 (18)	17	0.00
	FSIQ	85 (12)	13	100 (26)	17	0.00
Visual-motor integration	VMI	91 (8)	13	101 (10)	17	0.01
	Discs (RAKIT)	10.5 (8)	12	15.5 (8)	14	0.03
Fine motor function	MD-total	5.0 (6.5)	11	2.5 (6.3)	17	0.21
Speed	Baseline speed (ANT)	356.5 (128)	12	332 (152)	17	0.91

VIQ = verbal intelligence quotient; PIQ = performance intelligence quotient; FSIQ = full-scale intelligence quotient; VMI = visuomotor integration; MD-total = manual dexterity total score; SBM = spina bifida myelomeningocele.

^a p -values are two-tailed.

Table 4 – Odds ratios (ORs) for impaired performance outcomes, kinetic tremor, dysmetria and involvement of cerebral malformations (SBM) for the nonretarded (FSIQ ≥ 70) group.

		VIQ < 85	PIQ < 85	VMI < 85	MD-total impaired	Kinetic tremor	Dysmetria	SBM
VIQ < 85	OR	–						
	95%CI							
PIQ < 85	OR	3.33	–					
	95%CI	0.47–29.31						
VMI < 85	OR	1.4	4.8	–				
	95%CI	0.04–16.19	0.43–133.56					
MD-total impaired	OR	0.87	2.2	0.64	–			
	95%CI	0.09–6.94	0.44–10.97	0.02–9.46				
Kinetic tremor	OR	3.08	2.27	1.33	0.33	–		
	95%CI	0.31–83.24	0.44–14.04	0.09–43.05	0.06–1.77			
Dysmetria	OR	0.73	1.71	0.54	0.43	3.81	–	
	95%CI	0.08–5.84	0.35–8.41	0.02–8.08	0.09–2.09	0.71–22.36		
SBM	OR	3.33	41.25	1.36	1.19	2.37	3.21	–
	95%CI	0.47–29.31	4.79–373.79	0.12–14.68	0.24–5.79	0.45–14.04	0.63–16.78	

OR = odds ratio; CI = confidence interval; VIQ = verbal intelligence quotient; PIQ = performance intelligence quotient; FSIQ = full-scale intelligence quotient; VMI = visuomotor integration; MD-total = manual dexterity total score; SBM = spina bifida myelomeningocele. Functioning is impaired when scores are in the 15th percentile or lower.

and motor impairments were significantly more frequent than they are in the healthy population. In the full cohort of 41 children cognitive and motor performance outcomes were strongly related. However, the results were confounded by 11 severely disabled children. After their exclusion the above-mentioned correlation disappeared and the subsequent odds ratio analyses supported this latter result.

The results further revealed that even after exclusion of the severely disabled children the children with cerebral malformation (SBM) were more severely impaired on performance IQ and visual-motor integration. The results showed associations between cerebral malformation and poor motor quality. No significant effects of SBM on fine motor performance or overall reaction speed were found, nor were there high correlations between IQ measures and motor performance. A closer examination of the three items measuring fine motor functions nevertheless revealed a possibly interesting result relating to movement speed for MD item 1: the children with cerebral malformation were slower performing the task (results not presented). These results suggest that in children with SBM speed (i.e. the time needed for information processing and motor initiation) is not typically compromised but does show deficits (i.e. prolonged movement times) for tasks requiring more complex movements.

The results of the current study are in line with the work of Jansen-Osmann and Wiedenbauer.²⁶ They found that children with spina bifida had difficulties in spatial cognition measured with tasks focused on spatial perception, mental rotation, spatial visualisation, and spatial working memory, which corresponds to our results on VMI and 'disks', measuring the same function domains. In their study gross motor development (in particular 'age of walking') appeared to be correlated with the development of these visuospatial abilities.²⁶ In the literature problems with fine motor skills such as writing and drawing, especially reduced speed and legibility, are reported.^{10,11} Barnes et al. found deficient writing skills in young adults suffering from SB with hydrocephalus (SBH), which coincided with the findings reported for children.^{13,27} Their SBH sample also showed poor coordination and failed on

motor tasks requiring rapid recruitment of temporally organized movements. The current results thus corroborate the findings of Dennis et al.¹⁴ on poor writing skills, although, interestingly, the associations between the children's IQ measures, their performance on the simple fine motor tasks and their rater-assessed motor quality were weak, whereas the associations with the more complex visual-motor performance outcomes were stronger.

The study of spina bifida is seriously hampered by the sheer complexity of the neuropathology throughout the neuraxis. Since in our study all the children with cerebral malformation had both ACM and hydrocephalus, we cannot definitively attribute the cause of any poor function to ACM or HC alone. In an earlier study of our group, we²⁸ suggested that a comparison between behavioural data and the literature on cognitive deficits associated with hydrocephalus and/or cerebellar disorders might allow the contributions of the different neuropathological aspects to the observed cognitive deficits to be disentangled. Research conducted in the past decade supports the idea that the cerebellum is not only involved in motor functions but also in higher-order cognitive (non-motor) functions such as visual-spatial abilities,²⁹ verbal fluency,³⁰ working memory, implicit and explicit learning, and language.^{31–34}

In conclusion, our findings underscore the role of cerebral malformation in spina bifida and its consequences for neuropsychological functioning; in particular we found a large effect on cognitive functions and visual-motor integration, and an association between cerebral malformation and motor quality. In contrast, the associations between cerebral malformation and fine motor skills and motor initiation as measured by simple reaction-time tasks, proved to be weak or non-existent. Thus, although fine motor functions are below average in children with SB (complete group) we found no evidence for a strong relationship with cerebral malformation. Moreover, the cognitive deficits in SB seem to be quite independent from fine motor functions. Accordingly, although as a group children with SB show both cognitive and fine motor deficits, for diagnostic and intervention purposes the two

function domains should be assessed and trained separately. Impaired fine motor performance is probably related to lower academic achievement, in particular to writing. This complicated developmental interaction further strengthens the need for an individualized management of children with SB.

Acknowledgements

This study is part of the Nijmegen interdisciplinary spina bifida (NISB) research programme dedicated to fostering the care of children with spina bifida and their families. We are grateful to all the parents and their children for their willingness to take part in our study. We thank Marlou Essink for her help in the assessment of the patients' motor functions and Professor Floor Kraaimaat for his comments on an earlier draft of the manuscript.

Completion of the manuscript was supported by a grant from the Jan Jongmans Foundation, Gennep, The Netherlands.

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