# Spina bifida: implications for cognitive functioning, disability and health in young adults

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Voor mijn ouders, Arend en Wimmy

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# Spina bifida: implications for cognitive functioning, disability and health in young adults

# Spina bifida: implicaties voor het cognitief functioneren, de participatie en het welbevinden van jong volwassenen

(met een samenvatting in het Nederlands)

**Proefschrift**

ter verkrijging van de graad van doctor aan de Universiteit Utrecht op gezag van de rector magnificus, prof. dr. J.C. Stoof, ingevolge het besluit van het college voor promoties in het openbaar te verdedigen op dinsdag 29 april 2008 des namiddags om 2.30 uur

door

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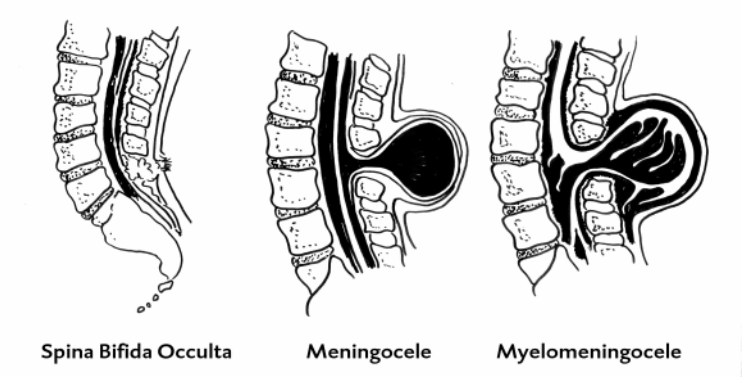
# Chapter 1

## INTRODUCTION

## SPINA BIFIDA AND HYDROCEPHALUS

Spina bifida (Latin for split spine) is a serious birth abnormality associated with a failure of closure of the neural tube. It is one of the most common congenital birth defects, occurring in the first month of the pregnancy. This developmental error may lead to a variety of spinal defects: myelomeningocele, meningocele or spina bifida occulta (see Figure 1).[1]

**Figure 1**




Spina bifida occulta or 'hidden' spina bifida is a very mild and common form of spina bifida. It results from a gap in one or more vertebral arches, but the spina cord and meninges remain entirely within the vertebral canal. Only rarely does it present itself with neurological defects. The prevalence of this type of spina bifida is unknown, but a rough estimate is that 10% to 20% of the general population is affected.[2,3] The defect is often found 'by accident' during radiographic studies or diagnoses, though sometimes visible signs are present, e.g., a dimple, birthmark or small patch of hair on the lower back.[1] Some persons with spina bifida occulta experience only in later life neurological deficit or problems with bladder and bowel control due to a tethered cord.

Spina bifida aperta or spina bifida cystica is the 'open form' of spina bifida, which may occur as meningocele and as myelomeningocele. In both forms a cele or cyst is present. The cele is covered by meninges and/or skin. In meningocele the neural tissue is not damaged. In myelomeningocele, the most common and more serious form of the two, the cele contains spinal cord, spinal roots and cerebrospinal fluid. The spinal cord is not properly developed, which results in some form of impairment in approximately 99% of the persons.[1,3]

The exact cause of spina bifida is still unknown. Spina bifida likely results from the interaction of genetic factors and environmental factors.[3] The incidence of spina bifida

varies across the different parts of the world and even across different parts of Europe.[3-5] There are some explanatory factors of the differences. One is different preventive policies, like the use of folic acid. The use of folic acid before the conception and during the pregnancy can reduce the incidence of neural tube defects.[1,5] In the Netherlands, the prevalence of neural tube defects was 6.4 per 10.000 live births in the period 1993-1995.[6] Some four years later, after a campaign promoting the use of folic acid, the prevalence of neural tube defects (of which 95% were spina bifida aperta) dropped to 3.8 per 10.000 live births.[7] Other possible explanations for differences in incidence are differences in antenatal screening policies between countries and differences in the acceptance of termination of pregnancy after detection of the neural tube defect.[1,8]

After birth, immediate treatment is focused on closure and replacement of the neural tissues to prevent further damage and infection. After closure of the defect roughly 85% of the newborns with spina bifida aperta develop hydrocephalus.[6,7] Newborns with spina bifida aperta very often also have a Chiari II (Arnold Chiari) malformation of the medullar-spinal junction. It is believed that hydrocephalus results from obstruction of the circulation of CSF due to Chiari II malformation, though underabsorption of CSF also might be present.[1,5] The treatment of hydrocephalus consists of placing a shunt releasing the pressure due to an excess of CFS.

## Outcome

In the 1950's roughly 75% of the newborns with spina bifida died within the first year of life.[9] Since then, the survival rate has increased dramatically, and in the early 1990's two-thirds of the live born babies survived because of the use of new shunting techniques and better renal management.[1,5,10-19] Also, long-term orthopaedic and urological management treatment have considerably improved.[6,10,17,20] Only recently a relatively large number of newborns have survived into adulthood.[17,21] However, a growing number of studies shows that the physical and cognitive functioning of these children is less favourable than initially anticipated.[11,13,14,18,20,22-30]

During the course of life the emphasis of the treatment shifts from (life saving) medical treatment at young age, followed by interventions focused on an optimal level of functioning at school age, towards more psychosocial care at adolescence and adulthood. The functional limitations persist into adulthood.

## Cognitive functioning

Cognitive functioning is affected in many children with spina bifida aperta. Hydrocephalus has been identified as the major cause of cognitive dysfunctioning. There is no standard neuropsychological profile of children with spina bifida and hydrocephalus. Some children are retarded as a result of hydrocephalus, whereas other children with seemingly similar health conditions only show specific or subtle cognitive dysfunction or no dysfunction at all. A variety of cognitive dysfunctions has been described in the literature, which can be present but not necessarily are present in a patient with spina bifida.[21,26,29-57] On average intelligence is lower and studies found intelligence favoured towards a higher verbal component compared to a lower performance component. Speech development is poorer, though phonological, semantic and syntax abilities are normal. Some children with spina bifida and hydrocephalus have hyperverbal behaviour, also referred to as cocktail party syndrome. Attention, planning and executive functioning are also likely to be affected in children with spina bifida and hydrocephalus. Few studies on memory suggest that memory deficits are not characteristic in these children, though verbal learning ability seems to be affected and children may exhibit poor learning for orientation in space. Visual-motor functioning and hand functioning tend to be impaired, which in turn can lead to difficulties in functional abilities like play skills, handwriting and driving throughout the life span. Reading comprehension, spelling, mathematics and handwriting are affected in part of the group of children.[21,26,29-57]

## Quality of life

When compared to their healthy peers, youngsters with chronic conditions such as spina bifida are confronted with additional barriers in the transition from adolescence to adulthood.[58-60] Cognitive functioning is considered to be a variable that mediates or moderates behavioural and social outcome.[61] Unrestricted participation has been shown to be difficult to obtain by persons with spina bifida. Level of education is lower than in the general population, unemployment rate is high, only few persons can live independently without extra help and persons with spina bifida seem to have more difficulty finding a partner.[17,19,62-64] On reaching adulthood, part of the population with spina bifida shows poor long-term achievement.[21] Surprisingly, the number of studies on the quality of life of persons born with spina bifida is still limited and their results are equivocal. Some authors[65] found that the health related quality of life of their study population was below normal, whereas others concluded that quality of life was good or comparable to persons without spina bifida.[58,59,66]

Although there is a growing attention for adults with spina bifida, the majority of spina bifida studies still concern children under the age of 16 years. Also, studies tend to focus on the physical impairments as determinants of outcome, and the influence of cognitive impairments seem underrated. Other limitations apply to most studies published to date, like small sample sizes, absence of controls or normative data and a narrow focus, e.g., on intelligence quotient scores (IQ) only. A comprehensive neuropsychological examination is required in the diverse population of persons with spina bifida. Such studies are lacking in the adult spina bifida population. Although IQ test scores seem to be relatively stable during school years[67], it remains to be established whether cognitive deficits remain stable throughout the lifetime. Hence, results from paediatric studies cannot be transferred easily to adults with spina bifida.

### Long-term care

Medical care for spina bifida patients in the Netherlands is currently organized in multidisciplinary spina bifida teams. The guidance given by these teams is mainly focused on the period of childhood and most have an upper age limit for patients. There is a general agreement that multidisciplinary care for patients with spina bifida needs to be continued into adulthood.[20,24,68-74] The transition from paediatric to adult health-care is however not well organized to date. A similar situation has been described for other countries and other chronic conditions.[23] In order to improve the transition of youngsters with spina bifida into adulthood and to adjust counselling by health care professionals to the needs of these youngsters, it is important to study determinants of social participation and hindrances for participation as perceived by young adults with spina bifida.

## THE ASPINE STUDY

The ASPINE study (Adolescents with SPina bifida In the NEtherlands) was started to examine the functioning of young adults with spina bifida in the Netherlands. The aim of the study was to identify care needs in order to provide recommendations for the improvement of care and guidance for this age group. For this reason, ASPINE was designed as a broad exploratory study of physical functioning, cognitive functioning, independence in daily living, functioning in the community, perceived health and life satisfaction, as well as the actual care and need for care. Because of this broad focus, the ASPINE study was performed by both a physician (Marjolein Verhoef) and a psychologist

(Hans Barf). They jointly set up the study and gathered the data, after which they wrote separate dissertations about the medical and psychological aspects of spina bifida in young adulthood, respectively.[75]

To assess the functioning of young adults with spina bifida, we used a model based on the International Classification of Functioning, Disability and Health (ICF).[76] The ICF model distinguishes between the outcome of diseases at three levels; (1) body functions and body structures, (2) activities and (3) participation. Factors that affect these outcomes are not only the disease itself, but also external factors (environmental factors) and personal factors (for example age, gender and personality). Apart from assessing health status and from objectifying the level of functioning, we measured level of functioning and 'well-being' as perceived by the participants. The measures used to assess the cognitive functioning, level of activities, performance in participation and overall well-being are described in more detail in the chapters 2-7.

## Participants

All 12 multidisciplinary spina bifida teams in the Netherlands were invited to participate in the ASPINE study, and all but one participated. Each spina bifida team listed all current and past patients who met the inclusion criteria of having spina bifida aperta or spina bifida occulta (ICD-codes 741 and 756.17[77]), being aged 16-25 years and living in the Netherlands. Patients received a written invitation from a member of their spina bifida team to participate in the study. We also used other ways of enlisting possible participants because the number of potential participants did not meet our initial expectations. Patients were invited by BOSK, the Dutch patients' association. Adverts were placed in magazines with an invitation to contact the researchers. Other organizations providing care to patients with spina bifida were also asked to participate in the study (13 rehabilitation centres, 91 special housing projects and 12 hospitals in areas where no spina bifida team was active). Six rehabilitation centres and 8 special housing projects responded, which resulted in 52 further potential participants. However, most of them were already known through the files of the spina bifida teams.

In total, of the 350 patients who were invited to participate in the ASPINE study, 181 were willing to participate (response 51.1%). Two patients were excluded afterwards because of co-morbidity causing more physical and/or mental problems than the neural tube defect itself. Some patients reported their reason for their refusal to participate. In majority, the reason was that the time required for the examination did not fit the busy daily schedule. Another reason was that after a childhood period of intensive medical

treatment, the invitee avoided contacts with the health care system. Demographic and medical characteristics were retrieved from medical files to detect possible non-response bias. Participants and non-participants did not significantly differ in terms of gender, age, type of spina bifida, level of lesion and hydrocephalus (Table 1).

**TABLE 1: COMPARISON BETWEEN PARTICIPANTS AND NON-PARTICIPANTS**

| | Participants (N = 179) | Non-participants (N = 171)* | Total (N = 350) | Significance (p-value) |
|---|---|---|---|---|
| **Gender** | | | | |
| % female | 58.7 | 50.9 | 54.9 | 0.163 |
| **Age** | | | | |
| mean (SD) | 20.4 (3.0) | 20.3 (3.1) | 20.3 (3.0) | 0.816 |
| **Type of spina bifida** | | | | |
| % aperta | 79.3 | 86.0 | 81.8 | 0.205 |
| **Level of lesion** | | | | |
| % … – L2 | 19.0 | 23.1 | 20.5 | 0.648 |
| % L3 – L5 | 65.9 | 64.4 | 65.4 | |
| % S1 – … | 15.1 | 12.5 | 14.1 | |
| **Hydrocephalus** | | | | |
| % shunted | 66.5 | 64.2 | 65.6 | 0.700 |

* Numbers can vary because not all data for non-participants were available from the medical records

All patients were examined once, in a session lasting about 3 hours, usually in the hospital where they had been treated in childhood. Some patients were visited at home. Data were collected by means of interviews, physical examinations (performed by Marjolein Verhoef, a physician), neuropsychological tests (performed by the psychologist Hans Barf, or by well-trained students under his supervision) and questionnaires. Medical history data were collected from patient records before the examination. Questionnaires were mailed to the participant together with the confirmation of the appointment. The returned questionnaires were checked for missing data during the examination.

## SCOPE OF THIS THESIS

The main aims of this thesis were (1) to obtain insight into the cognitive functioning of young adults with spina bifida, and (2) to obtain knowledge of the relationships of

hydrocephalus and cognitive dysfunction on educational career, participation and life satisfaction of young adults with spina bifida. The topics of this study are dealt with in the following chapters.

Chapter 2 describes the secondary health conditions in young adults with spina bifida. The prevalence of secondary impairments is related to type of spina bifida, level of lesion and hydrocephalus.

In chapter 3 the cognitive status is assessed. The effects of primary neurological parameters on the cognitive status are explored, as well as the effects of secondary parameters (complications of hydrocephalus) and associated pathology.

Chapter 4 reports on the educational career and predictors of attending special education. Predictive value of primary neurological parameters, as well as of cognition and secondary impairments was explored.

Chapter 5 reports on the restrictions in social participation in relation to health condition and activity limitations. It also reports on the perceived hindrances of the young adults with spina bifida.

Chapter 6 addresses the life satisfaction of young adults with spina bifida in comparison to that of the general population. Life satisfaction is related to the health condition as a possible determinant.

Chapter 7 further explores the relation between quality of life and several domains of cognitive functioning.

Finally, chapter 8 is a general discussion of the overall conclusions, strengths and limitations of this study, as well as possible implications for treatment, health care policy and directions for future research.

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# Chapter 2

## SECONDARY IMPAIRMENTS IN YOUNG ADULTS WITH SPINA BIFIDA

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## ABSTRACT

The aim of this study was to examine the prevalence of secondary impairments in young adults with spina bifida and to relate the prevalence to the type of spina bifida and the level of lesion. This cross-sectional study is part of the ASPINE (Adolescents with Spina Bifida in the Netherlands) study.

Data were collected on medical history, hydrocephalus (shunt: yes/no), neurological level of lesion (International Standards for Neurological and Functional Classification of Spinal Cord Injury), visual acuity (Landolt rings), spasticity (Modified Ashworth Scale), contractures (range of motion), scoliosis (deviation from perpendicular), ambulation (Hoffer criteria), pressure sores and blood pressure (physical examination), epilepsy, pain, incontinence and sexuality (questionnaire), and cognitive functioning (Raven Standard Progressive Matrices). In total, 179 patients with spina bifida participated (41% male, age range 16 to 25 years, mean 20 years 9 months, SD 2 years 11 months). These were 37 patients with spina bifida occulta, 119 patients with spina bifida aperta and hydrocephalus ($AHC^+$) and 23 patients with spina bifida aperta without hydrocephalus ($AHC^-$).

Of our patient group, 73 had a high-level lesion (L2 and above), 68 a mid-level lesion (L3 to L5), and 38 a low-level lesion (S1 and below). Both subdivisions were strongly related with patients with higher lesions more often having hydrocephalus. Most secondary impairments were found for patients with $AHC^+$, and patients with $AHC^-$ were mostly comparable to patients with spina bifida occulta. According to level of lesion, most medical problems were found in the high-level lesion group. However, all subgroups suffered from health problems.

## INTRODUCTION

With advances in medical treatment, care for people with spina bifida has become lifelong. However, knowledge about health conditions in spina bifida is based largely on studies in children. A few articles on medical problems in young adults with spina bifida have been published (Shurtleff and Sousa 1977, Hagelsteen et al. 1989, Hunt 1990, Steinbok et al. 1992, Exner et al. 1993, Farley et al. 1994, Hunt and Poulton 1995, Morgan et al. 1995, McDonnell and McCann 2000, Bowman et al. 2001, Hunt and Oakeshott 2003). The most important medical problems described are hydrocephalus and cognitive dysfunction, urinary and faecal incontinence, reduced mobility, renal failure, hypertension, pressure sores, obesity, epilepsy, and decreased visual acuity. Orthopaedic problems such as scoliosis, fractures, and contractures are also often mentioned (Swank and Dias 1992, Brinker et al. 1994). However, the prevalence of medical problems in adolescent patients with spina bifida varies between studies, which might be due to differences in age of the participants, definition problems, and differences in circumstance (e.g. cultural aspects, time of study).

Spina bifida has various manifestations. Most articles have focused on patients with myelomeningocele only. Only few have been published on patients with spina bifida occulta (Satar et al. 1995). The subdivision that is generally used in articles is based on the level of lesion, with different cut-off points and different ways of assessing the level of lesion (Shurtleff et al. 1975, Hunt 1990, Swank and Dias 1992, Hunt and Poulton 1995, Staal et al. 1996, McDonnell and McCann 2000). Most overview articles do not describe outcome for different subgroups at all (Hagelsteen et al. 1989, Steinbok et al. 1992, Exner et al. 1993, Farley et al. 1994, Morgan et al. 1995, Bowman et al. 2001).

The aim of the present study was to describe secondary health conditions in a large group of Dutch adolescents with spina bifida aperta as well as spina bifida occulta, to be able to give better information on secondary health conditions for parents and patients themselves and to be able to give sufficient care to patients with spina bifida while growing up. To obtain more specific information on subgroups, two relevant classifications based on primary characteristics of the condition were used. The first classification is based on the type of spina bifida (aperta and occulta) and the existence of hydrocephalus in patients with spina bifida aperta. The second classification used is based on the level of lesion.

This study is part of the ASPINE study (Adolescents with Spina Bifida in the Netherlands), a cross-sectional study on physical and cognitive disabilities, health care, participation in society, and life satisfaction of this group of adolescents.

## METHOD

### Participants

Young adults were included with any kind of spina bifida (myelomeningocele, meningocele) or spina bifida occulta (International Classification of Diseases, 9th revision codes 741 and 756.17 respectively; World Health Organization 2003), aged between 16 and 25 years, and living in the Netherlands. Excluded were non-Dutch-speaking patients or patients with comorbidity causing more physical and/or cognitive problems than the neural tube defect itself.

Patients were recruited from 11 of the 12 spina bifida teams in the Netherlands. The Dutch Association for Patients with Spina Bifida also invited members to participate and advertisements were placed in two national magazines and on the internet. In addition, rehabilitation centres, housing facilities, and special schools were approached to find potential participants.

In total, 350 patients were invited by mail to participate in this study, of whom 181 were willing to participate. For 20% of the non-participants the reasons for not participating are known. Unknown address and lack of time were mentioned in one-third of patients.

### Instruments

Data were collected by means of interview, a physical examination (performed by a physician), and neuropsychological tests (performed by a neuropsychologist). Data about medical history were collected from medical records.

Hydrocephalus was categorized as either having a shunt at the time of the physical examination or having had one previously. For six patients, hydrocephalus was mentioned in the medical record at some time, however no shunt was placed. We assume that this was only minor hydrocephalus and we categorized those patients as not having hydrocephalus. Level of lesion was defined in accordance with the International Standards for Neurological and Functional Classification of Spinal Cord Injury (Ditunno et al. 1994, Maynard et al. 1997) as the lowest completely unimpaired dermatome level on both sides measured with sensitivity to pin prick and light touch. Three lesion level groups were defined: high level (HLL; L2 and above), middle level (MLL; L3 to L5), and

low level (LLL; S1 and below; Shurtleff et al. 1975, Evans et al. 1985, Swank and Dias 1992, Staal et al. 1996).

Visual acuity was measured for both eyes with Landolt rings, using a stenopeic opening when needed. A visual acuity of 0.8 or more for at least one eye was defined as normal. Having both eyes with a visual acuity of less than 0.8 was defined as decreased visual acuity (Caines and Dahl 1997). Patients using medication for epilepsy at the time of the physical examination were classified as suffering from epilepsy. Cognitive functioning was determined with the Raven Standard Progressive Matrices (Raven et al. 1998). A 20-minute time limit was used for testing, for which recent Dutch norm scores are available (Bouma et al. 1996). The test result (T-score) was corrected for age and sex and converted to IQ. Mean IQ of a healthy population is 100, with a standard deviation (SD) of 15. Patients with an IQ of 70 or below are described as having learning disabilities* (Lezak 1995). Information about pain was requested in the interview. Patients were asked whether they suffered from pain in the head, neck, or back at least once a month and whether they had experienced increased pain in the last year. Spasticity was measured with the Modified Ashworth Scale for grading spasticity (Bohannon and Smith 1987). Patients with a slight increase in muscle tone, manifested by a 'catch', followed by minimal resistance throughout the remainder (less than half) of the range of motion in one or both legs, were defined as having spasticity.

Contractures were measured for hip and knee extension by using a goniometer. Patients with limitations in range of motion of more than 30° in one or both extremities were defined as having contractures. When one foot (or both) could not be put in a neutral position this was registered as a foot deformity. Scoliosis was ascertained during the physical examination when there was at least one curve that deviated more than 2cm from perpendicular. Patients were determined as suffering from lumbar lordosis when lumbar lordosis was obviously present in a sitting position.

For ambulation the Hoffer criteria were used (Hoffer et al. 1973). Community ambulators walk indoors and outdoors for most of their activities and may need crutches or braces, or both. They use a wheelchair only for long trips. Household ambulators walk only indoors and with an apparatus. They are able to get in and out of a chair and bed with little if any assistance. They may use a wheelchair for some indoor activities at home and school and for all activities in the community. Nonfunctional ambulators are patients who are able to walk in a therapy session. Afterwards they use their wheelchair. Nonambulators are patients who use a wheelchair only. In our study an extra category of normal ambulators

was added. Those are patients not using walking aids or a wheelchair at all. According to the Hoffer classification, it can be seen as a subgroup of the community ambulators.
Patients who had good sitting balance with both hands free were classified as having a 'no balance deficit'. Patients who needed at least one hand to maintain upright sitting position were classified as having a deficit (Swank and Dias 1992).
Information on bladder and bowel management was obtained by a review of the history. Incontinence was classified as spoilage with either urine or faeces requiring the need for change of clothes or napkin at least once a month (with or without use of condom-type, urethral or suprapubic catheter). Patients were also asked whether they experienced bladder or bowel incontinence as a problem.
Data on sexuality were gathered by means of a structured interview. Patients were classified as sexually active or not. 'Sexually active' was defined as having ever masturbated or had sexual contact with others, with sexual contact meaning French kissing or further sexual contact. The sexually-active patients were asked how often they experienced an erection, ejaculation, lubrication, and orgasm when masturbating or having sexual contact. Experiencing those functions sometimes or never was defined as subnormal in those who were sexually active.
Pressure sores were determined by physical examination describing four stages of pressure sores: redness that did not disappear on pressure, blisters, superficial wounds, and deep wounds (Haalboom and Bakker 1992). Having blisters or worse was classified as having pressure sores. Blood pressure was determined using an Omron HEM-705CP blood pressure monitor (O'Brien et al. 1996). Blood pressure was measured every hour during the examination; three times in total. The mean of the three values was calculated. Hypertension was defined as a mean diastolic blood pressure of more than 95mmHg and/or a mean systolic blood pressure of more than 160mmHg.

### Statistical analysis

Three subgroups based on type of spina bifida were defined: spina bifida occulta, spina bifida aperta without hydrocephalus ($AHC^-$), and spina bifida aperta with hydrocephalus ($AHC^+$). Three subgroups based on level of lesion were used, as defined above: HLL, MLL, and LLL. Data were analyzed with descriptive statistics by using SPSS (version 10). Percentages of patients with several conditions are presented in the tables. Data were dichotomized and presented in cross tabs, using $\chi^2$ to determine differences between subgroups of patients; $p<0.05$ indicates significant differences between subgroups.

Although some subgroups are small, percentages were used to make the results between subgroups easily comparable.

### Ethical approval

The medical ethics committee approved the ASPINE study. Informed consent from all participants was obtained.

## RESULTS

No significant difference between participants and non-participants was found with regard to age, sex, type of spina bifida, level of defect, and being shunted for hydrocephalus on the basis of information from medical records. Two invited participants were excluded because of comorbidity independently inducing serious physical and/or cognitive disorders.

A total of 179 patients participated in the ASPINE study. Almost all were examined in a hospital; in 10 participants the interview was performed by phone and in these cases data based on the physical examination and on neuropsychological testing are missing. In one case neuropsychological testing was impossible because of total visual impairment. Missing data might cause small differences in numbers of patients. Data were complete for 168 patients.

Mean age of the population was 20 years 9 months, with an SD of 2 years 11 months and a range of 16 to 25 years. Seventyfour patients (41%) were men. Of the 142 patients with spina bifida aperta, 109 were diagnosed as having myelomeningocele, 13 as having meningocele, and 20 as having aperta not specified. The 37 patients with spina bifida occulta included all 15 patients who were diagnosed as having a lipoma.

Level of lesion (divided into three subgroups) is related to the type of spina bifida (occulta versus aperta) with a Spearman's correlation of 0.36, and to the presence or absence of hydrocephalus with a Spearman's correlation of 0.55. Patients with $AHC^{-}$ were most comparable to patients with spina bifida occulta as regards level of lesion (Table I).

In the following result sections, percentages of patients suffering from different problems are described separately for the subgroups of each classification. This might suggest a causal relationship in some cases. However, spina bifida is a complex condition with different manifestations, which are often related. One should, therefore, be careful in presuming causality disregarding other aspects of the disease.

**TABLE I: INTERRELATIONSHIP BETWEEN TYPE OF SPINA BIFIDA AND LEVEL OF LESION**

| Lesion type | AHC$^+$ | AHC$^-$ | Occ | Total |
|---|---|---|---|---|
| **HLL** | 66 | 2 | 5 | 73 (41%) |
| **MLL** | 47 | 7 | 14 | 68 (38%) |
| **LLL** | 6 | 14 | 18 | 38 (21%) |
| **Total** | 119 (66%) | 23 (13%) | 37 (21%) | 179 (100%) |

Figures are numbers of patients except as noted. AHC$^+$, spina bifida aperta with hydrocephalus; AHC$^-$, spina bifida aperta without hydrocephalus; Occ, spina bifida occulta; HLL, high-level lesion (L2 and above); MLL, middle-level lesion (L3 to L5); LLL, low-level lesion (S1 and below).

**TABLE II: PERCENTAGES OF PATIENTS WITH SPINA BIFIDA WITH A SURGICAL HISTORY, RELATED TO TYPE OF SPINA BIFIDA AND LEVEL OF LESION**

| | AHC$^+$ (n=119) | AHC$^-$ (n=23) | Occ (n=37) | | HLL (n=73) | MLL (n=68) | LLL (n=38) | |
|---|---|---|---|---|---|---|---|---|
| **Neurosurgery, %** | 100 | 100 | 87 | ** | 100 | 97 | 92 | |
| Shunt revisions, % | 84 | 0 | 0 | ** | 78 | 57 | 11 | ** |
| Cervical decompression, % | 10 | 0 | 0 | * | 10 | 6 | 3 | |
| Tethered cord surgery, % | 18 | 39 | 68 | ** | 23 | 25 | 55 | ** |
| **Orthopedic surgery, %** | 77 | 39 | 46 | ** | 85 | 69 | 21 | ** |
| Scoliosis surgery, % | 27 | 4 | 8 | ** | 41 | 6 | 5 | ** |
| Lower extremities surgery, % | 68 | 39 | 38 | ** | 71 | 69 | 13 | ** |
| **Urological surgery, %** | 50 | 61 | 14 | ** | 55 | 40 | 32 | * |
| **Total number of neurological, orthopedic and urological operations** | | | | | | | | |
| Median | 8 | 3 | 2 | | 9 | 6 | 2 | |
| Range | 2-35 | 1-28 | 0-11 | | 3 - 35 | 0 - 33 | 0 - 15 | |

AHC$^+$, spina bifida aperta with hydrocephalus; AHC$^-$, spina bifida aperta without hydrocephalus; Occ, spina bifida occulta; HLL, high-level lesion (L2 and above); MLL, middle-level lesion (L3 to L5); LLL, low-level lesion (S1 and below). *Significant differences between three subgroups with Pearson's $\chi^2$ $p<0.05$. **Significant differences between three subgroups with Pearson's $\chi^2$ $p<0.01$.

## Medical history

The total number of occurrences of neurosurgery, orthopaedic surgery, or urological surgery varied from 0 to 35, being highest in the AHC$^+$ group and the HLL group. All except three patients with spina bifida occulta had undergone surgery in these categories in the past. Table II shows the percentages of patients having had one or more operations for certain problems. Shunt revisions had been performed in most patients with AHC$^+$, with a maximum of 24 revisions and a median of 3. Tethered cord surgery was performed mainly in patients with spina bifida occulta. Orthopaedic surgery was common in most

**TABLE III: PERCENTAGES OF PATIENTS WITH SPINA BIFIDA AND NEUROLOGICAL PROBLEMS, RELATED TO TYPE OF SPINA BIFIDA AND LEVEL OF LESION**

| Neurological problem | AHC$^+$ (n=119)[a] | AHC$^-$ (n=23)[a] | Occ (n=37) | | HLL (n=73)[a] | MLL (n=68)[a] | LLL (n=38)[a] | |
|---|---|---|---|---|---|---|---|---|
| **Visual acuity below 0.8 (both eyes), %** | 8 | 0 | 5 | | 10 | 5 | 3 | |
| **Epilepsy, %** | 9 | 0 | 3 | | 13 | 3 | 3 | * |
| **IQ =< 70, %** | 20 | 0 | 3 | ** | 22 | 9 | 6 | * |
| **Pain, %** | 35 | 26 | 25 | | 26 | 29 | 26 | |
| **Spasticity, %** | 13 | 0 | 0 | * | 13 | 3 | 3 | * |

AHC$^+$, spina bifida aperta with hydrocephalus; AHC$^-$, spina bifida aperta without hydrocephalus; Occ, spina bifida occulta; HLL, high-level lesion (L2 and above); MLL, middle-level lesion (L3 to L5); LLL, low-level lesion (S1 and below). [a]Numbers of observations: AHC$^+$, range 111 to 119; AHC$^+$, range 20 to 23; HLL, range 69 to 73; MLL, range 65 to 68; LLL, range 34 to 38. *Significant differences between three subgroups with Pearson's $\chi^2 p<0.05$. **Significant differences between three subgroups with Pearson's $\chi^2 p<0.01$.

patients with AHC$^+$ and had occurred in almost half of the patients with spina bifida occulta. The higher the level of lesion, the more patients had undergone orthopaedic surgery.

## Neurological problems

Neurological problems are shown in Table III. Epilepsy and abnormal visual acuity occurred mainly in the AHC$^+$ group, although no significant difference was found. One patient with a shunt had total visual impairment. An IQ below 70 was found only in patients with AHC$^+$ except for one patient with spina bifida occulta, and significantly more were found in patients with HLL. Altogether 49 patients reported an increase in pain in their head, neck, or back in the past year, with 29 patients reporting back pain, 25 with headache, and 19 with neck pain. One-third of patients with spina bifida occulta complained of pain, as did one-quarter in both the AHC$^+$ and AHC$^-$ groups. Similar percentages were found in the different groups defined by level of lesion. Spasticity in one or both legs was found in 13% of patients with AHC$^+$ and was not found in the LLL group.

## Orthopaedic problems and ambulation

Table IV identifies the most important orthopaedic problems and ambulatory status of the young adults with spina bifida. In more than one-third of the patients with AHC$^+$ and more than one-half of patients with HLL, obvious scoliosis could be diagnosed during the

**TABLE IV: PERCENTAGES OF PATIENTS WITH SPINA BIFIDA AND ORTHOPAEDIC PROBLEMS, AND AMBULATORY STATUS RELATED TO TYPE OF SPINA BIFIDA AND LEVEL OF LESION**

| Condition | AHC$^+$ (n=119)[a] | AHC$^-$ (n=23)[a] | Occ (n=37) | | HLL (n=73)[a] | MLL (n=68)[a] | LLL (n=38)[a] | |
|---|---|---|---|---|---|---|---|---|
| **Scoliosis** | 41 | 10 | 16 | ** | 59 | 19 | 6 | ** |
| **Lumbar lordosis** | 39 | 15 | 6 | ** | 50 | 22 | 0 | ** |
| **Sitting balance deficit** | 19 | 0 | 0 | ** | 29 | 2 | 0 | ** |
| **Contractures hip** | 18 | 0 | 0 | ** | 28 | 2 | 0 | ** |
| **Contractures knee** | 36 | 5 | 3 | ** | 46 | 15 | 0 | ** |
| **Foot deformities** | 85 | 45 | 49 | ** | 86 | 86 | 20 | ** |
| **Ambulation** | | | | ** | | | | ** |
| Normal ambulator | 14 | 70 | 84 | | 4 | 38 | 92 | |
| Community ambulator | 17 | 17 | 11 | | 11 | 27 | 5 | |
| Household ambulator | 13 | 4 | 0 | | 8 | 15 | 3 | |
| Non-functional ambulator | 7 | 4 | 0 | | 6 | 7 | 0 | |
| Non-ambulator | 49 | 4 | 5 | | 71 | 13 | 0 | |

AHC$^+$, spina bifida aperta with hydrocephalus; AHC$^-$, spina bifida aperta without hydrocephalus; Occ, spina bifida occulta; HLL, high-level lesion (L2 and above); MLL, middle-level lesion (L3 to L5); LLL, low-level lesion (S1 and below). [a]Numbers of observations: AHC$^+$, range 106 to 119; AHC$^-$, range 20 to 23; Occ, range 35 to 37; HLL, range 64 to 73; MLL, range 63 to 68; LLL, range 35 to 38. *Significant differences between three subgroups with Pearson's $\chi^2 p<0.05$. **Significant differences between three subgroups with Pearson's $\chi^2 p<0.01$.

physical examination. Problems with sitting balance were present only in the AHC$^+$ group and mainly in the HLL group. Contractures in hips and knees were most common in the AHC$^+$ group and the HLL group. Most patients with AHC$^+$ and almost half of the patients with AHC$^-$ and spina bifida occulta had foot deformities. These were present in most patients with HLL and MLL and in only one-fifth of the patients with LLL. All (but two) patients with spina bifida occulta were normal or community ambulators. In the AHC$^+$ group one-half of the patients were non-ambulators and about one-third were normal or community ambulators. Level of lesion was related to ambulation, with patients with HLL mainly being non-ambulators and patients with LLL mainly being normal ambulators. Walking devices were often used: orthopaedic shoes in 41% of patients, orthosis in 21%, and crutches, sticks, or a walking frame in 16%.

## Bladder and bowel management

Most patients (60%) used clean intermittent self-catheterization for bladder management, and only 10 patients were dependent on others for clean intermittent catheterization. Nine patients used Credé's method to urinate. Twenty-four patients had

**TABLE V: PERCENTAGES OF PATIENTS WITH SPINA BIFIDA AND URINARY AND FAECAL INCONTINENCE, RELATED TO TYPE OF SPINA BIFIDA AND LEVEL OF LESION**

| Condition | $AHC^+$ (n=119) | $AHC^-$ (n=23) | Occ (n=37) | | HLL (n=73) | MLL (n=68) | LLL (n=38) | |
|---|---|---|---|---|---|---|---|---|
| **Urinary incontinence, %** | 71 | 52 | 35 | ** | 70 | 68 | 32 | ** |
| **Experiencing urinary incontinence as problem, %** | 50 | 44 | 41 | | 50 | 59 | 24 | ** |
| **Fecal incontinence , %** | 46 | 13 | 8 | ** | 45 | 34 | 13 | ** |
| **Experiencing fecal incontinence as problem, %** | 47 | 30 | 14 | ** | 47 | 40 | 18 | * |
| **Obstipation, %** | 45 | 30 | 35 | | 53 | 34 | 30 | * |
| **Use of napkin, %** | 76 | 52 | 30 | ** | 78 | 68 | 30 | ** |

$AHC^+$, spina bifida aperta with hydrocephalus; $AHC^-$, spina bifida aperta without hydrocephalus; Occ, spina bifida occulta; HLL, high-level lesion (L2 and above); MLL, middle-level lesion (L3 to L5); LLL, low-level lesion (S1 and below). *Significant differences between three subgroups with Pearson's $\chi^2 p<0.05$. **Significant differences between three subgroups with Pearson's $\chi^2 p<0.01$.

a continent urostoma, two patients a suprapubic catheter, and one patient a urethral catheter.

Accidents of urine leakage never happened in one-quarter of the total group, 13% had accidents less than once a month, 24% had monthly accidents, 11% had weekly accidents, and a 26% of the total group had daily accidents. Napkins were used by two-thirds of patients. Almost one-half of all patients experienced urinary incontinence as a problem. About onethird of the patients with spina bifida occulta were incontinent (having accidents of urine spoilage more than once a month), whereas one-half of the $AHC^-$ group and almost threequarters of patients with $AHC^+$ did. Urinary incontinence was found twice as frequently in the HLL and MLL group than in the LLL group (Table V).

Many patients used medication for their bladder: 24% used oral bladder-spasmolytics, and 5% used intravesical oxybutynin. Almost one-third of patients used antibiotics at low doses continuously. In the 2 years before the study almost twothirds of all patients needed a course of antibiotics for urinary tract infections: 36% once or twice, 50% three to five times, and 14% more than five times.

**TABLE VI: PERCENTAGES OF PATIENTS WITH SPINA BIFIDA WHO ARE SEXUALLY ACTIVE AND HAVE PROBLEMS WITH SEXUAL FUNCTIONING, RELATED TO TYPE OF SPINA BIFIDA AND LEVEL OF LESION**

| Condition | AHC$^+$ (n=116) | AHC$^-$ (n=23) | Occ (n=37) | | HLL (n=71) | MLL (n=67) | LLL (n=38) | |
|---|---|---|---|---|---|---|---|---|
| **Males sexually active, %** | 60 | 93 | 100 | ** | 46 | 86 | 94 | ** |
| **Females sexually active, %** | 46 | 89 | 88 | ** | 44 | 66 | 82 | ** |
| **Sexually active men** | (n=28) | n=13) | (n=12) | | (n=13) | (n=25) | (n=15) | |
| Problems with erections, % | 29 | 8 | 0 | | 39 | 16 | 0 | * |
| Problems with ejaculations, % | 39 | 15 | 0 | * | 54 | 24 | 0 | * |
| Problems with orgasm, % | 43 | 8 | 0 | ** | 54 | 20 | 7 | * |
| **Sexually active women** | (n=32) | (n=8) | (n=22) | | (n=19) | (n=25) | (n=18) | |
| Problems with lubrication, % | 6 | 0 | 14 | | 5 | 12 | 6 | |
| Problems with orgasm, % | 47 | 38 | 23 | | 47 | 36 | 28 | |

AHC$^+$, spina bifida aperta with hydrocephalus; AHC$^-$, spina bifida aperta without hydrocephalus; Occ, spina bifida occulta; HLL, high-level lesion (L2 and above); MLL, middle-level lesion (L3 to L5); LLL, low-level lesion (S1 and below). *Significant differences between three subgroups with Pearson's $\chi^2 p<0.05$. **Significant differences between three subgroups with Pearson's $\chi^2 p<0.01$.

For bowel management 27% of patients made use of bowel lavage and 17% took oral laxatives. For 15% of patients, faeces were removed manually and only 6% used rectal-stimulating laxatives and 2% were on a special diet. Percentages of patients suffering from faecal incontinence were lower than percentages suffering from urinary incontinence in all groups. The highest percentage was found in the AHC$^+$ group and the HLL group. Differences were significant for both subdivisions.

## Sexual functioning

Sixty-five percent of patients (73% male, 60% female) indicated that they had been sexually active at some time (including French kissing and masturbating). Further data on sexual functioning were gathered only on those 115 sexually-active patients.

In Table VI the percentages of sexually-active males and females and their perceived sexual problems are described. The AHC$^+$ group and the HLL group contained significantly fewer sexually-active patients than the other groups. For males with AHC$^+$ and patients with HLL who were sexually active, significantly more problems with sexual functioning were described than for the other male subgroups. Male patients with spina bifida occulta did not report problems concerning these sexual functions at all. For female patients no significant difference was found between subgroups with regard to sexual functioning.

**TABLE VII: PERCENTAGES OF SPINA BIFIDA PATIENTS WITH OTHER HEALTH PROBLEMS, RELATED TO TYPE OF SPINA BIFIDA AND LEVEL OF LESION**

| Health problem | AHC$^+$ (n=112) [a] | AHC$^-$ (n=20) | Occ (n=37) | HLL (n=69) [a] | MLL (n=65) | LLL (n=35) | |
|---|---|---|---|---|---|---|---|
| **Pressure sores, %** | 18 | 5 | 14 | 19 | 20 | 0 | * |
| **Hypertension, %** | 3 | 5 | 3 | 3 | 3 | 3 | |

AHC$^+$, spina bifida aperta with hydrocephalus; AHC$^-$, spina bifida aperta without hydrocephalus; Occ, spina bifida occulta; HLL, high-level lesion (L2 and above); MLL, middle-level lesion (L3 to L5); LLL, low-level lesion (S1 and below). [a]Numbers of observations: AHC+, range 111 to 112; HLL, range 68 to 69; *Significant differences between three subgroups with Pearson's $\chi^2 p<0.05$.

### Other health problems

Pressure sores of stage two or more, meaning at least blisters, were present in 15% of the total group. Pressure sores on the buttocks were present only in patients with hydrocephalus. Furthermore, pressure sores were on the feet. No pressure sores were found at the time of the physical examination in patients with sacral level of lesion (Table VII). Deep wounds were present in one spina bifida occulta patient and three patients with AHC$^+$. About one-quarter of patients had suffered from pressure sores in the previous year. Prevention of pressure sores was actively performed by 64% of patients, mainly patients with AHC$^+$. Most important methods used were wearing orthopaedic shoes, using a special cushion for the wheelchair, daily skin inspection, and regularly changing their sitting position.

Hypertension was present in only five patients (3%), with no significant difference between subgroups.

## DISCUSSION

This study describes the complexity of the secondary impairments that young adults with spina bifida experience. A broad group of patients with spina bifida was studied. Therefore, we were able to study differences between subgroups. Nevertheless, a few limitations might limit the generalization of the results.

This study concerns a Dutch population. The Dutch population might differ from the population in other countries because of differences in medical care and cultural requirements. We tried to include all patients suffering from spina bifida in the Netherlands. Although one spina bifida team did not participate in this study, published data suggest that the patients of that team are comparable to those included in this study

(Staal et al. 1996). Almost one-half of patients did not take part in this study, which might have influenced the external validity. However, no significant difference was found for age, sex, level of lesion, hydrocephalus or type of spina bifida between participants and non-participants. We also tried to include patients with the most serious disabilities by approaching special housing facilities. As far as the patients with spina bifida occulta are concerned, we could inevitably only trace patients who were already known with spina bifida, as is true of other studies concerning that group (Satar et al. 1995).

The great diversity in earlier studies, in terms of age, type of spina bifida, definitions, and cut-off points, makes it difficult to compare their outcomes and the results of this study.

Most patients had an extensive history of neurosurgery, orthopaedic surgery, and urological surgery; the highest rates of those who had surgery were in the $AHC^+$ and HLL groups. Most patients with $AHC^+$ had undergone several shunt revisions, which is comparable to earlier studies (Hunt 1990, Steinbok et al. 1992, Bier et al. 1997, Bowman et al. 2001).

Our study confirms the results of Hunt and Poulton (1995) that IQ is related to hydrocephalus. The percentage of patients with IQ below 70 was comparable to that in earlier studies (Hunt and Poulton 1995, Bowman et al. 2001). Epilepsy was a problem requiring treatment in less than one-tenth of the total group, which is also comparable to earlier studies (Farley et al. 1994, McDonnell and McCann 2000). Pain was a common problem and was present in one-quarter to onethird of patients. It was not correlated with the level of lesion or the type of spina bifida. This should be a point for consideration in the care for these patients.

The percentage of scoliosis found for the patients with spina bifida aperta is comparable to that in an earlier study (Steinbok et al. 1992). Patients with $AHC^+$ and patients with HLL both have a significantly higher risk of developing spinal deformities such as scoliosis and lordosis. Almost all patients with $AHC^-$, spina bifida occulta, or patients with LLL were walkers, and more than one-half of the patients with $AHC^+$ and three-quarters of patients with HLL were wheelchair dependent. This is comparable to published figures (Steinbok et al. 1992, Staal et al. 1996, McDonnell and McCann 2000, Bowman et al. 2001).

Incontinence was a problem for most of our patients. Even one-third of patients with spina bifida occulta and one-third of patients with LLL suffered from incontinence, mostly from urinary incontinence. Because of problems of definition, comparison with data from the literature is difficult. It is a matter for concern that almost one-third of the patients used low-dose prophylactic antibiotics: from the literature it is known that antibiotics are not beneficial for the long-term prevention of urinary tract infections and that this might

lead to antibiotic-resistant urinary tract infections (Trautner and Darouiche 2002). This warrants further study into treatment requirements for this group.

Most patients indicated that they had been sexually active at some time, which is comparable to the population of Dutch school-going teenagers aged 16 years (Brugman et al. 1995). This is something that physicians should be aware of when consulting those patients. Results on sexual functioning showed that problems with erection, ejaculation, and orgasm in male patients were common in our patient group, mainly in the $AHC^{+}$ or HLL group. Problems with orgasm were common in female patients. Similar percentages were found in the literature for this population (Sandler et al. 1996, Sawyer and Roberts 1999). A detailed analysis of sexual problems and sex education in our patient group will be published elsewhere.

Pressure sores were present in 15% of patients, which is a lower percentage than found in most literature (Hagelsteen et al. 1989, Exner et al. 1993, Brinker et al. 1994, Hunt and Poulton 1995). This might be due to the cross-sectional design of this study, in which only pressure sores present at the time of the physical examination were counted. Farley et al. (1994) reported a similar percentage (18%) of patients with a pressure sore at the time of the interview.

Summarizing our results, in many domains the $AHC^{+}$group and HLL group showed significantly more problems than other groups. The $AHC^{-}$ group bore a strong resemblance to the spina bifida occulta group in most conditions. Most research is done on patients with spina bifida aperta, but this study shows that patients with spina bifida occulta also suffer from significant secondary health impairments. Further research on this spina bifida occulta group is therefore suggested.

Finally, a few other aspects of this study need attention. Assessing the level of lesion is complicated. Because of the irregularity of the defect in spina bifida, it is known that sensory loss and motor loss are not always equal and, moreover, differences between the right and left sides of the body are common in patients with spina bifida. This complicates the definition of level of lesion. Reviewing the literature, many different ways of assessing the level of lesion, different points in time, and different cut-off points for level of lesion are used, which makes it difficult to compare data (Shurtleff and Sousa 1977, Hunt 1990, Steinbok et al. 1992, Swank and Dias 1992, Brinker et al. 1994, Hunt and Poulton 1995, Staal et al. 1996, McDonnell and McCann 2000, Bowman et al. 2001).

In our multicentre study we found that information in medical records on the level of lesion was unreliable because lesion levels were determined at different ages, sometimes using the motor level and sometimes using the sensory level. At times there was no

description of the method at all. In our study the sensory level for assessing the level of lesion in accordance with the international standards for neurological and functional classification of spinal cord injury was used because it could be performed in a more standardized way and took little time (Ditunno et al. 1994, Maynard et al. 1997). Motor-level assessment proved difficult in some patients because of other medical problems such as arthrodeses and contractures. Using a standardized assessment of level of lesion at an early age is recommended, to enable prognostic research in the future. From school age the sensory level can be tested in a standardized way and should be documented regularly. Using similar cut-off points in research is also important to permit the comparison of data. Because of the problems with subdivision on the level of lesion we also used a different classification based on the presence of hydrocephalus. The results of our study show that it is easy to form subgroups based on these characteristics and it is important to separate patients with spina bifida aperta into a group with and without hydrocephalus.

Our study showed that secondary impairments are present in young adult patients with different kinds of spina bifida. Lifelong care for patients with spina bifida is therefore necessary. Secondary impairments are present in different domains, which makes multidisciplinary care recommended. Little is known about the effect of aging on patients with spina bifida, which makes it even more important to keep a review patients' progress. Further research on aging patients with spina bifida is recommended, to optimize the care for this group.

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# Chapter 3

## COGNITIVE STATUS OF YOUNG ADULTS WITH SPINA BIFIDA

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## ABSTRACT

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

## INTRODUCTION

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

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

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

paediatricians with a better understanding of the results of treatment that has been given to them.

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

## METHOD

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

### Participants

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

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

Ten of the 181 individuals who were willing to participate were only interviewed by telephone; they were not included in the present study. Two participants with comorbidity that could independently induce serious physical and/or cognitive

impairments were excluded: one had serious heart disease and one had a chromosome disorder. Participants with pathology associated with SB were not excluded from the analysis, apart from one participant who could not be examined neuropsychologically because of blindness which was acquired after having been shunted for HC early in life. Data for 168 participants were included for the analysis.

## Materials

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

### *General intelligence*

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

### *Memory and verbal learning*

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

The Verbal Learning Test (VLT; Mulder et al. 1996) is a recent Dutch version of the California Verbal Learning Test (Delis et al. 1987). Participants are orally presented with a 'shopping list' (list A) consisting of 16 common nouns drawn from four semantic categories (fruits, clothing, tools, and spices). The items are presented five times (trials) in identical order. In each trial participants recall as many nouns as they can think of. After list A, a second (interfering) list is presented, of which the participant is again asked to recall as many nouns as possible. This is followed by a free and cued recall of list A. In the latter, the participant is provided with the semantic categories of list A. After 20 minutes filled with distracting tests, free and cued recall of list A is again determined. Finally, the

participant is asked to identify the 16 nouns of list A among 28 distracting nouns. Thanks to elaborate psychometric studies in the Dutch population (Mulder et al. 1996), learning indices can be compared with population normative values. The indices are corrected for age and sex and normative scores are calculated. The normative scores (z scores multiplied by two), have a mean of 0 (SD 2). For the purpose of this study, we inspected the normative scores and selected the three most informative learning parameters: Total Recall of list A (the total number of recalled items across five trials), Learning slope (the rate of learning of additional items across five trials), and Consolidation (the ability to remember learnt items over a longer period of time).

*Executive functioning*

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

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

Word production according to lexical rules (UNKA test; Jennekens-Schinkel et al. 1990) addresses the interface of memory, language, and behavioural regulation. The participant is asked to generate as many words as they can think of that begin with the prescribed letters (U, N, K, A). Production time is 60 seconds per letter. The dependent variable is the total number of correct words. Data on the performance of the typically developing population is available (A Jennekens- Schinkel, personal communication 2001).

*Reaction time*

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

## Data collection

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

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

participants the missing values were substituted by the mean score of the total sample on that particular test.

## Data analysis

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

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

For case finding, the clinical significance of poor performances was defined by setting a cut-off point and considering scores from 2SD worse than the population mean of the variable as 'deviation scores' in the IQ, MQ, VLT, UNKA tests. In the absence of suitable population data, the reference value was assigned to the group with SB occulta for the TMT-B, TMT-(B-A), and reaction speed measures. For the Wisconsin Modified Card Sorting Test, the clinically significant cut-off point of five categories was used because failing to find all six categories is pathognomonic for impairment. Between the three clinical groups, group differences in the proportions of participants who obtained deviation scores were compared using the $\chi^2$ test. Furthermore, we summed the deviation scores to a 'total cognitive deviation' score, in order to explore the range (0 to 6) across cognitive domains. For analysis of the effect of number of drain revisions and of associated pathology on the cognitive status within the group of persons with SB aperta with HC ($AHC^+$), we used the cognitive measure that correlated most strongly with all other cognitive measures, i.e. IQ. Statistical analyses were performed using the independent samples t-test.

**TABLE I: DEMOGRAPHIC AND NEUROLOGICAL CHARACTERISTICS OF THE STUDY SAMPLE**

| | SB Occulta N = 37 N (%) | AHC- N = 20 N (%) | AHC+ N = 111 N (%) |
|---|---|---|---|
| **Gender** | | | |
| female | 25 (68%) | 7 (35%) | 69 (62%) |
| male | 12 (32%) | 13 (65%) | 42 (38%) |
| **Age**, mean (SD) | 20.6 (3.2) | 21.0 (3.1) | 20.7 (2.9) |
| **Hydrocephalus** | | | |
| shunted | 0 - | 0 - | 111 (100%) |
| compensated | 1 (3%) | 5 (25%) | 0 - |
| no | 36 (97%) | 15 (75%) | 0 - |
| **Level of Lesion** | | | |
| ≥ L2 | 5 (14%) | 2 (10%) | 62 (56%) |
| L3 – L5 | 14 (38%) | 7 (35%) | 44 (40%) |
| ≤ S1 | 18 (49%) | 11 (55%) | 5 (5%) |
| **Shunt Revisions** | | | |
| no shunt or no revision | 37 (100%) | 20 (100%) | 16 (14%) |
| 1 | - | - | 25 (23%) |
| 2 - 4 | - | - | 35 (32%) |
| more than 5 | - | - | 35 (32%) |
| **Corpus Callosum** | | | |
| malformation | 1 (3%) | 1 (5%) | 7 (6%) |
| **Bleeding/Ischaemia** | | | |
| yes | - | - | 1 (1%) |
| **Encephalitis** | | | |
| yes | 2 (5%) | - | 10 (9%) |
| **Epilepsy** | | | |
| yes | 2 (5%) | - | 11 (10%) |
| **Total number of surgical interventions**, mean (SD) | 4.9 (3.4) | 8.3 (7.8) | 8.7 (5.9) |
| **Ambulation** | | | |
| wheelchair dependent | 2 (5%) | 3 (15%) | 75 (68%) |
| **Continence** | | | |
| incontinence | 14 (38%) | 11 (55%) | 90 (81%) |
| **Living conditions** | | | |
| with parents | 21 (57%) | 11 (55%) | 54 (49%) |
| sheltered home | 1 (3%) | 2 (10%) | 42 (38%) |
| independently alone | 7 (19%) | 7 (35%) | 12 (11%) |
| independently together | 8 (22%) | - | 3 (3%) |
| **Vocational status** | | | |
| unemployed | 3 (8%) | 2 (10%) | 25 (23%) |
| student ordinary education | 21 (57%) | 11 (55%) | 32 (29%) |
| student special education | 2 (5%) | 2 (10%) | 23 (21%) |
| paid normal work | 11 (30%) | 5 (25%) | 18 (16%) |
| sheltered work | - | - | 13 (12%) |

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

**TABLE II: MEAN SCORE AND 95% CI* ON COGNITIVE TESTS FOR PARTICIPANTS WITH SPINA BIFIDA (SB) OCCULTA, APERTA WITHOUT (AHC-) AND APERTA WITH HYDROCEPHALUS (AHC+)**

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

* CI = confidence interval; [a] significant difference between Occ and AHC+, p<0.0167; [b] significant difference between AHC- and AHC+, p<0.0167; N, number; SPM, Standard Progressive Matrices; WMS, Wechsler Memory Scale; VLT, Verbal Learning Test; WmCST, Wisconsin modified Card Sorting Test; TMT, Trail Making Test; UNKA, word production according to lexical rules; RT, reaction times; IQ, intelligence quotient; MQ, memory quotient; sec, seconds; msec, miliseconds.

## RESULTS

Demographic and neurological characteristics of the groups of persons with SB occulta, SB aperta without HC (AHC$^-$), and SB aperta with HC (AHC$^+$) are summarized in Table I. Sixty percent of the participants were female. The AHC$^-$ had a significantly smaller

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

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

The majority of deviation scores (Table III, Fig. 1) were obtained by The AHC$^+$ participants: about one-fifth had an IQ or MQ below 70. Between one-third and one-half of AHC$^+$participants had a deviation score on the VLT. Almost half failed on the Wisconsin Modified Card Sorting Test. A quarter deviated on TMT-B; when corrected for completion time of part A, this percentage dropped to 12%. Only 7% had a deviation score on the UNKA. As far as reaction speed was concerned, deviating decision times were found in 44% of The AHC$^+$ participants and deviating motor times in one quarter of

**TABLE III: NUMBERS (%) OF DEVIATION SCORES FOUND IN GROUPS OF YOUNG ADULTS WITH SPINA BIFIDA (SB) OCCULTA, SB APERTA WITHOUT (AHC-) AND WITH HYDROCEPHALUS (AHC+)**

| | | SB Occulta | AHC- | AHC+ | |
|---|---|---|---|---|---|
| **Raven SPM** | IQ | 1 (3%) | - (0%) | 22 (20%) | [a] |
| **WMS** | MQ | - (0%) | - (0%) | 16 (14%) | [a] |
| **VLT** | list A total recall | 2 (9%) | - (0%) | 22 (41%) | [a] |
| | learning slope | 1 (4%) | - (0%) | 21 (39%) | [a] |
| | consolidation | 4 (17%) | - (0%) | 12 (22%) | [a] |
| **WmCST** | total categories | - (0%) | 3 (15%) | 50 (45%) | [b] |
| **TMT** | time part B | 2 (5%) | - (0%) | 26 (23%) | [c] |
| | difference B-A | 3 (8%) | - (0%) | 13 (12%) | [c] |
| **UNKA** | total number of words | - (0%) | - (0%) | 10 (9%) | [a] |
| **RT** | mean decision time | - (0%) | 3 (25%) | 34 (57%) | [c] |
| | mean motor time | - (0%) | 3 (25%) | 15 (25%) | [c] |
| **Total cognition deviation score** | number of deviation scores | | | | |
| | 0 | 27 (73%) | 14 (70%) | 24 (22%) | |
| | 1 | 9 (24%) | 6 (30%) | 38 (34%) | |
| | 2 | 1 (3%) | - (0%) | 21 (19%) | |
| | 3 | - (0%) | - (0%) | 12 (11%) | |
| | 4 | - (0%) | - (0%) | 10 (9%) | |
| | 5 | - (0%) | - (0%) | 4 (4%) | |
| | 6 | - (0%) | - (0%) | 2 (2%) | |

[a] 2*SD below population mean; [b] below maximum score; [c] 2*SD below mean Occulta; SPM, Standard Progressive Matrices; WMS, Wechsler Memory Scale; VLT, Verbal Learning Test; WmCST, Wisconsin modified Card Sorting Test; TMT, Trail Making Test; UNKA, word production according to lexical rules; RT, reaction times; IQ, intelligence quotient; MQ, memory quotient

AHC$^+$ participants. Participants of both other groups obtained considerably fewer deviation scores. Four participants with SB occulta had deviation scores in consolidation (VLT). Among those with AHC$^-$, three of 20 participants had a deviation score on the Wisconsin Modified Card Sorting Test and three of 12 had a deviation score on both decision time and motor time of the reaction speed task. When exploring generality versus specificity of the deviations, six persons with AHC$^-$ had one deviation score and

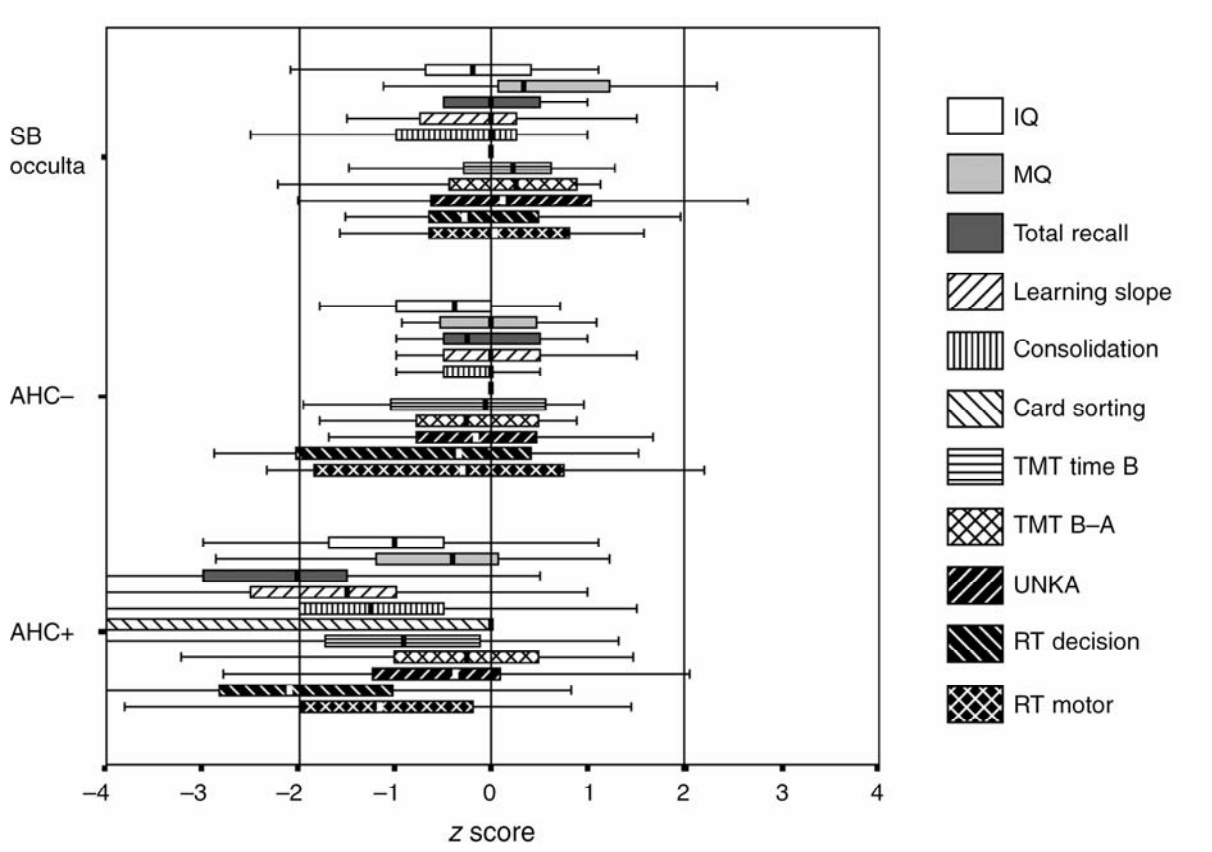


**Figure 1: Cognitive status in relation to type of spina bifida and hydrocephalus.**
For purpose of graphical display by means of boxplots, all scores were converted to z scores. Hinges mark 25th and 75th centiles and length of box corresponds with interquartile range. Whiskers show range of values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

none had 2 or more deviation scores. Similar to persons with $AHC^-$, of the persons with SB occulta, 24% had one or more deviation scores and one person had 2 deviation scores. In contrast, 79% of the persons with $AHC^+$ had one or more deviation scores, 44% had 2 or more deviation scores, and 15% had a more general deviation with 4 or more deviation scores.

IQ significantly correlated with all but one of the other cognitive scores. Pearson's r ranged from 0.21 to 0.73. The majority of the correlations (7 of 10) were higher than 0.40. IQ was found to be the most robust measure of general cognitive status. Within the group with $AHC^+$, mean IQ decreased with increasing number of shunt revisions: 0 or 1

revision (mean IQ=86), 2 to 4 revisions (mean IQ=86), and 5 or more revisions (mean IQ=77). Post-hoc analysis showed the difference between the groups with no or one shunt revision and with five or more shunt revisions to be significant ($p<0.0167$).

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

## Discussion

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

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

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

As mentioned earlier, the group with $AHC^-$ performed similarly to individuals with SB occulta on the cognitive tests. Few deviation scores were found and none of the persons

with AHC$^-$ had 2 or more deviation scores. Together with Friedrich et al. (1991), we conclude that those with AHC$^-$ do not have serious cognitive problems. Considering the small number of participants in this subgroup, further research is warranted.

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

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

Three features of the present study have to be mentioned. Every effort was made to approach all those with SB, including those with SB occulta, and to examine every individual we were able to trace. Non-response analysis showed that the participant group was similar to the non-participant group on important demographic and illness characteristics. Therefore, we feel that our findings can be generalized over the population of young adults with SB in the Netherlands. Of course, only those patients were included who were diagnosed with SB occulta at birth or later on in life when the defect caused physical complaints. There certainly are many persons with undiscovered SB occulta. This selection bias will probably make little difference to cognitive outcome, in which we found the full breadth of normality in this group.

The second feature to be mentioned is that we did not exclude patients with an IQ below 70. We insisted on including these participants for the reason that learning disability (US

usage: mental retardation) is a possible outcome of the neural tube defect. Excluding participants with learning disability would possibly lead to a too optimistic view of cognitive status.

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

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

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

## Conclusion

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

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

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# Chapter 4

## EDUCATIONAL CAREER AND PREDICTORS OF TYPE OF EDUCATION IN YOUNG ADULTS WITH SPINA BIFIDA

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## ABSTRACT

Children with spina bifida (SB) often require special education. To date, little information is available about the educational career of these children. This study focuses on educational career and predictors of attending special education of young adults with SB, using a cross-sectional study including 178 young Dutch adults with SB aged from 16–25. The main outcome was attending regular versus special education. For searching predictive power we selected age, gender, type of SB, level of lesion, hydrocephalus (HC), number of surgical interventions, ambulation, continence and cognitive functioning. Chi-square tests and binary logistic regression were used in the data analysis. Participants with HC attended special primary education more often (59%) than participants without HC (17%). For those participants with HC, the necessity of special primary education was associated with below average intelligence (75% versus 35%), wheelchair dependence (82% versus 39%) and surgical interventions (74% versus 44%). Only half of the participants with HC followed regular secondary education, whereas for participants with SB without HC, the outcome in secondary education was similar to that of the general population (92%). Intelligence was the main predictor of attending special secondary education (odds 5.1:1), but HC (odds 4.3:1) and wheelchair dependence (odds 2.6:1) were also a significant. Other variables were not significant predictors of special secondary education.

## INTRODUCTION

Survival rate of children with spina bifida (SB) continues to increase because of the use of improved shunting procedures and better renal management (Lorber, 1971; McDonnell and McCann, 2000; Bowman et al., 2001). A challenge for rehabilitation is to support these children, given their individual abilities, towards adult life and successful integration into society. Education plays an important role in this process.

In The Netherlands, in cases of children who need extra help, the aim is to provide individual support that is tailored to the needs of the child (OC&W, 2001). As in most other Western countries, this can be in the form of attending an ordinary school with individual support. But for children for whom extra help is not sufficient because of physical disabilities, mental disabilities and/or behavioural problems, schools for special education are available. In addition to the general aim of providing regular education, teaching in special schools is geared towards enabling as many pupils as possible to change to a regular primary or secondary school (OC&W, 2001). About 5% of all Dutch children attend a special primary school (age 4–12) and about 11% require special secondary education (age 12 and above). Of this population of children in secondary special education roughly, one-third are children from cultural minorities who have recently arrived in The Netherlands and who have not yet a sufficient command of the Dutch language (OC&W, 2002).

Physical and/or cognitive limitations are common in children with SB (Steinbok et al., 1992; Baron & Goldberger, 1993; Wills, 1993; Hunt & Poulton, 1995). Many SB children with or without hydrocephalus (HC) require special education, the number being roughly estimated at 50% (Tew & Laurence, 1975; Laurence & Beresford, 1976; Hunt, 1981; Kokkonen et al., 1991; Wasson et al., 1992; Staal-Schreinemachers et al., 1996; Bowman et al., 2001) However, little is known about the educational career of these children and about the predictors of educational outcome (Wasson et al., 1992). From the few studies on the subject it would seem that mental and physical impairments as well as school attendance play a role, but these studies suffer from small sample size and the fact that the composition of the samples spans several decades in which schooling might have changed considerable (Laurence & Beresford, 1976; Carr et al., 1981; Wasson et al., 1992; Hurley & Bell, 1994).

On reaching adulthood, part of the population with SB shows poor long-term achievement (Hunt et al., 1999). Better insight into educational career and its predictors might provide a basis for improving the support system for children with SB. This paper

focuses on the following research questions: (1) to what extent are young adults with SB able to follow regular education throughout the course of their educational career and (2) what are the predictors of attending special education?

## METHODS

This study is part of the ASPINE project, a crosssectional multi-centre study of physical and cognitive abilities, health care, social participation and life satisfaction of Adolescents with Spina bifida In the Netherlands. The ethics and research committees of the participating institutions have approved ASPINE.

### Participants

Participants were young adults who had spina bifida (SB) aperta or occulta, within the age range 16 to 25 years and with a sufficient command of the Dutch language. Two participants with co-morbidity that could independently induce physical and/or mental impairments were excluded: one had a cardiac affection and the other had a chromosome-13 disorder. Participants were recruited by 11 of the 12 Dutch Spina Bifida Teams, in co-ordination with the Dutch Spina Bifida Patients Association, organizations for sheltered homes and rehabilitation centres. The written invitations to participate were sent by the controlling physicians or the management of the sheltered home to 350 young adults. Of these, 178 (51%) participated in this study, written informed consent being obtained from them or from their parents if applicable. Basic patient characteristics were collected from medical records to facilitate a non-response analysis. There were no significant differences at an alpha level of 5% between the response group and the non-response group with regard to age, gender, type of SB, level of lesion, or being shunted for hydrocephalus (HC). Of the 178 participants, nine were interviewed by telephone.

### Variables

Medical records were examined according to a fixed protocol and participants underwent a physical and neuropsychological examination and a semi-structured interview.

Demographic variables taken into account were age (years) and gender. Data on neurological variables were also gathered with respect to type of spina bifida (occulta or aperta), level of lesion (defined as the lowest completely unimpaired level on both sides measured with sensitivity to pin prick and light touch) and hydrocephalus (defined as

having been shunted early in life to reduce intracranial pressure). Participants were grouped in level of lesion L2 or above, L3–L5 or S1 and lower. For the purpose of logistic regression analysis the variable was dichotomised into two variables; one with a cut-off point of L2 and above and the other with a cut-off point of S1 and below.

As educational outcome may be affected by school absenteeism, as medical interventions are usually accompanied by school absenteeism and as the number of medical interventions varies widely in children with SB, we calculated the mean number of surgical interventions per life year to summarize medical history of all surgeries of any kind. For the purpose of analysis, this variable was dichotomised into a 'many surgical interventions' group and a 'few surgical interventions' group, using the median value of the study group as the cut-off point.

Physical disabilities measured were ambulation and continence. Ambulation was assessed using an adaptation of the scale of Hoffer et al. (1973), in which people can be classified into five groups. For the purpose of this study we dichotomised the variable and classified normal and community ambulators as 'walkers', and the other categories as 'wheelchair-dependent'. Incontinence was defined as having a faecal and/or urinary accident at least once a month.

Cognitive functioning was assessed using the Standard Progressive Matrices (SPM) (Raven, 1996; Raven et al., 1998) The SPM is a multiplechoice test requiring accuracy of discrimination and evaluation of logical relations in visual displays. We used a twenty-minute time limit for the results, as recent Dutch norm scores are available for comparison (Bouma et al., 1996). The dependent variable was the number of adequate solutions, corrected for age and gender, and converted to the intelligence quotient (IQ). Mean IQscore of the healthy population is 100 (standard deviation [SD]=15). For the purpose of this study a cut-off point of 1 SD was taken, resulting in a group with an average IQ (IQ>85) and a group with a below average IQ (IQ≤85).

Participants were questioned about their educational career; the type of education (a) at the start of primary school (age approximately four years), (b) at the end of primary school, (c) at the start of secondary school (age approximately 12 years) and (d) when leaving secondary school were recorded. Each change in type of education during primary or secondary school was also recorded. If a participant had not yet completed his/her education, the type of education being currently followed at the time of the interview was taken as the educational outcome. The criterion for having a certain type of educational outcome was obtainment of a certificate or completion of at least 75% of that type of education.

### Data analysis

Statistical analysis was performed using SPSS 10.0.7 for Windows. All analyses comparing proportions between groups were done using the Chi-square test. Conditional backward binary logistic regression was used to explore predictors of educational outcome. Associations between predictors were expressed using Kendall's Tau. Alpha was set at 0.05.

## RESULTS

### Demographic and neurological characteristics

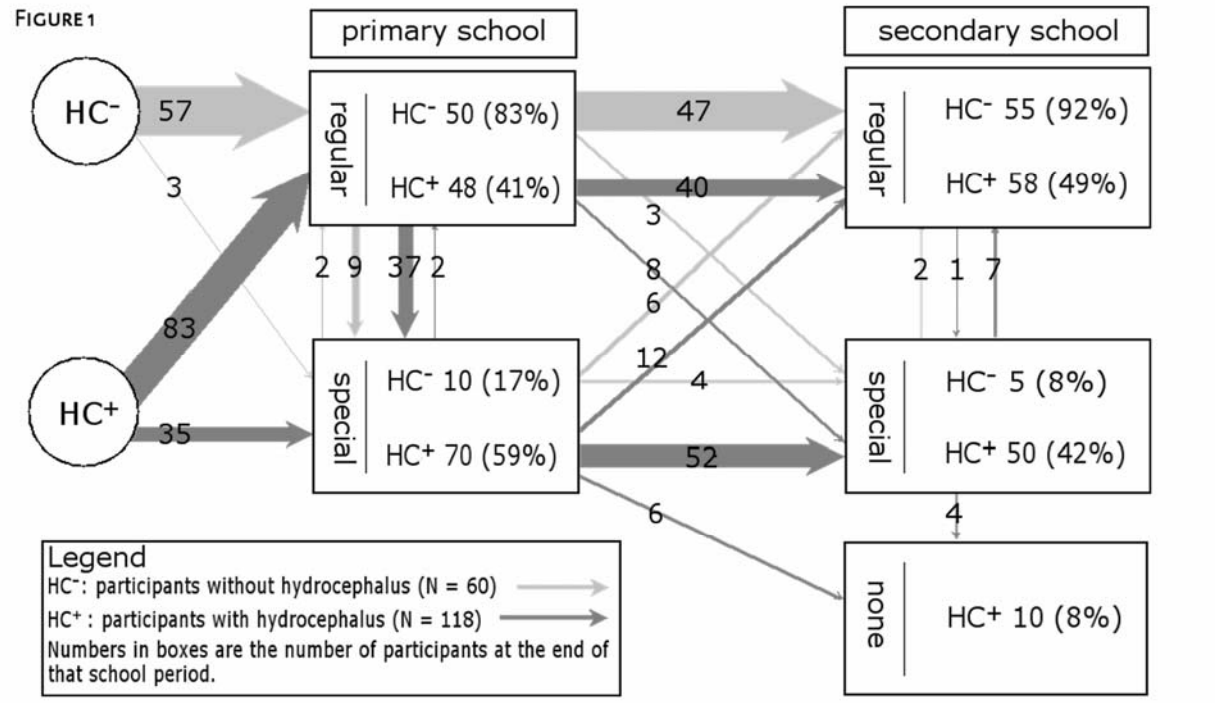
Demographic and neurological characteristics of the participants are summarized in Table 1. All participants with hydrocephalus (HC) had spina bifida (SB) aperta. There were no significant differences in age or gender between the groups of participants with and

**TABLE 1: CHARACTERISTICS OF PARTICIPANTS (N = 178)**

| Characteristics | | HC- | HC+ | Total |
|---|---|---|---|---|
| **Gender** | Female | 34 (57%) | 71 (60%) | 105 (59%) |
| | Male | 26 (43%) | 47 (40%) | 73 (41%) |
| **Age (years)** | Mean (SD) | 20.8 (3.12) | 20.7 (2.85) | 20.7 (2.93) |
| **Type of spina bifida *** | Aperta | 23 (38%) | 118 (100%) | 141 (79%) |
| | Occulta | 37 (62%) | | 37 (21%) |
| **Level of lesion *** | L2 or higher | 7 (12%) | 66 (56%) | 73 (41%) |
| | L3 – L5 | 21 (35%) | 46 (39%) | 67 (38%) |
| | S1 or lower | 32 (53%) | 6 (5%) | 38 (21%) |
| **Ambulation *** | (Community) walker | 55 (92%) | 37 (31%) | 92 (52%) |
| | Wheelchair dependent | 5 (8%) | 81 (67%) | 86 (48%) |
| **Continence *** | Continent | 33 (55%) | 23 (20%) | 56 (32%) |
| | Incontinent | 27 (45%) | 95 (81%) | 122 (69%) |
| **Surgical interventions per life year *** | Less than 0.33 | 50 (83%) | 39 (33%) | 89 (50%) |
| | More than 0.33 | 10 (17%) | 79 (67%) | 89 (50%) |
| **Intelligence (n=168) *** | IQ>85 | 46 (81%) | 51 (46%) | 97 (58%) |
| | IQ≥85 | 11 (19%) | 76 (54%) | 71 (42%) |

* Statistically significant difference between HC- and HC+, $p < 0.001$

FIGURE 1




without HC. The level of the lesion was significantly higher in participants with HC than in those without HC. A significantly larger proportion of participants with HC than without HC were wheelchair dependent (69% versus 8%), were incontinent (81% versus 45%), had undergone many surgical interventions (67% versus 17%) and had an IQ below 85 (54% versus 19%).

## Educational career

Educational career is represented in Figure 1. Almost all participants without HC and two-thirds of participants with HC had started off in regular primary education. The participants who had started their educational career in special education were more seriously disabled than those who had started in regular education: more of them had a below average IQ (75% compared to 35%), were wheelchair dependent (82% compared to 39%) and had undergone many surgical interventions (74% compared to 44%). During the course of primary education 31% of the participants with HC and 15% of those without who had started off in regular education had changed to special education.

Subsequent analyses on the subgroups of participants with HC showed: (1) participants who, at some stage in primary school, had changed from regular to special education, were more seriously disabled than those who had continued in regular school, as exemplified by wheelchair dependence (70% compared to 57%) and below average IQ

(62% compared to 33%). Significantly more participants who had changed the type of education were female (81% compared to 57%). (2) Comparing the group of participants who in the past had changed from regular to special primary schools with those who had started off in special primary schools, the former group was found to be less disabled, as exemplified by wheelchair dependence (70% compared to 82%) and below average IQ (62% compared to 75%). Again, the group who shifted contained more girls than the group of participants who had started off in special primary schools (81% compared to 42%). Age, continence and surgical interventions per life year did not influence transfer from regular to special school during primary education.

A significantly smaller proportion of participants with HC (41%) had completed regular primary education than those without HC (83%). Most participants (80%) had continued the same type of education when moving from primary to secondary school (Figure 1), but in both the group of participants with and in that without HC, participants had moved from regular to special education or vice versa. Six participants with HC had not embarked on secondary education. During secondary education a few participants (6%) had changed from regular to special education or vice versa. A further four participants had dropped out of special education prematurely. Of those who had completed regular primary school, 90% also completed regular secondary education or were still in regular secondary school at the time of the study. However, of those who had completed special primary education, 31% changed to regular secondary education. Overall, significantly more participants without HC (92%) had completed regular secondary education or were still in regular education at the time of the study than participants with HC (49%).

**Predictors of education outcome**

The groups of participants who had completed special secondary education or were attending special education at the time of the study were more disabled than the group of participants in regular education (Table 2). A significantly higher proportion had a below average IQ, was wheelchair dependent and was incontinent. Also, more participants in special education had undergone many surgical interventions per life year. Pathology was also worse for the group of participants in special education. A significantly larger proportion of participants with SB aperta went to special education, more participants had HC and/or higher levels of lesion. No differences between the two groups were found with respect to age and gender.

For further analysis of the predictor variables affecting type of education, conditional logistical regression using a backward elimination model was performed (Table 3).

**TABLE 2: PREDICTORS OF TYPE OF SECONDARY EDUCATION**

| | | Regular (n=113) | Special (n=65) |
|---|---|---|---|
| **Gender** | Female | 69 (61%) | 36 (55%) |
| **Age** | Older than 21 years | 55 (49%) | 30 (46%) |
| **Type of spina bifida** | Aperta | 78 (69%) | 63 (97%)** |
| **Shunted for hydrocephalus** | Yes | 58 (51%) | 60 (92%)** |
| **Level of lesion** | L2 and higher | 33 (29%) | 40 (62%)** |
| **Level of lesion** | S1 and lower | 33 (29%) | 5 (8%)** |
| **Intelligence** | IQ below average | 28 (26%) | 43 (71%)** |
| **Ambulation** | Wheelchair dependent | 37 (33%) | 49 (75%)** |
| **Continence** | Incontinent | 70 (62%) | 52 (80%)* |
| **Surgical interventions per life year** | More than 0.33 | 45 (40%) | 44 (68%)** |

* p < 0.01 ; ** p < 0.001

Associations between the predictor variables used in this regression analysis, being type of spina bifida, level of lesion, hydrocephalus, ambulation, continence, surgical interventions per life year and intelligence were all significant, Kendall's Tau ranged from 0.196 to 0.718. Age and gender were not significantly correlated to each other or to any other variable.

The analysis started using all predictor variables. Subsequently the following variables were dropped from the model: number of surgical interventions, continence, level of

**TABLE 3: BINARY LOGISTIC REGRESSION ON REGULAR VERSUS SPECIAL SECONDARY EDUCATION**

| | B | Wald | Odds ratio | 95% CI | |
|---|---|---|---|---|---|
| | | | | Lower | Upper |
| **Gender** | | 2.175 | | | |
| **Age** | | 0.768 | | | |
| **Type of spina bifida** | | 1.010 | | | |
| **Shunted for hydrocephalus** | 1.467 | 6.561* | 4.335 | 1.411 | 13.315 |
| **Level of lesion L2 and higher** | | 0.584 | | | |
| **Low level of lesion S1 and lower** | | 0.407 | | | |
| **IQ** | 1.634 | 17.701** | 5.126 | 2.394 | 10.977 |
| **Ambulation** | 0.963 | 5.166* | 2.620 | 1.142 | 6.013 |
| **Continence** | | 0.339 | | | |
| **Surgical interventions per life year** | | 0.449 | | | |

* p < 0.05 ; ** p < 0.001; B, Co-efficient; CI, Confidence interval; Wald, Wald statistic.

lesion S1 and lower, age, type of spina bifida, level of lesion L2 and higher and finally gender. Three significant predictors remained, the strongest predictor being IQ ($p<0.001$), showing that participants with a below average IQ had an odds of 5.1:1 for having attended special secondary education. Having HC ($p=0.010$) had an odds of 4.3:1 for having attended special secondary education. Being wheelchair dependent ($p=0.023$) had an odds of 2.6:1 for having attended special secondary education. The overall percentage of correctly classified participants was 77.4%. Further analysis on ambulation showed that ambulation was a predictor of type of education, not only in the groups of participants with a below average IQ, but also in the group of participants with an average IQ.

## Discussion

This study addresses educational outcome of young adults with spina bifida (SB). Educational career was described using the well-known landmarks of the start of primary education, the transition point from primary to secondary education and the highest received education/ training. Thereafter, possible predictors of attending special secondary education were analysed.

Three findings on school career pertain to the period of primary education. Firstly, this study shows that participants with SB and hydrocephalus (HC) more often started off in special primary education or were transferred to special primary education than participants with SB without HC, thus supporting earlier findings on educational outcome (Tew and Laurence, 1975; Kokkonen et al., 1991). Secondly, even for participants without HC, the proportion of those in special primary education (17%) was higher than that of the general population (5%), but at the end of secondary education the proportion was similar at 8% (OC&W, 2002). Thirdly, a large number of participants with HC (mostly female) started off in regular primary school but had to switch to special education during the course of primary school. These participants were less ambulant and had lower intelligence than those who where able to remain in regular education. These data on educational transfer have not previously been reported. It appears that the demands of regular education were too high for these participants during childhood. This is in line with a recent Dutch study on children with mental disabilities which reports that the problems of integration into regular education start about the third year at school (age six), because at that stage the children are required to learn to read and to work more independently and, along the way, these children cannot keep up with the pace and level

demanded (Poulisse, 2002). It might be that the physical limitations increased and that the school was no longer suited to the needs of the children (Hunt, 1981). Parental preference for regular education might have further increased the number of children with SB starting off in regular primary education (Lauder et al., 1979). Also, in The Netherlands it is not uncommon to advise the parents of disabled/handicapped children to start the education in regular preschool and kindergarten. When the child is older and educational demands increase, he/ she can at that point be transferred to a school more adapted to his/her special individual needs.

Of the participants who had completed regular primary school, 90% continued their school career in regular secondary schools. For participants without HC the outcome of secondary education was similar to that of the general population, whereas only half of the participants with HC managed regular secondary education. Nonetheless, it is encouraging that about one-third of the participants who had followed special primary education somehow found their way into regular secondary education. Perhaps these children suffered delayed development (Adams, 1969; Wills et al., 1990; Barnes et al., 2002) and at later age were able to cope with the demands of ordinary education, or maybe they missed too many of the first years of primary education due to surgical interventions to be able to stay in ordinary primary school. A more detailed study is required to reveal the mechanism behind this finding.

Not surprisingly, and similar to previous reports (Wasson et al., 1992), below average IQ was a strong predictor for a career in special education. However, in the present study, and contradictory to the findings of Wasson (1992), being wheelchair-bound was independently associated with attending special education. In principle, wheelchair dependence should not be a cause of leaving regular education for those with normal intelligence and this is a matter of concern. However, wheelchair dependence was associated with both incontinence and surgical interventions. The association between these variables might be the reason why in the bivariate analysis these predictors were significantly related to the type of education, whereas in the logistic regression analysis only wheelchair dependence was a significant predictor. Wheelchair dependence is known to be associated with pain and fatigue and these may also be the cause of real limitations in educational career (Vogels, 2002).

Of the neurological variables, only HC proved to be a predictor of type of education. Again, colinearity between pathology and the associated impairment might have obscured the predictive value of type of spina bifida and level of lesion. For a child with spina bifida occulta or a child with a level of lesion of S1 or lower, the chance for a need to

go to special education is minimal. For a child with hydrocephalus and a level of lesion above L3 this chance is nearly 60%.

Four features of the present study should be mentioned. Every effort was made to approach all SB persons, including those with spina bifida occulta, and to examine every patient whom we were able to trace. Non-response analysis showed that the participating group was comparable to the non-response group on important demographic and illness characteristics. Hence, the findings can be generalized with regard to the population of young adults with SB in The Netherlands. Of course, within the group of participants without hydrocephalus, the only patients included were those diagnosed with SB occulta at birth or later on in life, mostly because the defect caused physical complaints. There are certainly many individuals with an undiscovered SB occulta. This selection bias will probably make little difference to the educational outcome in the group of participants without HC, which we found to be similar to the general population.

The second feature to be mentioned is that young adults with an IQ below 70 were not excluded. We insisted on including these participants for the reason that mental retardation is a possible outcome of the neural tube defect. Excluding mentally retarded participants would possibly lead to a too optimistic view of educational outcome.

Thirdly, only IQ was used as an indicator of cognitive status. It can be argued that even with an average IQ a person may still suffer from specific cognitive disorders, which may hamper their educational career and this may be especially true for patients with SB and HC. However, it has been repeatedly shown that IQ is a strong predictor of educational outcome (Raven et al., 1983; Sternberg et al., 2001) and we showed elsewhere that IQ was strongly related to specific cognitive disorders (Barf et al., 2003).

Finally, at the time of the study, 63% of the participants without HC and 50% of the participants with HC had not yet completed their education. It can be argued that those participants who had not yet completed their schooling might still change the type of education. However, based on data on the general population (OC&W, 2002) and on our own data of the participants who had completed their education, it seems very unlikely that a significant number of participants will later transfer to another type of education.

## CONCLUSION

Young adults with SB accompanied by HC are more likely to follow special education and to have a lower educational outcome than those without HC, whereas young adults with SB without HC have an educational outcome similar to their healthy peers. Intelligence

level was the main predictor of educational outcome, but HC and wheelchair dependence were also significant factors. Carers of children with SB should be aware of the fact that wheelchair dependence and associated physical and social problems may restrict children in their educational career. Their challenge is to help create conditions for attending regular education to ensure optimal opportunities in later life.

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# Chapter 5

## RESTRICTIONS IN SOCIAL PARTICIPATION OF YOUNG ADULTS WITH SPINA BIFIDA

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## ABSTRACT

Purpose: To determine participation restrictions of young adults with spina bifida (SB) in relation to health condition and activity limitations.

Method: A total of 179 persons aged 16 to 25 years and born with SB participated in a cross-sectional study. The main outcome on four domains of participation (independent living, employment, education and partner relationships) was assessed using a structured questionnaire.

Results: At the mean age of 21 years only 16% were living independently, more than one-third of the participants went to special secondary education, 53% of those who finished education did not have a regular job and 71% did not have a partner. Health condition variables (type of SB, hydrocephalus and level of lesion) and to a lesser extent activity limitations (wheelchair dependence and incontinence) were significant determinants for having participation restrictions. Perceived hindrances in participation included long-distance transportation (19%-36%), accessibility (10%-42%), physical impairments (22%-40%), emotional barriers (20%-32%) and financial limits (3%-17%). More severe SB, defined as hydrocephalus, high level of lesion and wheelchair dependence, was related with more experienced hindrances due to long-distance transportation accessibility of buildings.

Conclusions: Many young adults with spina bifida experience participation restrictions. Severity of SB was negatively related to participation. Social integration should be a major focus in the professional guidance of youngsters with physical disabilities.

## INTRODUCTION

Spina bifida (SB) is a health condition that is caused by a congenital neural tube disorder. Some 50 years ago the life expectancy of babies born with SB was poor.[1,2] The use of shunting for hydrocephalus and intermittent catheterization have hugely increased chances of survival and now life expectancy for babies born with SB is near to normal.[2-6]

A wide variety of physical impairments and delay of cognitive development are however common in persons born with SB.[3,4,7-13] The first few years of a child with SB are the most endangered. At school age a somewhat quieter period starts, the focus mainly lying on education. New problems arise in the period of transition from childhood to adolescence and adulthood. Challenges in this period include (in)dependent living, work, social relationships, partnership and child wish.

One earlier Dutch study of functioning of young adults with SB aperta concluded that chances for living independently, attending regular education, and having a regular job was small and dependent on the level of neurological deficit.[14] International studies also show that unrestricted participation is difficult to reach for persons with SB. Level of education is lower than in the general population,[3,15,16] unemployment rate is high,[3,4,10,15-20] only few can live independently without extra help,[3,15,18] and persons with SB seem to have more difficulty finding a partner.[3,15] In order to improve the transition of youngsters with SB into adulthood and to adjust counseling by health care professionals to the needs of these youngsters, it is important to study determinants of social participation and hindrances for participation as perceived by young adults with spina bifida.

The aim of this study is to examine participation restrictions of a large group of young adults born with spina bifida in relation to disease characteristics, activity limitations and perceived hindrances for participation.

## METHODS

### Participants

The present study was part of the Aspine project.[7,11] Participants were persons with SB aperta or occulta (International Classification of Diseases, 9th revision codes 741 and 756.17 respectively)[21] who were between 16 and 25 years of age and who had sufficient command of the Dutch language. Participants with co-morbidity that could independently induce serious physical and/or mental impairments were excluded. Participants were recruited through 11 of the 12 Dutch Spina Bifida Teams in co-

ordination with the Dutch Spina Bifida Patients Association, organizations for sheltered homes, and rehabilitation centers. The ethics and research committees of the respective institutions approved the study. Written informed consent was obtained from the participants or their parents if applicable. Invitations were sent to 350 persons, of whom 179 participated in this study. Patient characteristics (age, gender, type of SB, level of lesion, presence of hydrocephalus), collected from medical records, were not significantly different (alpha = 0.05) between the participating and the non-participating group.

**Instruments**

Medical records were examined according a fixed protocol and participants underwent a physical and neuropsychological examination. The participants were interviewed between August 1999 and August 2001. Seven participants had the questionnaires filled in on their behalf by their parents.

*Demographic variables* were age and gender. *Health condition characteristics* included type of spina bifida (occulta or aperta) and the presence of hydrocephalus, which was defined as having been shunted early in life to reduce intracranial pressure. Both were retrieved from medical files. Further, level of lesion was defined as the lowest completely unimpaired dermatome level on both sides with sensitivity to pin prick and light touch and was assessed by physical examination.

*Activity limitations* were assessed during the physical examination. Ambulatory status was categorized using an adaptation of the Hoffer scale for ambulation.[22] Persons without walking problems and community ambulators were considered 'walkers' and persons using a wheelchair for shorter or longer distances were considered 'wheelchair dependent'. Incontinence was defined as having fecal and/or urinary accidents at least once a month and was dichotomized as continent or incontinent. As part of the neuropsychological examination intelligence was assessed using the Raven Standard Progressive Matrices[23] ,the number of correct answers was converted into an intelligence quotient (IQ) score with mean=100 and SD=15 in the population. An IQ below 85 was considered 'below average' and an IQ equal to or higher than 85 was considered 'at least average'.

Participants were interviewed about their *social participation*, in terms of their educational career, vocational status, marital status and residential status using structured questions. *Perceived problems in relation to social participation* were collected as part of a self-report questionnaire. Participants were asked to quantify the occurrence of five types of hindrances with respect to school or work, visiting family or friends and spending

leisure time: long distance transportation, accessibility of buildings, physical impairments, emotional distress and costs. Participants quantified the occurrence of problems as 'never', 'sometimes', 'regularly' or 'very often'. For the purpose of this study the last three quantifications were combined and the scores were dichotomized.

### Statistics

Descriptive statistics were used and differences in the proportions between subgroups were tested using the Chi-square test or the Fisher's exact test in case of two by two tables. All results were taken to be significant at a p-level below 0.05 two-sided. In case of multiple testing a Bonferroni correction was applied.

## RESULTS

The characteristics of the study group are shown in Table 1. Most participants were born with SB aperta and 84% of these participants were shunted because of hydrocephalus early in life. Mean age was 20.7 years (SD 2.9); half of the participants were 21 years of age or older. Moderate to strong correlations existed between type of SB, having hydrocephalus and level of lesion (Kendall's Tau 0.34 - 0.72; $p < 0.001$). These health conditions were also significantly related to wheelchair dependence, continence and IQ (Kendall's Tau 0.25 - 0.61; $p < 0.003$). The intercorrelations between wheelchair dependence, continence and IQ were weak (Kendall's Tau 0.17 - 0.27; $p < 0.03$).

### Mobility

As shown in Table 1, 39% of the participants were wheelchair user for household ambulation. An even larger proportion made use of an electric or self propelled wheelchair (47%) or other aids like a tricycle or hand-bike (17%) for longer-distance transportation. Of all participants aged 18 years or older, 26% had a driver's license. Participants who were wheelchair dependent had less often a driver's license (12%) than participants who were not wheelchair dependent (36%; $p = 0.002$) and depended more often on special transportation (89% against 32%; $p < 0.001$).

### Social participation

In Table 2 participation outcomes are summarized and related to demographic characteristics, health condition and activity limitations. Half of the participants were still living with their parents. Thirty-one percent were living in an institution, independent

**TABLE 1: CHARACTERISTICS OF PARTICIPANTS**

| | | spina bifida total | spina bifida without HC | spina bifida with HC |
|---|---|---|---|---|
| | N | 179 | 60 | 119 |
| **Gender** | Male | 41% | 43% | 40% |
| | Female | 59% | 57% | 60% |
| **Age** | 21 or older | 49% | 48% | 49% |
| | younger than 21 | 51% | 52% | 51% |
| **Spina bifida** | aperta | 79% | 38% | 100%* |
| | occulta | 21% | 62% | 0% |
| **Level of lesion** | L2 or higher | 41% | 12% | 55%* |
| | L3 – L5 | 38% | 35% | 39% |
| | S1 or lower | 21% | 53% | 5% |
| **Ambulation** | wheelchair dependent | 39% | 7% | 55%* |
| | independent | 61% | 93% | 45% |
| **Continence** | incontinent | 69% | 45% | 81%* |
| | continent | 31% | 55% | 19% |
| **Intelligence** | IQ below 85 | 39% | 19% | 49%* |
| | IQ 85 or higher | 61% | 81% | 51% |

HC = hydrocephalus; * = significant difference between participants with and without hydrocephalus; $p < 0.001$

living centre or sheltered home and 16% were living completely independent. More than one-thirds of all participants went to special secondary education or had no education beyond primary school and only 16% had a higher level of education. At the time of the study, half of the participants were still in full-time education. Of the 92 participants who finished their education, 47% had a regular job, 15% were working at a sheltered workplace and 38% were unemployed. Most participants (71%) did not have a partner at the time of the study. One-fifth had a steady relation with a boy or girl and 7% was married or living together.

**TABLE 2: SOCIAL PARTICIPATION OF YOUNG ADULTS WITH SB (N=179)**

| | | Independent living | | | Education (secondary) | | | Employment[a] | | Relationships | |
|---|---|---|---|---|---|---|---|---|---|---|---|
| | | parents | special care | independent | none or special | regular low and medium level | regular high level | unemployed or sheltered workplace | regular employment | no partner | partner |
| | N | 94 | 56 | 29 | 65 | 84 | 29 | 49 | 43 | 127 | 52 |
| | % | 53% | 31% | 16% | 37% | 47% | 16% | 53% | 47% | 71% | 29% |
| **Demographic** | | | | | | | | | | | |
| Age | => 21 | 37% | 36% | 28%** | 35% | 48% | 17% | 51% | 49% | 68% | 32% |
| | < 21 | 67% | 27% | 5% | 38% | 47% | 15% | 60% | 40% | 74% | 26% |
| Gender | male | 58% | 30% | 12% | 40% | 44% | 16% | 44% | 56% | 74% | 26% |
| | female | 49% | 32% | 19% | 34% | 50% | 16% | 59% | 41% | 69% | 31% |
| **Health condition** | | | | | | | | | | | |
| Spina bifida | aperta | 51% | 39% | 10%** | 45% | 45% | 10%** | 62% | 38%* | 76% | 24%* |
| | occulta | 57% | 3% | 41% | 5% | 54% | 41% | 13% | 88% | 51% | 49% |
| Hydrocephalus | with | 50% | 45% | 5%** | 51% | 42% | 8%** | 69% | 31%** | 80% | 20%** |
| | without | 57% | 5% | 38% | 8% | 58% | 33% | 18% | 82% | 53% | 47% |
| Lesion level | L2 or above | 48% | 45% | 7%**[b] | 55% | 41% | 4%** | 69% | 31% | 77% | 23% |
| | L3 – L5 | 50% | 31% | 19% | 30% | 49% | 21% | 47% | 53% | 75% | 25% |
| | S1 or lower | 66% | 5% | 29% | 13% | 55% | 32% | 29% | 71% | 53% | 47% |
| **Activity limitations** | | | | | | | | | | | |
| Wheelchair | dependent | 43% | 51% | 6%** | 58% | 41% | 1%** | 68% | 32% | 80% | 20% |
| | independent | 59% | 18% | 23% | 23% | 51% | 26% | 43% | 57% | 65% | 35% |
| Continence | incontinent | 50% | 37% | 12%*[b] | 43% | 47% | 11%* | 64% | 36%* | 77% | 23%* |
| | continent | 57% | 18% | 25% | 23% | 48% | 29% | 27% | 73% | 57% | 43% |
| Intelligence | low IQ | 51% | 35% | 14% | 65% | 35% | 0%** | 73% | 27%* | 74% | 26% |
| | average IQ | 43% | 41% | 15% | 17% | 57% | 25% | 35% | 65% | 70% | 30% |

* = significant difference at $p < 0.01$; ** = significant difference at $p < 0.001$ ; [a] = only participants who finished education (N = 92); [b] = only significant different for age 21+ at $p < 0.01$

### Demographic characteristics in relation to social participation

There were no differences in social participation between men and women and between younger and older participants, except for place of residence. Participants aged 21 or older were more often living independently (28%) than participants aged younger than 21 (5%; $p < 0.001$).

### Health condition in relation to social participation

Having SB aperta, having hydrocephalus and having a high level of lesion were all associated with more participation restrictions: higher frequency of special care living arrangements, special secondary education, not having a regular job and not having a partner (Table 2). Level of lesion seemed to be related to higher participation in employment and relationships, but these differences were not significant after correction for multiple testing. As correlates of participation might be age-specific, all analyses were also performed for both age groups (>21 and <21) separately. Only one difference occurred: the association between level of lesion and residence was not significant in the group of participants younger than 21 years. In general, relationships between health condition and participation variables were somewhat stronger in the group of participants older than 21 years than in the group younger than 21 years.

### Activity limitations in relation to social participation

Wheelchair dependence was related to type of residence and type of education, but not to employment or to having a partner. Being incontinent was related to restrictions in all participation domains. IQ was related to type of education and employment, but not to type of residence and having a partner. Corrected for age, the association between continence and residence was not significant for participants younger than 21 years. In general, associations between activity limitations and participation were less pronounced than associations between health condition and participation.

### Perceived hindrances in participation

A considerable number of participants reported hindrances in participation (see Table 3). Most hindrances were reported in the domain of leisure (range 17% - 42%), but problems were also reported for visiting family and friends (range 9% - 32%) and attending work or school (range 3% - 37%). Problems were most frequently related to physical impairments, followed by accessibility of buildings, long-distance transportation and emotional distress. Costs were not often mentioned as hindrance in participation.

**TABLE 3: PROPORTION OF YOUNG ADULTS WITH SB PERCEIVING HINDRANCES IN PARTICIPATION (N=179)**

| | work or education | visiting friends and family | leisure |
|---|---|---|---|
| **long distance transportation** | 19% [e] | 26% [c,d,e] | 36% [c,d,e] |
| **accessibility of building** | 10% [d] | 32% [b,c,d,e] | 42% [c,d,e] |
| **physical impairments** | 37% | 22% [a] | 40% |
| **emotional distress** | 20% | 20% | 32% |
| **costs** | 3% | 9% | 17% [d,e] |

Significance level for differences set at a corrected alpha of $p < 0.003$
[a] = significant difference for subgroups of age
[b] = significant difference for subgroups of type of SB
[c] = significant difference for subgroups of HC
[d] = significant difference for subgroups of level of lesions
[e] = significant difference for subgroups of wheelchair dependence

Problems with long-distance transportation and accessibility of buildings were more often reported by persons with hydrocephalus, with a high level of lesion and who were wheelchair dependent. Participants who were wheelchair dependent reported two to four times more often problems with long distance transportation and accessibility of buildings ($p<0.005$) and three times more often financial problems hindering leisure activities ($p<0.001$). Age, gender, type of SB, continence and low IQ were not consistently related to perceived hindrances in participation. Also, occurrence of problems due to own physical impairments, emotional distress or financial restraints were not consistently related to either health condition or activity limitations.

## DISCUSSION

This study addresses social participation and its determinants in young adults with spina bifida. The results indicate considerable participation restrictions. Healthy young adults leave their parents' homes at a mean age of 21 years,[24] whereas only 28% of the participants older than 21 years in this study were living independently. One-third of the young adults with spina bifida needed special education, against 5% of the general population.[25] The unemployment rate of 53% in this study was also higher than that of disabled persons between 15 and 64 years of age (28%)[26] and far higher than the 8% in the general population aged 15 to 24 years.[24] The domain of relationships however

seemed to be less affected by having spina bifida. Participation was related to type of spina bifida, having hydrocephalus, being wheelchair-dependent and being incontinent.
Participants reported most perceived hindrances in participation in the domain of leisure, but they also reported problems regarding going to school or work and visiting family or friends. Physical impairment was most often mentioned as hindrance in participation, but accessibility of buildings, long-distance transportation and own emotional barriers were also frequently mentioned to hinder participation.
The present results with respect to independent living, education and employment are similar to those of previous studies on social participation.[3,4,10,14-20] There was a clear relation between wheelchair dependence and perceived hindrances in participation in society, especially accessibility of buildings and long-distance transportation. This finding is not specific for young adults with spina bifida. A recent study in a large representative sample of persons with chronic illness or physical disabilities showed that one-thirds of persons with disabilities reported problems with steep slopes while entering (or leaving) public buildings, one-fifth reported problems with entering a bus or train, one-third reported that high thresholds and curbstones were reasons for them not to leave the house and 16% reported to experience problems with tight doorways. Furthermore, only 44% of those using special transportation were satisfied with this facility.[26]
Contrary to expectations, no relation was found between severity of spina bifida and perceived hindrances due to physical impairments or emotional distress. Apparently, young adults with a less serious spina bifida are, in their own perception, just as restricted in their participation due to pain, fatigue, incontinence and emotional stress (shame, sadness) as young adults with severe spina bifida. This finding confirms the results of Minchom et al., who refuted the assumption that spina bifida would have less psychological impact in mildly disabled young persons.[27] An explanation might be that participants with mild spina bifida more often have a socially active lifestyle or have higher expectations with respect in participation and consequently more acutely perceive hindrances than participants with severe spina bifida.
Some features of the present study should be mentioned. Every effort was made to approach all persons with spina bifida, including those with spina bifida occulta, and to examine every patient whom we were able to trace. Non-response analysis showed that the participating group was comparable to the non-response group on important demographic and illness characteristics. Hence, the findings may be generalized with regard to the population of young adults with spina bifida in The Netherlands. Unavoidably, the group of participants with spina bifida occulta included only patients

diagnosed at birth or later in life, mostly because the defect caused physical complaints. Most certain there are many persons with an undiscovered spina bifida occulta. Yet, this study showed that even persons with a less serious spina bifida experienced restrictions in participation.

The second feature to be mentioned is that young adults with an IQ below 70 were not excluded, unlike most other studies. We insisted on including these participants because mental retardation is a possible outcome of the neural tube defect and may lead to serious participation problems. Excluding mentally retarded participants would possibly lead to a too optimistic view of the outcome in terms of social participation.

Our study showed that social participation of persons born with spina bifida is far from optimal. We found that participation restrictions were related to severity of spina bifida and, although to a lesser extent, to activity limitations like wheelchair dependence and incontinence.

## IMPLICATIONS

Much more focus should be directed to reduce participation restrictions, especially with respect to leisure activities and mobility for both wheelchair dependent and non wheelchair dependent young adults with spina bifida. Course of actions must be undertaken like diminution of impact of physical impairments and abolish emotional distress. Leveling of barriers to social participation of wheelchair using persons with spina bifida (and, for that matter, other physically disabled persons) due to inaccessibility of buildings should become a priority of counselors and governmental authorities.

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# CHAPTER 6

## LIFE SATISFACTION OF DUTCH YOUNG ADULTS WITH SPINA BIFIDA

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## ABSTRACT

This study concerns life satisfaction and its determinants in Dutch young adults with spina bifida (SB). Data on life satisfaction (Life Satisfaction Questionnaire [LiSat-9]) were related to hydrocephalus, lesion level, disabilities, and demographic variables. In total, 179 young adults with SB participated (41% male, age range 16–25y; 79% SB aperta, 67% hydrocephalus [HC], 39% wheelchair-dependent). Most were satisfied with their life as a whole (24% dissatisfied). No difference was found from a population reference group (28% dissatisfied). Highest proportions of dissatisfaction were found for financial situation (44%), partnership relations (49%), and sex life (55%). Least dissatisfaction was found for contact with friends (17%) and families (15%). Young adults with SB and HC were more satisfied with their financial situation and family life but were less satisfied with self-care ability and partnership relations than those without HC and the reference group. However, except for self-care ability, relationships between life satisfaction and having SB were weak. In conclusion, self-care ability and partnership relations were rated least favourable and may need more attention from care providers. Overall, SB does not seem to be an important determinant of life satisfaction.

## INTRODUCTION

Spina bifida (SB) is a congenital condition that may result in a wide variety of physical and cognitive limitations.[1–7] After the initial hazardous years, the survival rate for a person with SB is near normal,[8,9] although hydrocephalus (HC), bladder management, and kidney-sparing always remain an issue.[7,9] Several authors have proposed that chronic conditions such as SB confront youngsters with additional barriers in the transition from adolescence to adulthood compared with their typically developing peers.[10–12] They may find themselves, for example, disadvantaged with regard to career opportunities or finding a partner. Better insight into the quality of life (QoL) of young adults with SB might identify support needs and might, thereby, provide a basis for improving the support system. Knowledge of the QoL of persons with SB is also relevant with regard to today's policies of aborting pregnancies early in gestation, the use of new operation techniques such as in utero closure of the neural tube,[13] and deciding whether or not to treat early after birth. Such decisions by medical professionals are based on, among other considerations, the expected long-term QoL of the patients involved. However, professionals tend to rate the QoL of their patients much lower than the patients themselves do.[14] Only limited research into the QoL of persons born with SB is available, and the results are equivocal. Some authors[15] found that the healthrelated QoL of their study population was below normal, whereas others concluded that QoL was good[10] and comparable to that of persons without SB.[11,16]

QoL is a notion with a rather broad meaning, but it is usually associated with well-being or life satisfaction.[17] Defining life satisfaction as an element of QoL, rather than equating it to QoL, is an effective approach.[18] This study addressed life satisfaction, the more subjective perceptions of the quality of one's own existence,[19] and focuses on the following research questions: (1) what is the life satisfaction of young adults born with SB?; (2) how does this compare with the life satisfaction of the general population?; and (3) does having SB affect life satisfaction?

## METHOD

### Participants

Participants were people who had SB aperta or occulta[20] who were within the age range 16 to 25 years, and who had sufficient command of the Dutch language. Participants with comorbidity that could independently induce serious physical and/or cognitive

impairments were excluded. Participants were recruited by 11 of the 12 Dutch Spina Bifida Teams, in coordination with the Dutch Spina Bifida Patients Association, organizations for sheltered homes, and rehabilitation centres. The ethics and research committees of the participating institutions approved the study. Written informed consent was obtained from the participants, or their parents if applicable. Patient characteristics (age, sex, type of SB, level of lesion, presence of HC) were collected from medical records to facilitate a non-response analysis. Invitations were sent to 350 persons, of whom 179 participated in this study. There was no significant difference ($\alpha$=0.05) between the response group and the nonresponse group with regard to age, sex, type of SB, level of lesion, or having a shunt for HC. Participants were interviewed orally and filled in questionnaires as part of a larger project focusing on the physical and cognitive disabilities of young adults with SB.[5,7] Participants were interviewed between August 1999 and August 2001. Seven participants had the questionnaires filled in on their behalf by their parents.

## Population

Data on the general population were taken from Post et al.[21] For this study a random sample of 1200 persons between 18 and 65 years of age was drawn from the municipal register of the city of Utrecht. These persons received a questionnaire by mail, which was completed and returned by 507 persons. From this reference group all 132 persons between 18 and 25 years of age were selected.

## Instruments

The Life Satisfaction Questionnaire (LiSat-9)[22,23] contains one question about satisfaction with life as a whole, and eight questions about domain-specific life satisfaction (shown in Table II). Each question was answered on a 6-point scale (1=very dissatisfied, up to 6=very satisfied). As described by Fugl-Meyer et al., persons with scores of 1 to 4 were classified as 'dissatisfied' and persons with scores of 5 and 6 as 'satisfied'. [22,23] The Dutch LiSat-9 has previously been used and showed satisfactory internal consistency reliability (Cronbach's alpha 0.74–0.85).[24,25]

Demographic variables taken into account were age and sex. Disease characteristics taken into account were type of SB (occulta or aperta), the level of the lesion (defined as the lowest completely unimpaired level on both sides with sensitivity to pin prick and light touch), and the presence of HC (defined as having been shunted early in life to reduce

intracranial pressure). Ambulatory status (Hoffer scale[26]), incontinence, and intelligence (Raven standard progressive matrix[27]) were determined.

### Statistics

Descriptive statistics were used and differences in the percentage of satisfied persons were tested with the $\chi^2$ test or Fisher's exact test for 2×2 tables. This was followed by two series of logistic regression analyses: first, to check the impact of lesion level and having HC on life satisfaction, and second, to analyze differences in life satisfaction between persons with SB and the reference group, controlled for age and sex. Collinearity between type of SB, HC, and level of lesion on the one hand, and self-care ability, wheelchair dependence, incontinence, and intelligence on the other, unfortunately prohibited the use of all these variables in the same regression analysis. Therefore, only the disease characteristics were used. In addition to the odds ratios and their confidence intervals, Nagelkerke $R^2$ are reported as a proxy for percentage-explained variance. All results are taken to be significant at $p<0.05$ two-sided. For multiple testing, a Bonferroni correction was applied.

## RESULTS

Table I shows characteristics of the study group. Most participants had SB aperta and 84% of this group developed HC early in life, requiring a shunt. Participants with HC more often had a higher level of lesion than participants without HC. Mean age did not differ across the three groups. Except for the group of participants with SB aperta without HC, more females than males participated. Participants without HC showed fewer disabilities and better social integration than participants with HC.

Because of the minimum age of 18 years, the reference group comprised a smaller proportion of persons younger than 21 years than the group of persons with SB (Table I). The proportion of female participants was similar in both groups. In terms of intelligence, marital status, and vocational status, the groups of persons with SB but without HC were similar to the reference group.

### Life satisfaction of persons with SB

In Table II, the proportions of dissatisfied persons (LiSat-9 item score below 5) are displayed. Of all persons with SB, 21% were dissatisfied with their life as a whole. The highest proportions of dissatisfaction were found in the domains of financial situation

TABLE I: DESCRIPTIVE CHARACTERISTICS OF PARTICIPANTS AND POPULATION

| | | SB occulta | SB aperta without HC | SB aperta with HC | reference group |
|---|---|---|---|---|---|
| **N** | | 37 | 23 | 119 | 132 |
| **Age** | % 15-20 | 46 | 48 | 45 | 22[b] |
| | % 21-26 | 54 | 52 | 55 | 78 |
| **Gender** | % male | 68 | 39 | 60 | 68 |
| **Level of lesion** | % L2 or higher | 14 | 9 | 56[a] | - |
| | % L3 – L5 | 38 | 30 | 40 | - |
| | % S1 or lower | 49 | 61 | 5 | - |
| **Ambulation** | % wheelchair dependent | 5 | 9 | 56[a] | - |
| **Continence** | % incontinent | 38 | 57 | 81[a] | - |
| **Intelligence** | % below average IQ (<85) | 16 | 25 | 54[a] | 16 |
| **Living status** | % with parent | 57 | 57 | 50[a] | - |
| | % sheltered home | 3 | 9 | 45 | - |
| | % independently | 41 | 35 | 5 | - |
| **Marital status** | % married / living together | 22 | 4 | 3 | 18 |
| | % partner but not living together | 27 | 39 | 15 | 47 |
| | % no relationship | 51 | 57 | 82[a] | 35 |
| **Vocational status** | % unemployed | 8 | 9 | 26[a] | 2 |
| | % student | 60 | 65 | 49 | 64 |
| | % paid work | 32 | 26 | 25 | 32 |
| | % housekeeping | 0 | 0 | 0 | 2 |

[a] significant difference ($p < 0.05$) between the group of persons with SB with HC and the merged group of persons SB without HC; [b] significant difference ($p < 0.05$) between the reference group and the total group of persons with SB; SB, spina bifida; HC, shunted for hydrocephalus; IQ, intelligence quotient

(44%), partnership relations (49%), and sex life (55%). The lowest proportion of dissatisfaction was found for contact with friends (17%) and families (15%).

To analyze differences between groups of persons with different types of SB, first the groups of persons with SB occulta and those with SB aperta without HC were compared. There was no difference in life satisfaction between these two groups on any item. We,

TABLE II: PROPORTIONS OF DISSATISFIED PERSONS (%) WITH SPINA BIFIDA (SB) AND IN THE POPULATION

| | SB total (N=179) | SB occulta (N=32) | SB aperta without HC (N=23) | SB aperta with HC (N=119) | reference group (N=132) |
|---|---|---|---|---|---|
| **Life as a whole** | 21 | 14 | 17 | 24 | 28 |
| **Self care ability** | 22 | 14 | 0 | 28 [a] | 5 [b] |
| **Leisure situation** | 25 | 16 | 4 | 32 [a] | 25 |
| **Vocational / educational situation** | 31 | 29 | 30 | 32 | 38 |
| **Financial situation** | 44 | 60 | 50 | 38 [a] | 66 [b] |
| **Sex life** | 55 | 46 | 41 | 61 [a] | 45 [b] |
| **Partnership relations** | 49 | 35 | 41 | 56 [a] | 34 [b] |
| **Family life** | 15 | 19 | 22 | 12 | 32 [b] |
| **Contacts friend and acquaintances** | 17 | 8 | 9 | 21 [a] | 17 |

[a] significant difference between persons with HC and persons without HC; $p \leq 0{,}003$; [b] significant difference between persons with SB and the reference group; $p \leq 0{,}01$; Dissatisfaction was defined as a score below 5 on each LiSat-9 item; HC, shunted for hydrocephalus

therefore, merged these two groups into one group as participants without HC. Comparing the persons with and without HC, persons with HC were more often dissatisfied with their self-care ability (p=0.002) and leisure situation (p=0.003).

### SB versus reference group

Of the reference group, 28% were dissatisfied with life as a whole (Table II). The highest proportions of dissatisfaction were found with respect to financial situation (66%) and sex life (45%), whereas the lowest proportions of dissatisfaction were found for contacts with friends (17%) and self-care ability. When compared with the reference group, persons with SB were more often dissatisfied with their self-care ability (p<0.000) and partnership relations (p=0.008). However, they were less often dissatisfied with their financial situation (p<0.000) and family life (p=0.001) than persons in the reference group. These differences can mostly be attributed to the effect of HC. The group of persons with HC differed significantly from the reference group on all four items (p<0.001), whereas no significant difference was found between the group of persons without HC and the reference group.

**TABLE III: DETERMINANTS OF LIFE SATISFACTION FOR PARTICIPANTS WITH SPINA BIFIDA**

| Determinant | Life as a whole | Self-care ability | Leisure situation | Vocational/ educational | Financial situation | Sex life | Partnership relations | Family life | Contacts friends |
|---|---|---|---|---|---|---|---|---|---|
| **Gender** | 1.10 (0.52-2.33) | 0.44 (0.19-1.03) | 1.07 (0.52-2.20) | 0.77 (0.39-1.53) | 1.53 (0.82-2.85) | **3.05[b] (1.53-6.10)** | 1.93 (0.99-3.74) | 0.71 (0.29-1.73) | **0.34[a] (0.13-0.89)** |
| **Age** | 1.34 (0.63-2.84) | 1.15 (0.52-2.54) | 0.80 (0.39-1.62) | 0.96 (0.49-1.88) | 0.99 (0.53-1.83) | 1.41 (0.73-2.75) | 0.98 (0.51-1.87) | 0.80 (0.34-1.86) | 0.82 (0.35-1.88) |
| **Hydrocephalus** | 1.28 (0.47-3.50) | 1.68 (0.53-5.29) | 2.15 (0.78-5.92) | 0.53 (0.22-1.28) | **0.45[a] (0.20-0.99)** | 1.11 (0.49-2.55) | 2.01 (0.87-4.64) | 0.71 (0.24-2.10) | **4.70[a] (1.25-17.63)** |
| **High lesion** | 1.73 (0.75-4.00) | **3.97[b] (1.65-9.53)** | 1.39 (0.65-2.97) | **2.22[a] (1.03-4.81)** | 1.13 (0.56-2.31) | **2.84[b] (1.31-6.15)** | 1.25 (0.59-2.63) | 1.18 (0.41-3.37) | 0.85 (0.34-2.18) |
| **Low lesion** | 1.28 (0.37-4.47) | 4.49 (0.50-40.15) | 2.55 (0.62-10.52) | 2.27 (0.75-6.83) | 1.09 (0.43-2.75) | 1.64 (0.62-4.35) | 0.94 (0.35-2.49) | 0.51 (0.15-1.66) | 0.43 (0.11-1.72) |
| **Nagelkerke $R^2$** | *4.4%* | *25.4%* | *10.7%* | *7.7%* | *5.2%* | *18.3%* | *7.4%* | *3.9%* | *11.7%* |

Results are shown as odds ratios (95% confidence interval) unless otherwise defined. Condition per determinant: sex (male); age (21y or older); high lesion (L2 or higher); low lesion (L5 or higher). Results in bold are significant at the following levels: [a]p<0.05; [b]p<0.001

**TABLE IV: ASSOCIATIONS BETWEEN HAVING SPINA BIFIDA, AGE AND GENDER WITH LIFE SATISFACTION**

| Determinant | Life as a whole | Self-care ability | Leisure situation | Vocational/ educational | Financial situation | Sex life | Partnership relations | Family life | Contacts friends |
|---|---|---|---|---|---|---|---|---|---|
| **Gender** | 0.88 (0.51-1.54) | 0.52 (0.25-1.07) | 0.98 (0.57-1.68) | 1.25 (0.76-2.05) | 1.61 (0.99-2.62) | **1.69[a] (1.04-2.73)** | 1.63 (0.99-2.68) | 1.07 (0.59-1.93) | **0.40[a] (0.2-0.83)** |
| **Age** | 1.45 (0.80-2.61) | 1.07 (0.54-2.12) | 0.76 (0.44-1.31) | 1.15 (0.68-1.94) | 1.11 (0.67-1.81) | 1.30 (0.78-2.17) | 0.82 (0.49-1.37) | 0.81 (0.44-1.50) | 0.84 (0.44-1.59) |
| **Spina Bifida** | 0.76 (0.44-1.30) | **6.27[b] (2.52-15.61)** | 0.94 (0.54-1.61) | 0.74 (0.45-1.21) | **0.40[b] (0.24-0.65)** | 1.57 (0.96-2.55) | **1.76[a] (1.06-2.91)** | **0.35[b] (0.19-0.63)** | 0.96 (0.52-1.80) |
| **Nagelkerke $R^2$** | *1.8%* | *13.2%* | *0.5%* | *1.2%* | *7.7%* | *3.9%* | *5.3%* | *6.5%* | *3.9%* |

Results are shown as odds ratios (95% confidence interval) unless otherwise defined. Condition per determinant: sex (male); age (21y or older). Results in bold are significant at the following levels: [a]p<0.05; [b]p<0.001

### Determinants of life satisfaction of persons with SB

The impact of demographic and disease characteristics on life satisfaction of persons with SB is shown in Table III. No significant determinant was found between these variables and satisfaction with life as a whole, leisure situation, partnership relations, and family life. A high level of lesion was a single significant determinant for dissatisfaction with self-care ability and vocational/educational situation. HC was a single significant determinant of satisfaction with financial situation. Both sex (males) and high level of lesion predicted dissatisfaction with sex life. Finally, dissatisfaction concerning contact with friends was predicted by sex (females) and HC.

Apart from self-care ability (25.4%) and to some extent sex life (18.3%) only a minimal amount of variance was explained by the determinants used.

A series of logistic regression analyses were performed to explore the effect of having SB on life satisfaction (Table IV). Age was not a significant determinant of life satisfaction in any logistic regression analysis. Sex (male) was a significant determinant for dissatisfaction with sex life. Sex (female) was also a significant determinant for contact with friends. Group membership (SB versus reference group) was a significant determinant for dissatisfaction with self-care ability and partnership relations as well as a determinant for satisfaction with financial situation and family contact. However, except for self-care ability (13.2%), the amount of variance explained by age, sex, and having SB was again very small.

## Discussion

One of the major findings of this study is that life satisfaction of young adults born with SB is more or less similar to that of their typically developing peers, and that severity of SB has only a minor impact on life satisfaction. This finding confirms the results of most of the previous smaller studies, using similar or different conceptualizations of QoL. Query et al.[10] concluded that, despite reports of physical and emotional stress, most children and adolescents with HC and SB judged the quality of their lives and family relationships to be good. Similarly, Lindstrom and Kohler[11] found only small differences in QoL between adolescents with myelomeningocoele and typically developing youths. However, self-esteem was lower in the group of persons with myelomeningocoele. Sawin et al.[16] found a moderately high QoL in families with adolescents with SB. In addition, they found no relationship between QoL and shunt status, level of lesion, and severity of the SB. Padua et al.[28] concluded from their study on adolescents with SB that there is no

linear inverse correlation between disability and QoL in patients with SB and that patients with mild disabilities needed as much psychological support as patients with severe disabilities.

The finding that persons with severe disabilities are more satisfied with their lives than others might expect, is known from groups of patients with different aetiologies. For example, Bach et al. showed that professionals underestimated the QoL of persons with high tetraplegia.[14] Post et al. reported that persons with paraplegia due to spinal cord injury had an overall life satisfaction similar to that of the general population.[21,24]

Despite similar overall life satisfaction, differences in satisfaction between persons with SB and typically developing persons were found for several domains, although these differences were not always in the expected direction. The group of persons with SB and HC were more satisfied with their financial situation and family life than the general population. This might possibly be explained by different ways of living. Whereas most typically developing young adults were students, had left their parental homes to live on their own, and many of them having a low income, persons with SB and HC were mostly living with their parents or in a sheltered home. In both circumstances, financial limitations might be a burden mainly for their caregivers. In the present study, persons with SB and HC were more often dissatisfied with their sex life and partnership relations than the general population. For partnership relations, having SB was a significant factor, but for sex life the significant determinant was sex; males were less satisfied with their sex life or with not having a sex life at all. The topic of sexuality has been explored in greater detail elsewhere.[29] More persons with SB were dissatisfied with their self-care ability than persons in the general population. This was to be expected, considering the number of persons with SB being wheelchair-dependent or incontinent, and being dependent on others for help.

However, except for self-care, differences in life satisfaction between persons with and without SB were generally small and were probably more strongly related to other factors, such as living arrangements, than to having SB by itself. Thus, having SB does not in general seem to be an important determinant of life satisfaction for the persons involved.

With regard to the differences between subgroups of persons with SB, some findings are worthy of discussion. One finding is that the proportions of satisfied persons with SB aperta without HC and the persons with SB occulta were similar across all domains. In addition, these two groups taken together (SB without HC) showed a level of well-being

fairly similar to that of the reference group, even though a considerable number suffered from incontinence.

Within the group of young adults with SB, those with HC were more dissatisfied with five out of eight life domains. This was most obvious for self-care ability and leisure activities. However, it must be emphasized that the large majority of persons with HC were satisfied with their lives as a whole, and with six out of eight life domains. Despite a few high odds ratios in the regression analyses, the amount of variance explained by HC and level of lesion was minimal for all domains of life, apart from self-care ability and sex life. This supports the statements by Sawin et al.[16] that conditions specific for SB, such as shunt status, level of lesion, and severity of the SB, are not major determinants of QoL. Differences in life satisfaction may be more strongly associated with factors not included in the present study, e.g. parental hope and other family factors.[16] Other factors might contribute as well, e.g. coping, social support, living status, or differences in personality.[11] More research is needed into the other determinants of life satisfaction to permit the development of interventions for those persons with SB at risk of poor life satisfaction.

Our study adds to the available evidence on several aspects. First, a large number of persons were included. Second, contrary to earlier studies,[10,15,16] we used reference data and were able to perform statistical testing of differences between both groups. Third, the standardized measure of life satisfaction used in the present study enabled us to differentiate between several important domains of life. Last, most earlier studies included children and/or adolescents,[15,16] whereas our study reports on a sample of adolescents and young adults (age 16–25y).

The study has some limitations. The cut-off point for the difference between satisfied and dissatisfied is rather strict, meaning that the proportion of satisfied persons might have been underestimated. In addition, seven parents filled in the questionnaires on behalf of their child. They were, however, instructed to fill in the questionnaire in a way they felt it would best reflect their child's own perception of life satisfaction, and not the parent's perception. The small number of parental reports most probably did not bias the results. Sawin et al.[16] showed that parents are accurate in supplying overall quality-of-life data for their adolescent. Another point is that the lack of persons aged 16 and 17 years in the population sample might have influenced the results. However, the analyses showed no relationship between age and life satisfaction, and in the regression analysis the comparisons were corrected for age. Furthermore, multiple comparisons were made, which increases the chance of false positives. Because the influence of SB on life satisfaction was positive for some domains and negative for others, adding up item scores

and analyzing a total LiSat-9 score only would obscure influences of interest. To minimize the chance of false positives, the alpha coefficient was corrected with the Bonferroni method.

In general, satisfaction with life seems to be fairly similar for young adults with or without SB, which in itself is good news. However, young adults with SB are at risk for dissatisfaction with important aspects of life, such as self-care, partnership relations, and sex life, even when this does not seem to be reflected in their satisfaction with life as a whole. Professional care providers should pay special attention to issues of sexuality and autonomy.

Finally, the participants in our study are in the middle of a period of transition and integration into society. Further longitudinal research is needed to study their development of life satisfaction over time, as well as research into personal and environmental factors that might influence life satisfaction.

## CONCLUSION

In a large group of Dutch young adults born with SB, a good to fair level of satisfaction with life was found. Self-care ability, sex life, and partnership relations were domains of life that need the attention of professional care providers. However, SB itself does not seem to be an important determinant of life satisfaction.

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# Chapter 7

## COGNITIVE FUNCTIONING IS A DETERMINANT OF SUBJECTIVE QUALITY OF LIFE IN YOUNG ADULTS WITH SPINA BIFIDA AND HYDROCEPHALUS

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## ABSTRACT

This study concerns cognitive functioning as a determinant of subjective quality of life of Dutch young adults with spina bifida and hydrocephalus (SBHC). In total, 110 young adults with SBHC participated (16 - 25 years, 63% female). Correlations and hierarchical regression models controlling for age, gender and functional independence were used to examine the association between cognitive domains such as executive functioning and subjective quality of life. Quality of life was measured with a two-item measure of happiness and satisfaction with life and a Visual Assessment Scale for overall quality of life. Executive functioning was associated with subjective quality of life (standardized Beta values 0.331 and -0.278; $p<0.05$). Intelligence, memory and verbal fluency were not associated. Functional independence was associated with subjective quality of life, but the association was weaker than that with executive functioning.

In conclusion, in young adults with SBHC executive functioning was associated with subjective quality of life. This finding underlines the importance of adding cognitive functioning to medical and functional status as a determinant for outcome.

## INTRODUCTION

Spina bifida (SB) is a complex congenital disorder that may result in a wide variety of neurological deficits. Major progress in medical treatment in the first years after birth has enlarged life expectancy of persons with SB to nearly normal or even normal.[1-4] It was only recently, however, that scientific interest focused on health conditions of adolescents and young adults with SB and that timely knowledge of secondary impairments and cognitive functioning of young adults with SB became available.[3,5-12]

SB does have a definite impact on the quality of life of SB patients.[13-15] Although there is some confusion about the concept of quality of life, the scientific community tends to acknowledge the existence of two main conceptualizations of quality of life. One concept is the objective approach, based on one's characteristics that can be objectively measured by an external appraiser. The second concept is the subjective approach whose focal point is the person's emotional or cognitive assessment of the congruence between his/her life expectations and achievements.[16,17] This second concept can be further inventoried in several ways.[16-18] One approach is that of health related quality of life (HRQOL), in which subjectively experienced health in physical, mental, functional and social domains of life are measured multidimensional. The second approach is that of well-being, or the subjective evaluation of life as a whole or of aspects of life. Equivalent terms are happiness, global well-being and subjective well-being.[18] A third approach conceives of quality of life as a super-ordinate construct, incorporating both HRQOL and well-being in a broader definition of quality of life. Research into quality of life of persons with SB has mainly focussed on the HRQOL.[19-26] It is important not to focus only on HRQOL, but also on wider subjective approaches of quality of life if one wants to account for the full range of consequences of a condition such as SB.[18,27]

A number of studies have examined the association between secondary health conditions of SB and (mostly) health related quality of life. Several determinants of HRQOL were examined, amongst which degree of ambulation[21], dependence in daily activities[19], urinary/faecal incontinence and medical management of urological problems[22,23], pain[25] and parental hope.[26] Not all of these variables appeared to be associated with (health related) quality of life and quality of life appeared to be not necessarily lower for persons with more severe disability as a result of SB.[15,19,20,24]

One characteristic of persons with SB, especially those with SB and hydrocephalus (SBHC), has only rarely been related to quality of life, namely lower cognitive functioning. Cognition of persons with SB occulta and those with SB without hydrocephalus cannot be distinguished from that of healthy peers.[11] However, persons with SB and HC (SBHC)

are, generally, functioning on a lower than average cognitive level when compared with the normal population,[11,28-31] although the effect of HC and associated illness parameters on cognition varies greatly between individuals.[11,29] Cognitive functioning is considered to be a variable that mediates or moderates behavioural and social outcome.[32,33] Studies on patients with cognitive impairments of different etiology have shown a distinct relation between cognition and (health related) quality of life.[34,35] Therefore, the relation between cognitive functioning and quality of life is relevant to be examined in persons with SBHC. Yet, to our knowledge, only one study reported on this relationship in persons with SBHC. Hetherington et al.[36] found no relation between intelligence and subjective quality of life, but they did find the latter to be related to functional math skills. However, their sample size was small and only participants with an IQ over 70 were included.
The aim of the present study is to investigate the association of several domains of cognitive functioning with the well-being approach of subjective quality of life in SB persons with hydrocephalus.

## METHODS

### Participants

Data were gathered from 110 young adults (aged 15 – 25 years) with SB aperta who were shunted early in life for hydrocephalus (HC). The subjects participated in a larger cross-sectional multi-centre study that examined physical and cognitive abilities, health care, social participation and life satisfaction.[11,12]
The ethics and research comities of the participating institutions have approved of the study. Informed consent was obtained from all participants. There were no significant differences between the response group and the non-response group with regard to age, gender and level of lesion (alpha = 0.05).

### Variables

Medical records were examined according to a fixed protocol. Participants underwent a physical and neuropsychological examination and a semi-structured interview and they filled in questionnaires.
Hydrocephalus was defined as being shunted early in life for hydrocephalus. In accordance with the 'International Standards for Neurological and Functional Classification of Spinal Cord Injury'[37] the 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 three subgroups based on their level of lesion: high (L2 or above), middle (L3 – L5) and low (S1 and below).[12]

The motor score of the Functional Independence Measure (FIM) was used to rate independence in activities of daily living. The FIM scores were based on observations during the preceding physical examination, supplemented by information from the participants if necessary.[37,38] The FIM motor score covers self care, sphincter control, transfer and locomotion.

A trained psychologist assessed cognitive functioning by means of a battery of tests that covered major cognitive domains such as intelligence, memory, attention and executive functioning. The assessment lasted 90 minutes with a short break. More detailed information on the instruments is provided elsewhere.[11]

The Standard Progressive Matrices was applied to measure 'fluid' intelligence.[39] The score used was the number of correct items, converted to intelligence quotient (IQ).

The updated Wechsler Memory Scale[40] 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 three different tests. The Wisconsin modified Card Sorting Test (WmCST)[41] assessed the ability to maintain and shift a cognitive set. The number of correct categories (range 0-6) was used as the score. The Trail Making Test[42] (TMT) parts A & B evaluated speed, divided attention and switching ability. The difference in execution time (seconds; B-A) needed to complete parts A and B was used as the score. To address the interface of memory, language, and behavioural regulation, word production according to lexical rules (UNKA test[43]) was used. The total number of correct words was scored.

A clinical significance of poor performances was defined by setting a cut-off point and considering scores from 2SD worse than the population mean of the variable as 'deviation scores' in the IQ, MQ and UNKA tests. In the absence of suitable population data, the reference value was taken from data on the TMT for a group of young adults with spina bifida occulta.[11] For the WmCST, the clinically significant cut-off point of five categories was used because failing to find all six categories is pathognomonic for impairment. More information on 'deviation scores' is provided elsewhere.[11]

To assess subjective quality of life, two different measures were used. First, participants were asked to rate how happy they were the past month and how satisfied they were the past month.[44] The answer was given on a seven point verbal rating scale (VRS). The

summed score of both questions was taken as an indication of the life satisfaction in the past four weeks (range 2 – 14; 14 being the best possible satisfaction). Cronbach's alpha for the two items was 0.82. The summed score is referred to as the VRS-score.
Secondly, participants were asked to rate their current quality of life on a continuous visual analogue scale (range 1 – 100 mm; 100 being the best possible quality of life). The rating is referred to as VAS-score.

### Statistics

Data were analysed using SPSS for Windows version 14. The relation between cognition and measures for subjective quality of life were examined using Pearson correlation and a hierarchical regression analysis, controlling for age, gender and functional independence. Alpha was set at 5%.

## RESULTS

Demographic and neurological characteristics are summarized in Table 1. The majority of the 110 participants were female (63%) and half of the participants was aged 21 years or older. 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% were suffering from faecal and/or urinary incontinence. The mean FIM motor score was 72.8 (SD 14.6; range 7-91).
Mean scores on memory function (MQ) and interface of memory, language, and behavioural regulation (word production task) and divided attention (TMT time B-A) were within the normal range compared to the population. Yet, the proportions of persons scoring 2 SD below the population means, indicating an impairment on the particular tests, were higher than expected in the population (9% to 14% against an estimated 2.5% in the general population). The mean IQ was 83, roughly one SD below the population mean. The IQ of 30% of the participants was below average (between 85 and 70) and of 20% it was equal to or lower than 70. The group of 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 mean summed score on the VRS was 10.8 on a 2-14 scale (Table 1). For the quality of life scale ranging from 1 to 100, the mean VAS score was 68. If the summed satisfaction score would be converted to a scale ranging from 1 to 100, the mean score on both outcome measures would be roughly 70. The linear association between both outcome measures was strong (Pearson correlation 0.68; $p < 0.001$).

**TABLE I: DEMOGRAPHIC AND NEUROLOGICAL CHARACTERISTICS, COGNITIVE FUNCTIONING AND QUALITY OF LIFE OF THE STUDY GROUP**

| Characteristics | | mean (SD) | n (%) |
|---|---|---|---|
| | N | | 110 |
| **Sex** | | | |
| | Male | | 41 (37%) |
| | Female | | 69 (63%) |
| **Age** | | 20.8 (2.88) | |
| **Level of lesion** | | | |
| | L2 or higher | | 61 (55%) |
| | L3 – L5 | | 44 (40%) |
| | S1 or lower | | 5 (5%) |
| **Ambulation** | | | |
| | wheelchair dependent | | 60 (55%) |
| **Continence** | | | |
| | Incontinence | | 89 (81%) |
| **Functional independence** | | | |
| | FIM motor score, mean (SD) | 72.8 (14.6) | |
| **Cognition** | | | (impaired*) |
| | IQ | 83 (14.7) | 22 (20%) |
| | MQ | 91 (15.8) | 16 (15%) |
| | WmCST | 4.9 (1.58) | 50 (45%) |
| | Trail making test (time B-A) | 46 (30.1) | 13 (12%) |
| | Word production task | 38 (13.8) | 10 (9%) |
| **Happiness and satisfaction** | VRS | 10.8 (1.95) | |
| **Quality of life at this moment** | VAS | 68 (21.1) | |

* individual scores below 2SD from the population means and in the case of WmCST failing to find all 6 categories were considered to indicate impairments

Age and gender were not related to either cognitive functioning or subjective quality of life (Table 2). The FIM motor score was moderately strongly (0.24 - 0.44) related to all cognitive scores, but only weakly to the VAS score (0.23) and not significantly to the VRS score. The WmCST score was, weakly but significantly, related to both subjective quality of life measures (0.22 - 0.23), and the TMT score was moderately related to the VAS score (-0.33), but not to the VRS score. Intelligence, memory and word production were not significantly related to quality of life. All cognitive test scores were significantly related to each other (0.23 - 0.74). Various non-linear associations were also explored, but these analyses did not reveal stronger relationships than the linear correlations.

**TABLE 2: ASSOCIATIONS (PEARSON CORRELATION) BETWEEN DEMOGRAPHIC VARIABLES, FUNCTIONAL INDEPENDENCE AND COGNITIVE FUNCTIONING IN RELATION TO QUALITY OF LIFE (N = 110)**

| | *Life satisfaction* (VRS) | *Quality of life* (VAS) | Gender# | Age | Functional independence (FIM) |
|---|---|---|---|---|---|
| **Gender** | 0,100 | - 0,002 | | | |
| **Age** | - 0,024 | - 0,093 | 0,042 | | |
| **FIM** | 0,167 | 0,234* | 0,019 | - 0,065 | |
| **IQ** | - 0,026 | 0,075 | 0,121 | 0,047 | 0,437** |
| **MQ** | - 0,073 | 0,028 | 0,010 | 0,026 | 0,298** |
| **WmCST** | 0,216* | 0,231* | 0,114 | - 0,103 | 0,368** |
| **Trail making test (time B-A)** | - 0,086 | - 0,330** | 0,028 | 0,005 | - 0,240* |
| **Word production task** | - 0,007 | 0,126 | - 0,008 | 0,080 | 0,239* |

* p < 0.05; ** p < 0.001; # Kendall's Tau B

To further investigate the relationship between cognition and subjective quality of life, a hierarchical regression analysis was performed, controlling for age, gender and functional independence (Table 3). When analysing the model for the VRS score, only the WmCST was found to be significantly associated with life satisfaction (B = 0.441; p = 0.005). The proportion of variance explained by the model was, however, small (14.5%). When analysing the model for the VAS score, the FIM motor score was significantly related to quality of life, when added into the model (B = 0.327; p = 0.029). However, after adding the cognition scores to the model, the association with the FIM motor score was no longer significant. The TMT was a significant determinant of the VAS score (B = -0.124; p = 0.030). Again however, the proportion of explained variance by the added variables was small (17.1%).

## DISCUSSION

The association between major domains of cognitive functioning and the well-being approach of subjective quality of life of young adults with spina bifida and hydrocephalus (SBHC) was investigated. It was the domain of executive functioning, rather than the domains of intelligence, memory and verbal fluency, that was significantly associated with subjective quality of life. In the hierarchical analyses, measures of executive functioning were also the strongest determinants of subjective quality of life, although the

**TABLE 3: HIERARCHICAL REGRESSION MODELS FOR DEMOGRAPHIC VARIABLES, FUNCTIONAL INDEPENDENCE AND COGNITIVE FUNCTIONING IN RELATION TO QUALITY OF LIFE**

| | | Life satisfaction (VRS) | Quality of Life (VAS) |
|---|---|---|---|
| | N | 102 | 99 |
| **block 1** | | B (Beta) | B (Beta) |
| | **Gender** | 0.586 ( 0.144) | 1.635 ( 0.038) |
| | **Age** | -0.015 (-0.021) | -0.299 (-0.041) |
| | $R^2$ change | 2.1% | 0.3% |
| | model F | 1.071 | 0.149 |
| **block 2** | | B (Beta) | B (Beta) |
| | **FIM** | 0.020 (0.149) | 0.327 (0.226) |
| | $R^2$ change | 2.2% | 4.9% * |
| | model F | 1.462 | 1.752 |
| **block 3** | | B (Beta) | B (Beta) |
| | **IQ** | -0.021 (-0.154) | -0.196 (-0.134) |
| | **MQ** | -0.020 (-0.149) | -0.135 (-0.097) |
| | **WmCST** | 0.441 ( 0.331) *** | 3.166 ( 0.228) |
| | **Trail making test (time B-A)** | -0.003 (-0.078) | -0.124 (-0.278) *** |
| | **Word production task** | 0.000 (-0.003) | 0.151 ( 0.098) |
| | $R^2$ change | 10.2% | 11.8% * |
| | model F | 1.964 | 2.315 * |
| | total $R^2$ | 14.5% | 17.1% |

* sign change of $R^2$; $p < 0.05$; ** sign F; $p < 0.05$; *** sign unstandardized B; $p < 0.05$
note: only the unstandardized B of the last step of the model is shown

associations were weak and the variance explained by the model was low. Even so, it is noteworthy that the predictive value of executive functioning was higher than that of functional independence.

The present study was part of a larger investigation of outcome in SBHC. We already reported functional independence, which refers to self care, sphincter control, transfer and locomotion, to be closely related to level of lesion, an indicator of severity of the condition.[38] It has been demonstrated that severity of health condition is not necessarily associated with health related quality of life.[19,20] We could specify elsewhere that in the group of participants of the present study severity of health condition was associated with specific domains of life satisfaction like self care ability, but that it was minimally associated with overall satisfaction with life.[24]

Similar to Hetherington et al.[36] we did not find an association between intelligence and subjective quality of life. Whereas Hetherington et al. found everyday mathematics to be related to subjective quality of life, we found an association with executive functioning. Our study adds to the available knowledge, because the study sample was much larger than that of Hetherington, we did not exclude persons with an IQ below 70 and perhaps on an explanatory level because mathematics have been shown to be dependent on executive functioning.[45] Furthermore, our study shows the importance of measuring cognitive function not solely by intelligence tests, as argued by others.[29] Even in persons with SBHC and normal intelligence, subtle cognitive deficits may exist.[11,29]

Executive functioning is a badly defined cognitive domain but nonetheless an important domain for daily living. 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 is related to self-care independence and social participation[46] and might also be related to everyday physical activity.[47] Studies in other diagnostic groups confirm a relation between executive functioning and quality of life. In persons with traumatic brain injury, *e.g.* executive (dis)functioning was uniquely associated with identifying emotions, which in turn was associated with overall quality of life.[48] Some found executive functioning to be a good predictor of health related quality of life after stroke[34] and in children with epilepsy.[35] Also in schizophrenia patients the executive function appeared to have a direct impact on perceived quality of life of the social domain[49] and to play a major role in moderating the relationship between subjective experience of self esteem and life satisfaction and psychological functioning.[50] The findings of this study suggest that, in addition to medical and functional status, it is important to examine cognitive ability of children and young adults with chronic health conditions in order to establish determinants of outcome.

The question why executive functioning, rather than domains such as intelligence or memory, is related to measures of well-being 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 every more complicated or more articulated act. In this respect it is of note that executive functioning and working memory are closely related concepts. However, our results are of a preliminary nature, not in the least because the domain of executive functioning is not yet clearly demarcated and as executive functioning is a plural and quickly changing theoretical concept.[51]

It is important to note some limitations of the study. First, this explorative study is not intended to determine causality. Causality might go both ways between predictors and outcome and any evidence for causality clearly needs further study.

Secondly, in this study only the well-being approach of subjective quality of life was addressed. There is a widespread agreement that quality of life is a multidimensional concept and a single-item approach might not do justice to a broad concept such as quality of life. Results regarding health status and social participation of this study group were reported elsewhere.[15,24,52] The present study focused mainly on the domain of well being or subjective quality of life.[18] We chose to measure subjective quality of life with two simple measures, a verbal rating scale and a visual analogue scale consisting of two and one item respectively. There is evidence that a single-item approach of quality of life is an acceptable method for a global assessment.[20,53-55] In addition, it has been argued that for children and adolescents it is preferable not to use diseases-specific measures of quality of life and that a self-report questionnaires should be tailored to the level of expressive and receptive language abilities as well as time perception and memory.[27] We feel that our single-item approach suffices for a global assessment of subjective well being.

Finally, a poor outcome on the TMT and WmCST could be caused by other factors than impairment of executive functioning alone. To determine a more thorough understanding of the association between executive functioning and quality of life, executive functioning should be assessed in more detail.

## CONCLUSION

There was an association between executive functioning of young adults with SBHC and their rating of their quality of life. This association needs further investigation, but it underlines the importance to add cognitive functioning to medical and functional status

as determinants for outcome and to include a wider variety of neuropsychological tests than solely intelligence scales.

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# Chapter 8

## GENERAL DISCUSSION

This chapter discusses the main conclusions of our study, as well as its strengths and limitations. It also discusses implications with respect to health care programmes for persons with spina bifida. Finally, suggestions are made for future research.

## MAIN FINDINGS

Secondary *physical* impairments were present in all participants of our study. Most frequent impairments were incontinence (especially for urine), constipation, foot deformities and scoliosis. Many persons used devices for their mobility (Chapter 2). Among all participants, those with hydrocephalus and /or with a high level of lesion were most likely to suffer secondary impairments.

Young adults with spina bifida aperta with hydrocephalus had a lower cognitive status than respondents with spina bifida aperta without hydrocephalus and those with spina bifida occulta (Chapter 3). The cognitive status of the two groups of persons without hydrocephalus was similar to that of the healthy general population. Almost half of the participants with spina bifida aperta with hydrocephalus had cognitive impairments (defined as a score two standard deviations below the population mean). Most impairments pertained to the domain of executive functioning, but more general impairments involving most domains of cognitive functioning occurred as well. Cognitive status appeared to be negatively influenced by multiple shunt revisions and by pathology associated with hydrocephalus.

Persons with spina bifida and hydrocephalus had more often started off in special primary education or had been transferred to special primary education. They had a lower educational outcome than persons with spina bifida without hydrocephalus, whose educational career was similar to that of healthy peers (Chapter 4). Intelligence level was the strongest predictor of educational outcome, but hydrocephalus and wheelchair dependence were also significant predictors.

The results on social participation and its determinants in young adults with spina bifida indicate considerable restrictions to participation (Chapter 5). Compared to healthy peers, young adults with spina bifida were less often living independently, more often in need of special education and more often unemployed. 'Starting relationships', however, appeared to be less affected by having spina bifida. Social participation was related to type of spina bifida, hydrocephalus, wheelchair-dependence and urinary incontinence. Young adults with spina bifida perceived their social participation to be particularly hampered in the domain of leisure, but also in going to school or work and visiting family

or friends. Physical impairment was mentioned as the foremost impediment in social and societal participation, but accessibility of buildings, problems in long-distance transportation and own emotional barriers were also frequently mentioned. No relation was found between severity of spina bifida and perceived hindrances in relation to physical impairments or emotional distress. The fact that those with relatively minor dysfunctions experience social barriers, might be understood from previous findings in spina bifida research that those who are capable of functioning in surroundings of healthy people feel marginal, because of their functional impairments and have to exert undue effort to maintain their position.[1]

With respect to life satisfaction, one of the major findings of the study is that the overall satisfaction with life of young adults born with spina bifida is more or less similar to that of their healthy peers and that severity of spina bifida had only a minor impact on life satisfaction (Chapter 6). Nevertheless, when focusing on particular domains, differences between persons with spina bifida and healthy persons were apparent, although not always in the same direction. The only larger difference concerned self-care.

With respect to relationships between cognitive functioning and quality of life as perceived by young adults with spina bifida and hydrocephalus, we observed an association between executive aspects of cognitive functioning and perceived quality of life (Chapter 7). The more executive dysfunction, the less quality of life was perceived. Importantly, cognitive domains such as intelligence and memory were not associated with self-perceived quality of life. However, the relationship between executive function needs further research, because lexical word production (currently conceived of as tapping executive function) was also not associated with quality of life.

## Our findings in relation to previous studies

There is a growing research focus on adults with spina bifida, but the majority of spina bifida studies still concern children under the age of 16 years. Also, studies tend to focus on the physical impairments as determinants of outcome. It is well established that part of the population with spina bifida and hydrocephalus is suffering from impaired cognitive functioning.[2-6] Yet, cognitive functioning as a determinant of outcome is as yet insufficiently recognised. Our study adds to the available knowledge in several ways.

With the ASPINE-study[7-9] we were able to describe the condition of young adults with spina bifida and of important subgroups within that population with respect to physical functions, activities and participation.[10] The findings on secondary impairments

supplemented earlier studies, which pertained to smaller groups of participants and either did not describe subgroup differences or only in relation to level of lesion.[11-22]
The findings on cognitive functioning of our study group add to the available literature on the heterogeneity of persons with SB and highlight the necessity of comprehensive assessment rather than relying solely on IQ.[2-6] The educational outcome for persons with SB and hydrocephalus support earlier findings[23,24], but also add to the knowledge of the educational career. Many children with hydrocephalus switched between normal and special education and 8% dropped out after special primary school. The results on social participation confirmed considerable restrictions to social participation in young adults with spina bifida[14,21,24-31], but also revealed that, from the perspective of the young adult with spina bifida, restrictions are not related to the severity of spina bifida. The finding that severity of SB has only a minor impact on life satisfaction confirms the results of most of the previous but smaller studies, using similar or different conceptualizations of quality of life from ours.[32-35] Our study adds to the available evidence, because in the ASPINE-study not only the concept of health related quality of life was included[36], but also that of life satisfaction (Chapter 6). Furthermore, our data are from a nationwide cohort rather than small groups, pertain to young adults rather than children, and are controlled by reference data of healthy peers. The exploration of the association between cognitive functioning and quality of life supports findings previously reported.[37] It also adds to the available knowledge, because the study sample was a nationwide cohort and because we did not exclude persons with an IQ below 70. Furthermore, our study shows the importance of including not only intelligence a measure of cognitive status, but a wider variety of neuropsychological tests, as argued for by others.[2] Hydrocephalus, or rather early shunting to reduce intracranial pressure, has proven to be a strong predictor for several domains of outcome, such as cognitive functioning, educational outcome and several domains of quality of life and participation in society. The number of shunt revisions was associated with poorer cognitive functioning. Other studies also showed that, despite management of hydrocephalus and intracranial pressure early in life, cognitive development may be far from normal.[38,39] The hypothesis is warranted that hydrocephalus is not solely responsible for cognitive dysfunction and that brain malformations contribute to poor outcome.[40,41] Obviously, adequate monitoring of recurrent hydrocephalus or shunt malfunction is still needed. Patients (young or adult) with assumed arrested hydrocephalus might also benefit from monitoring of the intracranial pressure and shunting may prove helpful for preserving or improving neuropsychological functioning.[42]

An alarming conclusion from our study is that wheelchair dependence is still a barrier to social participation. Contrary to previous finding[43], it is at least in The Netherlands, a significant determinant of educational outcome. Wheelchair dependence has to be conceived of as a factor associated with activity limitations like pain or fatigue, rather than as a cause of educational outcome.[44] Similarly, long distance transportation and accessibility of buildings pose restrictions in the domains of work, education, visiting people and leisure time for a considerable number of young adults with spina bifida. These findings are not specific for persons with spina bifida.[45] Dismantling barriers to social participation of physically disabled persons due to inaccessibility of buildings should become a priority of counsellors and governmental authorities.
For young adults with spina bifida without hydrocephalus, we were able to confirm the findings of a very small previous study[46] that the cognitive status of persons without hydrocephalus is similar to that of the healthy general population. Also, young adults with spina bifida without hydrocephalus have an educational outcome similar to their healthy peers, which supports earlier finding.[23,24] Contrary to expectations, although in line with limited research till date, the type of spina bifida appears to have limited impact on overall life satisfaction or on the perceived hindrances in relation to physical impairments or emotional distress.[32-35] The young adults with spina bifida occulta or spina bifida aperta without hydrocephalus may use healthy peers as a reference group and therefore perceive less quality of life and more obstacles in participation. Suboptimal quality of life and restrictions in participation for apparently less severely handicapped young adults with spina bifida should not be underestimated by caregivers.

## STUDY POPULATION

In the initial stages of the study the study population was estimated at roughly 800 living persons with spina bifida in the age range of 16 to 25 years. In the absence of a national registration system for spina bifida patients, the number was based on prevalence numbers for the Netherlands.[47,48] An estimated 125 live births per 195.000 births a year, multiplied by 10 years and then corrected for a survival rate of 65%[48], led to the estimated 800 potential participants. Based on those numbers and considerations for the study (to obtain a representative study sample and generalizable results) and the purpose to analyse subgroups in relation to outcome variables, the aim was to have a study sample of 350 participants. To be able to facilitate a non-response of roughly 25% to 30%, we estimated that a total of 500 young adults had to be invited to participate in the study.

In the following stage of the study it became gradually clear that the number of spina bifida sufferers was overestimated. When checking medical records we found fewer young adults than anticipated and numerical misstatements, because of duplicates (persons with spina bifida were known to more than one spina bifida team). In addition, one hospital-based spina bifida team in the northern part of The Netherlands did not participate in the study, which further reduced the potential number of participants. Every effort was made to enlarge the number of participants as much as possible. Patients were invited by BOSK, the Dutch patients' association. Adverts were placed in magazines with an invitation to contact the researchers. Other organizations providing care to persons with spina bifida were also asked to participate in the study (13 rehabilitation centres, 91 special housing projects and 12 hospitals in areas where no spina bifida team was active). Six rehabilitation centres and 8 special housing projects responded, which resulted in 52 further potential participants. However, most of the latter were already known through the spina bifida teams. Furthermore, the response rate was lower than expected. Some persons reported their reason for refusing to participate. The most frequent reason was that the time needed for the examination did not fit the busy daily schedule of the person with spina bifida. Another reason was avoidance of contacts with the health care system, after a childhood period of intensive medical treatment. In the end, out of a total of 350 contacted young adults with spina bifida, 179 participated in the study. Analysis showed that participants and non-participants did not significantly differ in terms of gender, age, type of spina bifida, level of lesion and hydrocephalus (Chapter 1).[7] And as we, despite our efforts, found hardly any persons not already known to one or more spina bifida teams, we conclude that the present study group adequately represents the Dutch population of young adults with spina bifida. Hence, we concluded that our results can be taken to be representative for this population. The 95% confidence interval (CI) for a dichotomy outcome of, e.g., 50% in our finite population is still only ±5,1%, as opposed to a CI of ±3,9% for the initial numbers of 350 participants and a population of 800. Because of the representativeness of the study group, the group of participants can viewed as a nation wide cohort of Dutch young adults with spina bifida.

## METHODOLOGICAL CONSIDERATIONS

The over-all aim of the study was to identify care needs of young adults with SB in order to provide recommendations for the improvement of care and counselling. For this reason, ASPINE was designed as a broad explorative study of physical functioning,

cognitive functioning, independence in daily living, functioning in the community, perceived health and life satisfaction, as well as of actual care and need of care.
The number of participants was large enough to allow analysis of several domains of functions, activities and participation in relevant subgroups. As already discussed, it was possible to study the groups of participants with spina bifida aperta without hydrocephalus and with spina bifida occulta. In addition, whereas previous studies on older persons with SB had not been able to concentrate on a specific age range because of limited number of patients, we had the opportunity to focus on the period of life following childhood for a study of the relationship between the domains of functions, activities and participation.
In agreement with the International Classification of Functioning, Disability and Health (ICF)[10] we gathered data on physical and functional impairments, limitations in activities and restrictions in social and participation. Our extensive set of measures included a physical examination, a neuropsychological examination, an interview, several questionnaires and data taken from the medical records. Examinations we performed by a physician and a psychologist. The dual setup of the study allowed us to gather data on medical status and physical functioning, as well as neuropsychological functioning and to study the relations between these domains and their joint relationships with measures of activities and participation.
We used standardized measures if available, preferably with reference data for the general population, which is a strength compared to previous studies that often used non-standardized or disease-specific measures and therefore lack the value of generalisation for groups of patients with different disorders. The use of multivariate analysis techniques, so far not very common in the literature, provided new insights into the relationship between the different domains.

Despite the strengths of this study there are also limitations and methodological considerations. A cross-sectional retrospective design limits the possibility to infer causality from the relationships found. For information on medical history, including for example level of lesion, shunting and number of shunt revisions we relied on medical records. We often encountered unclear or incomplete information. An advantage of our study was that we determined level of lesion in a standardized way in the physical examination and also that the participant could supplement incomplete data. However, signs and symptoms may change over time and recollections of the course of the disease may undergo adaptations and this limits the prognostic value of our results.

Also, despite not having found differences between participants and non-participants on important variables, we cannot rule out a non-response bias on important outcome variables. The association between type of spina bifida, level of lesion and the presence of hydrocephalus was high. Given the strong association between health condition, secondary impairments and activity limitations, we assume that the results for cognitive functioning and educational career are representative for the whole population. However, severity of spina bifida was only weakly associated with quality of life and therefore we are less certain about how representative those results are.

Longitudinal studies such as those by Hunt have contributed much to the knowledge about long term outcome for with persons spina bifida.[13,18,22,27,28,49-51] However, such results cannot easily be translated into prognoses for children born with spina bifida today. Medical treatment has improved and policies for treating newborns with spina bifida change over time. On the one hand improved medical treatment may improve the outcome for a group of patients and data from previous cohorts may therefore underestimate the outcome for current generations. On the other hand, improved medical treatment may also result in better chances of survival for more severe cases of spina bifida with relatively worse long term prognosis. Even when our cohort of persons born between the late 1970's and beginning of the 1980's is of a more recent date, inferring a possible outcome for newborn with spina bifida should be done with some caution.

The present study deliberately included young adults with spina bifida occulta. It is often thought that spina bifida occulta is a mild, unproblematic form of spina bifida. The results of our study show that persons with spina bifida occulta can experience various medical problems of which especially the effect on quality of life should not be underestimated by caregivers. However, in the introduction it was already pointed out that the prevalence of this type of spina bifida is roughly estimated at 10% to 20% of the general population.[52,53] The persons with spina bifida occulta included in our study were known and registered in the hospital administrations. They either had visual skin marks or had neurological problems. Therefore the included group is presumably not representative of the total group of young adults with spina bifida occulta.

To keep respondent burden within reasonable limits and for other practical reasons, the neuropsychological examination was allowed to take a maximum of 90 minutes for each participant. We examined the feasibility of the test battery in a pilot study of ten participants. Despite positive earlier experiences with more or less the same set of neuropsychological tests, the available 90 minutes for the neuropsychological

examination turned out to be too short. One option was to drop one or two tests. However, the battery was already a minimum set to cover most domains of cognitive functioning and tailored to what we expected to be most informative in young adults with spina bifida. Therefore it was decided to keep the complete set of neuropsychological tests for the final study, but with the compromise that half of the participants would have the test for verbal learning and the other half would have the test for reaction timing and tapping. This solved the 90 minute time limit problem, but complicated running multivariate analysis with the data of all neuropsychological tests.

One other problem we encountered was collinearity. Collinearity occurs when independent variables are so highly correlated that it becomes difficult or impossible to distinguish their individual influences on an outcome variable. It does not affect the ability to predict an outcome in a regression analysis, but in practical terms, it means that there is a degree of redundancy or overlap among the predicting variables. Collinearity within the variables for health condition (type of spina bifida, hydrocephalus and to some extent level of lesion) slightly complicated the interpretation of the contribution of the individual predictors in multivariate analysis. However, the emphasis of our study being on description of the outcome in relevant subgroups, the classification of type of spina bifida with or without hydrocephalus proved to be a satisfactory solution for the collinearity between both variables.

It is impossible to measure all relevant factors thoroughly in one study. Given the aim of the study we preferred to provide an extensive description of the functioning of young adults with spina bifida in ICF-terms of functions, activities and participation, rather than to search for causal relations between the variables. This meant that personal factors, such as coping style and personality, were not included in the study and that the examination of external factors was not extended to, e.g., domestic situation, attitudes of parents and social support. Other studies already outlined the importance of these other factors.[34,54-61]

## IMPLICATIONS FOR HEALTH CARE POLICY ON YOUNG ADULTS WITH SPINA BIFIDA.

The care for patients with spina bifida has been focusing mainly on children. From a historical point of view this is understandable. However, since the majority of the persons now survive into adulthood[13,18,21,22,28,61-67], the care for these persons should change as well.[15,20,64,68-74]

To be able to make recommendations for future care for young adults with spina bifida, we interviewed respondents about actual care and care shortcomings. We also interviewed all multidisciplinary spina bifida teams in the Netherlands.[7,8] At the time of the study 53% of the participants were still under the care of a spina bifida team. Of the other participants 39% had been under the care of a spina bifida team in the past and 14 participants (8%) had never been seen by a team. Whether or not someone was still under the care of a spina bifida team was closely related to the policy of a particular team with respect to when to stop multidisciplinary care. Some teams stop care at the age of 16 whereas other teams continue to care in the form of a follow-up 'adult team'. From the interviews with the spina bifida teams a difference in emphasis on particular aspects of care transpired. Some teams focus on medical issues, whereas others have a more holistic approach, including psychosocial issues and participation. Asked which problems they considered of particular importance for (young) adults with spina bifida, the spina bifida teams emphasized the ability to maintain self care, (lack of) insight of the persons in their own health condition as well as lack of initiative seeking help and an increase of psychosocial problems. Furthermore, the spina bifida team members agreed that structural medical care for adolescents and (young) adults was lacking. Also, they indicated a lack of attention and expertise for the psychosocial care.

From our study it can be inferred that many young adults with spina bifida have either a specific or global cognitive impairment. Professional caregivers should be aware that even in young adults with apparently normal cognition such impairments may exist and can affect activities and participation. Considering the effect of hydrocephalus on cognitive functioning an adequate monitoring of recurrent hydrocephalus, shunt malfunction or assumed arrested hydrocephalus is needed, even in adult persons.

Carers of children with spina bifida should be aware of the fact that, apart from deficient learning abilities, wheelchair dependence and associated physical and social problems may also restrict children in their educational career. It is encouraging that half of the young adults with spina bifida manage a fair to good level of education. The challenge is to continue to help create conditions for attending regular education to ensure optimal opportunities in later life.

Spina bifida has an effect on the health status and social participation of all young adults with spina bifida and the effect should not be underestimated for persons with milder forms of spina bifida or with lesser impairments.

More effort should be directed to reduce participation restrictions, especially with respect to leisure activities and mobility for both wheelchair dependent and non

wheelchair dependent young adults with spina bifida. To diminish impact of physical impairments and to abolish illness-related emotional distress, barriers to social participation of wheelchair dependent persons should be broken down; *e.g.*, accessibility of buildings should become a priority of counsellors and governmental authorities.
The researchers discussed the results of the present study with the coordinating physicians of the various multidisciplinary spins bifida teams. There was agreement about the view that the care for (young) adults with spina bifida should also be done in the form of a multidisciplinary spina bifida team. Such a team could consist of a ' core team' of a rehabilitation doctor and a urologist.[7,8] If screening by the core team would reveal specific problems, a specialist should be readily available for consulting. Teams indicated a lack of knowledge for specific domains like psychosocial care for those in the transition period from child to adult. Given the fact that individuals with spina bifida vary greatly in physical and cognitive functioning, it might be needed to concentrate multidisciplinary care for (young) adults with spina bifida in a limited number of teams. Though, care should be available in all parts of The Netherlands. Persons with spina bifida often have cognitive impairments and teams indicated that lack of insight of the persons in their own health condition as well as lack of initiative seeking help was one of the main concerns for adult care. Therefore we would like to emphasize the need for an active role by caregivers in prevention of psychosocial problems and regular medical check-ups for treatment if necessary.

## PROPOSALS FOR FUTURE RESEARCH

Participants of our study are in the middle of a period of transition and integration into society. Further follow-up assessments are needed to study their development of participation and life satisfaction over time, as well as to study personal and environmental factors that influence participation and life satisfaction. Such studies should include psychological factors like coping self-efficacy and family factors.
Future research into the cognitive status of persons with spina bifida is desired, preferably using specific measures for various domains of cognitive functioning. Relating cognitive functioning to (recent) data of brain scans may provide new insights into the association of brain malformations and hydrocephalus and cognitive functioning. Dennis et al.[40] proposed a model of neurocognitive functioning that describes the complex association of spina bifida in association with neurological and environmental factors which have a

moderating and mediating effect on cognitive phenotype. Such a model helps identifying research lacunae, as well as suggesting possible approaches for enhancing outcome.
Research into functioning of older persons with spina bifida is needed to explore effects of ageing. Physical problems may become more prominent with aging. Also, in persons with hydrocephalus the effects of aging might affect cognitive functioning more or sooner than in the general population and thus affect social participation and self care ability[40] Knowledge about aging in this group of persons may help to prevent future problems.
Longitudinal cohort studies in persons with spina bifida are necessary to provide better insight into the relation between the severity of spina bifida measured at birth and the outcome regarding all aspects of life (medical, functional, participation and well being). These studies should include the generation of young adults, but also younger and if possible older cohorts.

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# SUMMARY

This thesis is based on the results of the ASPINE-study (Adolescents with SPina bifida In the NEtherlands), a cross-sectional descriptive study which was started in 1998 to gain insight into functioning and health of young adults with spina bifida. This thesis is focussed on cognitive functioning, social participation and life satisfaction of young adults with spina bifida. Other results have been described in the thesis by Marjolein Verhoef (2005), rehabilitation physician and co-researcher of the ASPINE-study.

Chapter 1 provides a brief introduction about spina bifida. Spina bifida (Latin for "split spine") is a developmental birth defect involving the neural tube. An incomplete closure of the embryonic neural tube results in an incompletely formed spinal cord. Spina bifida malformations fall into three categories: spina bifida occulta, spina bifida cystica (myelomeningocele), and meningocele. The most common location of the malformations is the lumbar and sacral areas of the spinal cord. In the Netherlands the incidence of spina bifida aperta is roughly 4 per 10.000 live births. Spina bifida can result in a variety of problems, amongst them incontinence and restrictions of mobility, depending on the severity of the defect. In the most severe form of spina bifida, myelomeningocele, approximately 85% of the children also develop hydrocephalus, which in turn may lead to restrictions of cognitive functioning. In The Netherlands, care for children with spina bifida is organised in multidisciplinary spina bifida teams. Due to improvements in medical care, the life expectancy of children with spina bifida has improved considerably and nowadays most children reach the adult age. However, too little is known about the problems regarding the medical condition and the quality of life of adolescents and young adults with spina bifida and about their needs for medical care.
The aim of the study was describe the functional limitations, social participation and subjective quality of life of young adults with spina bifida in order to make recommendations concerning the care and support for this group of young adults. Participants were persons with spina bifida aperta or spina bifida occulta and were between 16 and 25 years of age. Invitations were sent to 350 persons, of whom 179 (51%) participated in this study, including 37 patients with spina bifida occulta and 142 patients with spina bifida aperta. The group of participants did not significantly differ from the non-participants on demographic and spina bifida characteristics. The selection of measurements of the ASPINE-study was based on the ICF model (International Classification or Functioning, Disability and Health). Medical records were examined according a fixed protocol and participants underwent a onetime physical and

neuropsychological examination. Participants were also interviewed and filled in questionnaires.

Chapter 2 reports on the investigation of secondary impairments in young adults with spina bifida. Participants with spina bifida were subdivided into participants with spina bifida occulta (n=37) and spina bifida aperta (n=142), the latter being subdivided into 119 participants with hydrocephalus and 23 participants without hydrocephalus. Results showed that type of spina bifida was strongly associated with the level of lesion (L2 and above, L3-L5 and S1 and below). Participants with a high level of lesion mostly had spina bifida aperta and hydrocephalus.
Medical history and secondary impairments were reported groups of participants according to both type of spina bifida and level of lesion. Health problems were present in all subgroups. Most frequent secondary impairments were incontinence (especially urinary incontinence), foot deformities, scoliosis and constipation. Pain in the head, neck and back and sexual problems were also common. Many patients needed mobility devices. Participants with spina bifida aperta and hydrocephalus and participants with a high lesion level showed the largest numbers of surgical procedures and secondary impairments. Participants with spina bifida aperta without hydrocephalus and participants with spina bifida occulta were similar with respect to the prevalence of secondary impairments.

Chapter 3 reports on the cognitive status of 168 young adults with spina bifida in relation to type of spina bifida (aperta or occulta) and the presence of hydrocephalus. Cognitive status was assessed using a set of neuropsychological tests covering the domains of intelligence, memory, verbal learning, executive functioning, verbal fluency and reaction timing. Our results indicated that the cognitive status of the group of young adults with spina bifida occulta and the group of spina bifida aperta without hydrocephalus was comparable to each other and to that of the general population. Almost half of the group of young adults with hydrocephalus had cognitive impairments of some sort, indicating that the other half young adults with hydrocephalus had a cognitive status comparable to that of the general population.
For the group of young adults with hydrocephalus, impairments (defined as a score 2 SD below the population mean) were found across all domains of cognitive function, but were most noticeable on the domains of executive functioning, decision timing and verbal learning. Roughly 70% had a domain specific impairment and 30% had a global

impairment across most or all domains. Of the young adults with hydrocephalus 20% had an IQ equal or lower than 70. Intelligence was found to be the most robust measure of overall cognitive status. Poor intelligence was associated with a high frequency of shunt revisions as well as with co-morbid conditions like epilepsy and corpus callosum malformations within the group of young adults with hydrocephalus.

Chapter 4 reports on the educational career and predictors for type of primary and secondary education in 178 young adults with spina bifida. The results indicated that participants without hydrocephalus had an educational career comparable to healthy children. Participants with hydrocephalus more often started in special primary school or switched from regular to special primary school. The group of participants who switched to special primary school more often had a disability or was female. Nearly 60% of participants with hydrocephalus finished primary school in special education. However, a considerable number of them managed to switch from special primary school to regular secondary school. On the other hand, another 8% did not continue education at all after primary school.
We also reported on the predictors of educational outcome (regular or special education). Several predictors were associated with educational outcome. A logistic regression analysis showed intelligence as the main determinant. However, hydrocephalus and wheelchair dependence were additional independent determinants of educational outcome.

Chapter 5 reports on participation restrictions of 179 young adults with spina bifida in relation to disease characteristics, activity limitations and perceived hindrances for their educational career, vocational status, residential status and marital status, and problems they encountered with respect to school or work, visiting family and friends and leisure time. Results showed that young adults with spina bifida had a lower level of education and a far higher unemployment rate compared to health peers. They were also less often living independently. Marital status seemed less affected by spina bifida. Poorer outcomes across most domains of participation were associated with type of spina bifida (occulta or aperta), having hydrocephalus, being wheelchair dependent and being incontinent. Intelligence was most strongly associated with educational outcome.
Most hindrances regarded leisure, but hindrances were also reported for visiting family and friends and for attending school or work. Problems were most frequently related to physical impairments, followed by accessibility of buildings, long-distance transportation

and emotional distress. Costs were not often mentioned as hindrance in participation. Problems with long-distance transportation and accessibility of buildings were more often reported by participants who were wheelchair user, as well as participants with hydrocephalus or with a high level of lesion. Occurrence of problems due to physical impairments (e.g. pain or fatigue), emotional distress (e.g. shame or sadness) or financial restraints were not consistently related to either health condition or activity limitations, indicating that these hindrances were also perceived as problems by persons with milder forms of spina bifida or less severe activity limitations.

Chapter 6 reports on life satisfaction of 179 young adults with spina bifida. The aim of the study was to determine levels and predictors of life satisfaction of young adults with spina bifida and to compare life satisfaction of this group to life satisfaction of an age-matched population group. The results showed that 21% of the young adults with spina bifida were dissatisfied with their life as a whole, which proportion is not different from their healthy peers. Comparing satisfaction with various life domains, we found that a higher proportion of young adults with spina bifida was dissatisfied with self care ability and partnership relations. But more were satisfied with the domains of financial situation and family life. Examining the effects of age, gender, hydrocephalus and level of lesion on life satisfaction within the group of young adults with spina bifida, it was found that having a high level of lesion (L2 and higher) was associated with more dissatisfaction on some domains, but there were no predictors found for (dis)satisfaction with life as whole. This finding might perhaps be attributed to their domestic situation, young adults with spina bifida more often still living with their parents.

Chapter 7 reports on the association between cognitive functioning and subjective quality of life in 110 young adults with spina bifida and hydrocephalus. Relationships were examined between two scores for subjective quality of life, a numerical rating scale and a visual assessment scale, and scores for neuropsychological functioning, intelligence, memory and executive functioning. To control for an effect of physical limitations in activities of daily living, a measure for functional independence was added to the analysis. The results showed that the young adults with spina bifida and hydrocephalus rated their subjective quality of life at roughly 70 on a 0-100 scale. Moderate associations were found were between measures for executive functioning and subjective quality of life, but not for other domains of cognitive functioning. Further analysis showed that when adding all variables to a linear regression model, associations between executive functioning and

subjective quality of life remained significant, and were stronger than association between functional (in)dependence and subjective quality of life. However, a lower subjective quality of life was still only to a minimal degree explained by poorer executive functioning. On the other hand, these results showed the importance of not only using intelligence as an overall measure of cognitive functioning when exploring the effect of cognitive functioning as a possible determinant for outcome.

Chapter 8 is the general discussion of our findings. It also presents the main conclusions of the study about young adults with spina bifida:

- Many spina bifida patients suffer from secondary impairments such as incontinence, constipation, foot deformities, scoliosis and loss of mobility. The groups most likely to suffer from these impairments are young adults with hydrocephalus or a high level of lesion.
- Persons without hydrocephalus have a cognitive status similar to that of healthy peers. The cognitive status of the group of young adults with hydrocephalus is lower than that of the general population. Roughly half of the young adults with hydrocephalus suffer from specific or global cognitive impairments.
- Persons without hydrocephalus have an educational outcome similar to that of healthy peers. The educational outcome of the group of young adults with hydrocephalus is lower than that of the general population. Only half of the young adults with hydrocephalus attend regular secondary school. Intelligence level was the main predictor of educational outcome, but hydrocephalus and wheelchair dependence were additional significant predictors.
- Many young adults with spina bifida experience participation restrictions for the domains of educational career, vocational status and residential status. Severity of spina bifida and to a lesser extent activity limitations were negatively related to participation. Perceived restrictions in participation in relation to accessibility of buildings and long distance transport were most noticeable for wheelchair dependent patients. Perceived restrictions in relation to physical impairments and emotional stress were found for all patients with spina bifida, regardless of the severity of the spina bifida.
- The overall life satisfaction of young adults with spina bifida is similar to that of healthy peers. The domains of self care ability and partnership relations are negatively affected by spina bifida, but overall spina bifida does not seem to be an important determinant of life satisfaction.

- Executive functioning was associated with how young adults with spina bifida and hydrocephalus rated their happiness and satisfaction in life as well as their quality of life. It underlines the importance of adding cognitive ability as a determinant for outcome in addition to medical and functional status and to include a wider variety of neuropsychological tests than solely intelligence.

This chapter also discusses the strengths and limitations of our study, as well as possible implications for treatment and health care policy for patients with spina bifida. Our study showed that spina bifida is a health condition with a variety of manifestations. The size of the study sample allowed to us the describe the outcome for important subgroups, including the often neglected milder forms of spina bifida (occulta and aperta without hydrocephalus) as well as previously often omitted patients with an IQ below 70. The most important recommendation is that the medical care for young adults with spina bifida should be available in every part of The Netherlands, preferable by specialized multidisciplinary teams. A core team should at least consist of a rehabilitation physician and an urologist. The core team can screen for specific problems in all domains and should be able to consult a specialist if needed.

The chapter ends with suggestions for future research. Longitudinal cohort studies are important to determine long term outcome of spina bifida patients.

# NEDERLANDSE SAMENVATTING

Dit proefschrift is gebaseerd op de resultaten van de ASPINE-studie (Adolescenten met SPina bifida In NEderland). Het in 1998 gestarte onderzoek had tot doel om meer inzicht te krijgen in de gezondheidstoestand en het functioneren van jonge volwassenen met spina bifida. Het cross-sectionele onderzoek is daarom beschrijvend van aard. Dit proefschrift gaat voornamelijk over cognitief functioneren, maatschappelijke participatie en tevredenheid met het leven van deze jonge volwassenen. In het proefschrift van Marjolein Verhoef (revalidatiearts en mede-onderzoeker) dat in 2005 verscheen, werden o.a. functiestoornissen op orgaanniveau, beperkingen in het dagelijkse leven en ervaren gezondheid beschreven.

In hoofdstuk 1 worden de verschillende vormen van spina bifida kort toegelicht. Spina bifida (of 'open rug') is een aangeboren afwijking van het ruggenmerg. Er zijn verschillende vormen: spina bifida aperta (myelomeningocele en meningocele) en spina bifida occulta. Spina bifida aperta komt in Nederland voor bij ongeveer 4 op de 10.000 levend geboren baby's. Afhankelijk van de ernst van de spina bifida kan deze leiden tot een grote variëteit aan lichamelijke beperkingen, waaronder incontinentie en rolstoelafhankelijkheid. Bij de meest ernstige vorm van spina bifida ontstaat bij ongeveer 85% van de kinderen ook een hydrocefalus (waterhoofd), wat bij veel kinderen leidt tot beperkingen in het cognitieve functioneren.
De behandeling en begeleiding van kinderen met spina bifida is in Nederland georganiseerd in multidisciplinaire spina bifida werkgroepen. Deze werkgroepen zijn doorgaans gericht op kinderen en jongeren tot ongeveer 18 jaar. Door de ontwikkelingen in de medische behandeling in de laatste decennia is de levensverwachting van kinderen met spina bifida duidelijk verbeterd. Het merendeel van deze kinderen bereikt hierdoor nu de volwassen leeftijd, maar blijft in meer of mindere mate afhankelijk van medische zorg. Zorg en begeleiding voor deze groep is echter nog onvoldoende ontwikkeld, mede doordat nog te weinig bekend is over problemen met betrekking tot gezondheidstoestand en functioneren van adolescenten en jonge volwassenen met spina bifida.
Het doel van de studie was het verkrijgen van inzicht in functiestoornissen, maatschappelijke participatie en tevredenheid met het leven van jonge volwassenen met spina bifida, om gefundeerde adviezen te kunnen geven over de behandeling en begeleiding van deze groep jonge volwassenen.
Er werden 350 personen (leeftijd tussen 16 en 25 jaar) met spina bifida aperta of occulta opgespoord en uitgenodigd om deel te nemen aan het onderzoek. Uiteindelijk werkten

179 personen (51%) mee. Op demografische en voor spina bifida specifieke kenmerken bleek de groep jonge volwassenen die niet deelnam aan het onderzoek niet significant af te wijken van de groep deelnemers.
De selectie van meetinstrumenten in de ASPINE-studie was gebaseerd op het ICF model (International Classification of Functioning, Disability and Health). Gegevens op functieniveau werden verzameld door middel van een statusonderzoek en een eenmalig lichamelijk en neuropsychologisch onderzoek. Betreffende activiteiten en sociale en maatschappelijke participatie leverden deelnemers gegevens via interview en vragenlijsten.

Hoofdstuk 2 beschrijft de secundaire stoornissen bij jonge volwassenen met spina bifida. In totaal werden 179 patiënten geïncludeerd, van wie 37 bekend met spina bifida occulta en 142 met spina bifida aperta. De deelnemers met spina bifida aperta werden in twee groepen verdeeld: 119 met en 23 zonder hydrocefalus. Het type spina bifida (occulta, aperta zonder hydrocefalus en aperta met hydrocefalus) bleek sterk gerelateerd aan de hoogte van de laesie (L2 en hoger, L3-L5, S1 en lager). Deelnemers met een hoge laesie waren bijna allemaal gekenmerkt door spina bifida aperta met hydrocefalus.
Medische voorgeschiedenis en in het onderzoek vastgestelde secundaire stoornissen werden geclassificeerd naar zowel type spina bifida als hoogte van de laesie. De meest voorkomende stoornissen waren incontinentie (vooral voor urine), voetmisvormingen, scoliose en obstipatie. Ook pijn in het hoofd, de nek en de rug en problemen met seksueel functioneren kwamen frequent voor. Daarnaast waren veel deelnemers voor hun mobiliteit afhankelijk van hulpmiddelen.
Secundaire stoornissen kwamen in alle groepen in meer of mindere mate voor. De aanwezigheid van secundaire stoornissen was het meest uitgesproken bij deelnemers met spina bifida aperta en hydrocefalus en deelnemers met een hoge laesie. Bij laatstgenoemde categorie deelnemers was ook het aantal chirurgische interventies opvallend groter. Het percentage secundaire stoornissen bij deelnemers met spina bifida aperta zonder hydrocefalus verschilde niet van dat bij deelnemers met spina bifida occulta.

Hoofdstuk 3 beschrijft het cognitieve functioneren van 168 jonge volwassenen met spina bifida. Het voornaamste doel van dit onderdeel van het project was verkrijgen van kennis over de relatie van het cognitieve functioneren met het type spina bifida (aperta of occulta) en hydrocefalus. De gebruikte neuropsychologische testbatterij bestreek de

belangrijkste domeinen van het cognitieve functioneren. Die domeinen waren intelligentie, geheugen, verbaal leren, executief functioneren en reactiesnelheid. Onze resultaten lieten zien dat het cognitieve functioneren van jonge volwassenen met spina bifida occulta en van jonge volwassenen met spina bifida aperta zonder hydrocefalus noch van elkaar noch van het functioneren van gezonde leeftijdgenoten verschilde. Ongeveer de helft van de jonge volwassenen met hydrocefalus had enigerlei stoornis in cognitief functioneren. Dit betekent dat de andere helft van deze groep deelnemers met hydrocefalus in cognitief functioneren niet afweek van gezonde leeftijdgenoten.
Wanneer stoornis werd gedefinieerd als twee maal de standaarddeviatie beneden het gemiddelde van de populatie, werden binnen de groep jonge volwassenen met hydrocefalus stoornissen gevonden in alle cognitiedomeinen, maar vooral op de gebieden van executief functioneren, reactiesnelheid en (verbaal) leren. Bij 70% van de jonge volwassenen betrof de stoornis één of enkele specifiek(e) domein(en). Dertig procent had een globale, dat wil zeggen de meeste of alle onderzochte domeinen betreffende, stoornis. Van de jonge volwassenen met hydrocefalus had 20% een IQ gelijk aan of lager dan 70. Intelligentie bleek in deze groep een goede maat voor het algehele cognitieve functioneren. Lage intelligentie bleek voorts geassocieerd met een hoge frequentie van drain revisies en ook met aan hydrocefalus veel voorkomende pathologie zoals epilepsie en afwijkingen van het corpus callosum.

In hoofdstuk 4 wordt het gevolgde onderwijstraject in primair en voortgezet onderwijs besproken en wordt ingegaan op voorspellers van onderwijstype. Van 178 jonge volwassenen met spina bifida werden de gegevens verzameld over hoe zij het onderwijs doorliepen vanaf de basisschool tot aan het moment van het onderzoek. Het onderwijstraject van jonge volwassenen zonder hydrocefalus was gelijk aan dat van gezonde leeftijdsgenoten. Jonge volwassenen met hydrocefalus waren vaker begonnen op een school voor speciaal basisonderwijs, of stapten ergens in het basisschooltraject over van regulier naar speciaal onderwijs. De personen in deze laatste groep waren vaker meisjes en ze hadden vaker een beperking dan jongeren die in het reguliere basisonderwijs waren gebleven. Ongeveer 60% van de jonge volwassenen met hydrocefalus kreeg aan het eind van de basisschool speciaal onderwijs. Een aanzienlijk deel daarvan stroomde vervolgens in bij het reguliere vervolgonderwijs. Acht procent van de jonge volwassenen met hydrocefalus stopte na de basisschool met alle onderwijs.
Het hoofdstuk rapporteert ook over voorspellers van regulier of speciaal onderwijstype. Het type onderwijs hing met veel factoren samen, maar in een logistische regressie

analyse bleek vooral intelligentie een goede voorspeller. Daarnaast waren hydrocefalus en rolstoelafhankelijkheid voorspellers van onderwijstype.

Hoofdstuk 5 beschrijft de uitkomst voor jonge volwassenen met spina bifida op het gebied van problemen bij de maatschappelijke participatie. De gegevens werden verkregen uit vragenlijsten en interviews van 179 jonge volwassenen met spina bifida. De verzamelde informatie betrof onderwijsniveau, werk, woonsituatie en burgerlijke staat. Daarnaast werden gegevens verzameld over ervaren problemen in relatie tot school of werk, tot bezoek aan vrienden of familie en tot vrijetijdsbesteding. In vergelijking tot gezonde leeftijdsgenoten waren jonge volwassenen met spina bifida vaker lager opgeleid, hadden vaker geen arbeidsbetrekking hebben en woonden minder vaak zelfstandig. Wel hebben ongeveer even veel jonge volwassenen een langdurige relatie als gezonde leeftijdsgenoten. Een slechtere uitkomst was vaak geassocieerd met spina bifida aperta, hydrocefalus, rolstoelafhankelijkheid en incontinentie. Een lager onderwijsniveau was vooral geassocieerd met een lagere intelligentie.
Jonge volwassenen met spina bifida rapporteerden de meeste problemen op het gebied van vrijetijdsbesteding, maar een aanzienlijk deel van de respondenten rapporteerde ook problemen bij schoolgang of werk en bij bezoeken van vrienden of familie. De respondenten relateerden de problemen vooral aan fysieke beperkingen (zoals pijn of vermoeidheid), maar ook aan toegankelijkheid van gebouwen, vervoer over lange afstanden en emotionele problemen zoals schaamte of verdriet. Financiële omstandigheden vormden voor de meeste respondenten geen probleem. Rolstoelafhankelijke respondenten en respondenten met hydrocefalus of met een hoge laesie rapporteerden de meeste problemen vanwege toegankelijkheid van gebouwen en vervoer over lange afstanden. De gerapporteerde problemen waren niet geassocieerd met de ernst van de spina bifida. Kortom, ook personen met een mildere vorm van spina bifida of minder ernstige beperkingen bleken problemen te ervaren bij de maatschappelijke participatie.

Hoofdstuk 6 beschrijft de resultaten over de tevredenheid met het leven van 179 jonge volwassenen met spina bifida. Het doel van de studie was de tevredenheid met het leven te vergelijken met die van gezonde leeftijdsgenoten en te onderzoeken in welke mate spina bifida effect heeft op de tevredenheid met het leven. Uit de resultaten bleek dat 21%, niet meer of minder dan gezonde leeftijdsgenoten, globaal ontevreden is met het leven. Het aantal respondenten dat tevredenheid was met de eigen zelfredzaamheid en

met partnerrelaties was echter lager dan bij gezonde leeftijdsgenoten, terwijl juist meer jonge volwassenen met spina bifida tevreden waren ten aanzien van financiële situatie en familieleven. Bij nader onderzoek binnen de groep jonge volwassenen met spina bifida naar de effecten van leeftijd, geslacht, hydrocefalus en hoogte van de laesie bleek de laatste wel gerelateerd aan een hogere proportie ontevreden respondenten op bovengenoemde domeinen, maar bleek geen relatie tussen spina bifida kenmerken en globale tevredenheid met het leven.

In hoofdstuk 7 wordt ingegaan op de relatie tussen cognitief functioneren en kwaliteit van leven van 110 jonge volwassenen met spina bifida en hydrocefalus. De respondenten gaven aan hoe tevreden en gelukkig zij waren geweest de afgelopen maand en hoe zij hun algehele kwaliteit van leven beoordeelden op het moment van het onderzoek. De gegevens werden gerelateerd aan de uitkomsten op de neuropsychologische tests betreffende de domeinen intelligentie, geheugen, executief functioneren. Om te controleren voor het effect van fysieke beperkingen op de activiteiten van het dagelijks leven is ook de mate van functionele afhankelijkheid meegenomen in de analyses. De jonge volwassenen met spina bifida en hydrocefalus bleken hun kwaliteit van leven te beoordeelden als 70 op een schaal van 0 tot 100. Een matig sterke associatie werd gevonden tussen de scores op de tests van executief functioneren en de ervaren kwaliteit van leven. Er was geen significant verband tussen kwaliteit van leven en de andere domeinen van cognitief functioneren, inclusief de gebruikte maat voor intelligentie. Bij toetsing van een model waarin alle variabelen waren opgenomen bleek dat het verband tussen executief functioneren en kwaliteit van leven bleef bestaan en dat dit verband zelfs sterker was dan het verband tussen functionele afhankelijkheid en kwaliteit van leven. Echter, een lagere subjectieve kwaliteit van leven werd slechts in beperkte mate verklaard door slechter executief functioneren. Dit resultaat toont toch dat men zich bij onderzoek naar gevolgen van cognitief disfunctioneren niet dient te beperken tot intelligentie als algemene maat voor cognitief functioneren, maar cognitie breder moet inventariseren.

Hoofdstuk 8 is de algemene discussie van de resultaten. In dit hoofdstuk worden de belangrijkste conclusies beschreven:

- Veel jonge volwassenen met spina bifida hebben secundaire stoornissen zoals incontinentie, obstipatie, voetmisvormingen, scoliose en verminderde mobiliteit. Bij personen met hydrocefalus en/of een hoge laesie zijn deze stoornissen het meest frequent.

- Personen zonder hydrocefalus functioneren cognitief niet anders dan gezonde leeftijdsgenoten. Het cognitieve functioneren van jonge volwassenen met hydrocefalus is lager dan van gezonde leeftijdsgenoten. Ongeveer de helft van de jonge volwassenen met hydrocefalus heeft specifieke of globale cognitieve beperkingen.
- Personen zonder hydrocefalus volgden een onderwijstraject dat zich niet onderscheidde van dat van gezonde leeftijdsgenoten. Het niveau van laatst genoten onderwijs is bij jonge volwassenen met hydrocefalus lager dan dat bij gezonde leeftijdsgenoten. Slechts de helft van de jong volwassenen met hydrocefalus volgt regulier voortgezet onderwijs. Intelligentie is de beste voorspeller voor type onderwijs, maar ook hydrocefalus en rolstoelafhankelijkheid zijn significante voorspellers.
- De maatschappelijke participatie van veel jonge volwassenen met spina bifida is geringer dan van gezonde leeftijdsgenoten: het onderwijsniveau is lager, de deelname aan het arbeidsproces en de zelfstandigheid van wonen is geringer. Ernst van de spina bifida en in wat mindere mate ook beperkingen van de activiteiten hangen samen met de geringere participatie. Problemen in de participatie vanwege slechte toegankelijkheid van gebouwen en van vervoer over lange afstanden werden meer ervaren door jonge volwassenen die rolstoelafhankelijk waren. Problemen als gevolg van fysieke beperkingen en emotionele problemen werden ervaren en gerapporteerd door alle patiënten met spina bifida, ongeacht de ernst van de spina bifida.
- De globale tevredenheid met het leven van jonge volwassenen met spina bifida is gelijk aan die van gezonde leeftijdsgenoten. De tevredenheid met het leven is echter minder dan van gezonde leeftijdsgenoten als aspecten zoals zelfredzaamheid en partnerrelaties worden beschouwd.
- Executief functioneren houdt verband met hoe gelukkig en tevreden jonge volwassenen met spina bifida en hydrocefalus zijn met hun leven en hoe ze hun kwaliteit van leven beoordelen. Deze samenhang benadrukt het belang van cognitief functioneren als determinant voor uitkomst, naast determinanten als medische status en fysiek functioneren, als mede het belang om niet alleen intelligentie mee te nemen als maat voor algemeen cognitief functioneren.

Dit hoofdstuk beschrijft verder de sterke en minder sterke kanten van de studie. Ook worden implicaties besproken voor de behandeling en zorg van patiënten met spina bifida. Onze studie toont aan dat jonge volwassenen met spina bifida het hoofd moeten

bieden aan een grote verscheidenheid van fysieke en cognitieve functiestoornissen en aan belemmeringen in de maatschappelijke participatie. Ze toont ook dat in geval van condities zoals spina bifida de globale kwaliteit van leven zich niet onderscheidt van die van gezonde gelijken, terwijl ze op relevante onderdelen bepaald verminderd is.
Door de grote steekproefomvang konden de uitkomsten voor belangrijke subgroepen worden beschreven, waaronder de in het verleden vaak vergeten groepen met lichtere vormen van spina bifida zoals spina bifida occulta en spina bifida aperta zonder hydrocefalus. Ook vonden wij het belangrijk om voorheen vaak van onderzoek uitgesloten personen met lagere intelligentie (criterium wordt dikwijls gelegd bij IQ lager dan 70) in de studie op te nemen.
De belangrijkste aan de studie ontleende aanbeveling is om de medische zorg voor jonge volwassenen met spina bifida te continueren in de vorm van multidisciplinaire spina bifida werkgroepen. Een werkgroep zou kunnen bestaan uit een kernteam van revalidatiearts en uroloog met goede mogelijkheden voor doorverwijzen naar andere specialisten. Gespecialiseerde zorg voor jonge volwassenen met spina bifida moet in alle regio's van Nederland beschikbaar zijn.
Tenslotte worden aanbevelingen voor toekomstig onderzoek gegeven. Het is belangrijk om het onderzoek naar het functioneren in brede zin op lange termijn te continueren in longitudinale cohort studies.

# LIST OF ABBREVIATIONS

## List of abbreviations

| | |
|---|---|
| AHC– | spina bifida aperta without hydrocephalus |
| AHC+ | spina bifida aperta with hydrocephalus |
| ANOVA | analysis of variance |
| ASPINE | Adolescents with SPina bifida In the Netherlands |
| BOSK | Dutch patient organisation |
| CI | confidence interval |
| CSF | cerebrospinal fluid |
| FIM | Functional Independence Measure |
| HC | hydrocephalus |
| HLL | high level of lesion; L2 and above |
| HRQOL | health related quality of life |
| ICF | International Classification of Functioning, Disability and Health |
| IQ | intelligence quotient |
| ICD | International Classification of Diseases |
| LiSat | Life Satisfaction Questionnaire |
| LLL | low level of lesion; S1 and below |
| MLL | middle level of lesion; L3 to L5 |
| MQ | memory quotient |
| N | number |
| Occ | spina bifida occulta |
| OR | odds ratio |
| RT | reaction times |
| QoL | quality of life |
| SB | spina bifida |
| SD | standard deviation |
| SPM | Standard Progressive Matrices (Raven) |
| TMT-A | Trail Making Test part A |
| TMT-B | Trail Making Test part B |
| UNKA | test of word production according to lexical rules |
| VAS | visual analogue scale |
| VLT | verbal learning test |
| VRS | verbal rating scale |
| WMS | Wechsler Memory Scale |

# DANKWOORD

Bijna tien jaar geleden begon ik mijn promotieonderzoek op het revalidatiecentrum De Hoogstraat, een periode van mijn leven die ik nu afsluit met een titel en dit boekje. De periode laat zich goed opdelen in een periode óp De Hoogtsraat en een periode ná De Hoogstraat. Het is al even geleden, maar ik kijk nog steeds met veel plezier terug op de eerste periode. Ik heb er veel geleerd en De Hoogstraat was een stimulerende en prettige omgeving. Ik denk dat menigeen, waaronder soms ook ik, twijfelde of er aan de periode ná De Hoogstraat ooit een eind zou komen, maar het is na pieken en dalen wel gelukt. Veel mensen hebben in meer of mindere mate bijgedragen aan een goede afloop en aan een prettige tijd op de De Hoogstraat.

Promotor Arie Prevo, beste Arie, dank je wel voor je steun en toegankelijke begeleiding de afgelopen jaren. Je gaf de moed niet op en bleef informeren en stimuleren, waarbij je ook duidelijk oog bleef houden voor de menselijke kant. Ik heb je steun enorm gewaardeerd.
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De periode ná De Hoogstraat is nu al jaren de periode UMCG. Daar werd ik verwelkomd door de fijne collega's van de 'blauwe patio', namelijk Carien, Anneriek, Annet, Anneke, Marijke, Marjan en Juanita. Dank jullie wel voor de gezelligheid, vriendschap, collegialiteit en inzichten in de vrouwelijke psyche en verzorging. Carien, dank je voor de mogelijkheid om aan het proefschrift te werken als het 'dagelijkse werk' het toeliet. Anneke, wat fijn dat jij mijn paranimf wil zijn!

De (ex)BVL-ers van de 10e verdieping, ik kwam werken op een moment dat BVL zich zou splitsen. Ik ben blij dat het allemaal niet zo'n vaart liep en dat ik jullie toch beter heb mogen leren kennen; niet alleen vanwege de nuttige band met 'centraal' en laatste nieuwtjes, maar vanwege ook de gezellige lunches, uitjes en beroemde quizen.

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# CURRICULUM VITAE

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Hans Barf is geboren op 26 september 1970 in Groningen. Hij volgde het voortgezet onderwijs aan de scholengemeenschap De Waezenburg in Leek en behaalde achtereenvolgens zijn Mavo-, Havo- en Vwo-diploma. In 1990 startte hij met de studie Psychologie aan de Universiteit Utrecht. Tijdens zijn studie was hij een jaar lang parttime student-assistent bij de vakgroep Psychonomie. In 1998 rondde hij zijn studie af, met als afstudeerrichting Cognitieve Functiestoornissen. Aansluitend begon hij een opleidingstraject tot onderzoeker bij het Revalidatiecentrum De Hoogstraat in Utrecht.
De laatste acht maanden van zijn opleidingstraject was hij parttime research-coördinator bij het Revalidatiecentrum De Hoogstraat. Sinds 2003 is hij werkzaam als stafadviseur Patiëntenraadpleging in het Universitair Medisch Centrum Groningen, aanvankelijk bij Bureau Voorlichting van de beleidsstaf en na de reorganisatie in 2007 bij de sector Oncologie.
Hans is getrouwd met Edith Kämink en heeft drie kinderen, Karlijn (1998), Ilse (2000) en Wouter (2002).