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

Hans A. Barf, PhD1, Marcel W. M. Post, PhD2,3, Marjolein Verhoef, MD, PhD2,3, Rob H. J. M. Gooskens, MD, PhD3,4 and Arie J. H. Prevo, MD, PhD2

From the 1University Medical Centre Groningen, Groningen, 2Rehabilitation Centre De Hoogstraat, 3Rudolf Magnus Institute for Neuroscience and 4Department 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.

ORIGINAL REPORT

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 the 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\textsuperscript{TM}) 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 Associate 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 \((\text{sec}; B – A)\) 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 \((33)\) and because cognitive test performance might be 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 \((\text{TMT time }B – A)\) 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 \((\text{WmCST})\). 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 \((34, 35)\). 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 \((\text{Beta} = 0.23; p = 0.029)\). 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 the model is presented in Table II.

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

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

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

J Rehabil Med 42
dissatisfied with life as a whole and no significant difference. 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.
REFERENCES