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Quality of life of children with spina bifida in Kenya is not related to the degree of the spinal defects

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Summary

OBJECTIVES To assess the relationship between the degree of symptoms and defects caused by spina bifida and the quality of life (QoL) of children with spina bifida in Kenya.

METHODS Children with spina bifida at BethanyKids at Kijabe Hospital, at mobile clinics throughout Kenya and a control group of healthy children were interviewed regarding their QoL. The SEIQoL-DW was used as an instrument to measure the QoL. This instrument can transfer well across cultures by using open questions clustered in six domains chosen based on the study group responses. Symptoms and impairment caused by spina bifida were obtained from the medical files.

RESULTS Sixty-nine per cent of 102 patients with spina bifida had a myelomeningocele, 59% had hydrocephalus, and of 49 incontinent children older than 2.5 years, 67% used clean intermittent catheterization. Quality of life of children was 65.1 on a scale from 0 to 100, compared to 78.0 in the healthy control group. Not one single determinant made a significant difference in the overall QoL. The most important domains were 'development' and 'self-actualization'. The lowest scores were found in the domain 'health and disease'.

CONCLUSION This study showed no significant influence of the degree of defects of spina bifida on the QoL, but identified domains that can be targeted for improvement in children with spina bifida.

keywords quality of life, spina bifida, disability, Africa

Introduction

Spina bifida (SB) is a congenital spinal dysraphism (disorder of fusion of the neural tube). Neural tube defects have an incidence around the world of 0.79–6.39 per 1000 live births (Bowman *et al.* 2009). SB can be divided into spina bifida *occulta* and spina bifida *aperta*. In SB occulta, the defect is covered by a layer of skin and the defect is not visible immediately, which may cause it not to be discovered until it becomes symptomatic, while in SB aperta, the defect is immediately visible. Spina bifida aperta can be further subdivided into myelomeningocele, meningocele and lipomeningocele. The most common form of SB aperta is myelomeningocele, consisting of a sac containing meninges, cerebrospinal fluid, nerves and/or dysplastic spinal cord. Both the brain and spinal cord are often dysplastic, causing additional pathology such as hydrocephalus and Chiari II malformation (McLone & Dias 2003; Kaufman 2004). The spectrum of malformations results in various neurological symptoms, including paralysis and/or spasticity of extremities, urinary incontinence, faecal incontinence and neurocognitive retardation (Shurtleff 2000; Bowman *et al.* 2001; Davis *et al.* 2005). Surgical closure of the spinal defect soon after birth is the standard of management of SB (Charney *et al.* 1991). Complications of SB include renal failure, kyphosis, latex allergy and pressure sores. The medical care of patients with SB therefore is complex and multi-faceted, focused on preventing complications. Clean intermittent catheterization (CIC) is commonly used to prevent renal failure and to control urinary incontinence.

Spina bifida is a chronic disease, and its multiple symptoms and complications are likely to influence the children's quality of life (QoL). In recent years, several studies have investigated the QoL of children with SB in the western world (Kirpalani *et al.* 2000; Cate *et al.* 2002; Padua *et al.* 2002; Lemelle *et al.* 2006). These studies have highlighted the negative impact of mobility limitation, urinary incontinence and level of the lesion on QoL. A study by Bier *et al.* (2005) concludes that 'maximizing

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functional independence should be a priority in improving HRQOL in individuals with myelomeningocele'. Interestingly, however, children with SB rate their own QoL less negatively than the persons treating them (Rasmussen *et al.* 1993; Parkin *et al.* 1997). More surprisingly, a study by Padua *et al.* (2002) concluded that there was 'no linear inverse correlation between disability and QOL in patients with spina bifida', and another study by Muller-Godeffroy *et al.* (2008) confirmed this by showing that medical factors and disability were not significantly related to the health-related QoL. A Dutch study by Barf *et al.* (2007) concluded that SB did not seem to be an important determinant of life satisfaction.

To the authors' knowledge, there are no validated QoL studies of children with SB from African settings. This is important, as social and economic factors would be expected to play a significant role in QoL, thus leading to different results than in Western studies. Consequently, the aim of this study is to assess the relationship between degree of symptoms and defects caused by SB and the QoL of children with SB in Africa.

Patients and methods

Study design and patients

This cross-sectional study was undertaken at BethanyKids at Kijabe Hospital (BKKH) and its several mobile clinics throughout Kenya. BKKH is a faith-based non-governmental referral unit for children with surgical disabilities, primarily spina bifida and hydrocephalus. The management of SB at BKKH is non-selective, i.e. all children are considered surgical candidates regardless of the motor level or presence of hydrocephalus. Children were included in the study if they had a diagnosis of SB and were not older than 18 years when interviewed. During the interviews, it became clear that parents who spend a maximum of 6 months with the SB child were not able to reflect about QoL with SB; therefore, the exclusion criterion was age <6 months.

Methods

All children with SB at BKKH or in the mobile clinics were invited to participate in the study. A control group of healthy children in the same age category was recruited from the local community of Kijabe. Parents of the children were given consent forms in English and Kiswahili, and interviews followed the standard consenting process. Preferably, the child was interviewed, though in some cases this was impossible because of the age of the child or because of his/her cognitive retardation. In such cases, the parent of the child was interviewed regarding the child's QoL. Analysis of paired parent–child interviews, taken at the same time, in 23 patients with SB in our study group showed no significant differences in total QoL score nor in the scores of QoL domains. The only significant difference was found in weight, 'health and disease' was significantly more important to parents than children. This finding supports the decision to use both child and parent interviews in the analysis, where the difference in weight is considered in the interpretation of the results. After the interview, further medical data were obtained from the patient's medical file. The study was approved by the Ethics Committee of Kijabe Hospital.

Interview and medical data

For QoL assessment, the Schedule for the Evaluation of Individual Quality of Life-Direct Weight (SEIQoL-DW) tool was used. This is an open method tool, validated and reliable to detect the aspects of the individual QoL (Moons *et al.* 2004), consisting of three phases. In phase 1, the patient or his/her parent was asked to name the five most important areas in the patient's life. This was an open question, not a multiple choice one, and they could name anything they found important. In phase 2, subjects were asked to score these five areas, using a visual analogue scale from 0 to 100 – 0 being 'it could not be worse' and 100 being 'it could not be better'. In phase 3, the named areas were ranked and weighted using a coloured rotating sliced disc. The areas were attached to one colour on that disc and were enlarged or reduced based on their importance to the patient. All five areas added up to 100%.

Additional data were extracted from the patients' medical files, including motor level of the SB, presence of hydrocephalus, history of VP shunting and incontinence. A formal classification system was used to score functional urinary incontinence (Lemelle *et al.* 2006), with or without catheter (Table 1).

Table 1 Classification systems for urine incontinence

	Leakage	Urinary incontinence
Incontinent	Permanent	Leakage from continuous to several times a day
	Frequent	Leakage from several times a day to several times a week
Continent	Occasional	Leakage from several times a week to several times a month
	Rare	Leakage less than once a month
	Never	No leakage at all

Clustering

The SEIQoL-DW can transfer well across cultures by using open questions (Joyce *et al.* 2003). For analysis purposes, these were clustered into six domains chosen based on the study group responses: (i) health and disease; (ii) development; (iii) resources; (iv) basic needs; (v) self-actualization; and (vi) social environment. 'Health and disease' included answers such as complications of SB, health, SB symptoms, pain and urinary problems. 'Development' consisted both of physical development (such as being able to walk and grow) as well as psychological and communicative development. 'Resources' consisted of answers such as financial situation and toys. 'Basic needs' included references to food, clothes, shelter, sleep, education and health care. 'Self-actualization' consisted of topics like playing, hobbies, trips, independence and religion. 'Social environment' was defined by topics such as friends, family and environment.

Statistical analysis

Individual scores (0–100) and weights (0–100% = 0.00–1.00) were multiplied, and all five areas received in six clusters as follows: (i) health and disease, (ii) development, (iii) resources, (iv) basic needs, (v) self-actualization and (vi) social environment. Naturally, with this as with any other artificial clustering, two or more named topics may have ended up being placed into the same cluster. These combined topics were then used to calculate a weighted mean score per domain.

Statistical analysis included the Mann–Whitney *U* test, with significance level set at 0.05. For testing within-group differences with more than two categories, the Kruskal–Wallis test was used, with significance level set at 0.05. All statistical analyses were performed using SPSS® v15.

Results

Patients

During the period December 2007–February 2008, a total of 125 children with SB and 64 healthy children, or their parents, were interviewed and their medical charts reviewed. Twenty-three children with SB were excluded because of being younger than 6 months of age (*n* = 20) or missing key information from the chart (*n* = 3). The healthy group comprised 24 parent interviews and 40 child interviews. The group of 102 patients with SB consisted of 63 parent interviews and 39 child interviews. The demographic and other baseline data of the SB study group are listed in Table 2. Myelomeningocele was the most frequent lesion. The percentage of SB children with hydrocephalus was 59%. Of 49 children with documented incontinence, 33 (67%) used CIC.

Table 2 Baseline characteristics of 102 children with spina bifida

	n = 102 (%)
Gender	
Male	56 (55)
Female	46 (45)
Age	
0–2 years	35 (34)
2–6 years	25 (24)
6–12 years	21 (21)
12+ years	21 (21)
Lesion type	
Meningocele	14 (19)
Myelomeningocele	50 (69)
Lipomeningocele	8 (11)
Missing	30
Hydrocephalus	
Yes	59 (59)
No	41 (41)
Missing	2
Movement lower extremity	
Yes	62 (61)
No	39
Missing	1
*CIC use**	
Catheter	33 (67.3)
No catheter	16 (32.7)
*Urine continence**	
Continence	22 (34)
Incontinence	29 (45)
Missing	14 (21)

CIC, clean intermittent catheterization.
*In children >2.5 years, the age they should be continent documented.

Quality of life

Mean SEIQoL-DW of children with SB was 65.1 ± 16.6, on a scale of 0–100. In healthy Kenyan children, this score is 78.0 ± 14.2, predictably significantly higher ($P < 0.001$). Table 3 shows the different important domains of QoL in children with SB and healthy children. In the SB group, 'development' was the most important domain, and it received a fair score (63), although the SB group scored significantly lower than the healthy control group (81) on this domain. Next, in order of importance in the SB group came 'self-actualization', which scored very well (73). 'Health and disease' received predictably the lowest score (51), but it was not given much weight. Next to the significant

Table 3 The six SEIQoL-DW domains with their weight and score in patients with spina bifida

	Spina bifida group (*n* = 102)		Control group (*n* = 64)		*P*-value	
Domain	Mean weight (SD)	Mean score (SD)	Average weight (SD)	Average score (SD)	Weight	Score
Development	24.2% (20.2)	62.9 (26.2)	20.8% (14.9)	80.8 (19.3)	0.386	<0.001
Self-actualization	21.4% (21.0)	72.8 (19.3)	31.0% (25.5)	78.2 (14.6)	0.017	0.202
Social environment	19.2% (20.5)	65.8 (27.4)	17.1% (18.0)	69.7 (31.6)	0.663	0.304
Basic needs	15.8% (19.2)	62.1 (23.6)	17.7% (21.2)	74.6 (22.2)	0.648	0.009
Health and disease	10.8% (16.0)	51.0 (25.2)	6.0% (10.6)	83.8 (15.2)	0.059	<0.001
Resources	8.6% (13.2)	58.0 (26.7)	7.4% (16.9)	74.1 (32.8)	0.141	0.041

SD, standard deviation.

difference of ‘development’ scores between SB and control group, the SB group differed also significantly in scores of ‘health and disease’, ‘basic needs’ and ‘resources’. ‘Self-actualization’ was significantly more important in the healthy control group.

QoL and SB determinants

Several SB variables were tested for their influence on QoL, both through the overall SEIQoL-DW score and the subscore for each domain. Table 4 shows that none of the variables made a significant difference to the overall QoL, nor to the subscores of the QoL domains ‘development’, ‘self-actualization’, ‘health and disease’ and ‘resources’. The only significant differences were found within the scores of ‘social environment’ and ‘basic needs’, where incontinent children scored higher than continent children.

Discussion

Study design, methods and clustering

The strength of the SEIQoL-DW as an instrument for assessing QoL in our study group is the fact that it translates easily across cultures, as the questionnaire consists of open questions (Joyce *et al.* 2003); it is validated and reliable (Moons *et al.* 2004); and it is a quantitative measurement making it possible to compare different study groups (Joyce *et al.* 2003). A weakness of the SEIQoL-DW is the variety of answers that can be given. In the administration manual of the SEIQoL-DW, the authors caution in interpreting total QoL scores as it is the sum of the products of individual cue scores by cue weights, each of which may vary independently (O’Boyle *et al.* 1993). The large variety of answers needs to be clustered. The difficulty lies in correctly assigning answers that can fit into more than one cluster, or answers that are hard to place into any cluster. For instance, the domain ‘health and disease’ includes replies related to SB, its consequences and complications. Mobility problems, however, although often a consequence of SB, were clustered in the domain ‘development’ rather than ‘health and disease’. Such ambiguities and inconsistencies are inherent to any clustering of real-life replies and must be borne to mind in interpreting the results of the current study. In addition, by using the SEIQoL-DW, there is the pitfall that the interviewer will use suggestive questions in an attempt to understand the interviewee (Joyce *et al.* 2003).

In our analysis, there were no significant differences in total SEIQoL-DW scores between patients and proxies. In another research, there was also no significant difference between the total SEIQoL-DW scores obtained from patients and proxies (Pearcy *et al.* 2008). These findings underline the ability of using a third party in assessing the SEIQoL-DW. However, data published on this subject are scarce.

Patients

The percentage of hydrocephalus in our SB group was 59%, well below the usual range of 78–86% in developed countries (Stein & Schut 1979; Olsson *et al.* 2007; Talamonti *et al.* 2007), but well within the more conservative range of 54–59% reported in African and Asian studies (Alatise *et al.* 2006; Kumar & Singhal 2007). One likely explanation for this geographical difference is a sampling distortion – the Western studies appear to include only myelomeningoceles, while the African and Asian reports include all spinal dysraphisms. Indeed, the percentage of hydrocephalus in our study among the SB subtype myelomeningocele group was 81%. As one can read in the above-named studies, our study population is very similar to other studies.

Table 4 Impact of patient variables on overall QoL and QoL domains

	SEIQoL-DW (SD)	*P* (*n*)	Development (SD)	*P* (*n*)	Self-actualization (SD)	*P* (*n*)
(a)						
Gender						
Male	67.9 (15.3)	0.090 (102)	66.8 (25.2)	0.169 (79)	76.3 (15.8)	0.154 (70)
Female	61.7 (17.5)		58.7 (27.0)		68.2 (22.7)	
Lesion type						
Meningo	64.8 (17.7)	0.706 (72)	60.1 (28.4)	0.912 (57)	73.0 (16.8)	0.878 (47)
Myelomeningo	62.8 (17.8)		60.2 (28.2)		69.5 (23.5)	
Lipomeningo	58.7 (62.7)		57.9 (23.6)		68.4 (19.7)	
Hydrocephalus						
Yes	66.6 (16.8)	0.410 (100)	64.8 (26.7)	0.392 (77)	74.2 (19.9)	0.477 (69)
No	63.7 (16.4)		62.7 (23.8)		72.0 (18.8)	
Movement lower extremity						
Yes	64.6 (16.9)	0.870 (101)	65.1 (24.9)	0.539 (78)	71.1 (19.6)	0.328 (69)
No	65.5 (16.2)		58.9 (28.5)		75.2 (18.7)	
*Urine continence**						
Continence	63.1 (19.3)	0.494 (51)	69.1 (27.1)	0.367 (43)	68.8 (21.3)	0.489 (39)
Incontinence	67.2 (15.2)		62.1 (27.5)		74.9 (14.9)	

	Social environment (SD)	*P* (*n*)	Basic needs (SD)	*P* (*n*)	Health and disease (SD)	*P* (*n*)	Resources (SD)	*P* (*n*)
(b)								
Gender								
Male	67.6 (27.0)	0.600 (70)	64.4 (22.5)	0.339 (56)	53.9 (21.2)	0.454 (41)	60.8 (29.1)	0.613 (38)
Female	64.1 (28.0)		59.0 (25.2)		45.6 (31.8)		55.5 (24.8)	
Lesion type								
Meningo	75.2 (16.5)	0.829 (46)	65.5 (15.4)	0.283 (43)	59.3 (25.0)	0.119 (29)	67.3 (30.2)	0.470 (26)
Myelomeningo	67.4 (24.7)		64.3 (23.0)		49.7 (20.3)		51.3 (27.6)	
Lipomeningo	57.2 (37.2)		35.3 (34.5)		22.5 (3.5)		57.5 (24.7)	
Hydrocephalus								
Yes	66.5 (26.7)	0.975 (69)	63.2 (23.9)	0.629 (56)	56.7 (19.9)	0.066 (40)	51.3 (24.6)	0.130 (37)
No	65.0 (29.6)		59.4 (23.5)		42.5 (30.1)		65.7 (28.4)	
Movement lower extremity								
Yes	63.5 (28.8)	0.457 (70)	62.6 (23.5)	0.793 (55)	49.4 (29.7)	0.500 (41)	58.5 (26.3)	0.889 (38)
No	68.4 (25.9)		61.0 (24.5)		53.6 (16.5)		56.9 (28.7)	
*Urine continence**								
Continence	46.7 (32.6)	0.036 (36)	48.6 (26.1)	0.019 (20)	51.9 (32.0)	0.500 (21)	55.1 (28.0)	0.573 (14)
Incontinence	68.3 (25.3)		75.0 (13.6)		42.2 (26.6)		64.8 (19.5)	

SD, standard deviation.
*In children >2.5 years of age with or without catheter.

SB determinants

Several SB determinants were purposefully omitted in the baseline Table 2, because there were too few data for reliable statistical analysis. Examples of such omitted variables include shunt infections, shunt malfunction, shunt revision and use of continence drugs. Some of these factors may well play a role, but a larger sample size would be required for their analysis. Some determinants were purposefully omitted in the baseline Table 2, because there was a high co-linearity between the determinants, for example between motor level of lesion and movement lower extremity. The most practical and functional determinants were selected for analysis.

For the determinant 'lesion type', there were 30 missing values. This is a large number and mainly caused by incomplete documentation in the medical files. Another determinant with missing values was 'urine continence', some children were not able to differentiate between continent and incontinent or it was not translated very well, and sometimes, there was no parent available to add information.

Quality of life and SB determinants

A key finding of our study is the significant difference in QoL between children with SB and healthy children. Concerning overall QoL and QoL within the domain of 'health and disease' (comparable to the health-related QoL used in many other studies), not one analysed SB-related variable made a significant difference. As previously mentioned, previous literature supports this finding (Parkin *et al.* 1997; Padua *et al.* 2002; Muller-Godeffroy *et al.* 2008). The 'health and disease' domain had the lowest scores, but this domain was not given much weight. Even so, this domain is more important to the parent than to the child, and therefore appears falsely more prominent in the dominated parent-interviewed SB study group. 'Health and disease', with SB as a part of it, is of little importance for children. This may be a reason why their level of SB has no significant influence on the overall QoL. Medical determinants may not make a significant difference, perhaps because of the fact that the children are born with the disease and its complications and become used to it. Or a sample-size problem may still be at play here. To test this, a power calculation was performed for the variable 'continence', yielding the impractical required sample size of 1741 children.

For every domain, the mean scores were calculated. It is important to keep in mind with interpreting these results that not every interviewee gave answers in every single domain, for example some named three answers in one domain. The number of scores per domain is therefore lower than the total number of interviewed patients with SB and has less power to find significant differences. The only significant differences were found within the scores of 'social environment' and 'basic needs', where incontinent children scored better than continent children. Lemelle *et al.* (2006) concluded that they 'did not find a strong relationship between incontinence and health-related QoL'. However, continent children scoring lower than incontinent children were completely unexpected and are impossible or at least difficult to explain. The numbers of scores per domain were low (36 and 20), thus it is likely that the outcomes are not very reliable. 'Basic needs' is a domain, which has not much interference with continence, but social environment could have.

Assuming although that none of the analysed SB determinants have a significant impact on overall QoL, this study begs the question: What *does* make a significant difference on the QoL of children with SB? 'Self-actualization' as a domain scored best in QoL and second in importance. This in fact could be one reason why children with SB appear to have a good life. This domain includes playing, hobbies, trips, independence and religion. Often children with SB can play very well, have hobbies and do many things, which give them joy in life. It is very important for them to have peers and family who recognize their ability in this area, as this may improve their score in other domains such as 'social environment'. As children with SB are able to share with others the very activities that improve their QoL, their overall QoL also increases.

The 'development' domain, which received average scores, was the most important domain for SB children. This domain consists of items such as physical development, ability to walk and grow, and psychological and communicative development. The SB group scored significantly lower on this domain than the healthy control group. Acceptance and stimulation of the child by the people around him/her can play a significant role in the psychological and communicative development. Such acceptance comes from community and caregiver education regarding SB and its consequences. Indeed, a widespread education process – as initiated by our institution – can make a long-lasting impact on the QoL and future of children with spina bifida.

Conclusion

Spina bifida is a disease with many symptoms and potential complications. The QoL of children with SB is 65 compared to 78 in the healthy control group. As found in Western studies, no SB-related clinical factor, including the level and type of spinal lesion, made a significant difference on the QoL of children with SB in our study.

The most important domains of QoL were 'self-actualization' and 'development'. The lowest scores were found in the domain 'health and disease'. 'Self-actualization' was the key domain, which gave the children a better QoL. The most important domain for children with SB was 'development', with significantly lower scores than for healthy children. This domain could be improved by family, caregiver and community education regarding the condition of SB and its consequences.

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