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Self-reported health-related quality of life in children and adolescents with myelomeningocele

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The aim of the study was to investigate self-reported health-related quality of life (HRQOL) in children and adolescents with myelomeningocele (MMC) and to examine the impact of clinical impairments and limitations in activities of daily living (ADL). Fifty patients (28 females, 22 males) between 8 and 16 years of age (mean age 12y 1mo [SD 2y 4mo]) with MMC from three German paediatric centres and their mothers completed standardized measures on HRQOL (KINDL-R) and limitations in ADL (Childhood Health Assessment Questionnaire). Lesion level was thoracic in nine, sacral in 25, and lumbosacral in 11 patients. Twenty-one patients were community walkers, 17 were able to walk in the home, and seven used a wheelchair. Two-thirds had health problems related to the central nervous system causing current difficulties (eight patients had a shunt, six had hydrocephalus, and 10 had a tethered cord). Patients in the study group reported diminished overall HRQOL compared with norm data, specifically in the dimensions of emotional well-being, self-esteem, and peer relations. Adolescents reported diminished HRQOL in the dimension of peer relations. Most medical parameters as well as limitations in ADL were not significantly associated with HRQOL. Our findings confirm the results of studies which dispute a linear inverse association between condition severity and HRQOL and emphasize the importance of peer relations in young patients with MMC.

Myelomeningocele (MMC) is a serious congenital condition with an incidence of 1 per 1000 births worldwide; the prevalence in Germany is 2.6 per 1000 children.[1] MMC is caused by incomplete neural tube closure during the third and fourth weeks of gestation and is frequently associated with other health conditions such as hydrocephalus, Arnold-Chiari syndrome, and tethered cord syndrome. Children with MMC experience a variety of orthopaedic, urological, and neurological problems, various degrees of limited mobility, and self-care capability, as well as restrictions in participation which threaten their health-related quality of life (HRQOL).[2] HRQOL is a broad concept, which refers to the assessment of various aspects of health from the patient's point of view and includes physical, mental, and social well-being and functioning.[3]

The results of some studies in children with MMC indicate that self-reports from children differ from proxy-reports (parents, physicians) of their children's HRQOL.[4,5] HRQOL measures should elicit data from the child wherever possible. Self-report measures of HRQOL have been shown to be reliable in children aged 8 years and above when using instruments targeted on the cognitive development of the respective age group.[6]

Two descriptive studies showed that young people with MMC rated their overall HRQOL and their condition-specific QOL high or moderately high.[7,8] Few studies compared children with MMC to either a norm population or a healthy control group. Lindström and Eriksson[9] reported a lower subjective QOL in young people with MMC in five Nordic countries compared with healthy controls. A study of children with MMC and/or hydrocephalus from the UK found that the mean overall QOL score of these patients was similar to that of children with learning disabilities* and much lower than that of children with other chronic conditions.[2]

Several studies in this field investigate determinants of HRQOL, particularly impairments and limitations in activities. Some studies have suggested an inverse relationship between lesion level and HRQOL.[10,11] Others reported no[2,7,8,12] or only partial[13] associations between the type of MMC or lesion level and HRQOL. Other medical aspects such as presence of other chronic conditions or psychological aspects such as parental hope[13] also appear to be associated with HRQOL.[2,13]

Physical impairments may have a direct impact on HRQOL, but also may indirectly affect HRQOL by limiting the child's activities. Limitations in ambulation and self-care activities have been reported to be important determinants of HRQOL in children and adolescents with MMC.[12,14]

Young people with MMC have an increased risk of psychosocial maladjustment, particularly lower self-esteem and more internalizing problems.[15,16] From a conceptual view, the constructs of psychosocial adjustment and HRQOL present some similarities. Both are patient (or proxy) reported outcomes, and both refer to well-being and everyday role functioning. However, the construct of psychological adjustment aims at psychosocial functioning and psychiatric assessment from an expert point of view. The construct of HRQOL is strictly patient oriented and reflects a lay model of subjective health. However, the prevalence of psychiatric problems may well be associated with diminished subjective HRQOL. The relationship has not been investigated in this population.

See end of paper for list of abbreviations.

*North American usage: mental retardation.

In general, studies on HRQOL in young people with MMC showed inconsistent results, which may depend on different definitions of HRQOL, different instruments, different responders, and different age ranges. The impact of severity of the condition and physical limitations has been demonstrated in some but not all studies. The objectives of our study were to investigate the self-reported physical, emotional, and social aspects of HRQOL of children and adolescents (8–16y) with MMC compared with healthy children and adolescents and to identify factors associated with HRQOL. We hypothesized that a higher degree of impairments would be associated with lower HRQOL. We also hypothesized that a higher degree of limitation in activities of daily living (ADL) and a higher degree of emotional and behaviour problems would be associated with lower HRQOL.

Table I: Sociodemographic and clinical characteristics of study sample

Mean age (SD), range 8–16y	12y 1mo (2y 4mo)	
	n (%)	
Sex (*n*=50)	28 females (56.0)	
	22 males (44.0)	
Type of school (*n*=50)	23 primary school (46.0)	
	17 secondary school (34.0)	
	9 special needs school (learning disabilities) (18.0)	
	1 special needs school (physical disabilities) (2.0)	
Family structure (*n*=47)	7 (14.9) single parents	
Parental education		
Mother (*n*=41)	≤10y of school: 31 (75.7)	
	>10y: 10 (24.4)	
Father (*n*=33)	≤10y of school: 22 (66.7)	
	>10y: 11 (33.3)	
Level of lesion (*n*=45)		
Sacral L5, S1/2	25 (55.6)	
Lumbar L3/4	11 (24.4)	
Thoracic L1/2	9 (20.0)	
Operation past 12 months (*n*=43)	12 (27.9)	
CNS-related health problems[a] (*n*=33)		
Hydrocephalus	6 (18.2)	
Shunt	8 (24.2)	
Tethered cord	10 (30.3)	
Chiari malformation	6 (18.2)	
Vision problems	3 (9.1)	
Ambulation (*n*=45)		
Community walker	21 (46.7)	
Household/near environment walker	17 (37.8)	
Wheelchair user	7 (15.6)	
Management of neurogenic bladder (*n*=42)		
Spontaneous control or SCIC[b]	23 (54.8)	
CIC[c] with need of assistance	14 (33.3)	
No bladder control	5 (11.9)	
	Mean (SD)	*% t>70[d]*
CHAQ Disability Index (*n*=50)	1.0 (0.8)	
Emotional/behavior problems (*n*=48)		
(CBCL syndrome scale scores[a])		
Withdrawn	4.4 (3.3)	21.0
Anxious/depressed	5.7 (4.8)	20.9
Social problems	4.0 (3.1)	23.0
Attention problems	5.2 (4.1)	27.2
Aggressive behavior	6.4 (6.2)	10.5

[a]Multiple responses were possible. [b]SCIC, self-CIC. [c]CIC, clean intermittent catheterization. [d]*t*-score >70, 2 SD above mean of the norm population. CNS, central nervous system; CHAQ, Childhood Health Assessment Questionnaire; CBCL, Child Behavior Checklist.

Method

Patients with MMC in the age range 8 to 16 years attending the outpatient services of three German paediatric centres and their mothers were eligible to participate in the study. The criteria for inclusion were age and a diagnosis of MMC. Children and adolescents with cognitive disabilities who were not able to complete self-report questionnaires were excluded from the study. The ethics committees of the participating centres approved the study. Parents received information on the study by mail and gave written consent for themselves and the child. The children were also informed and gave consent to their participation.

Sociodemographic data included questions on the child's age, sex, type of school, parents' marital status, and education.

Clinical data included level of spinal cord lesion, number of operations in the past 12 months, ambulation ability, and management of neurogenic bladder. Physicians reported associated health problems related to the central nervous system (CNS) such as shunt, tethered cord, and Arnold-Chiari malformation if the problems caused current difficulties.

Limitations in ADL were measured with the Childhood Health Assessment Questionnaire (CHAQ).[17] Developed for children with juvenile arthritis, the instrument has also been used to measure disability in MMC.[18] It provides a disability index: higher scores indicating more limitation in ADL. The instrument has shown good internal reliability (Cronbach's α=0.93) and convergent validity.[17]

To measure emotional and behavioural problems, we used the Child Behavior Checklist (CBCL; German version/maternal report).[19] Following critical comments on the applicability of the CBCL to children with a chronic physical condition,[20] we used only five of the eight syndrome scales (withdrawn, anxious/depressed, social problems, attention problems, aggressive behaviour). Scale consistency has been reported between 0.70 and 0.92 for the respective scales (German version). Higher scores indicate more emotional or behaviour problems. German norm data are available from a representative population sample of 1600 children.[20]

HRQOL was measured using the Revidierter Kinder Lebensqualitätsfragebogen (KINDL-R) questionnaire, a generic quality of life measure developed in Germany by Ravens-Sieberer and Bullinger. The internationally well established[3] instrument provides six dimension scores as well as an index: higher scores indicating better HRQOL. The instrument has been psychometrically tested with good internal consistency (Cronbach's α>0.80) and convergent and discriminant validity. Population norms are available from a reference group of 1500 schoolchildren.[21]

Differences in HRQOL scores of the study sample compared with population norms were evaluated using a one sample *t*-test. Results are presented via norm means and SDs and mean differences of the study sample and 95% confidence intervals. We used Cohen's effect sizes to describe the impact of HRQOL differences, with d≥0.2 being classified as small, d≥0.5 as medium, and d≥0.8 as large effect.[22]

The analysis of clinical determinants (independent variables) on HRQOL (dependent variable) was conducted via

analysis of variance (ANOVA) or partial correlation. As age and sex are reported to affect KINDL-R scores,[21] we included age and sex as possible confounders (covariates) in the analysis of clinical determinants on HRQOL.

Results

Eighty families were contacted. Thirty (38%) refused to participate. The main reasons for non-participation were involvement in too many studies, time constraints, and an unwillingness to make an extra appointment.

A total of 50 children and adolescents (28 females, 22 males) and 48 mothers were included in the study. The children's age range was 8 to 16 years (mean age 12y 1mo [SD 2y 4mo]). Of the study participants, 46% of children attended primary school, 18% attended a special school for children with learning disabilities, and one attended a special school for children with physical disabilities. The number of single parents (15%) was comparable to the German population. About one-third of parents (24% mothers, 33% fathers) had received more than 10 years of education (Table I).

Patients were ranked in three groups according to the highest level of the spinal cord lesion: in 25 (56%) patients the lesion was classified as sacral, in 11 (24%) as lumbosacral or lumbar, and in nine (20%) as thoracic. Two-thirds of the sample (66%) had current CNS-related health problems (six [18%] had hydrocephalus, eight [24%] had a shunt, and 10 [30%] had a tethered cord). Twelve patients (28%) were reported to have had a surgical procedure in the previous 12 months. Twenty-one (47%) of the patients were able to walk in the wider community, 17 (38%) were able to walk in the home and nearby environment, and seven (16%) used a wheelchair primarily for ambulation.

Over 50% of our study sample had functional bladder control or were continent using clean intermittent self-catheterization, 14 (33%) children and adolescents needed assistance with clean intermittent catheterization, and five (12%) had no bladder control.

The mean CHAQ disability-index was 1.0 (SD 0.8).

The results from the CBCL scales (mothers' report) displayed high levels of emotional and behavioural problems in our sample. Eighteen children (36%) showed a *t*-score of >70 (2SDs or more above the mean of the norm population) in at least one of the CBCL syndrome scales, thus indicating serious psychiatric problems. At least 20% showed signs of social isolation (withdrawn), anxiety/depression, or social problems, and 27% scored high on attention problems.

Table II displays the KINDL-R scores of our study sample compared with norm data (six dimension scores and total score). Results of children and adolescents are presented separately due to how the norm data were published.[21] Compared with healthy schoolchildren, the patients with MMC (n=33) scored significantly lower in overall HRQOL (KINDL-R total score) with medium to large effect size (d=0.7), and in several dimensions of HRQOL: with medium effect size in the scales emotional well-being (d=0.6), and self-esteem (d=0.5), and large effect size in the friends scale (d=0.8). In the adolescent subgroup (n=17), mean scores were not statistically different from norm data in most scales, except in friends, with much lower HRQOL (d=1.0) in the study sample. Both children and adolescents reported similar HRQOL in the scale physical well-being compared with controls. Females reported significantly (p<0.05) lower physical well-being (mean 70.3 [SD 17]) compared with males (mean 79 [SD 14.5]). Other socioeconomic characteristics were not related to any areas of HRQOL or total HRQOL in our sample.

Surgery in the previous 12 months was associated with lower physical well-being and lower self-esteem (ANOVA, F=4.2, p<0.05; F=5.5, p<0.05). Other medical parameters (CNS-related comorbidity, lesion level, ambulatory status, management of neurogenic bladder) were not associated with any area of HRQOL or overall HRQOL.

The CHAQ disability index was neither significantly associated with the KINDL-R total score nor any KINDL-R scales (partial correlation).

Emotional and behaviour problems (proxy-report by mother) were only weakly associated with patient's self-reported HRQOL. The CBCL anxiety/depression score was associated with the KINDL-R total score and the KINDL-R scales self and school. The CBCL aggressive behaviour score was associated with the KINDL-R total score (partial correlation, r=–0.4, p<0.01). Children and adolescents with higher scores in the CBCL symptom scales reported lower HRQOL.

Table III displays the relation between several parameters

Table II: Health-related quality of life (HRQOL) in children (n=33) and adolescents (n=17) with myelomeningocele

HRQOL	*Children (8–12y)*			*Adolescents (13–16y)*		
KINDL-R	*Norm data*	*Study sample*		*Norm data*	*Study sample*	
scale[a]	*Mean (SD)*	*Mean Difference*[b]	*95% CI*	*Mean (SD)*	*Mean Difference*[b]	*95% CI*
Physical well-being	75.6 (13.6)	–3.6	–10.0 to 2.8	72.8 (15.2)	5.5	–0.6 to 11.6
Emotional well-being	83.0 (11.0)	–7.3	–12.2 to –2.2	79.5 (12.4)	–0.4	–9.6 to 8.7
Self-esteem	66.6 (18.4)	–10.4	–17.9 to –2.9	60.8 (19.2)	–2.0	–15.0 to 11.0
Family	84.0 (13.0)	–6.3	–13.4 to 0.7	77.6 (17.4)	3.3	–4.0 to 10.6
Friends	78.2 (13.3)	–14.5	–22.0 to –6.9	78.3 (12.7)	–19.1	–32.5 to –5.7
School	73.2 (12.6)	–1.2	–8.9 to 6.6	64.4 (13.6)	–2.2	–12.2 to 7.8
Total score	76.8 (8.7)	–7.2	–11.8 to –2.6	72.2 (9.4)	–2.5	–10.1 to 5.1

[a]Higher scores represent better HRQOL. [b]Comparison via one sample *t*-test. KINDL-R, Revidierter Kinder Lebensqualitätsfragebogen; CI, confidence interval.

of physical impairment and CHAQ disability index/KINDL-R total score. Patients were ranked in three levels of severity with regard to CNS-related health problems: lesion level; ambulation; and management of neurogenic bladder. The ranked scores demonstrated a linear relationship between most parameters of physical impairment and self-reported limitations in ADL (CHAQ disability index). Children with more CNS-related health problems (ANOVA, $F=4.5, p<0.05$), higher lesion level ($F=12.8$, $p<0.001$), and more ambulation problems ($F=29.6$, $p<0.001$) reported significantly higher disability in the CHAQ.

There was no statistically significant association between parameters of condition severity and HRQOL. The distribution of the KINDL-R total score indicated a U-shaped rather than linear relation between these variables, with patients with an intermediate level of either recent CNS-related health problems, lesion level, ambulation, or continence reporting somewhat lower HRQOL compared with those with a lower or higher level of severity.

Discussion

In this study, patients with MMC reported poorer HRQOL compared with the norm population. Patients aged 8 to 12 years reported lower overall HRQOL and particularly low scores in the dimensions of psychological well-being, self-esteem, and peer relations. Medium- to large-effect sizes emphasized the clinical importance of these results.

In general, our results confirm other studies which have reported reduced HRQOL in children with MMC.[9,10,12] However, these studies relied on parental reports to assess their children's HRQOL, whereas this study provides children's and adolescents' self-reports on their HRQOL.

Adolescents (13–16y) in our study showed less impact of their condition on HRQOL. This seems remarkable because healthy adolescents tend to report poorer HRQOL compared with pre-adolescent children, while this decline in HRQOL was not seen in this study. Adolescents with MMC did not decline further from the (already quite low) scores reported by children. We cannot explain this result from our data. Apart from the small sample size within the adolescent age group which limits the power of finding significant differences, we speculate that adolescents may have developed more elaborate coping strategies compared with younger children which enable them to adjust more adequately to their condition. Furthermore, transition through puberty is a difficult developmental milestone for all children. Children with MMC may have met the challenge of 'being different' at an earlier age and thus developed a more resilient attitude to the challenge of adolescence.

In adolescence, the individual's focus shifts from family to peers. In our study, adolescents with MMC reported much lower HRQOL compared with norm data in the domain peer relations, with even larger effect size than in the younger age group. Our findings indicate that it can be difficult for young people with MMC to participate in peer activities and maintain friendships.

Interestingly, children and adolescents with MMC did not report impaired HRQOL in the dimension of physical well-being. This may be due to the comparable stability of the condition: children perceive their condition as a regular state rather than an illness.

Our findings indicate that MMC may not affect all domains of HRQOL equally. From the patient's point of view, peer relations seem to be the domain in which HRQOL is particularly poor, independently from the patient's age.

Similarly to the results of other studies, we only found a weak direct association between physical impairments and children's and adolescents' HRQOL. Only surgery in the past 12 months, indicating an unstable episode of the condition, contributed to an explanation of variance in HRQOL.

Our data do not confirm the expectation of a linear inverse association between physical impairment and HRQOL in young patients with MMC. Other studies reported similar results. Rendeli et al.[11] and Padua et al.[23] found an inverse association between severity of impairment and physical aspects of HRQOL, but not between severity of impairment and emotional aspects of HRQOL. Adolescents with *less* physical disability reported even lower emotional HRQOL. While medical parameters often are not significantly related to HRQOL, the association between limitations in ADL and HRQOL is quite a robust finding in the existing literature.[12,14] We found a strong linear association between condition variables indicating severity of impairment and the CHAQ disability index, which indicates the validity of the CHAQ in this clinical group. However, contrary to our expectations we found no association between limitations in ADL as measured by the CHAQ disability index and HRQOL.

Apparently, other factors determine HRQOL in this population. When ranked in three levels of severity with regard to current CNS-related health problems, lesion level, ambulation, and management of neurogenic bladder, distribution of the HRQOL total score indicated that the underlying rela-

Table III: Childhood Health Assessment Questionnaire Disability Index (CHAQ-DI), Revidierter Kinder Lebensqualitätsfragebogen (KINDL-R) Total Score, and clinical variables

	CHAQ-DI (range 0–3)[a] *Mean (SD)*	*KINDL-R Total Score (range 0–100)*[b] *Mean (SD)*
Clinical variables		
Current CNS-related health problems		
No	0.6 (0.4)	72.1 (11.2)
One	0.9 (0.6)	62.5 (8.6)
Two or more	1.2 (1.0)	71.9 (24.8)
Lesion level		
Sacral	0.6 (0.5)	71.7 (12.3)
Lumbar	1.0 (0.8)	63.9 (10.9)
Thoracic	1.8 (0.6)	69.0 (19.7)
Ambulation		
Community walker	0.4 (0.4)	72.9 (9.9)
Household/near environment walker	1.1 (0.6)	64.1 (18.2)
Wheelchair user	2.1 (0.6)	71.0 (18.0)
Neurogenic bladder		
Spontaneous control or SCIC[c]	0.9 (0.8)	71.3 (8.2)
CIC[d] with need of assistance	1.0 (0.7)	66.4 (13.9)
No bladder control	0.9 (0.8)	71.0 (14.9)

[a]Higher scores represent a higher grade of disability. [b]Higher scores represent better health-related quality of life. [c]SCIC, self-CIC. [d]CIC, clean intermittent catheterization. CNS, central nervous system.

tionship between severity and HRQOL might be U-shaped rather than linear. This potential relationship needs to be investigated by further studies and in larger populations.

A psychological line of argument may be given by the marginality concept. Marginality is a situation in which one's self-perception as healthy or disabled is uncertain or ambiguous. Children with less severe impairments may experience more psychosocial problems because they have difficulties with identifying themselves with either healthy or more severely impaired peers.[24] Family research in MMC also supports this hypothesis: mothers of children with a higher degree of physical impairment reported more attachment to the children, less family conflict, and a greater willingness to include them in decision-making processes.[25] Parents of children with high lesion levels may accept more fully the extent of their child's disability compared with parents of children with intermediate level of lesions, who may try to 'normalize' functioning as much as possible and place considerable strain on their child. This aspect may directly concern the child's healthcare providers. The strain of what is expected from the child in way of 'normal' or 'best possible' functioning (e.g. as much upright ambulation as possible) may lead to treatment decisions which can place a substantial burden on the child. Self-reported HRQOL may be an important, and so far unappreciated, source of information within this context.

Restrictions in mobility and functional independence have been defined as key parameters of participation as classified in the International Classification of Functioning, Disability and Health (ICF)[26] and shown to be important predictors of HRQOL in young people with MMC.[13,14] Functional independence may be more easily achieved both by independent walking or using a wheelchair than using braces and being a slow walker needing assistance to move around the community. Questionnaires which measure ADL in young people should cover the dimension of functional independence, even when achieved by using medical devices. The CHAQ does not measure functional independence in this way, and this may be one reason why the CHAQ disability index was not associated with HRQOL in our study.

The results from the CBCL scales demonstrated high levels of psychiatric problems in our sample. More than 20% were rated by their mothers to have serious problems in the CBCL syndrome scales social withdrawal, anxiety/depression, social problems, and attention problems. There was little overlap between the concept of HRQOL from the children's and adolescents' point of view and the more psychopathological concept of emotional and behaviour problems from the mothers' point of view in our sample. These findings underline the distinction between the two concepts and emphasize the importance of using self-report questionnaires for HRQOL assessment.

The main limitations of the study are the lack of a comparison group and the small sample size. Effects tend to be larger compared with norm data than to matched comparisons, and the sample size limits the precision of the results and the statistical power of the tests. We did not collect data on families that were approached but who did not participate in the study; we are, therefore, unable to report on non-responders and potential recruitment bias. The three centres involved in this study are located in large cities and generally provide care for a population of at least 500 000 inhabitants. They are large tertiary care centres with outpatient and in-patient services for children and adolescents with a variety of chronic conditions. The majority of children with MMC in Germany attend such specialized centres, although we have no information about those not attending.

Our data on the distribution of HRQOL scores which displays patients with the intermediate level of condition severity reporting lower HRQOL compared with those with less or more impairments are not statistically significant and have an observational character. However, the results may be of clinical interest and warrant further prospective studies to shed light on the relation between functional independence, mobility, parents' and clinicians' expectations, and HRQOL in children and adolescents with MMC.

Conclusion

Self-reported HRQOL may be a relevant outcome measure in the care of children and adolescents with MMC. Clinical decisions on treatment options should consider the perspective of the patient and aim for independence, a maximum of mobility, and increased opportunities for peer relations.

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List of abbreviations

ADL	Activities of daily living
CBCL	Child Behavior Checklist
CHAQ	Childhood Health Assessment Questionnaire
HRQOL	Health-related quality of life
KINDL-R	Revidierter Kinder Lebensqualitätsfragebogen
MMC	Myelomeningocele