Secondary impairments in young adults with spina bifida

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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.

See last page for list of abbreviations.

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 χ^2 to determine differences between subgroups of patients;

*US usage: mental retardation.

$p<0.05$ 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. Seventy-four 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.

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

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

Surgical history	*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	**
Orthopaedic 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, orthopaedic, 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 χ^2 $p<0.05$.
**Significant differences between three subgroups with Pearson's χ^2 $p<0.01$.

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 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 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.

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 in both eyes <0.8	8	0	5		10	5	3	
Epilepsy	9	0	3		13	3	3	*
IQ≤70	20	0	3	**	22	9	6	*
Pain	25	26	35		26	29	26	
Spasticity	13	0	0	*	15	6	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 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 χ^2 $p<0.05$.
**Significant differences between three subgroups with Pearson's χ^2 $p<0.01$.

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)[a]*		*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 χ^2 $p<0.05$.
**Significant differences between three subgroups with Pearson's χ^2 $p<0.01$.

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 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 one-third 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 three-quarters 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 two-thirds 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.

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.

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	**
Faecal incontinence	46	13	8	**	45	34	13	**
Experiencing faecal incontinence as problem	47	30	14	**	47	40	18	*
Obstipation	45	30	35		53	34	29	*
Use of napkin	77	52	30	**	78	68	29	**

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 χ^2 $p<0.05$.
**Significant differences between three subgroups with Pearson's χ^2 $p<0.01$.

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 males	(n=28)	(n=13)	(n=12)		(n=13)	(n=25)	(n=15)	
Problems with erections	29	8	0		39	16	0	*
Problems with ejaculation	39	15	0	*	54	24	0	**
Problems with orgasm	43	8	0	**	54	20	7	*
Sexually active females	(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 χ^2 $p<0.05$.
**Significant differences between three subgroups with Pearson's χ^2 $p<0.01$.

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.

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 one-third 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

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)*	χ^2
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 χ^2 $p<0.05$.

**Significant differences between three subgroups with Pearson's χ^2 $p<0.01$.

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.

DOI: 10.1017/S0012162204000684

Accepted for publication 14th January 2004.

Acknowledgements

We thank Olga de Wit and Maaike Dekker for their contribution in collecting the data. We also thank the BOSK patients' association, the organizations for sheltered homes, and the following centres for their participation in this study: AMC Amsterdam, Franciscusoord mytylschool, Isala clinics Zwolle, LUMC Leiden, Rehabilitation Centre Het Roessingh, Rehabilitation Centre Revalidatie Friesland, Rehabilitation Centre Tolbrug, St Elisabeth Hospital Tilburg, UMC Nijmegen, UMC Utrecht, University Hospital Rotterdam, University Hospital Maastricht, VUMC Amsterdam and Werkenrode. This study was supported by Johanna Children's Fund (Arnhem, the Netherlands) and Stinafo (The Hague, the Netherlands).

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List of abbreviations

AHC^-	Spina bifida aperta without hydrocephalus
AHC^+	Spina bifida aperta with hydrocephalus
ASPINE	Adolescents with Spina Bifida in the Netherlands
HLL	High level of lesion; L2 and above
MLL	Middle level of lesion; L3 to L5
LLL	Low level of lesion; S1 and below