J Rehabil Med 2010; 42: 56–59

ORIGINAL REPORT

IS COGNITIVE FUNCTIONING ASSOCIATED WITH SUBJECTIVE QUALITY OF LIFE IN YOUNG ADULTS WITH SPINA BIFIDA AND HYDROCEPHALUS?

Hans A. Barf, PhD[1], Marcel W. M. Post, PhD[2,3], Marjolein Verhoef, MD, PhD[2,3], Rob H. J. M. Gooskens, MD, PhD[3,4] and Arie J. H. Prevo, MD, PhD[2]

From the [1]University Medical Centre Groningen, Groningen, [2]Rehabilitation Centre De Hoogstraat, [3]Rudolf Magnus Institute for Neuroscience and [4]Department of Child Neurology, University Medical Centre Utrecht, Wilhelmina Children's Hospital, Utrecht, The Netherlands

Objective: **To test the hypothesis that cognitive functioning is associated with subjective quality of life of young adults with spina bifida and hydrocephalus (SBHC).**

Design: **Cross-sectional multi-centre study in The Netherlands.**

Subjects: **A total of 110 young adults with SBHC (16–25 years old, 63% female).**

Methods: **Cognitive domains measured were intelligence (Raven Standard Progressive Matrices), memory (Wechsler Memory Scale) and executive functioning (Wisconsin modified Card Sorting Test (WmCST), Trail Making Test A and B (TMT) and UNKA word production test). Subjective quality of life was measured with a visual analogue scale. Correlations and hierarchical regression analysis controlling for age, gender and functional independence were applied.**

Results: **The TMT score was significantly associated (–0.25) with subjective quality of life. In the hierarchical regression analysis both the WmCST and TMT scores were significant determinants of subjective quality of life (Beta values 0.24 and –0.31 respectively). Intelligence, memory and word production were not related to subjective quality of life. All 5 cognitive variables together explained a significant additional 14.6% of the variance of subjective quality of life (total explained variance 19.9%).**

Conclusion: **Executive functioning was associated with subjective quality of life in young adults with spina bifida and hydrocephalus. This finding underlines the importance of examining cognitive functioning of persons with SBHC in addition to medical and functional status in medical care and outcome research.**

Key words: cognition disorders, quality of life, congenital hydrocephalus, neural tube defects, neuropsychological tests.

J Rehabil Med 2010; 42: 56–59

Correspondence address: Marcel W. M. Post, Rehabilitation Centre De Hoogstraat, Rembrandtkade 10, NL-3583 TM Utrecht, The Netherlands. E-mail: m.post@dehoogstraat.nl

Submitted February 17, 2009; accepted September 30, 2009

INTRODUCTION

Spina bifida (SB) is a complex congenital disorder that may result in a wide variety of neurological deficits (1). The life expectancy of children with SB has increased, but they may experience permanent physical and cognitive problems associated with SB and hydrocephalus (HC) (1–4). Several studies into the quality of life (QoL) of adolescents and young adults with SB are available (5–18). The conceptualization and measures of QoL in these studies vary widely. There is a difference between health-related QoL (HRQoL), including experienced health in physical, mental, functional and social domains of life on the one hand, and subjective QoL (SQoL), including happiness, global well-being or subjective well-being, on the other hand (19). Research into QoL of persons with SB has focused mainly on HRQoL (6, 8–12, 14, 15, 18). Fewer studies have measured SQoL (13, 15–17) or included both approaches (7). This last study showed good overall QoL, despite clear health (e.g. continence) and participation (e.g. school) issues (7).

Cognitive functioning of persons with SB and HC (SBHC) is generally below that of persons with SB without HC and that of the normal population (4, 20–24), although the effect of HC and associated condition parameters on cognition varies greatly between individuals (4, 22). It is therefore relevant to examine the relationship between cognitive functioning and QoL. Several studies have found associations between executive functioning (21, 25) or overall intelligence (intelligence quotient (IQ)) (8, 18) on parameters such as ambulation, functional independence, everyday physical activity and HRQoL. However, we found only one study on the relationship between cognitive functioning and SQoL (20). In this study, a correlation was found between mathematics skills and SQoL, but not between IQ and SQoL. However, the sample size was small and only participants with an IQ over 70 were included.

The aim of the present study was to investigate the association of several domains of cognitive functioning with SQoL in persons with SBHC.

METHODS

Participants

Data were gathered from young adults with SB (aged 16–25 years) who participated in a larger cross-sectional multi-centre study (response rate 52%) (18, 22). There were no significant differences between the response group and the non-response group with regard to age, gender, level of lesion or being shunted for HC (alpha = 0.05). Of all

participants with complete data ($n = 168$), only participants with SB aperta who were shunted for HC early in life were included in the current study ($n = 110$). The study was approved by the ethics and research committees of all participating institutions. Informed consent was obtained from all all participants and their parents if participants were below 18 years of age.

Procedure

Medical records were examined according to a fixed protocol. Participants underwent a physical and neuropsychological examination, a semi-structured interview, and completed questionnaires.

Variables

HC was defined as being shunted early in life for HC. Level of lesion was defined as the lowest completely unimpaired dermatome level on both sides measured with sensitivity to pin-prick and light touch. Participants were divided into 3 subgroups based on their level of lesion: high (L2 or above), middle (L3–L5) and low (S1 and below) (18).

The motor score of the Functional Independence Measure (FIM™) was used to rate independence in activities of daily living (26). The FIM motor score covers self-care, sphincter control, transfer and locomotion and has a range of 13 (dependent) to 91 (independent).

A trained psychologist assessed cognitive functioning by means of a battery of tests that covered major cognitive domains. Extensive information on the instruments used is provided elsewhere (22). The Standard Progressive Matrices test was applied to measure "fluid" intelligence (27). The score used was the number of correct items, converted to IQ. The updated Wechsler Memory Scale (28) was used as a global measure of memory function. Subtests include Personal and Current Information, Orientation to Time and Place, Mental Control, Logical Memory, Digit Span, Visual Reproduction, and Associative Learning. The score used was the memory quotient (MQ).

Executive functioning was assessed using different tests. The Wisconsin modified Card Sorting Test (WmCST) (29) assessed the ability to maintain and shift a cognitive set. Four stimulus cards depict one of 4 different shapes, which also differ in colour and number. The response cards have one of 3 categories (shape, colour or number) in common with the standard stimuli. The respondent is asked to sort the cards. After 6 correct responses, the participant is asked to sort the cards differently. The number of correct categories (range 0–6) was used as the score for set shifting. The Trail Making Test (TMT) (30) parts A & B measured speed, divided attention and switching ability. The difference in time (sec; B – A) needed to complete parts A and B was used as the score for divided attention and switching ability. To address the interface of memory, language, and behavioural regulation, the UNKA test (31) for word production according to lexical rules was used. The respondent is asked to generate as many words as possible that begin with the prescribed letters (U, N, K, A). Production time is 60 sec per letter. The total number of correct words was used to score word production.

To rate SQoL, participants were asked to rate their current quality of life on a visual analogue scale (VAS) with a range of 0–100; 100 being the best possible QoL. Such a single-item approach of QoL is an acceptable method for a global assessment (7, 32).

Statistics

Descriptive statistics were computed. Poor performance on cognitive tests was defined as a score lower than 2 standard deviation (SD) below the mean score of the general population or, if population figures were not available, below the mean score of young adults with SB occulta without HC (22). Relations between cognition and SQoL were examined using Spearman correlations. Subsequently, hierarchical regression analyses was used to examine relationships between cognition and SQoL controlling for age, gender and functional independence, as functional independence was significantly related to both SQoL and cognitive functioning (see results) and because cognitive test performance might been influenced by motor problems. Data were analysed using SPSS for Windows version 14.

RESULTS

Demographic and neurological characteristics are summarized in Table I. The majority of the 110 participants were female (63%) and half of the participants were aged 21 years or older (mean 20.8; SD 2.9). More than half of the participants had a level of lesion at L2 or higher. Half of the participants (55%) were wheelchair-dependent and 81% had faecal and/or urinary incontinence. The median FIM motor score was 79 (interquartile range (IQR) 65.7–84).

Median scores on memory function (MQ), word production (UNKA) and divided attention (TMT time B – A) were within the normal range of the general population and only 9–15% of all participants showed a poor performance on these tests. The median IQ was 85, which is 1 SD below the population mean and 17.3% showed an IQ below 70. The participants also scored well below average on set shifting (WmCST). Persons without cognitive impairments score all 6 categories on this test, but only 55% of the participants with SBHC managed to do so. The median score on the VAS for SQoL was 73 (IQR 50–84), indicating generally positive SQoL.

All cognitive test scores were significantly related to each other (0.23–0.71). Age and gender were not related to either cognitive functioning or SQoL (Table II). The FIM motor score was significantly related to all cognitive scores and to SQoL. Out of all cognitive tests, only the TMT score was significantly related to SQoL in bivariate analyses.

To further investigate the relationship between cognition and SQoL, hierarchical regression analysis controlling for age, gender and functional independence was performed (Table III). The FIM motor score was a significant determinant of SQoL when added into the model (Beta = 0.23; $p = 0.029$). However, after adding the cognition scores to the model, the FIM motor score was no longer a significant determinant. The TMT and WmCST were significant determinants of SQoL. The extra proportion of variance explained after adding the cognitive test scores to

Table I. *Demographic and neurological characteristics, cognitive functioning and quality of life of the study group (n = 110)*

Characteristics	
Gender, male, *n* (%)	41 (37)
Age, median (IQR)	21 (18.3–22.8)
Level of lesion, *n* (%)	
≥L2	61 (55)
L3–L5	44 (40)
≤S1	5 (5)
Ambulation, wheelchair dependence, *n* (%)	60 (55)
Incontinence, *n* (%)	89 (81)
FIM motor score, median (IQR)	79 (65.7–84.0)
Cognition, median (IQR)	
Intelligence Quotient	85 (74.1–92.8)
Memory Quotient	94 (81–101)
WmCST	6 (4–6)
Trail Making Test	38 (24–57)
Word production	39 (28–46)
VAS SQoL, median (IQR)	73 (50–84)

FIM: Functional Independence Measure; WmCST: Wisconsin modified Card Sorting Test; VAS: Visual Assessment Scale; SQoL: Subjective quality of life; IQR: interquartile range.

Table II. *Spearman correlations between demographic variables, functional independence and cognitive functioning in relation to quality of life (n = 110)*

	Subjective quality of life (VAS)	Gender	Age	FIM motor score
Gender	–0.00			
Age	–0.08	0.05		
FIM motor score	0.24*	0.02	–0.14	
Intelligence Quotient	0.01	0.15	0.04	0.36**
Memory Quotient	–0.03	0.01	0.01	0.27**
WmCST	0.19	0.12	–0.07	0.30**
Trail Making Test	–0.25*	0.03	0.02	–0.22*
Word production	0.14	–0.01	0.09	0.17

*$p < 0.05$; **$p < 0.01$.
FIM: Functional Independence Measure; WmCST: Wisconsin modified Card Sorting Test; VAS: visual assessment scale.

the regression model was 14.6% and the total proportion of variance explained by the final model was 19.9%.

DISCUSSION

This study showed that executive functioning, rather than intelligence, memory and word production, was significantly associated with SQoL. Although these associations were not strong, it is noteworthy that executive functioning was a stronger determinant of SQoL than functional independence in the final regression model. Our study adds to the available literature because of the large study sample, standardized neuropsychological testing of a range of cognitive domains and inclusion of persons with an IQ below 70.

Young adults with SBHC experienced generally positive SQoL, indicated by a median score of 73 on a 0–100 scale and only 25% of participants scoring below the midpoint of this scale. Elsewhere, we reported data from the same study, but included young adults with SB without HC, to rate life satisfaction with the LiSat-9 (17). This measure consisted of 6-point verbal rating scales and we found similar figures, 24% being dissatisfied with life as a whole and no significant difference between young adults with spina bifida and an age-matched population group (17).

Similar to Hetherington et al. (20) we did not find an association between intelligence and SQoL. In our earlier publications using data from the same study, but included young adults with SB without HC, intelligence was related to functional outcome (26), but not to most HRQoL domains (33) and HC was not associated with overall life satisfaction (17). These results appear to be consistent with those found in the present paper.

To examine relationships between cognition and QoL, it is necessary to assess specific cognitive domains and not only to measure IQ, as executive functioning was associated with SQoL, but IQ was not. Even in persons with SBHC and normal intelligence, cognitive deficits may exist (22).

Two measures of executive functioning were significantly related to SQoL, correcting for age, gender and functional independence. Hetherington et al. (20) did not measure executive functioning directly, but found everyday mathematics to be related to SQoL and mathematics have been shown to be dependent on executive functioning (34). The question as to why executive functioning, and not domains such as intelligence or memory, is related to SQoL is difficult to answer. It might be conjectured that problems in executive functioning have a more direct or more tangible impact on daily activities as they hamper more complicated or more articulated activities. Poor planning, attention or mental flexibility affect a person's ability to function independently, especially in new or conflicting situations where a person cannot rely on earlier experiences or automated behaviour. Executive functioning was found to be related to self-care independence (21, 35) and social participation (35) and might also be related to everyday physical activity (25).

It is important to note some limitations of the study. First, this cross-sectional study is not intended to determine causality. Secondly, we measured SQoL with a 1-item VAS. Although generally validated multi-item instruments are preferred, there is evidence that a single-item approach of QoL is an acceptable method for a global assessment (7, 32). In addition, it has been argued that for children and adolescents it is preferable not to use disease-specific measures of QoL and that a self-report questionnaire should be brief and simple in order to be tailored to the level of expressive and receptive language abilities as well as time perception and memory (13). Finally, a poor TMT and WmCST score could be caused by factors other than impairment of executive functioning alone. These tests were part of a cognitive test battery and all cognitive test scores were significantly correlated with each other. It is possible that fatigue, loss of motivation or problems in other cognitive domains might have influenced performance on the tests for executive functioning.

In conclusion, this study showed an association between executive functioning of young adults with SBHC and their subjective QoL. This association needs further investigation, but it underlines the importance of adding cognitive functioning to medical and functional status in care and outcome research and of including a wider variety of neuropsychological tests than solely intelligence scales.

Table III. *Hierarchical regression models for demographic variables, functional independence and cognitive functioning in relation to quality of life (n = 99)*

	Step 1 Beta (*p*-value)	Step 2 Beta (*p*-value)	Step 3 Beta (*p*-value)
Gender	0.04 (0.710)	–0.01 (0.962)	0.01 (0.947)
Age	–0.04 (0.687)	–0.03 (0.767)	–0.02 (0.856)
FIM motor score		0.23 (0.029)	0.12 (0.291)
Intelligence Quotient			–0.11 (0.457)
Memory Quotient			–0.14 (0.370)
WmCST			0.24 (0.032)
Trail Making Test			–0.31 (0.005)
Word production			0.12 (0.267)
R^2 change (%)	0.3	4.9	14.6
R^2 model (%)	0.3	5.2	19.9

FIM: Functional Independence Measure; WmCST: Wisconsin modified Card Sorting Test.

REFERENCES

1. McDonnell GV, McCann JP. Issues of medical management in adults with spina bifida. Childs Nerv Syst 2000; 16: 222–227.
2. Hunt GM, Poulton A. Open spina bifida: a complete cohort reviewed 25 years after closure. Dev Med Child Neurol 1995; 37: 19–29.
3. Hunt GM, Oakeshott P. Outcome in people with open spina bifida at age 35: prospective community based cohort study. BMJ 2003; 326: 1365–1366.
4. Iddon JL, Morgan DJR, Loveday C, Sahakian BJ, Pickard JD. Neuropsychological profile of young adults with spina bifida with or without hydrocephalus. J Neurol Neurosurg Psychiatry 2004; 75: 1112–1118.
5. Appleton PL, Minchom PE, Ellis NC, Elliott CE, Boll V, Jones P. The self-concept of young people with spina bifida: a population-based study. Dev Med Child Neurol 1994; 36: 198–215.
6. Padua L, Rendeli C, Rabini A, Girardi E, Tonali P, Salvaggio E. Health-related quality of life and disability in young patients with spina bifida. Arch Phys Med Rehabil 2002; 83: 1384–1388.
7. Sawin KJ, Brei TJ, Buran CF, Fastenau PS. Factors associated with quality of life in adolescents with spina bifida. J Holist Nurs 2002; 20: 279–304.
8. Schoenmakers MA, Uiterwaal CS, Gulmans VA, Gooskens RH, Helders PJ. Determinants of functional independence and quality of life in children with spina bifida. Clin Rehabil 2005; 19: 677–685.
9. Lemelle JL, Guillemin F, Aubert D, Guys JM, Lottmann H, Lortat-Jacob S, et al. Quality of life and continence in patients with spina bifida. Qual Life Res 2006; 15: 1481–1492.
10. MacNeily AE, Morrell J, Secord S. Lower urinary tract reconstruction for spina bifida - does it improve health related quality of life? J Urol 2005; 174: 1637–1643.
11. Oddson BE, Clancy CA, McGrath PJ. The role of pain in reduced quality of life and depressive symptomology in children with spina bifida. Clin J Pain 2006; 22: 784–789.
12. Kirpalani HM, Parkin PC, Willan AR, Fehlings DL, Rosenbaum PL, King D, et al. Quality of life in spina bifida: importance of parental hope. Arch Dis Child 2000; 83: 293–297.
13. Wallander JL, Schmitt M, Koot HM. Quality of life measurement in children and adolescents: issues, instruments, and applications. J Clin Psychol 2001; 57: 571–585.
14. Padua L, Rendeli C, Ausili E, Aprile I, Caliandro P, Tonali P, et al. Relationship between the clinical neurophysiologic pattern, disability, and quality of life in adolescents with spina bifida. J Child Neurol 2004; 19: 952–957.
15. Parkin PC, Kirpalani HM, Rosenbaum PL, Fehlings DL, Van Nie A, Willan AR, et al. Development of a health-related quality of life instrument for use in children with spina bifida. Qual Life Res 1997; 6: 123–132.
16. Andren E, Grimby G. Dependence and perceived difficulty in activities of daily living in adults with cerebral palsy and spina bifida. Disabil Rehabil 2000; 22: 299–307.
17. Barf HA, Post MW, Verhoef M, Jennekens-Schinkel A, Gooskens RH, Prevo AJ. Life satisfaction of young adults with spina bifida. Dev Med Child Neurol 2007; 49: 458–463.
18. Verhoef M, Barf HA, Post MW, van Asbeck FW, Gooskens RH, Prevo AJ. Secondary impairments in young adults with spina bifida. Dev Med Child Neurol 2004; 46: 420–427.
19. Post M, Noreau L. Quality of life after spinal cord injury. J Neurol Phys Ther 2005; 29: 139–146.
20. Hetherington R, Dennis M, Barnes M, Drake J, Gentili F. Functional outcome in young adults with spina bifida and hydrocephalus. Childs Nerv Syst 2006; 22: 117–124.
21. Heffelfinger AK, Koop JI, Fastenau PS, Brej TJ, Conant L, Katzenstein J, et al. The relationship of neuropsychological functioning to adaptation outcome in adolescents with spina bifida. J Int Neurpsychol Soc 2008; 14: 793–804.
22. Barf HA, Verhoef M, Jennekens-Schinkel A, Post MW, Gooskens RH, Prevo AJ. Cognitive status of young adults with spina bifida. Dev Med Child Neurol 2003; 45: 813–820.
23. Hommet C, Billard C, Gillet P, Barthez MA, Lourmiere JM, Santini JJ, et al. Neuropsychologic and adaptive functioning in adolescents and young adults shunted for congenital hydrocephalus. J Child Neurol 1999; 14: 144–150.
24. Bier JA-B, Morales Y, Liebling J, Geddes L, Kim E. Medical and social factors associated with cognitive outcome in individuals with myelomeningocele. Dev Med Child Neurol 1997; 39: 263–266.
25. Roebroeck ME, Hempenius L, van Baalen B, Hendriksen JG, van den Berg-Emons HJ, Stam HJ. Cognitive functioning of adolescents and young adults with meningomyelocele and level of everyday physical activity. Disabil Rehabil 2006; 28: 1237–1242.
26. Verhoef M, Barf HA, Post MW, van Asbeck FW, Gooskens RH, Prevo AJ. Functional independence among young adults with spina bifida, in relation to hydrocephalus and level of lesion. Dev Med Child Neurol 2006; 48: 114–119.
27. Raven JC. Standard progressive matrices. Oxford: Oxford Psychologist Press; 1996.
28. Wechsler D. Wechsler memory scale. San Antonio, TX: The Psychological Corporation; 1974.
29. Nelson HE. A modified card sorting test sensitive to frontal lobe defects. Cortex 1976; 12: 313–324.
30. Reitan RM, Wolfson D. Trail Making Test. In: Grant I, Adams KM, editors. Neuropsychological assessment of neuropsychiatric disorders. 2nd edn. Oxford: Oxford University Press; 1996, p. 3–42.
31. Jennekens-Schinkel A, Lanser JB, van der Velde EA, Sanders EA. Performances of multiple sclerosis patients in tasks requiring language and visuoconstruction. Assessment of outpatients in quiescent disease stages. J Neurol Sci 1990; 95: 89–103.
32. de Boer AGEM, van Lanschot JJB, Stalmeier PFM, van Sandick JW, Hulscher JBF, de Haes JCJM, et al. Is a single-item visual analogue scale as valid, reliable and responsive as multi-item scales in measuring quality of life? Qual Life Res 2004; 13: 311–320.
33. Verhoef M, Post MW, Barf HA, van Asbeck FW, Gooskens RH, Prevo AJ. Perceived health in young adults with spina bifida. Dev Med Child Neurol 2007; 49: 192–197.
34. van der Sluis S, de Jong PF, van der Leij A. Inhibition and shifting in children with learning deficits in arithmetic and reading. J Exp Child Psychol 2004; 87: 239–266.
35. Tarazi RA, Mahone EM, Zabel TA. Self-care independence in children with neurological disorders: An interactional model of adaptive demands and executive dysfunction. Rehabil Psychol 2007; 52: 196–205.