Disability & Rehabilitation, 2012; 34(9): 740–746

ISSN 0963-8288 print/ISSN 1464-5165 online
DOI: 10.3109/09638288.2011.619617

informa
healthcare

RESEARCH PAPER

Problematic psychosocial adaptation and executive dysfunction in women and men with myelomeningocele

Jan Stubberud & Gunnar Riemer

TRS – A National Centre for Rare Disorders, Sunnaas Rehabilitation Hospital, Nesoddtangen, Norway

***Purpose*: Myelomeningocele (MMC) is caused by incomplete neural tube development, affecting physical, cognitive and adaptive functioning. The aim of this study was to characterize women and men with problematic psychosocial adaptation with respect to cognitive functions and psychological symptoms. It was expected that cognitive functions, especially the executive functions, were mostly impaired. In addition, the clinical history, the neurological and the psychological functioning was investigated. *Method*: Twelve participants (six men, six women; age range 24–41 years) with MMC were recruited from TRS National Resource Centre for Rare Disorders. Inclusion criteria were (1) uncompleted upper secondary school, (2) unemployment and (3) lack of social relationships. The participants underwent a systematic clinical history, a clinical neurological examination, a neuropsychological assessment and questionnaires regarding cognitive and psychological functioning. *Results*: All participants had major neurological deficits and most of them had hydrocephalus and Chiari-II malformation. Cognitive deficits were present in a range of domains, especially the executive functions. The neuropsychological findings showed no obvious difference between men and women. Symptoms of psychopathology were more pronounced in women. The men were less capable to structure daily living. *Conclusions*: The neuropsychological findings disclosed that the executive functions were most impaired. Future research should be directed towards cognitive rehabilitation.**

Keywords: Chiari-II malformation, cognitive function, executive function, hydrocephalus, myelomeningocele, psychological symptoms, psychosocial problems, spina bifida

Implications for Rehabilitation

- It is conceivable that executive deficits in patients with myelomeningocele are decisive for problematic psychosocial adaptation.
- Cognitive rehabilitation should be considered for neuropsychological impairments to improve psychosocial functioning and quality of life.
- Gender differences in psychological functioning are evident.

Background

Myelomeningocele (MMC) or spina bifida is a birth defect caused by incomplete neural tube development resulting in a protrusion of the spinal cord, nerve roots and meninges through an opening in the spine. The term MMC will be used in this article because it addresses the neurological anomaly. Motor, sensory and autonomous functions vary depending on the lesion level and the degree of involvement of neuronal structures [1].

Individuals with MMC have involvement of the entire central nervous system, causing a variety of brain abnormalities. Hydrocephalus occurs in 95%, with 80%–90% requiring shunting [1]. Shunting restores the brain volume but does not repair the axonal and neuronal damage. Chiari-II malformation, a deformity of the brainstem and cerebellum, occurs in almost all births and dysgenesis of the corpus callosum in more than half of the patients [2]. These brain abnormalities have persisting effects on development, including problems in the neurological, cognitive and behavioural domains [2–4].

Typical cognitive impairments include problems with attention and high distractibility, memory, numerical and mathematical capacity, executive function and problem-solving skills. Executive functions are high-level cognitive functions that activate, organize, integrate and manage other functions [5,6]. Both frontal lobe damage and disruption of white matter circuits between the prefrontal cortex and other areas of the brain can result in executive dysfunction in MMC.

Correspondence: Mr. Jan Stubberud, TRS – A National Centre for Rare Disorders, Sunnaas Rehabilitation Hospital, N-1450 Nesoddtangen, Norway. Tel: 0047 66969362. Fax: 0047 66969331. E-mail: jan.stubberud@sunnaas.no

(Accepted August 2011)

Many patients with MMC have been observed to exhibit executive deficits such as difficulties with distractibility, organizing material, initiation and staying on task, working memory, set shifting, self-monitoring and emotional control [1,6,7]. Planning, goal-directed behaviour and problem solving are also impaired [8].

Most of the studies addressing cognitive impairments have been conducted in children and adolescents [1,8–13]. The general consensus is that overall patients with MMC have reduced cognitive functioning in a wide range of cognitive domains compared to healthy children.

The knowledge about how executive dysfunction influences psychosocial functioning in the adult population is relatively sparse. However, a study by Zukerman et al. [14] demonstrated that executive functions could predict several adult milestones, such as leaving home, attending college and engaging in a romantic relationship. As such, executive functions seem to be essential abilities for living independently and managing social and academic demands. Executive functioning and memory have also been positively correlated with everyday physical activity and functional independence in adolescents and young adults with MMC [15]. In addition, clinical experience and studies show that many of the patients have reduced quality of life, are at risk of being unemployed, socially isolated and lack daily structures. Patients with MMC are at greater risk of depressed mood, low self-worth and suicidal ideation [16,17]. Findings from our previous study indicated that the visual–spatial and executive functions were especially important for psychosocial adaptation in young men [18].

The aim of this study was to characterize women and men with problematic psychosocial adaptation with respect to cognitive functions and psychological symptoms. It is expected that the executive functions are mostly impaired. In addition, it was investigated whether there are differences between men and women with regard to clinical history and neurological functioning.

Methods

Participants

All the participants were recruited from TRS National Resource Centre for Rare Disorders, Sunnaas Rehabilitation Hospital, which offers life-long services on medical, psychological, social and educational issues. The centre is often contacted by young and middle-aged adults with MMC because of sparse follow-up after the age of 18 years in the ordinary Norwegian health care system. Many of the patients live in the rural, sparsely populated districts where medico-social care systems are rare, do not finish school, are not able to find a job and live alone in apartments just receiving the basal health care by public nurses and general practitioners. In many instances, TRS can be an important place for these patients to receive medical and social input. By the start of the study in January 2007, a total number of 250 patients over the age of 18 years were registered. In the period between January 2007 and December 2008, 65 patients between the age of 18 and 45 years (40 women, 25 men) contacted TRS because of various medical and psychosocial reasons. Out of this population, a study group was selected by using the following inclusion criteria:

1. Uncompleted upper secondary school (education in Norway is mandatory for all children aged 6–16 years)
2. Unemployment (i.e. not having had a paid job, either full-time or part-time during the last 3 years)
3. Lack of social relationships (e.g. living alone, absence of close friends, deficiency to engage in any form of play, recreational or leisure activity)

All these criteria had to be fulfilled by the participants to be included into the study. These criteria were chosen because they seemed to be relevant for the quality of psychosocial adjustment. Twelve of these 65 patients (six men, six women; age range 24–41 years, mean 32 years, SD 5.7) fulfilled the inclusion criteria.

Procedure

The participants were divided into two groups, one with men and one with women. Both groups stayed for 5 days at the centre. The participants underwent a clinical history addressing mobility, sensory function, types of stoma, orthopaedic surgery, hydrocephalus and other neurological diagnoses, and living factors. A complete clinical neurological examination was done. All participants underwent 2 hours of neuropsychological assessment with a fixed order of the tests.

Measures

The neuropsychological tests were paper and pencil based. These tests are well described in the literature. A brief description is provided in Table I**,** which summarizes the neuropsychological test battery and the measured cognitive domains. The neuropsychological assessment intended to cover the following cognitive functions: verbal (vocabulary, abstract verbal reasoning) and performance skills (spatial perception, non-verbal abstract problem solving), visual-motor coordination, and a number of executive functions such as working memory, mental flexibility and inhibition. The executive functions were measured by different tests. Inhibition of automatic responding was assessed with Stroop

Table I. Neuropsychological test battery and cognitive domains measured.

Type of neuropsychological test	Cognitive function
Wechsler Abbreviated Scale of Intelligence (WASI)	Verbal (vocabulary, abstract verbal reasoning) and performance skills (spatial perception, nonverbal abstract problem solving)
Grooved Pegboard (Halstead Reitan)	Visual-motor coordination
Letter-number sequencing (WAIS-III)	Working memory
Trail making Test (Halstead Reitan)	Mental flexibility
Stroop Test (D-KEFS)	Inhibition
Behaviour Rating Inventory of Executive Function (BRIEF-A)	Executive functions (inhibition, shift, emotional control, self-monitoring, initiative, working memory, planning/organizing, task monitoring, organizing of materials)

D-KEFS, Delis-Kaplan Executive Function System; WAIS-III, Wechsler Adult Intelligence Scale III.

Inhibition Delis-Kaplan Executive Function System in which words are printed in dissonant ink color and participants are instructed to name the colour of the ink instead of the more automatic response of reading the word [19]. To assess attention and working memory, the letter-number sequencing from Wechsler Adult Intelligence Scale III was used [20]. Here the participant has to repeat a sequence (e.g. Q-1-B-3-J-2) but place the numbers in numerical order and then the letters in alphabetical order. Mental flexibility was assessed with Trails B (Trail making Test), requiring rapid alternation between letters and numbers to connect them in order [21]. In addition, the following test was used: Wechsler Abbreviated Scale of Intelligence (WASI) [22]. The WASI is designed to be a brief intelligence assessment that yields Full Scale IQ, Verbal IQ and Performance IQ estimates. The Grooved Pegboard [23] was also used. The Grooved Pegboard is a manipulative dexterity test consisting of 25 holes with randomly positioned slots. Pegs with a key along one side must be rotated to match the hole before they can be inserted. This test requires more complex visual-motor coordination than most pegboard tests.

Questionnaires were used to assess everyday function that is not typically captured by laboratory tests. Behaviour Rating Inventory of Executive Function (BRIEF-A) assesses cognitive functions associated with executive functioning in the areas of inhibition, shift, emotional control self-monitoring, initiative, working memory, planning/organizing, task monitoring and organization of materials [24]. To document self-reported psychological problems and symptoms of psychopathology, the Symptom Checklist 90-R was used [25].

Statistical analysis

Concerning the results from the neuropsychological tests, no statistical analyses other than frequencies have been conducted because of the small sample size. Features from the clinical history, the living factors, clinical findings and the self-reported executive difficulties and psychological problems have been tested using contingency tables and the Pearson chi-squared test for men and women to check whether there were significant gender-specific differences. A p value of 0.05 or less was considered statistically significant.

Ethics

The project was approved by the Regional Ethic Committee for Medical Research Ethics, South-Eastern Norway. Written consent was obtained from the participants.

Results

Clinical history

Features from the clinical history are summarized in Table II. MMC levels were between T10 and S1, indicating the upper level of the neurological deficit, either motor or sensory. Hydrocephalus was present in 92% of the subjects and Chiari-II malformation in 75%. All the participants were incontinent, either performing clean intermittent catheterization or needing urostomy. Pressure sores have at some point been a complication in 92% (grades 3 and 4). In all participants, at least one kind of orthopaedic surgery was performed. The features showed no significant differences for men and women.

Table II. Features from the clinical history.

Clinical history	Men (n = 6)	Women (n = 6)	Total n (%)
MMC level	T10 – S1	T11 – L5	T10 – S1
Tethered cord	5	4	9 (75)
Surgical treated	2	2	
Hydrocephalus	6	5	11 (92)
Shunt	5	4	9 (75)
Third ventriculostomy	1	0	1 (8)
≥3 shunt revisions	5	3	8 (67)
Arnold–Chiari malformation (ACM)	4	5	9 (75)
Surgical treated ACM	1	0	
Epilepsy	1	1	2 (17)
Clean intermittent catheterization	4	3	7 (58)
Urostomy	4	3	7 (58)
Colostomy	0	1	1 (8)
Appendicostomy	2	1	3 (25)
Reduced sexual function	4	5	9 (75)
Pressure sores	5	6	11 (92)
Latex allergy	3	3	6 (50)
Orthopaedic surgery	6	6	12 (100)
Orthopaedic aids	2	3	5 (42)

Table III. Clinically important living factors.

Living factors	Men (n = 6)	Women (n = 6)	Total n (%)	p value
Sleep period 11 pm to 6 am	1	6	7 (58)	0.003
Obesity (body mass index >30)	1	4	5 (42)	n.s.
Smoking	1	1	2 (17)	n.s.
Alcohol abuse	1	0	1 (8)	n.s.
Physiotherapy	3	3	6 (50)	n.s.
Driving	4	3	7 (58)	n.s.

n.s., not significant.
The last column presents p values for significant gender differences.

The clinically important living factors are given in Table III. Fifty-eight percent of the participants had a normal sleeping pattern (11 pm to 6 am), but five of the six men had a nearly inversed sleeping pattern, meaning that they were mostly sleeping during daytime. This finding was statistically significant. Forty-two percent of the participants had a body mass index above 30, which means obesity.

Clinical findings

Findings from the clinical examination are listed in Table IV. Half of the participants were unable to walk and a third could only walk a few steps with technical aids. All the participants had reduced or missing sensibility on the legs, nystagmus and impaired fine motor functioning of the hands. These findings were similar for men and women.

Neuropsychological findings

The neuropsychological test results are summarized in Table V. Executive functions and visual-motor coordination were most impaired. Ninety-two percent of the participants

had reduced mental flexibility, 58% had an impaired ability to inhibit impulses and 42% had reduced working memory. All the participants had difficulties with visual-motor coordination. Minor impairments were found in other cognitive domains: 25% had reduced spatial perception, 17% reduced abstract verbal reasoning and one participant had difficulties with non-verbal abstract problem solving. None of the participants had impaired vocabulary. Although not having been statistically analysed, the neuropsychological findings showed no obvious difference between men and women. To visualize deficits in the executive function in relation to the other cognitive domains, a ratio between executive performance and non-executive was calculated. The men and women below cut-off range had a ratio of 1.3 and 1.2, respectively. Above cut-off these ratios were 0.4 or lower.

The self-reported executive difficulties are listed in Table VI. The T-scores are based on comparison to the normative sample that comprised 1050 self-reports and 1200 informant reports, with higher scores reflecting greater difficulties experienced by the individual. Clinically elevated is defined as a T-score of 65 or greater in keeping with instructions in the BRIEF-A manual [24]. At least half of the participants described problems with shift (e.g. disturbed by unexpected changes in routine), working memory, organization of materials (e.g. lose things), initiative, planning/organization (e.g. trouble prioritizing activities) and task monitoring (e.g. trouble finishing tasks). These difficulties did not differ significantly for men and women.

Table IV. Clinical findings.

Clinical findings	Men (n = 6)	Women (n = 6)	Total n (%)
Ambulation			
Without technical aids	1	1	2 (17)
With technical aids	2	2	4 (33)
Unable	3	3	6 (50)
Reduced/missing leg sensibility	6	6	12 (100)
Scoliosis (>20 degrees)	3	3	6 (50)
Reduced visual field	2	0	2 (17)
Nystagmus	6	6	12 (100)
Impaired fine motor function	6	6	12 (100)

Self-reported psychological problems

Self-reported psychological problems and symptoms of psychopathology are shown in Table VII. Altogether, the men reported 10 symptoms and the women 16 symptoms. Some of the symptoms were more frequent than others: Three of six women had somatization symptoms, i.e. somatic distress, and phobic anxiety symptoms. This latter symptom was

Table VI. Self-reported executive difficulties for men and women (Behaviour Rating Inventory of Executive Function).

Executive functions	T-score >65 Men	T-score >65 Women	N (%) above cut-off
Inhibit	2	0	2 (17)
Shift	4	3	7 (58)
Emotional control	1	1	2 (17)
Self-monitor	0	0	0 (0)
Initiate	3	3	6 (50)
Working memory	3	4	7 (58)
Plan/organize	3	3	6 (50)
Task monitor	3	3	6 (50)
Organization of materials	4	3	7 (58)

Cut-off is defined when T-score is >65, indicating executive difficulties.

Table VII. Self-reported psychological problems and symptoms of psychopathology (Symptom Checklist-90-R).

Symptom scales	T-score >63 Men	T-score >63 Women	N (%) above cut-off	p value
Somatization	1	3	4 (33)	n.s.
Obsessive-compulsive	4	2	6 (50)	n.s.
Interpersonal sensitivity	1	2	3 (25)	n.s.
Depression	1	2	3 (25)	n.s.
Anxiety	1	1	2 (17)	n.s.
Hostility	0	0	0	n.s.
Phobic anxiety	0	3	3 (25)	0.05
Paranoid ideation	1	1	2 (17)	n.s.
Psychoticism	1	2	3 (25)	n.s.
Sum of symptoms	10	16		n.s.
Global severity index	1	2	3 (25)	n.s.

n.s., not significant.
Cut-off is defined when T-score is >65. The sum of symptoms describes how many of the above listed symptoms were totally present for men and women. The last column presents p values for significant gender differences.

Table V. Quantitative distribution of performance across different cognitive domains and gender.

Range	Below cut-off		Lower normal		Normal		Upper normal		
Percentile %	<5		5–34		35–65		>66		
Gender	M	W	M	W	M	W	M	W	Impaired N (%)
Cognitive domain									
Executive functions									
Inhibition	**4**	**3**	1	1	—	2	1	—	7 (58)
Mental flexibility	**5**	**6**	1	—	—	—	—	—	11 (92)
Working memory	**3**	**2**	1	3	2	1	—	—	5 (42)
Vocabulary	—	—	1	2	1	2	4	2	0 (0)
Abstract verbal reasoning	**1**	**1**	1	3	2	—	2	2	2 (17)
Spatial perception	**1**	**2**	5	1	—	3	—	—	3 (25)
Nonverbal abstract problem solving	**1**	—	3	3	2	2	—	1	1 (8)
Visual motor coordination	**6**	**6**	—	—	—	—	—	—	12 (100)
Ratio executive/non-executive performance	**12/91.3**	**11/91.2**	3/100.3	4/90.4	2/50.4	3/70.4	1/60.2	0/50	

M, men; W, women.
Cognition is defined as impaired when performance level is <5th percentile (see bold numbers).

statistically significant. Four of the six men had obsessive compulsive symptoms. The Global Severity Index, a measure of overall psychological distress, showed that two women and one man had scores above cut-off.

Discussion

The main purpose of this study was to examine adults with MMC with problematic psychosocial adaptation and to check whether this was associated with executive dysfunction. The participants were severely impaired by neurological deficits and cognitive difficulties. Features from the clinical history and clinical findings showed no significant differences between men and women.

As expected, our results revealed that executive functions such as inhibition, mental flexibility and working memory were reduced in both men and women. It can be argued that these findings explain why these participants experience difficulties with regard to education, work and social relationships.

Studies have shown that long-term deficits in adaptive functioning are common among children and adolescents with MMC and hydrocephalus. Poor adaptation during childhood and adolescence places individuals at risk for continued difficulties in adulthood, such as underemployment, unemployment, limited independence, reduced self-esteem and dependence on federal and state support programs [6]. Many of these adults are unable to live independently, and relatively few work in professional occupations [26–28].

Executive deficits are a frequent sequel of traumatic and other types of acquired brain injury and have detrimental effects in many domains, from impeding return to work, to straining social relationships and increasing dependence upon others [29]. Furthermore, specific cognitive, academic, vocational and linguistic skills are of little benefit if the individual cannot efficiently use them in real life [5]. Executive functions enable individuals to account for short- and long-term consequences of their actions and to plan for those results [30,31]. It allows individuals to make real-time evaluations of their actions and make necessary adjustments if those actions are not achieving the desired result [32]. Therefore, it is conceivable that also the executive deficits in patients with MMC are decisive for problematic psychosocial adaptation.

A recent study investigated executive function as a mediating variable for the functional independence of adolescents with MMC aged 12–21 years [6]. This study was based on an ecological model which suggests that multiple factors such as executive functioning, adolescent resilience and family resourcefulness influence the adaptation outcomes in adolescents with MMC [11]. One of the findings was that executive function was significantly correlated with the functional independence and self-management outcome in terms of self-care and mobility, and it was concluded that executive functioning at least partially mediated the impact of neurological severity on functional independence. The study was limited by the small sample and the cross-sectional design, which prevented further understanding of the causality suggested in the model. The authors recommended further studies, which should include a longitudinal component and direct training of the executive functioning. In addition, it was postulated that executive dysfunction is likely to become progressively more problematic in early adulthood because of the increased need for independent problem solving [6]. Our study had a somewhat opposite design with the psychosocial problems as the basis for the inclusion and not the effect of risk factors such as executive functioning – and it has the same shortcoming of being limited by the small sample and the cross-sectional design (see below) – but it supports that the association between psychosocial maladjustment and executive dysfunction is also present in adults.

With regard to the self-reported executive difficulties, both men and women reported difficulties in executive functioning. Other studies have also used BRIEF as a method of measuring executive functioning, suggesting that this rating scale is applicable for identifying executive functioning [14,33,34]. Mahone et al. [33] studied a small sample of adolescents with MMC and found high levels of parent-reported problems with initiation, working memory, planning/organization, self-monitoring, metacognition and emotional control. Zukerman et al. [14] found that mother or teacher behavioural reports of adolescents' executive functioning were significant predictors for leaving home, romantic relationship experience and number of friends.

In our study, the objective findings of executive dysfunction were even more pronounced compared to the self-reported problems. This may be a result of limited awareness of own difficulties. In literature, there is evidence that subjective evaluation of cognitive functioning does not always predict objective test performance [35]. Hence, in clinical practice it may not be sufficient asking the patients about their executive difficulties; they also have to be tested to develop realistic plans to improve psychosocial functioning.

Self-reported psychological problems

Internalizing symptoms such as depression and anxiety have so far mainly been investigated in children and adolescents with MMC [12,13]. It has been found that emerging adults, like their younger counterparts, are at risk for depressive symptoms and anxiety [12]. A longitudinal follow-up of children aged 8–15 years with MMC matched with controls revealed that pre-adolescent differences between groups were maintained for several adjustment variables, indicating that adolescents have enduring academic and attention problems and difficulties with social development. On the other hand, women with MMC were found to be at risk for an exacerbation of social difficulties and negative perceptions of their physical appearance [13]. The authors of this study assert that the developmental trajectories may change in early adulthood and emphasize that gender-based research is needed, because some components of intervention may need to be gender specific. Interestingly, the self-reported psychological problems and symptoms of psychopathology in our study also revealed some gender differences. Twice as much of the men reported obsessive compulsive symptoms when compared to the women. On the other hand, half of the women reported both somatization and phobic anxiety symptoms, whereas only one man reported somatization symptoms. The anxiety

symptoms showed a significant gender difference. From the general population, it is known that women tend to report more psychological distress than men [36]. However, the reason why the women reported more symptoms of phobic anxiety and the men obsessive compulsive symptoms is not known. It could be possible that a large number of medical interventions during lifetime create gender-specific reactions. The study design and the small number of participants do not permit to conclude on these matters, but this could be an interesting area for future investigation.

Living factors and clinical history

In the following, some aspects of the living factors and clinical history will be discussed with regard to executive dysfunction. Inversed sleeping pattern was only reported by men. This finding was significant. To our knowledge, little is known about this field and the reasons for this gender difference. Sleeping problems in MMC often present as sleep apnea as a consequence of Chiari-II malformation [37]. In our participants, Chiari-II malformation was more frequent among women, so this cannot explain the difference. The lacking ability to maintain a normal day rhythm could be caused by a deficiency in structuring and organizing. However, the test results of these executive functions did not reveal any differences between men and women. Hence, we have no plausible explanation for the gender difference of the inversed sleeping pattern. It is apparent that inversed sleep may cause a couple of problems, concerning both the biological rhythm and social adjustment.

Almost half of the participants, mostly women, were obese. Obesity may be caused by paresis and executive dysfunction, such as reduced inhibition. However, neither of these features showed a gender difference. Perhaps hormonal factors are playing a role. Elevated obesity rates among adults, particularly in women, have been reported before [38].

There was a high lifetime prevalence of pressure sores. Also here, executive dysfunction may play a role. A previous study reported among others memory problems as a significant risk factor to develop pressure sores [39].

Limitations and directions for future research

There are several limitations to the present study. The sample size is small. This limitation is hard to surmount because the number of persons with MMC in this target group is small in Norway. It would have been desirable to examine an age-matched control group with better psychosocial adaptation, but this was also difficult because of the widespread location of the persons with MMC. The neuropsychological tests and questionnaire were selected to focus on the executive functions. Because of time restrictions, a limited number of tests were allowed. As a consequence, the neuropsychological evaluation is somewhat limited, especially in the area of attention and memory. Nevertheless, the results are useful and in accordance with the clinical relevant deficiencies in this patient group. Neuropsychological assessment should be conducted more often to better understand clinical and psychosocial needs in the adult MMC population. Similar to most of the other studies conducted thus far, the present study also used a cross-sectional design that fails to establish a causal relationship between psychosocial problems and cognitive deficits.

Clinical implications

To date, research efforts have mainly focused on categorizing cognitive impairments in MMC and not on cognitive rehabilitation. Cognitive rehabilitation can be defined as a process whereby people with brain injury work together with professional staff to alleviate cognitive deficits to function as adequately as possible [40]. The effectiveness of cognitive rehabilitation is well documented for patients with stroke and traumatic brain injury [41–43]. Future research should be directed towards cognitive rehabilitation for these impairments to improve psychosocial functioning and quality of life. We are currently doing a study where we are examining the effects of a theoretically grounded cognitive rehabilitation programme on patients with MMC who experience executive difficulties.

Conclusion

The participants with problematic psychosocial adaptation were characterized by impaired executive functions, such as mental flexibility, inhibition of impulses and working memory. Self-reported executive difficulties were less pronounced than the objective findings. The men reported less psychopathological symptoms than women, but had more problems with structuring daily living. Future research should be aware of gender differences and directed towards cognitive rehabilitation.

Acknowledgements

We thank the nurses Karen Grimsrud, Astri Andersen and Pål-Erik Plaum for assistance in taking care of the participants and contributing to take the clinical history.

Declarations of interest: The authors report no conflict of interest.

References

1. Fletcher JM, Copeland K, Frederick JA, Blaser SE, Kramer LA, Northrup H, Hannay HJ, et al. Spinal lesion level in spina bifida: a source of neural and cognitive heterogeneity. J Neurosurg 2005;102:268–279.
2. Barkovich AJ. Paediatric neuroimaging. 3rd ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2000.
3. Hommet C, Cottier JP, Billard C, Perrier D, Gillet P, De Toffol B, Sirinelli D, et al. MRI morphometric study and correlation with cognitive functions in young adults shunted for congenital hydrocephalus related to spina bifida. Eur Neurol 2002;47:169–174.
4. Dicianno BE, Kurowski BG, Yang JM, Chancellor MB, Bejjani GK, Fairman AD, Lewis N, Sotirake J. Rehabilitation and medical management of the adult with spina bifida. Am J Phys Med Rehabil 2008;87:1027–1050.
5. Ylvisaker M, Szekeres SF, Feeney TJ. Cognitive rehabilitation: executive functions. In: Ylvisaker M, editor. Traumatic brain injury rehabilitation: children and adolescents. 2nd ed. Boston, MA: Butterworth-Heinemann; 1998.
6. Heffelfinger AK, Koop JI, Fastenau PS, Brei TJ, Conant L, Katzenstein J, Cashin SE, Sawin KJ. The relationship of neuropsychological functioning to adaptation outcome in adolescents with spina bifida. J Int Neuropsychol Soc 2008;14:793–804.
7. Dennis M, Barnes MA. The cognitive phenotype of spina bifida meningomyelocele. Dev Disabil Res Rev 2010;16:31–39.

8. Rose BM, Holmbeck GN. Attention and executive functions in adolescents with spina bifida. J Pediatr Psychol 2007;32:983–994.
9. Dennis M, Landry SH, Barnes M, Fletcher JM. A model of neurocognitive function in spina bifida over the life span. J Int Neuropsychol Soc 2006;12:285–296.
10. Iddon JL, Morgan DJ, Loveday C, Sahakian BJ, Pickard JD. Neuropsychological profile of young adults with spina bifida with or without hydrocephalus. J Neurol Neurosurg Psychiatr 2004;75:1112–1118.
11. Sawin KJ, Buran CF, Brei TJ, Fastenau PS. Correlates of functional status, self-management, and developmental competence outcomes in adolescents with spina bifida. SCI Nurs 2003;20:72–85.
12. Holmbeck GN, Devine KA. Psychosocial and family functioning in spina bifida. Dev Disabil Res Rev 2010;16:40–46.
13. Holmbeck GN, DeLucia C, Essner B, Kelly L, Zebracki K, Friedman D, Jandasek B. Trajectories of psychosocial adjustment in adolescents with spina bifida: a 6-year, four-wave longitudinal follow-up. J Consult Clin Psychol 2010;78:511–525.
14. Zukerman JM, Devine KA, Holmbeck GN. Adolescent predictors of emerging adulthood milestones in youth with spina bifida. J Pediatr Psychol 2011;36:265–276.
15. Dennis M, Jewell D, Drake J, Misakyan T, Spiegler B, Hetherington R, Gentili F, Barnes M. Prospective, declarative, and nondeclarative memory in young adults with spina bifida. J Int Neuropsychol Soc 2007;13:312–323.
16. 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.
17. Appleton PL, Ellis NC, Minchom PE, Lawson V, Böll V, Jones P. Depressive symptoms and self-concept in young people with spina bifida. J Pediatr Psychol 1997;22:707–722.
18. Stubberud J, Riemer G, Plaum PE, Grimsrud K. Psychosocial adaptation and cognitive functioning in young male adults with myelomeningocele. Cerebrospinal Fluid Res 2007;4:39.
19. Delis DC, Kaplan E, Kramer JH. Delis-Kaplan Executive Functioning System (D-KEFS). San Antonio, TX: The Psychological Corporation; 2001.
20. Wechsler Adult Intelligence Scale III. San Antonio, TX: The Psychological Corporation; 1997.
21. Reitan RM. Validity of the Trail Making Test as an indicator of organic brain damage. Percept Motor Skill 1958;8:271–276.
22. Wechsler Abbreviated Scale of Intelligence. San Antonio, TX: The Psychological Corporation. 1999.
23. Matthews CG, Kløve H. Instruction manual for the Adult Neuropsychology Test Battery. Madison, WI: University of Wisconsin Medical School; 1964.
24. Gioia GA, Isquith PK, Guy SC, Kenworthy L. Behavior rating inventory of executive function: professional manual. Lutz, FL: Psychological Assessment Resources, Inc; 2000.
25. Derogatis LR. Symptom Checklist-90 (Revised edition). Minneapolis, MN: Pearson Assessments; 1994.
26. Hunt GM. Open spina bifida: outcome for a complete cohort treated unselectively and followed into adulthood. Dev Med Child Neurol 1990;32:108–118.
27. Hunt GM, Poulton A. Open spina bifida: a complete cohort reviewed 25 years after closure. Dev Med Child Neurol 1995;37:19–29.
28. Barf HA, Post MW, Verhoef M, Jennekens-Schinkel A, Gooskens RH, Prevo AJ. Restrictions in social participation of young adults with spina bifida. Disabil Rehabil 2009;31:921–927.
29. Mazaux JM, Masson F, Levin HS, Alaoui P, Maurette P, Barat M. Long-term neuropsychological outcome and loss of social autonomy after traumatic brain injury. Arch Phys Med Rehabil 1997;78:1316–1320.
30. Tranel D, Anderson SW, Benton A. Development of the concept of "executive function" and its relationship to the frontal lobes. In: Boller F, Grafman J, editors. Handbook of neuropsychology. Amsterdam: Elsevier; 1994. pp 125–148.
31. Stuss DT, Levine B. Adult clinical neuropsychology: lessons from studies of the frontal lobes. Annu Rev Psychol 2002;53:401–433.
32. Levine B, Stuss DT, Winocur G, Binns MA, Fahy L, Mandic M, Bridges K, Robertson IH. Cognitive rehabilitation in the elderly: effects on strategic behavior in relation to goal management. J Int Neuropsychol Soc 2007;13:143–152.
33. Mahone EM, Zabel TA, Levey E, Verda M, Kinsman S. Parent and self-report ratings of executive function in adolescents with myelomeningocele and hydrocephalus. Child Neuropsychol 2002;8:258–270.
34. Burmeister R, Hannay HJ, Copeland K, Fletcher JM, Boudousquie A, Dennis M. Attention problems and executive functions in children with spina bifida and hydrocephalus. Child Neuropsychol 2005;11:265–283.
35. van Hooren SA, Valentijn SA, Bosma H, Ponds RW, van Boxtel MP, Levine B, Robertson I, Jolles J. Effect of a structured course involving goal management training in older adults: A randomised controlled trial. Patient Educ Couns 2007;65:205–213.
36. Almeida DM, Kessler RC. Everyday stressors and gender differences in daily distress. J Pers Soc Psychol 1998;75:670–680.
37. Hillan J, McCann JP: Hypoventilation and sleep apnoea in spina bifida–an unrecognised problem? Cerebrospinal Fluid Res 2009;6:39.
38. Dosa NP, Foley JT, Eckrich M, Woodall-Ruff D, Liptak GS. Obesity across the lifespan among persons with spina bifida. Disabil Rehabil 2009;31:914–920.
39. Plaum PE, Riemer G, Frøslie KF. Risk factors for pressure sores in adult patients with myelomeningocele–a questionnaire-based study. Cerebrospinal Fluid Res 2006;3:14.
40. Wilson BA. Neuropsychological rehabilitation. Annu Rev Clin Psychol 2008;4:141–162.
41. Cicerone KD, Dahlberg C, Kalmar K, Langenbahn DM, Malec JF, Bergquist TF, Felicetti T, et al. Evidence-based cognitive rehabilitation: recommendations for clinical practice. Arch Phys Med Rehabil 2000;81:1596–1615.
42. Cicerone KD, Dahlberg C, Malec JF, Langenbahn DM, Felicetti T, Kneipp S, Ellmo W, et al. Evidence-based cognitive rehabilitation: updated review of the literature from 1998 through 2002. Arch Phys Med Rehabil 2005;86:1681–1692.
43. Rees L, Marshall S, Hartridge C, Mackie D, Weiser M; Erabi Group. Cognitive interventions post acquired brain injury. Brain Inj 2007;21:161–200.