# Quality of life and functional disability in skeletally mature patients with myelomeningocele-related spinal deformity

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**The purpose of the study was to assess the quality of life, physical function, self-motivation, and self-perception of skeletally mature patients with spina bifida and scoliosis. This is a prospective study on 19 skeletally mature patients with a mean age of 21.4 years. Several questionnaires were used for the study: Activities Scale for Kids, Quality of Life in Spina Bifida Questionnaire, The Health Self-Determinism Index for Children, Harter's Self-Perception Profile for Adolescents, and the Spina Bifida Spine Questionnaire. This study found no association between spinal deformity or other features related to spina bifida and self-perception, motivation, and overall physical function. More severe scoliosis affects quality of life and is related to the degree of pelvic obliquity and the age of the patients. Individuals with motor-level dysfunction below L3 had significantly better overall physical function compared with those with a higher level of lesions. This was the only factor found to affect physical function. Our findings suggest that most limitations in patients with spina bifida are not related to the degree of scoliosis but to other associated disabilities.** *J Pediatr Orthop B* **00:000–000 © 2012 Wolters Kluwer Health | Lippincott Williams & Wilkins.**

**Journal of Pediatric Orthopaedics B** 2012, **00**:000–000

**Keywords: function, quality of life, scoliosis, sitting, spine deformity, walking**

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## Introduction

Traditionally, spinal deformity in children with a myelomeningocele has been considered to cause severe disability [1,2]. Surgical treatment of spinal deformity in children with a myelomeningocele is a challenging procedure but can provide good correction of spinal deformity and pelvic obliquity. Surgical treatment is, however, associated with a high complication rate, in particular pseudoarthrosis and deep wound infection [3,4]. In addition, poor soft tissue coverage, associated contractures, loss of sensation, weak bone, and absence of posterior elements have been reported [5,6].

The advantages of surgical treatment are less clear. Rodgers *et al.* [7] have reported that not all children with spina bifida develop problems related to their spinal deformity. In addition, Wai *et al.* [8] found that there is no relationship between spinal deformity and self-perception or physical function in these children. It is our practice to adopt a largely conservative approach in the management of these children and few are referred for surgery.

The main purpose of this study was to assess the quality of life, physical function, self-motivation, and self-perception of skeletally mature patients with spina bifida and scoliosis. We also aimed to determine the relationship of these factors with spinal deformity and other clinical features of spina bifida. We propose that the degree of spinal scoliosis is not necessarily associated with an increased level of functional disability in these patients.

## Materials and methods

This is a prospective study on 19 patients (nine male patients, 10 female patients) presenting to the institution of the first author. All patients had spina bifida cystica, were skeletally mature, and had scoliosis measured on anteroposterior (AP) radiographs (fixed lateral curve more than 20°). The age of our patients ranged from 13 to 35 years (mean 21.4 years). None of our patients underwent operative treatment for spinal deformity. We received ethical approval for the study. Informed consent was obtained from all patients. Only individuals who were able to complete questionnaires, either independently or with parental assistance, were included in the study. Patients who had undergone operative treatment (hip, foot surgery, etc.) 12 months before the study were excluded from the study.

We used methods similar to those previously published by Wai *et al.* [8]; however, for this study, we selected only skeletally mature children and excluded those treated operatively for scoliosis. The degree of scoliosis was assessed on AP radiographs by measuring the Cobb angle [9]. On the same radiographs, pelvic obliquity was measured according to the method used by Osebold *et al.* [10]. Skeletally mature patients were considered to be those who had Risser V on the same AP radiographs.

Neurologic motor level was determined using the International Myelodysplasia Study Protocol [11]. Ambulation was assessed using the four-grade ordinal scale described by Hoffer *et al.* [12] for classifying children with

1060-152X © 2012 Wolters Kluwer Health | Lippincott Williams & Wilkins DOI: 10.1097/BPB.0b013e32835c2a65

spina bifida as community ambulators; household ambulators; nonfunctional ambulators; and nonambulators.

Patients were divided into three groups according to the stability of sitting:

(1) Stable sitting – independent sitting for at least 3 s without any support.
(2) Poor sitting stability – stable sitting only possible with support.
(3) No sitting stability – impossible to sit even with support.

### Questionnaires

Several questionnaires were used to assess different aspects of physical function and quality of life.

To measure the overall physical function, Activities Scale for Kids performance version (ASKp38) was used, as it is a generic measure of overall physical disability and it represents disability that was clinically important to children and their families [13]. ASKp have established reliability and validity for children with physical disabilities [14]. Bagley *et al.* [15] found that ASKp38 could be used to measure the frequency of activity performance on two corresponding subscales: activities of daily living and play.

The Quality of Life in Spina Bifida Questionnaire (QLSBQ) is a tool to assess quality of life specifically in patients with spina bifida and scoliosis [16].

The Health Self-Determinism Index for Children (HSDI-C) was used to determine patients' motivation. HSDI-C consists of twenty items divided over four subscales representing the multidimensionality of intrinsic motivation. The HSDI-C was validated on a sample of 501 children. Factor analysis confirmed the presence of a general factor, conceptualized as overall intrinsic motivation for health, and four separate theory-consistent subcategories. Further construct validity was shown by HSDI-C's ability to discriminate an extreme group of exceptionally motivated children. The reliability of the HSDI-C was shown with good internal consistency and on test–retest correlation [17].

Harter's Self-Perception Profile for Adolescents (HSPPA) self-administered scale was used to determine self-perception. There are seven subscales assessing perceived academic competence, social competence, athletic competence, physical appearance, behavioral conduct, and global self-worth. HSPPA showed a moderate correlation with teacher's global assessment. Factor analysis confirmed a general factor of self-perception and the separations of each subcategory. The reliability of the athletic competence subscale was shown with good internal consistency and on test–retest correlation for a group of children from the school sports teams [18].

We also determined the function of our patients specifically related to the spine using the Spina Bifida Spine Questionnaire (SBSQ). Twenty-five items are formulated into a self-administered questionnaire. Questions are formatted into an ordinal five-category adjectival scale with scores for each item ranging from 0 (I can't do it myself) to 4 (Not hard at all). The item scores are summed and converted into an overall score out of 100. The SBSQ has excellent test–retest reliability and validity that was proven on the group of 30 children [19].

Statistical analysis was carried out by a professional statistician using Statgraphics Plus for Windows 5.1. (StatPoint Technologies Inc., Warrenton, Virginia, USA) Statistical analyses of the results obtained from questionnaires (ASK, QLSBQ, HSDI-C, HSPPA, SBSQ) and Cobb angle, pelvic obliquity, or age were carried out using Spearman's rank correlations. Spearman's rank correlations were also used for statistical analysis of Cobb angle and pelvic obliquity or age. The relationship between the Cobb angle, questionnaire results (ASK, QLSBQ, HSDI-C, HSPPA, SBSQ) and ambulation status, neurological motor level, grouped neurological motor level, sex, or sitting stability were calculated using the Kruskal–Wallis test. A *P*-value of less than 0.05 was considered to be statistically significant.

## Results

The results obtained from questionnaires and Cobb angle are presented in Table 1.

Stable sitting was found in 11 patients and poor sitting stability in eight patients. No patients were classified as having no sitting stability. According to the Hoffer criteria, six individuals were classified as community ambulators, one as a household ambulator, and 12 as nonambulators. There were no nonfunctional ambulators in our group. Only two patients had the absence of pelvic obliquity. In 17 patients, the obliquity ranged from 5 to 20° (mean 11.6°). During clinical examination, motor-level dysfunction was found as follows: T12 ($n = 7$), L1 ($n = 4$), L2 ($n = 0$), L3 ($n = 2$), L4 ($n = 4$), and L5 ($n = 2$). For a more detailed analysis, patients were divided into two groups according to dysfunction of the motor level: group 1 (neurologic level T12–L3) and group 2 (neurologic level L4–L5). Statistical relationships between demographic data, Cobb angle, selected clinical features related to spina bifida, and the results obtained from questionnaires are presented in Table 2.

## Discussion

Scoliosis is common in children and adolescents with spina bifida and affects around 50% of patients [20]. Furthermore, several other clinical features may be a challenge for patients and their physicians. Two-thirds of our study population were classified as nonambulators. We found that all except two of the children with scoliosis

**Table 1 Basic statistics obtained from questionnaires and the Cobb angle**

| | The Health Self-Determinism Index for Children [12] | Harter's Self-Perception Profile for Adolescents [13] | QLSBQ [11] | SB spine questionnaire [14] | ASK [10] | Cobb angle (deg.) |
|---|---|---|---|---|---|---|
| Mean | 77.4 | 110.4 | 193.3 | 0.37 | 44.6 | 77.5 |
| Median | 76 | 112 | 199 | 0.37 | 41.4 | 70 |
| SD | 11.1 | 17.8 | 29.1 | 0.1 | 15.9 | 28.6 |
| SE | 2.5 | 4.1 | 6.7 | 0.02 | 3.6 | 6.6 |
| Minimum | 60 | 72 | 107 | 0.13 | 11.2 | 30 |
| Maximum | 105 | 160 | 232 | 0.56 | 78.9 | 120 |

ASK, Activities Scale for Kids; QLSBQ, Quality of Life in Spina Bifida Questionnaire; SB, spina bifida.

**Table 2 Statistical relationships between demographic data, Cobb angle, selected clinical features related to spina bifida, and results obtained from questionnaires**

| | The Health Self-Determinism Index for Children [12] | Harter's Self-Perception Profile for Adolescents [13] | QLSBQ [11] | SB spine questionnaire [14] | ASK [10] | Cobb angle (deg.) |
|---|---|---|---|---|---|---|
| Age | 0.4 | 0.2 | −0.2 | −0.2 | 0.1 | 0.6[a] |
| Sex | 0.6 | 0.5 | 0.5 | 0.03[a] | 0.4 | 0.7 |
| Neurologic motor level | 0.9 | 0.1 | 0.8 | 0.3 | 0.1 | 0.7 |
| Motor level L3 | 0.6 | 0.7 | 0.5 | 0.02[a] | 0.03[a] | 0.6 |
| Ambulation status | 0.8 | 0.9 | 0.9 | 0.5 | 0.6 | 0.3 |
| Sitting stability | 0.7 | 0.9 | 0.4 | 0.038[a] | 0.4 | 0.2 |
| Pelvic obliquity | 0.1 | −0.2 | −0.3 | −0.3 | −0.3 | 0.7[a] |
| Cobb angle (deg) | 0.2 | −0.05 | −0.5[a] | −0.1 | −0.1 | |

ASK, Activities Scale for Kids; QLSBQ, Quality of Life in Spina Bifida Questionnaire; SB, spina bifida.
[a]Clinically significant correlations. Motor level L3 – patients were divided into two groups: group 1 (neurologic level T12–L3) and group 2 (neurologic level L4–L5).

in our study had pelvic obliquity, and eight of 19 had poor sitting stability. Other consequences of scoliosis include trochanteric sitting or skin problems manifesting as pressure sores. We found that the degree of pelvic obliquity was positively related to the severity of scoliosis, as measured by the Cobb angle. In addition, we found that older children tended to have more severe scoliosis compared with younger ones, and these findings may confirm progression of spine deformity after skeletal maturity. Despite previous findings that scoliosis is more common at higher neurologic levels [20], our results show no correlation between the severity of scoliosis and motor level. This is probably related to the selection criteria used for our study group. Our study included patients from the more significantly affected end of the spectrum of those with this condition.

It has been proposed that patients with more severe scoliosis will have more functional limitations [1,2]. Our analysis showed that although more severe scoliosis affects the quality of life (measured using QLSBQ), there was no correlation between the magnitude of scoliosis and the overall physical function, self-perception, and self-motivation. Our findings suggest that limitations in patients with spina bifida may not be related to scoliosis but to other associated clinical features of the condition.

Despite the fact that a variety of other disorders related to spina bifida are well understood, the relationship of spina bifida with the overall physical function and quality of life in skeletally mature patients has not been reported before. We found that individuals with motor-level dysfunction below L3 had significantly better overall physical function compared with those with a higher level of lesion. This was the only factor found to affect physical function and is likely to be related to the ability to walk in individuals with lower neurological involvement. Flanagan *et al.* [21] have reported previously that neurologic level and the presence of a shunt are related to quality of life in the opinion of parents. We were unable to find an association between any factors examined in our study and patients' self-motivation or quality of life. Several of our findings were in agreement with those of Wai *et al.* [8], who found no significant relationship between any aspect of spinal deformity with self-perception and overall physical function as measured by the ASK. However, major differences between the studies were a younger age of patients in the Wai and colleagues' study and the inclusion of those treated both operatively and conservatively [8].

Can we improve quality of life in children with spina bifida with surgery? Mazur and colleagues found considerable improvement in sitting balance after spinal fusion, but walking ability was affected in two-thirds of children. The best results in terms of sitting balance were obtained in the patients after anterior and posterior fusions, and ambulation status was adversely affected in this group [22]. In addition, Wai *et al.* [8] concluded that surgical treatment may at best improve sitting balance. Garg *et al.* [23] reported that kyphectomy improves sitting and skin problems in patients with myelomeningocele. Osebold [10] confirmed the fact of affected walking ability after surgical treatment of scoliosis. The complication rate of surgical procedures may

affect up to half of the patients [4]. Mercado and colleagues have proposed recommendations for treatment of children with neuromuscular scoliosis. They analyzed 198 publications and on the basis of a systematic review of the literature they concluded that spinal fusion improves quality of life in patients with muscular dystrophy and cerebral palsy, but not in spina bifida patients (grade C recommendation) [24]. Furthermore, Wright, on the basis of an evidence-based review, stated that the benefits of scoliosis surgery are uncertain. If the surgery is performed, both the anterior and the posterior approach should be used; in addition, all-pedicule surgery may be effective [25].

### Conclusion

This study failed to find a relationship between spinal deformity or other clinical features related to spina bifida and self-perception, motivation, and overall physical function. Our findings suggest that more severe scoliosis may affect quality of life in skeletally mature patients.

## Acknowledgements

### Conflicts of interest

There are no conflicts of interest.

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