

REGULAR ARTICLE

Health-related quality of life and ambulation in children with myelomeningocele in a Swedish population

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ABSTRACT

Aim: The aim was to study health-related quality of life (HRQL) in Swedish children with myelomeningocele (MMC) with respect to ambulatory function.

Methods: A physical examination of the lower limbs was performed, and occurrence of orthopaedic deformities and shunted hydrocephalus was documented.

A questionnaire on general health-related quality of life Child Health Questionnaire-50 Parent Form (CHQ-PF50) was answered by the parents of 62 children, mean age 12.5 (3.1) years.

Results: The non-ambulatory children had significantly more frequent spasticity in the lower limbs, more often joint contractures as well as hip dislocation or spine deformity compared with ambulating patients. Thirty-two per cent of the ambulators managed without wheelchair use. All non-ambulators were wheelchair users, of which 60% used both a manual and a powered wheelchair. The children with MMC perceived significantly lower HRQL of all subscales of CHQ compared with the healthy control group. Physical function was significantly higher in ambulatory patients, PF = 57.1 compared with 22.2 for non-ambulatory patients.

Conclusion: A Swedish population of children with MMC perceived lower HRQL compared with healthy children, but similar HRQL irrespective of ambulatory function except for the physical domain was reported.

INTRODUCTION

During the last decade, several studies about health-related quality of life (HRQL) in children and adolescents with myelomeningocele (MMC) have been published regarding both European and North American populations. Some authors emphasize improved HRQL by high physical function and independence. Being functionally ambulant as a community or household ambulator and independent with regard to mobility has been reported to be significantly associated with HRQL (1). Persons requiring less assistance for activities of daily living reported themselves to have higher HRQL than those with larger needs of assistance (2). Also children with MMC with reduced functional mobility and self-care experienced lower physical HRQL compared with children with functional ambulation (3). Other authors stress the mental aspects of HRQL, which were found to have no correlation with disability; patients with normal walking or total autonomy presented the same emotional problems as patients with severe disability (4). Furthermore, emotional health in young adults with spina bifida did not differ from the age-matched population without disability (5) and no associations to HRQL and limitations in ADL were reported (6). Authors have also reported level of

parental hopefulness to be essential for mental health in children and adolescents with spina bifida, suggesting caution when attempting to predict an individual child's HRQL based on physical symptoms and signs only (7).

At Karolinska University Hospital, a multidisciplinary team approach to carry on and evaluate a treatment protocol on children with spina bifida has been practised since early 1990's (8). One of the aims is to focus on enhancing the ambulatory function with active, early and individually planned physiotherapy, orthotic support and orthopaedic

Key notes

- Health-related quality of life (HRQL) was reported in a Swedish population of 62 children with myelomeningocele (MMC) by their parents. The non-ambulating children had significantly greater neurological and orthopaedic symptoms than the ambulating children. Similarly, perceived HRQL was found in both ambulation groups irrespective of ambulatory function except for the physical domain, but the children with MMC perceived lower HROL compared with healthy children.

surgery. Facilitation of motor development by sensorimotor stimulation, orthoses-supported stretching exercises in cases of muscle imbalance or presence of joint contractures as well as muscle function-sparing surgery were included. In a previous follow-up study of the active treatment concept in a young patient group (8), it was shown that not all children achieved the team's expectations of ambulatory function according to their muscle function level, if additional neurological deficiencies were present. The orthoses support also deviated from the protocol in cases when major joint contractures and spasticity occurred (8).

Our research has led to a practice comprising of a close analysis of the neurological and orthopaedic status to discriminate the factors important for relevant ambulatory function (9), but more knowledge is still needed to study prognostic factors for ambulation. It is also important to study whether intensive focus on motor development has an impact on other fields of development and social life. Our objectives were to evaluate the current ambulatory level and use of orthoses in a group of patients with MMC treated according to the current protocol and to investigate the perception of their HRQL with respect to their achieved ambulatory functions.

METHODS

Participants

A consecutive series of children with MMC, 6–17 years of age, who were treated at the neuro-orthopaedic department at Karolinska University Hospital, Stockholm, Sweden, between May 2004 and May 2007, were invited to participate in the study. One child who was found to have only meningocele was excluded, and three families refused to participate. One participant died during the study period; thus, 62 children, 29 girls and 33 boys, with a mean age of 12.5 (SD 3.1) years, were included.

Ethical approval for the study was obtained from the Regional Ethical Board at Karolinska Hospital, Stockholm, Sweden. Written informed consent from all participants' guardians was obtained.

Physical examination

Muscle function was defined according to five classes (10) based on manual muscle strength testing of the lower limbs (10). Presence of spasticity (9) was grouped as no spasticity, spasticity in ankle joint muscles only, or in hip and/or knee joint muscles. Joint flexion contractures (11) were defined for ankle, knee and hip.

Information on hydrocephalus, shunt surgery, orthopaedic as well as neurosurgical procedures, dislocation of the hip(s) or spinal deformities and radiographs were obtained from the medical records. The hip was defined as dislocated if the femoral head was totally dislocated, that is with Reimers index of 100%. Spine deformity was defined as either scoliosis with Cobb angle $>30^\circ$ and/or kyphosis $>70^\circ$ or a previously surgically treated spine deformity with no curvature at the present examination.

Ambulation

The children's ambulatory function was classified according to ambulation classes (10) (Table S1). Children who walked indoors but could use a wheelchair for long distances outdoors ($n = 37$) were defined as the ambulation group. Children with some walking function indoors or standing ability ($n = 25$) were defined as the non-ambulation group.

The type of orthoses used for walking and standing and the time of usage were documented: daily use >5 h/day, >2 h/day, <2 h/day or during therapy session or only occasionally. The usage and type of wheelchair were also documented.

Health-related quality of life

A validated questionnaire on general health-related quality of life, Child Health Questionnaire-50 Parent Form (CHQ-PF50) (12), which has also been validated for use in Sweden (13,14), was answered by the parent(s). The questionnaire has 12 subscales, each of which is scored from 0 to 100, with 100 reflecting the best function (Table S1). It also offers an opportunity to calculate a physical (PhS) and a psychosocial (PsS) summary score, with a norm value of 50, SD 10 (12) (Table S1). Results from 60 healthy Swedish children with a similar age span (6–18 years) were used for comparison (13).

The questionnaire was responded by mothers in 43 cases, by fathers in 11, by both parents in seven and by a grandmother in one case.

Statistical analysis

Distributions of variables are given as means, standard deviations (SD) and ranges.

When comparing the two groups, Fisher's exact test for dichotomous variables, chi-square test for non-ordered categorical variables and Mantel-Haenszel chi-square test for ordered categorical variables were used. Mann-Whitney's *U*-test was used for continuous variables.

All tests were two-tailed and conducted at the 5% significance level.

RESULTS

Demographic and clinical characteristics

Results in the entire group of children with MMC and according to the ambulation groups are presented in Table S2.

The included patients had various degrees of muscle strength, with 35% presenting no spasticity. The majority had shunted hydrocephalus (87%) and had previously undergone orthopaedic surgery (84%).

Age of the ambulation and non-ambulation groups did not differ (12.7 and 12.4 years, respectively), whereas need of special school teaching, shunted hydrocephalus, spasticity, joint contractures, hip dislocation and spine deformity occurred significantly more frequently in the non-ambulation than in the ambulation group.

The orthoses usage was equally common in the ambulation groups. The ambulatory children used more often than non-ambulatory children ankle-foot or knee-ankle-foot

orthoses (AFO or KAFOs), which were the most frequently used orthoses, at least 5 h/day. The most common orthoses in the non-ambulating group were Reciprocating gait orthosis and Standing frame, both including the lower limbs as well as the pelvis and trunk.

Twelve ambulatory patients (32%) did not use a wheelchair. Manual and powered wheelchairs were used among all children; a manual more often by the ambulators (56% of wheelchair users) and a powered more often by the non-ambulators (60%).

Participation in spare time physical activities beside sports in school was seen significantly more often in the ambulation than in the non-ambulation group.

Health-related quality of life (CHQ)

Children with MMC had significantly lower scores in all subscales of CHQ compared with healthy children, Figure S1. The reduction in scores varied between 56.7 for physical functioning (PF) to the least reduction of 12.3 for mental health (MH). The physical (PhS) and psychosocial (PsS) summary scores in the MMC group were also significantly reduced, 33.3 and 43.6, respectively, as compared with 55.6 and 53.3 in the healthy control group, $p < 0.0001$ for both (Fig. S2).

Ambulatory children had a significantly better physical function (PF) than the non-ambulatory children (57.1 and 22.2, respectively, $p < 0.001$), Fig. S3. No other significant differences were found in any other subscale of the CHQ. The physical summary (PhS) score was significantly higher in the ambulating group (36.0) compared with those who were non-ambulators (29.3, $p = 0.0102$).

DISCUSSION

The use of quality of life questionnaires as outcome tools reveals insights into the child's emotional, physical and social well-being (12). Self-reported HRQL is considered an important source of information from a child (6). Some of the children in our study did not visit the mainstream school and therefore could not fully answer the questions. Parental reports (CHQ-PF) have been reported to confirm similar outcome in HRQL for children with MMC older than 8 years (7) and was therefore used for retrieving reliable responses representing the child's opinion.

This study group of children with MMC had significantly lower physical as well as mental quality of life in comparison with the healthy control group. This might indicate that the higher morbidity including neuropathic pain, disturbing spasticity and contractures with related immobility in MMC children play a significant role for quality of life, also previously reported (1,3).

The impact of ambulatory function was significantly different between the two ambulation groups only regarding the PF subscale, which measures the presence and extent of physical limitations because of health problems. This finding is in accordance with that of Buffart et al. (15) in their studies on adolescent and young adult MMC-persons. Overall health, illness and resilience (GH subscale) were scored

similarly in both ambulation groups. Intensity and frequency of general pain and discomfort (BP subscale) were scored slightly but not significantly higher in the non-ambulators than in ambulators, possibly reflecting the more intact sensory function in the ambulators. Apparently, in this study group, neither health impression nor bodily pain was significantly affected by the degree of mobility.

The similar scoring of both negative and positive emotional states (MH subscale) and satisfaction with school, athletic ability/outlook, ability to get along with others and family/life overall (SE) indicates that being wheelchair-dependent does not reduce self-esteem. Müller-Godeffroy et al. (6) emphasized that children with less severe impairments may experience more psychosocial problems because of difficulties to identify themselves with either healthy or more severely impaired peers (6). They also proposed that children with MMC perceive their condition as a regular state rather than an illness, which may reflect the same results by the non-ambulating as by the ambulating children in this study. Regarding anxiety and concern with regard to the child's emotional, behavioural or physical well-being, the parents scored similar in both ambulation groups.

A powered wheelchair was used by children in both ambulation groups, but more frequently among the non-ambulators. This fairly high degree of use reflects the access to compensatory technical aids in rehabilitation of today not only in the high neurological levels, but also to enhance existing ambulation, and furthermore to prevent overloading problems in joints (16). Children with mid- and low lumbar lesion levels have increased heart rate during walking (17) and higher walking-related energy cost than peers without disability (18), which supports wheelchair use to enhance participation in society. An energy restoring carbon fibre material in ankle joints can improve walking biomechanics (19); though, their possible benefit of less energy consumption still remains to be evaluated. The present study was performed in a social environment with high acceptance of early wheelchair use, even for children with expected ambulatory function. This fact may partly explain similarly perceived HRQL in ambulators and those with wheelchair dependence.

Children and adolescents with MMC have gone through several surgical interventions, physiotherapy training and efforts to use orthoses with the aim to enable walking and to prevent joint contractures and deformities. Surprisingly, the parents of both ambulation groups scored a similar amount of limitations in personal time because of child's situation (PT subscale). One reason for this might be that orthosis usage is a time-consuming procedure, in particular for children who need orthoses spanning the trunk, hips and knees.

On the basis of comparisons between early gait training versus early wheelchair programme in children matched for age, sex, level of lesion and intelligence, Mazur et al. (20) concluded that patients with thoracic and high lumbar lesion levels should be given an opportunity to walk during childhood and early adolescence, even though very few will continue to walk as adults. Liptak et al. (21) did not find

any difference in activities of daily living related to early treatment programs and suggested a mix of the two regimes. The latter approach seems to be generally accepted in the current rehabilitation practice, as indicated by the similar ratings of HRQL by the ambulators and non-ambulators in all subdomains except for the physical domain.

On the basis of the various perspectives on being ambulatory, a big challenge is to recognize a child's potential ability to achieve functional walking and to set realistic goals. A detailed analysis and understanding of neurological and orthopaedic deficiencies that influence ambulation will help to establish a support level for an efficient ambulation.

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SUPPORTING INFORMATION

Additional Supporting Information may be found in the online version of this article:

Figure S1 Illustration of health-related quality of life as measured by the Child Health Questionnaire for all subscales in the entire group with myelomeningocele and the children in the healthy control group (HC) (mean/SD). Swedish norms from Andersson Gäre et al., *Clin Exp Rheumatol* 2001, healthy controls mean age 12.9 year. For explanations of subscales, see Table S1.

Figure S2 Health-related quality of life for the component summary scores in the entire group with myelomeningocele and the children in the healthy control group.

Figure S3 Health-related quality of life for all subscales as measured by the Child Health Questionnaire in children with myelomeningocele with respect to ambulatory (AMB) and non-ambulatory (Non-AMB) function.

Table S1 Definition of muscle function class, classification of ambulatory function and the physical and psychosocial dimensions of Child Health Questionnaire-50 Parent Form (CHQ-PF50)

Table S2 Demographic and clinical characteristics of the entire group of participants with myelomeningocele and according to ambulatory and non-ambulatory groups. Statistically significant differences are indicated in bold text. Mean (SD)/median (min-max) or n (%)

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