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SHORT COMMUNICATION

PAIN, ESPECIALLY NEUROPATHIC PAIN, IN ADULTS WITH SPINA BIFIDA, AND ITS RELATION TO AGE, NEUROLOGICAL LEVEL, COMPLETENESS, GENDER AND HYDROCEPHALUS

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Study design: **Cohort study.**
Objective: **To investigate the prevalence of neuropathic pain in adults with spina bifida, to study the relationship between neuropathic pain, age at examination, gender, completeness of injury, neurological level and presence of hydrocephalus.**
Methods: **A total of 110 patients with spina bifida who visited the spinal cord injury outpatient clinic Spinalis were included. At the yearly check-up they underwent examination by a physiotherapist and a neurologist and were interviewed about pain character, temporal profile and localization. The patients were divided into 2 groups; spina bifida with ($n = 57$) and without hydrocephalus ($n = 53$). Pain was classified as neuropathic when it was in an area of decreased sensibility and had no correlation to movement and/or inflammatory signs. Results were analysed by χ^2 analysis and Fisher's exact test.**
Results: **Twenty-two patients (20%) experienced nociceptive pain. Neuropathic pain was present in 11/110 (10%) patients, of these 62% experienced below level neuropathic pain. Neuropathic pain was present in 13% of male patients and 7% of female patients, 12% of patients with a lumbar level and 10% of patients with a thoracic level. Neuropathic pain was present in 9% of patients with a complete spinal cord injury, 14% of those with an incomplete spinal cord injury, 1,7% with hydrocephalus and 19% without hydrocephalus.**
Conclusion: **The results suggest that neuropathic pain is present in spina bifida. Careful analysis and classification of a patient's pain is clinically important. Neuropathic pain is more common in patients without hydrocephalus and in older patients. Presence of neuropathic pain was not related to gender, completeness of injury, or neurological level.**

Key words: spina bifida; hydrocephalus; neuropathic pain.

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INTRODUCTION

Spina bifida (SB) is a developmental defect with an incomplete closure of the vertebral column that is usually associated with a similar anomaly of the spinal cord. The rate of SB among newborns in Sweden has diminished gradually, probably due to increased widespread prenatal diagnostic techniques and the use of folic acid (1, 2). SB occurs within the first 6 weeks of pregnancy, possibly caused by a combination of genetic and environmental factors (3, 4). The most common site of SB is the lumbosacral region, but it is sometimes found in the neck.

Patients with SB have a varying degree of paresis in the lower extremities and a higher than usual occurrence of learning disabilities and problems with attention (5–9). Furthermore, patients with SB also tend to be less adaptable, easily distracted, less attentive and persistent, and less predictable.

SB may be associated with other congenital abnormalities, such as hydrocephalus (HC) (4). Approximately 90% of newborns with SB also have HC, an accumulation of fluid in and around the brain caused by the Arnold-Chiari malformation. SB represents a wide range of disability, ranging from patients walking independently without cognitive impairment to those with a complete spinal cord injury (SCI) combined with severe cognitive dysfunction.

Neuropathic pain (NP) is caused by dysfunction in the peripheral or central nervous system without peripheral nociceptor stimulation. Imbalance between pain signal transduction and the pain control system produces NP. Research into NP has been carried out in patients with acquired damage to the nervous system, but not in patients with congenital diseases like SB. In professional rehabilitation science the goal is to achieve and maintain optimal functioning in society (11). In the present study it is therefore more important to study the consequences of NP.

NP is common after both traumatic and non-traumatic SCI, but appears to be rare in patients with SB (12–14). Increasing numbers of patients with SB now survive for longer periods and it is thus important to improve the patient's quality of life. NP is known to have a great impact on daily life. It is therefore essential to study the prevalence of NP in adults with SB and the impact of pain on daily life. To our knowledge there have been no systematic examinations of the nature and prevalence of NP in patients with SB. Thus, the primary aim of the present study was to analyse the prevalence of NP in adults with SB. A secondary aim was to study the relationship of NP with age, gender, occurrence of HC, neurological level, completeness

of the spinal cord lesion, the impact of pain on daily life and when NP started.

METHODS

All adults over the age of 18 years with SB who visited the Spinalis SCI outpatient clinic during 5 consecutive years were included in the study. The Spinalis SCI post-acute unit provides yearly check-ups for patients with traumatic and non-traumatic SCI and for adults with SB living in the greater Stockholm region. The medical records from the paediatric clinic were studied in order to determine whether the patients had HC and if they had been shunted during their first year of life.

At the yearly check-up the patients were examined by a physiotherapist and by the same experienced neurologist, one of the authors (LW), and a thorough neurological and general examination was performed including classification according to the American Spinal Injury Association (ASIA) Impairment Scale (AIS) (10).

AIS A corresponds to complete SCI with no motor and sensory functions preserved in the lower sacral segments. AIS B–D corresponds to an incomplete SCI with various loss of motor and sensory functions. AIS E means normal motor and sensory functions. The examination also included the use of a cotton swab and needles to map sensory disturbances. The examination by the physiotherapist included joint movements.

The interview included analysis of the character, temporal profile and localization of the pain. In order to study the impact of pain in daily life the patients were questioned as to whether the pain was a problem in their daily life and when the pain started. Pain was considered neuropathic according to IASP criteria when burning and shooting in an area with decreased sensibility and with no correlation to movement or inflammatory signs (11).

Furthermore, pain was divided according to neurological level, as above level, at level and below the neurological level.

Pain was diagnosed as nociceptive when in an area with signs of inflammation and/or painful joint movements.

Of 115 patients, 5 were excluded due to other diagnoses, such as myelitis/encephalitis infections post-partum (1 case), tumours in the first year of life (1 case), cerebral paresis type diplegia (2 cases), and traumatic SCI at partus (1 case).

Analysis of data

Groups and subgroups were presented as absolute numbers and percentages. Comparisons between groups and trends were made using χ^2 test, and when the numbers were too small Fisher's exact test was used. A p-value > 0.05 was considered significant.

RESULTS

Patient data

Included in the study were 110 adults (over the age of 18 years) with SB living in the greater Stockholm area. Of the included patients 52 (47%) were male and 58 (53%) female. Most of the included patients were young; the mean age at the time of examination was 28.7 years (age range 18–64 years). Sixty-seven (61%) patients had a lumbar level, and 54% had a complete SCI. Four patients were not testable due to cognitive dysfunction. HC was present in 57 patients (1,7%).

Pain in general

Thirty-five patients (32%) experienced pain. In 22 patients (20%) the pain was classified as pure nociceptive pain. Eight patients had pure NP and 3 patients experienced both NP and nociceptive pain.

Neuropathic pain

In total 11 out of 110 patients (10%) fulfilled the criteria for NP. Two patients who were AIS E described pain with no correlation to movement or inflammatory signs, but they had no decreased sensibility as they were AIS E. Six of the patients with NP (55%) stated that pain was a problem in their daily life. Most of the patients could not remember when their pain started, but it was many years ago. Two of the patients with NP reported that their pain had got worse during the last year due to medical complications such as urinary tract infection and tethered cord.

Four patients experienced at level NP, 1 both at level and below level NP, and 6 patients experienced NP below the neurological level. Demographic data for the 115 patients is shown in Table I.

Age and neuropathic pain

NP was present in 2/65 (3%) of the patients aged 18–29 years at examination, in 3/24 (13%) aged 30–40 years at examination, and in 6/21 (29%) aged 40 years or more at examination. This difference reached statistical significance (Table I).

Gender and neuropathic pain

In the material there was a slight female predominance. Seven out of 52 males (13%) and 4 of 58 females (7%) experienced NP (not significant).

Neurological level and neuropathic pain

Eight of 67 patients (12%) with lumbar level and 3 of 29 patients (10%) with thoracic level experienced NP (not significant).

Completeness of injury and neuropathic pain

Five of 54 patients (9%) with complete SCI (AIS A) and 6 out of 43 (14%) with incomplete SCI (AIS B–D) had NP.

Hydrocephalus and neuropathic pain

One out of the 57 patients (1,7%) with HC and 10 out of the 53 patients (19%) without HC experienced NP. This difference reached statistical significance.

Table I. *Neuropathic pain and its relation to gender, neurological level, ASIA A–E classification, age and the presence of hydrocephalus (HC) in 110 patients with spina bifida*

Gender	Neurological level	AIS A–E	Age at examination, years	Presence of hydrocephalus
Male: 7/52, 13%	Thoracic: 3/29, 10%	A: 5/54, 9%	18–29: 2/65, 3%	HC: 1/57, 1,7%
Female: 4/52, 7%	Lumba:r 8/67, 12%	B–D: 6/43, 14%	30–40: 3/24, 13%	Not HC: 10/53, 19%
			Over 40: 6/21, 29%	

AIS: American Spinal Injury Association (ASIA) Impairment Scale.

DISCUSSION

The main finding of this study was that the prevalence of NP was low in patients with SB.

One of 10 patients with SB experienced NP. NP was more common in patients without HC and in individuals aged over 40 years at examination. Presence of NP was not related to gender, neurological level, and completeness of injury. In this study the patients were examined by the same neurologist and by several physiotherapists. The results of the examination may thus differ with different physiotherapists; however, they were all experienced physiotherapists and used the same manual for examination. The prevalence of NP in patients with SB was low compared with data from studies in traumatic and non-traumatic SCI with a prevalence of NP of approximately 40%. In traumatic SCI NP was more common in individuals injured late in life (13). In non-traumatic SCI age had no influence on the development of NP (14). The low prevalence of NP in patients with SB might be explained by the fact that SB is an early developmental defect that occurs during the first 6 weeks of pregnancy, i.e. related to plasticity in the developing nervous system. On the other hand, NP was more common in older persons. Older patients with SB often have other medical problems, e.g. diabetes mellitus or hypertension, which might influence their pain situation. One might speculate that concomitant disorders may be the background for pain or may facilitate NP due to afferent input.

In a study by Clancy et al. (15) half of the adolescents with SB were found to have pain. Pain was, however, not divided into neuropathic or nociceptive. The higher prevalence of pain in their study is probably due to the fact that patients were aged 9–19 years, and in the present study patients were aged 18 years and above.

This study focused on NP regardless of its intensity. However, it is well-known that many patients with SB have cognitive dysfunction, i.e. problems with memory and concentration and have a lesser ability to learn (5–9). Cognitive dysfunction is believed to have a close connection with HC. However, in a study by Vinck et al. (16) the conclusion was that HC is not the only explanation for the cognitive dysfunction in patients with SB. Cognitive dysfunction, and possibly other cerebral dysfunction, may make it difficult to gain a reliable description of the pain (8). This leads to uncertainty about the description of the pain and about the distribution and exact neurological level of the pain. The fact that in the present study patients without HC experienced NP more often supports this hypothesis.

Furthermore, due to cognitive dysfunction and difficulty in determining an exact neurological level, the AIS impairment scale (10) becomes less adequate. The AIS impairment scale was initially created for traumatic SCI. In the present study we used the ASIA scale because it is well known and has an established validity and, to our knowledge, there is no better scale. However, it also has to be validated for SB patients. In 6 patients NP was found to be a problem in daily life. Two of the patients with NP reported that their NP had got worse during the last 2 years. It is important to classify pain accurately, as treatments differ. When NP is present and is a problem in daily life it is important to determine the cause of the NP in order that a suitable treatment can be chosen and the patient's quality of life improved.

In summary, the results of the present study indicate that NP is not a common finding in SB patients. Neuropathic pain is more common in patients without HC and in older patients. Careful classification and analysis of the pain is of clinical importance.

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