# **Original** Article

# Assessment of health status in children with spina bifida

C Rendeli\*<sup>,1</sup>, E Ausili<sup>1</sup>, F Tabacco<sup>1</sup>, P Caliandro<sup>2</sup>, I Aprile<sup>2,3</sup>, P Tonali<sup>2</sup>, E Salvaggio<sup>1</sup> and L Padua<sup>2,3</sup>

<sup>1</sup>Spina Bifida Center of the Paediatric Department, Università Cattolica, Rome, Italy; <sup>2</sup>Institute of Neurology, Università Cattolica, Rome, Italy; <sup>3</sup>Istituto Don C. Gnocchi Fondazione Pro-Iuventute, Rome, Italy

**Study design:** Prospective multidimensional study by means of: (1) clinical assessment, (2) parental-administered questionnaire for general health (CHQ-PF50), and (3) standardised disability measurements.

**Objectives:** To assess the health-related quality of life (QoL) and disability in children with spina bifida (SB) and to correlate them with the clinical picture and our previous study on adolescents with SB.

Setting: SB Centre at a University Hospital in Italy.

**Methods:** A total of 29 consecutive children with SB (mean age 11.4, range 4–14 years) were evaluated through Child Health Questionnaire Parental Form (CHQ-PF50), the FIM instrument, and the Barthel index.

**Results:** Disability was inversely related only (r=0.49; P=0.007) to the physical aspect of the QoL of children. Similarly, the disability was inversely related (r=0.37; P=0.005) to the emotional aspect of QoL of patient's parents. Unexpectedly, for the mental aspects of QoL of patients, major disability was not associated with higher psychological distress and severe role disability due to emotional problems. At clinical examination, findings especially for continence and number of catheterisations were usually related to deterioration of physical aspects of QoL (r=-2.28; P=0.02) in children.

**Conclusion:** The multiperspective assessment showed that there is a linear inverse correlation between disability and QoL in children with SB only for physical aspects. Conversely, there is linear inverse correlation between disability and QoL in patient's parents regarding only emotional aspects. Moreover, this study provided useful information for clinical practice underlining that continence problems are those that most affect QoL in children with SB and their parents.

Spinal Cord (2005) 43, 230-235. doi:10.1038/sj.sc.3101707; Published online 18 January 2005

Keywords: spinal dysraphism; outcome research; disability; quality of life; CHQ-PF50

#### Introduction

Since the life expectancy of patients with spina bifida (SB) has increased over the last decades, studies on quality of life (QoL) have become necessary and essential<sup>1</sup> especially because SB involves the general status of the patient.<sup>2,3</sup> Children with SB, because of loss of motor function related to the primary lesion, are restricted in the performance of daily activities such as standing, ambulation and the voluntary control of bladder and bowel functions. Consequently, these neurological deficits may greatly impair the QoL of patients and objective information is scarce. Moreover, it has recently been suggested that more widespread use of standardised health measures may improve clinical practice.<sup>4–8</sup>

In order to assess the QoL in children with SB and to correlate it with conventional disability measures and clinical picture, we performed a prospective multidimensional study, according to the outcome research roles, by employing the most used general health status questionnaire for children (the Child Health Questionnaire-CHQ) and validated conventional clinical and disability measurements.<sup>9</sup> The results presented here examine the relationship between aspects of physical disability as experienced by an Italian sample of children with SB and their QoL as reported by parents.

#### Methods

We enrolled 33 patients with SB aged between 5 and 14 years of approximately 350 patients referred to the Spina Bifida Center of the Catholic University of Rome.

<sup>\*</sup>Correspondence: C Rendeli, Spina Bifida Center of the Paediatric Department, Università Cattolica, Largo F Vito 1, 00168 Rome, Italy

Four patients were excluded from the study because their parents did not give permission to the study. Therefore, we enrolled 29 consecutive patients (mean age 11.4 ranging from 5 to 14 years). Patients and their parents were exhaustively informed about the study and they gave written permission.

### Personal data and clinical examination

Before examination, we acquired personal data by asking patients or their parents, if the patient was younger than 10 years, to fill in a case form. Questions specifically concerned the following issues: educational level (eg, school attendance, presence of assistant teacher for handicapped), urological aspects (incontinence, urethral catheterisation, self-made catheterisaurine loss between the catheterisations, tion, pharmacological therapy). Medullar and brain magnetic resonance imaging were acquired (eg, site of lesion, type of lesion). Detailed clinical history, careful clinical examination (from the neurological and physical rehabilitation point of view) were always performed (eg, tendon reflexes of the four limbs, cutaneous sensitivity, muscle strength assessed according to the British Medical Research Council scale, joint function, trophism, dysmetria).

## Patient-oriented evaluation

The Child Health Questionnaire (CHQ-PF50) is a generic health-related quality-of-life (HRQL) tool that measures the physical and psychosocial well-being of children with or without disability above the age of 5 years and consists of domains representing the most essential components of a child's HRQL. The CHQ-PF50 is a parent-completed questionnaire. The Official CHQ-PF50 cross culturally adapted and validated Italian version, was administered to patient's parents in agreement with standardised methodologies.<sup>10</sup>

CHQ-PF50 is a disease-specific measure that consists of 50 items covering the general health status of patients with the purpose of being a quick and easy HRQL measure used in conjunction with other more functionally based outcome measures, to assess the effects of intervention. This questionnaire provides 15 specific categories of physical and emotional scores (four specific categories regarding patient's parents: parental time -PT, parental emotional – PE, family activity – FA, family cohesion – FC and others concerning children: physical functioning – PF, role physical – RP, bodily pain – BP, global general health – GGH, general health – GH, change health – CH, role emotional behaviour – REB, behaviour emotional – BE, mental health – MH, global behaviour emotional – GBE, self emotional – SE) summarised into two main scores: physical score (PhS) and psychosocial score (PsS). Very low scores for PhS indicate severe physical dysfunction, distressful bodily pain, frequent tiredness and unfavourable evaluation of the health status. Very low scores for PsS indicate frequent psychological distress, and severe social and

role disability due to emotional problems.<sup>11</sup> Higher CHQ-PF50 scores (0–100) indicate better health.

For patient-oriented assessment, the following main measurements were considered: PhS, PsS. In some cases, we also reported the subscore results to better assess the main physical and mental patterns. With regard to the subscores, RP and RE were found to be very important in this study. We have therefore specified the difference between RE, which assesses limitations of various kinds in everyday role activities *due to emotional problems*, and RP, which assesses similar limitations due *to physical health problems*. Note that CHQ-PF50 is normally used for patients within 5 and 14 years of age.

## Disability assessment

To assess the patients' physical disability, we used three common measurements. The Barthel index (BI) is considered the best scale for daily activities measurement. The BI measures performance of personal care (feeding, dressing, hygiene) and mobility (transferring, walking/wheeling) activities. In other words, BI measures what an individual 'can do' providing a measure of capacity.<sup>12,13</sup>

Moreover, we used the functional independence measure (FIM), that is the most widely accepted functional measure.<sup>14</sup> FIM consists of 18 scales scored from 1 to 7; higher numbers mean greater ability. We used the total score (sum of all the scales).<sup>15</sup>

Finally, to evaluate walking ability, we used an adapted form (8-point scale) of the physical therapy portion of the Patient Evaluation Conference System (Deambulation Index).<sup>16</sup>

For disability assessment, the following main measurements were considered: FIM, BI and Deambulation Index.

# Statistical analysis

Statistical analysis was performed using the STAT-SOFT (OK, USA) package.

As ordinal (such as CHQ-PF50) scale was used for measurement, nonparametric analysis of the correlation was assessed by Spearman's rank correlation coefficient; group comparisons were assessed by the Mann–Whitney *U*-test.

# Results

Results from the CHQ-PF50 are reported in Table 1. With regard to ambulation, seven patients proved to be totally autonomous, while three proved autonomous with the help of a tutorial aid, six were autonomous but had some limitations in the velocity of walking while 13 required assistance. Regarding the urinary picture, 18 patients were submitted to urethral catheterisation, one patient had continence but used urethral catheterisation, and three patients had no urological problems. With regard to the 18 patients who were submitted to catheterisation, all patients but two sometimes had

Table 1 Results of CHO	)-PF50
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	GGH	PF	REB	RP	BP	BE	GBE	MH	SE	GH	СН	PE	PT	FA	TOT-PhS	TOT-PsS	FC
Mean	65.3	63.4	86.6	83.3	78.3	76.5	73.8	73.4	76.0	54.1	3.9	61.4	79.7	79.3	39.8	49.5	66.2
SD	18.7	34.8	21.3	24.8	18.5	14.8	21.6	16.2	10.6	12.8	0.9	20.6	26.8	19.9	11.9	8.8	21.4

SD, standard deviation; GGH, global general health; PF, physical functioning; REB, role emotional behaviour; RP, role physical; BP, bodily pain; BE, behaviour emotional; GBE, global behaviour emotional; MH, mental health; SE, self emotional; GH, general health; CH, change health; PE, parental emotional; PT, parental time; FA, family activity; TOT-PhS, physical score; TOT-PsS, psychosocial score; FC, family cohesion



Figure 1 Correlation between site of lesion and Deambulation Index. SD, standard deviation; SE standard error

urine loss between the catheterisations; no patients did self-made catheterisation. A total of 26 patients used a pad (17 of them were submitted to catheterisation). In total, 21 patients had abnormal bowel function, across the spectrum from constipation, through urgency, frequency and occasional accidents and had to use pads or anal plugs and stomas, while eight described normal bowel function.

#### Relationship between lesion and disability/QoL

Site of lesion was related to all the main disability scores used: BI (P=0.018, r=0.44), Deambulation Index (P=0.0007, r=0.59) (Figure 1) and FIM (P=0.003, r=0.54) (Figure 2). Similarly, some QoL measurements were related to the site of lesion: PF (P=0.04, r=0.37), REB (P=0.02, r=0.41), RP (P=0.007, r=0.49). The more proximal the site of the lesion, the higher the disability and the lower the QoL.

Relationship between QoL and disability measurements The Deambulation Index (r = 0.75, P = 0.02) (Figure 3) was related with physical aspects of QoL: PhS (P = 0.007, r = 0.49), PF (P = 0.005, r = 0.50), RP



Figure 2 Comparison of disability between patients with lumbar, lumbosacral and sacral lesions



Figure 3 Comparison between Deambulation Index and TOT-PhS

(P=0.003, r=0.52). Similarly, the Deambulation Index was correlated with parent's life domains: PE (P=0.005, r=0.37), PT (P=0.003, r=0.54). The BI was correlated with physical aspects of QoL: PF (P=0.03, r=0.40), RP (P=0.047, r=0.37) and with parents' life domains:

PE (P=0.02, r=0.43), PT (P=0.007, r=0.49), FA (P=0.03, r=0.41). FIM was correlated with physical aspects of QoL and with parent's life domains too: PF (P=0.04, r=0.37), RP (P=0.047, r=0.37), PE (P=0.02, r=0.45). Unexpectedly, mental aspects of QoL (PsS) were not related with disability measurements.

Conversely, high physical disability was associated with frequent psychological distress, and severe role disability due to emotional problems only in patients' parents (P = 0.01, r = 0.44).

Comparing the physical picture between patients who were supported by an assistant teacher for the handicapped at school and those who were not, we observed a significant difference with higher scores in PhS and RP in patients without an assistant teacher: their mean PhS score  $\pm$  standard deviation was  $45.1 \pm 8.5$  while patients with an assistant teacher had a mean PhS score of  $37 \pm 12.4$  (P = 0.03). Similarly, in patients without an assistant teacher, there is a mean RP with higher scores:  $95.5 \pm 11.5$  versus  $74.5 \pm 26.5$  (P = 0.02). Moreover, patients who were supported by an assistant teacher had higher disability: mean DI score of  $3.6 \pm 1.8$  versus  $5.3 \pm 1.9$  (P = 0.02), mean FIM score of  $76.4 \pm 28.3$ versus 101.6 + 22.1 (P = 0.04). Comparison of the mental picture between patients who were supported by an assistant teacher for the handicapped at school and those who were not, did not show any significant difference. Patients supported by an assistant teacher for impaired children at school had higher disability and lower physical scores of QoL.

As for continence problems, we have observed that continent children had higher scores only for physical aspects of QoL (PhS) (continent children  $52.7 \pm 1.3$ ; noncontinent children  $38.3 \pm 11.7$ ; P = 0.02), but no differences with respect to noncontinent children were noted for psychological and mental aspects. If we analyse parents' involvement, the picture is quite different. Parents with noncontinent children had lower scores of mental aspects of QoL (PE) ( $58.2 \pm 18.9$ ) comparing with continent children' parents ( $88.9 \pm 12.7$ ; P = 0.02) (Figures 4 and 5).

Comparing mental and physical aspects between children who assumed anticholinergic drug and those who did not and their parents, we observed that those who assumed anticholinergic drug and their parents had a deterioration of QoL: mean PsS score of patients who assumed anticholinergic drug was  $46.5 \pm 26.9$  versus  $53.5 \pm 6.7$  of patients who did not assume anticholinergic drug (P = 0.03), mean PE score of their parents was  $53.5 \pm 19.7$  versus  $72.2 \pm 16.7$  (P = 0.02) and mean FA score was  $71.3 \pm 20.7$  versus  $90.5 \pm 12.3$  (P = 0.08). (Figures 6 and 7).

#### Conclusion

SB is among the most common birth defects that results in severe mortality and morbidity rates, with an incidence of 4.7 in 10 000 live births showing marked geographic, ethnic and temporal variations.<sup>15</sup> It consists



Figure 4 Comparison of physical aspects of QoL for ToT-PhS subscale for patients with and without continence



Figure 5 Comparison of mental aspects of QoL for PE subscale for parents who had children with or without continence

of a fault in the spinal column in which one or more vertebrae fail to fuse properly and consequently child's functions depend on the level of the lesion and on the nerve involved. The most common form of open neural tube defect is myelomeningocoele (MMC) developing during the fourth week of gestation characterised by protrusion of meninges through the bony defect. The goal of neurosurgical intervention is to repair the lesion within the first 24 or 48 h after birth and to prevent hydrocephalus with ventriculoperitoneal shunting. After shunt improvements and more aggressive treatment, survival increased resulting in a new generation of adolescents with chronic illness and disability.



Figure 6 Comparison of mental aspects of QoL for PsS subscale for patients who utilise or not anticholinergic drugs



Figure 7 Comparison of mental aspects of QoL for PE subscale for parents of children who utilise or not antic-holinergic drugs

Literature data reported a survival rate of 60% in 1963 which had increased to 90% by 1974.<sup>16</sup> It is estimated that at least 50–70% of children born with SB today will survive into adulthood.<sup>17</sup> Many literature data report that young people with SB are found to be at risk of social isolation and lower self-worth and this is strictly connected to a combination of functional loss, such as walking problems or incontinence, and dependency on others such as care givers and parents.

Traditional outcome assessment in neurological and rehabilitation fields has always been based on physicianderived and instrumental findings and recently, clinical researchers have underlined the need for a standardised evaluation of concepts such as QoL.<sup>18</sup> Sometimes physicians hypothesise the existence of a strict relationship between patients' QoL and their disability: the higher the disability, the more impaired the QoL.

Studies documenting outcomes in children and young people with chronic health conditions and disabilities are necessary and are a public health priority.<sup>19–22</sup> We performed a study to assess QoL and disability in children with SB, and to correlate it with the clinical picture to underline the importance of measuring the needs of parents and children with disability in order to improve the psychological and social well-being of the child.

In our SB sample, because walking ability is correlated with neurological levels, which is compatible with our previous report,<sup>23</sup> disability is strongly associated with clinical impairment of the proximal muscles of the lower limbs; this is because proximal muscles are mainly involved in walking and generally in the motor functions of the lower limbs. This relationship underlines the importance of clinical examination. The assessment of the severity of the disease through clinical examination, provides a measure of the muscle deficit. It also appears to be related to the disability of the patient.

The pattern of QoL is nonetheless more complex: expectedly, the higher the child's physical disability and severe motor deficit of the lower limbs, the higher the reduction in the physical aspect of QoL. On the contrary, unexpectedly, mental aspects of QoL have no correlation with disability: patients with lower disability (eg normal walking ability or total autonomy), presented the same emotional problems as patients with severe disability. This result partially confirms our previous observations detected on adolescents with SB aged between 14 and 18 years in which we noted that patients with higher disability (unable to walk, sphincterial problems and total necessity of assistance) presented even fewer emotional problems in daily activity than patients with lower disability.<sup>23</sup> In that occasion, we hypothesised that our observations were probably related to the greater feelings of global self-worth and self-esteem in physical appearance that is typical of adolescence.

Regarding the needs of families of children with disabilities we saw that there is a correlation between disability and emotional aspects of parents' QoL. We hypothesise that parents of children with severe physical disabilities have a greater psychological and emotional impairment, based on the uncertain future of their children and the availability of resources. This particular aspect underlines the necessity of transforming family needs into goals for intervention. Interesting data are obtained analysing QoL and necessity of an assistant teacher at school.

In our previous study, we have observed that the presence of an assistant teacher at school is associated with a better mental QoL probably because the assistant teacher may have a positive role for the emotional aspect of QoL.<sup>23</sup> In the current study, we have observed that children with an assistant teacher have more physical impairment as observed in the adolescent sample but children with an assistant teacher have higher physical aspect of QoL.

Moreover, analysing the correlation between QoL and clinical aspects, such as bladder and bowel dysfunction, we observed an interesting aspect in SB children population with respect to the adolescent one. In children, sphincterial problems and related clinical consequences cause a deterioration only for physical aspect of QoL. Conversely, in adolescents with SB we observed that incontinence (more than deambulation) was the most important factor in the deterioration of the mental and emotional aspect of QoL.<sup>23</sup>

We hypothesise that in children because there is still no social involvement, incontinence is seen like a nonrelevant problem; on the other hand in SB adolescent group, incontinence becomes a crucial problem in social life to hide from parents and friends.

With regard to the emotional behaviour and every day limitation in family activities, incontinence, is the most important problem for parents; also the number of catheterisation per day is an important factor for the deterioration of the physical aspect of QoL in children and of the emotional aspect of QoL in parents. We hypothesise that this particular aspect is due to the time that parents must devote to their children every day and everywhere.

Also regarding the assumption of anticholinergic drugs this study has underlined that everyday assumption of oral drugs may affect emotional and physical aspects both in children and their parents. This correlation is probably related to the concept of a possible psychological drug dependence, but more data are necessary to confirm this hypothesis.

With regard to the emotional picture, we believe that parents may greatly influence this aspect of patient QoL. Some studies assess parents' perception of QoL and perception of youth but the relationship is complex and still matter of debate. A study focused on this issue is under way.

Our study showed that there is a linear inverse correlation between disability and QoL in children with SB only for physical aspects. Conversely, there is a linear inverse correlation between disability and QoL in patient's parents regarding only emotional aspects. The data here reported, provided useful information for clinical practice underlying that continence problems and drug administration are the most important aspects of the QoL in children with SB and their parents. They also underline the importance of measuring the needs of children with disabilities in order to improve the physical, psychological and social well-being of the SB children.

#### Acknowledgements

Thanks to Mr Nicola Ruperto and Ms Selena Andrisani for their technical support.

### References

 Andresen EM, Meyers AR. Health-related quality of life outcome measures. Arch Phys Med Rehabil 2000; 81: S30–S45.

- 2 Hobart JC, Freeman JA, Lamping DL. Physicians and patient-oriented outcomes in progressive neurological disease: which to measure? *Curr Opin Neurol* 1996; **9:** 441–444.
- 3 Tonali P, Padua L, Sanguinetti C, Padua R, Romanini E, Amadio P. Outcome research and patient-oriented measures in the multiperspective assessment of neurological and musculoskeletal disorders. *Ital J Neurol Sci* 1999; **20**: 139–140.
- 4 American College of Physicians. Comprehensive functional assessment for elderly patients. *Ann Int Med* 1988; **109**: 70–72.
- 5 Apolone G, Mosconi P, Ware JE. *Questionario sullo stato di salute SF-36*. Guerini e associati: Milano 1997.
- 6 Andresen EM. Criteria for assessing the tools of disability outcomes research. Arch Phys Med Rehabil 2000; 81(Suppl 2): S15–S20.
- 7 Ware JE, Sherborn CD. The MOS 36-item short form health survey (SF-36). I. Conceptual framework and item selection. *Med Care* 1992; **30**: 473–483.
- 8 Ware JE. *SF-36 Physical and Mental Health Summary Scales: A User's Manual.* New England Medical Centre: Boston 1994.
- 9 Andresen EM, Lollar DJ, Meyers AR. Disability outcomes research: why this supplement, on this topic, at this time? *Arch Phys Med Rehabil* 2000; 81(Suppl 2): S1–S4.
- 10 Ruperto N *et al.* The Italian version of the Childhood Health Assessment Questionnaire (CHAQ) and the Child Health Questionnaire (CHQ). *Clin Exp Rheumatol* 2001; **19**(4 Suppl 23): S91–S95.
- 11 Landgraf JM, Abetz L, Ware JE. *The CHQ User's Manual. Second printing*. Health Act: Boston, MA 1999.
- 12 Guide for the Uniform Data Set for Medical Rehabilitation (Adult FIM). State University of New York at Buffalo: Buffalo, NY 1993.
- 13 Komer-Bitensky N, Mayo N, Cabot R, Becker R, Coopersmith H. Motor and functional recovery after stroke: accuracy of physical therapists' predictions. *Arch Phys Med Rehabil* 1989; **70:** 95–99.
- 14 Mc Donnell GV, McCann JP. Issues of medical management in adults with spina bifida. *Child's Nerv Syst* 2000; 16: 222–227.
- 15 Botto L, Moore CA, Khoury MJ, Erickson JD. Neural tube defects. *N Engl J Med* 1999; **341**: 1509–1519.
- 16 Deyo RA *et al.* Outcome measures for studying patients with low back pain. *Spine* 1994; **19**(Suppl): 2032–2036.
- 17 Minchom PE *et al.* Impact of functional severity on self concept in young people with spina bifida. *Arch Dis Child* 1995; **73:** 48–52.
- 18 Kozinetz CA *et al.* Health status of children with special health care needs: measurement issues and instruments. *Clin Pediatr* 1999; **38**: 525–533.
- 19 Kirpalani HM et al. Quality of life in spina bifida: importance of parental hope. Arch Dis Child 2000; 83: 293–297.
- 20 Lollar DJ, Simeonsson RJ, Nanda U. Measures of outcomes for children and youth. *Arch Phys Med Rehabil* 2000; 81(Suppl 2): S46–S52.
- 21 Juniper EF. How important is quality of life in pediatric asthma? *Pediatric Pulmonology* 1997; **15**(Suppl): 17–21.
- 22 Wake M, Hesketh K, Cameron F. The Child Health Questionnaire in children with diabetes: cross-sectional survey of parent and adolescent- reported functional health status. *Diabet Med* 2000; **17:** 700–707.
- 23 Padua L et al. Health-related quality of life and disability in young patients with spina bifida. Arch Phys Rehabil 2002; 83: 1384–1388.