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SYMPOSIUM: MYELOMENINGOCELE

Activity Level, Functional Health, and Quality of Life of Children with Myelomeningocele as Perceived by Parents

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Abstract

Background To provide the best health care for individuals with myelomeningocele (MM), clinicians and researchers need to understand their health and functional status as well as quality of life. The literature is mixed regarding the relationship between motor level and health-related quality of life (HRQOL) for these individuals.

Questions/purposes We compared the HRQOL of children with MM at the L2 and above and L3-5 motor level and to demonstrate how having a shunt, age, and body mass index affect HRQOL instruments for these two motor level groups.

Methods We recruited 50 patients with MM (24 male, 26 female) with a mean age of 11.5 years (range, 5–18 years) and motor levels of L2 and above (n = 15) and L3-5 (n = 35). Guardians were interviewed using standardized functional and HRQOL tools (the Pediatric Quality of Life and the Pediatric Outcomes Data Collection Instrument); height, weight, presence of a shunt, ambulatory level, and body mass index were also collected.

Results We found a decreased HRQOL score for children with MM in the L2 and above motor level compared with those in the L3-5 motor level group. HRQOL had no correlation with body mass index and limited correlation with age. The presence of a shunt correlated with a decreased HRQOL.

Conclusions Children with MM had deficits in HRQOL and that was associated with neurologic level and presence of a shunt.

Level of Evidence Level IV, prognostic study. See Guidelines for Authors for a complete description of levels of evidence.

Introduction

Improvement in quality of life and physical function and prevention of secondary complications should be major goals of medical care for children with myelomeningocele (MM). Across their lifespan, individuals with MM have complex impairments of the musculoskeletal system, skin issues, sensory deficits, obesity, hydrocephalus, bowel and bladder incontinence, depression, and cognitive dysfunction among other deficits [13]. As clinicians, we need to assess the contribution of these impairments to activity limitations and quality of life for individuals with MM.

Multiple studies [4, 6, 15–22] have looked at quality of life for persons with MM using a variety of outcome tools for parents and for youth. Several studies have suggested an inverse relationship [18, 20, 22] between health-related quality of life (HRQOL) and the lesion level, whereas others report no [4, 6, 17] or only a partial [15] relationship between the two. Using the Spina Bifida Health Related Quality of Life Questionnaire, Schoenmakers et al. [21] found

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This work was performed at Shriners Hospitals for Children, Chicago, IL, USA.

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independent mobility was the most important determining factor for HRQOL. Leger [16] used the Quality of Life in Spina Bifida P2 and found subjects had a high level of quality of life, but the number of reported secondary health conditions was concerning. Müller-Godeffroy et al. [17], using the KINDL-R, reported children with MM scored lower in the areas of emotional well-being, self-esteem, and peer relations.

The Pediatric Quality of Life Inventory™ (PedsQL™) [7, 24–27] is a HRQOL measure with 23 items in four Generic Core Scales encompassing Physical/Emotional/Social/School Functioning. This tool was used by Oddson et al. [18] in a study of HRQOL and depressive symptoms in children with spina bifida. Their findings showed a low HRQOL for these children compared with other chronically ill children [18]. Another common instrument that measures HRQOL is the Pediatric Outcomes Data Collection Instrument Version 2.0 (PODCI) [10], which was developed by a joint effort of the American Academy of Orthopaedic Surgeons and the Pediatric Orthopaedic Society of North America. It has 114 questions and can be broken down into four functional assessment scores (upper extremity functioning, transfers and basic mobility, sports and physical function, comfort/pain), a happiness score, and a global function score, which is the average of the four functional assessment scores. Each score has a possible range of 0 to 100. The scores were validated on children with moderate to severe orthopaedic disabilities [8, 10].

In addition to the effect of neurologic level on HRQOL, it is important to understand how demographics such as presence of a shunt, age, and obesity may also affect HRQOL. Children with more than four shunt revisions also have a lower functional level [2, 11, 12, 28]. Transition from childhood to young adulthood can be an especially difficult developmental period for those with MM [14]. Johnson et al. [14] found 72% of teens and young adults with MM had very limited participation in structured activities and most required assistive technology to aid in their mobility. Those with higher neurologic lesions often had associated decreased motor independence [11, 16]. Obesity is a pervasive problem for youth with MM [1, 12, 23] and research has not adequately evaluated whether there is a relationship with HRQOL measures.

The purpose of this study was twofold. First, to compare the scores on the HRQOL tools of children who have MM in the L2 and above motor level group versus those in the L3-5 motor level group. Second, to determine if there is a correlation among presence of a shunt, age, and body mass index (BMI) on quality of life for these two motor level groups.

Patients and Methods

As a sample of convenience, we recruited 50 English-speaking patients aged 5 to 18 years old with T10-L5 myelomeningocele who were seen during outpatient clinic visits at Shriners Hospitals for Children–Chicago from March to August 2008. One hundred ninety active charts from our MM clinic database were reviewed and the first 50 guardians/children who met the eligibility criteria and agreed to participate were given questionnaires. Ten eligible families declined to participate for various reasons. The average age of participants was 11.5 ± 3.9 years with 24 males and 26 females. Additional demographic information was gathered, including age, gender, presence of a shunt, neurologic level, ambulatory level, height, weight, and BMI (Table 1). We obtained prior Institutional Review Board approval and informed consent was obtained for each participant.

Neurologic level was categorized as (1) L2 and above describing a child with absent or less than fair strength of the quadriceps and musculature below on functional muscle testing; or (2) L3-L5 describing a child with normal quadriceps strength and no plantar flexor strength. Ambulatory status was classified into four levels: (1) independent ambulation with no assistive device; (2) walking full-time using an assistive device; (3) walking with an assistive device at home but uses a wheelchair in the community; and (4) uses a wheelchair full-time. BMI was calculated using the formula (www.cdc.gov): weight (pounds)/stature (inches) × stature (inches). BMI was calculated as a percentile and the subject was placed into one of four categories: (1) healthy weight (5th–85th percentile); (2) overweight (85th–95th percentile); (3) obese (greater than or equal to 95th percentile); and (4) underweight (less than the 5th percentile).

Parents of the 50 subjects were interviewed in person during the MM clinic using two HRQOL outcome tools: the PedsQL™ Version 4.0 [7, 24–27] parent version for young children (ages 5–7), children (ages 8–12), and the teen report (ages 13–18); and the PODCI Version 2.0 [10] parent form for children (ages 2–18). Because of concerns about time and length of the instrument, 22% of the parents declined to complete the PODCI questionnaire.

Differences in outcome scores between neurologic levels and between children with and without shunts were determined using Wilcoxon-Mann-Whitney U tests. Correlations between age and BMI with outcome scores were assessed using Pearson correlation coefficients.

Results

Scores on the PODCI and PedsQL™ were lower for those in the L2 and above motor level group versus those in the L3-5 group (Table 2). This was most apparent for the categories of transfers and basic mobility, sports and physical function, global function, and health.

Table 1. Demographics for subjects with myelomeningocele (n = 50)

Demographics	Category	Frequency	Percent
Gender	Male	24	48%
	Female	26	52%
Neurologic level	L2 and above	16	32%
	L3-5	34	68%
Ambulation level	Walks full-time with no assistive device	10	20%
	Walks full-time with an assistive device	21	42%
	Assistive device at home and wheelchair in community	5	10%
	Full-time wheelchair use	14	28%
Shunt	Shunt	44	88%
	No shunt	6	12%
Body mass index category	Healthy weight (5–85th percentile) L2 and above	8	16%
	Healthy weight (5–85th percentile) L3-5	10	20%
	Overweight (85–95th percentile) L2 and above	3	6%
	Overweight (85–95th percentile) L3-5	11	22%
	Obese (> 95th percentile) L2 and above	5	10%
	Obese (> 95th percentile) L3-5	13	26%
	Underweight (< 5th percentile) all children	0	0%

Table 2. HRQOL for different neurologic levels of children with myelomeningocele

Outcome questionnaires	Maximum scores	Means for L2 and above	Means for L3-5	p Value
PODCI subscales				
Upper extremity and physical function	100	84.17	87.16	0.2582
Transfers and basic mobility	100	37.67	75.32	0.0064*
Sports and physical function	100	15.33	47.80	0.0047*
Comfort	100	88.00	81.76	0.2595
Happiness	100	81.67	74.80	0.4825
Global function	100	55.50	73.08	0.017*
PedsQL™ quick scores				
Health	800	321.88	445.59	0.0512*
Feelings	500	348.44	350.00	1.0000
Getting along	500	353.13	304.41	0.0734
School	500	254.69	301.47	0.1314
Total	2300	1278.13	1401.47	0.2622

* $p < 0.05$; HRQOL = health-related quality of life.

Those subjects who had the presence of a shunt, older age, and higher BMI scored lower on the PODCI measures of transfers and basic mobility, sports and physical function, happiness, and global function and symptoms (Table 3). There was no association among the PedsQL™ scores and presence of a shunt. We found a negative correlation between age and sports and physical function scores on the PODCI with older subjects having lower scores (Table 4). There was no association between HRQOL measures and BMI or BMI risk category (Table 4).

Discussion

Individuals with MM have multiple medical issues, impairments, decreased activity levels, and reduced functional health throughout their lifespan and require intervention from a multidisciplinary medical team. The literature is mixed regarding a positive or negative relationship between motor level and HRQOL for these individuals and what demographics may affect quality of life. The dual purpose of this study was to first to compare

Table 3. HRQOL scores for subjects with and without a shunt

Outcome questionnaires	Shunt (n = 44)	No shunt (n = 6)	p Value
PODCI subscales (each out of 100)			
Upper extremity and physical function	85.27	93.40	0.1318
Transfers and basic mobility	64.27	87.60	0.0498*
Sports and physical function	36.65	66.80	0.0375*
Comfort	81.19	92.20	0.4812
Happiness	72.50	95.00	0.0242*
Global function	66.77	84.80	0.0232*
PedsQL™ quick scores			
Health (out of 800)	391.48	512.50	0.2680
Feelings (out of 500)	344.32	387.50	0.2010
Getting along (out of 500)	320.45	316.67	0.6981
School (out of 500)	289.20	266.67	0.6444
Total (out of 2300)	1345.45	1483.33	0.5729

* $p < 0.05$; HRQOL = health-related quality of life.

Table 4. Correlation of HRQOL scores with age and body mass index

Outcome questionnaires	Age		BMI	
	Correlation	p Value	Correlation	p Value
PODCI subscales				
Upper extremity and physical function	0.2919	0.1117	0.0291	0.8765
Transfers and basic mobility	−0.1971	0.2906	−0.1976	0.2867
Sports and physical function	−0.3772	0.0358*	−0.2543	0.1675
Comfort	−0.0031	0.9870	−0.1683	0.3655
Happiness	0.0793	0.6743	0.1147	0.5388
Global function	−0.1782	0.3404	−0.2216	0.2309
PedsQL™ quick scores				
Health	−0.0484	0.7396	−0.1832	0.2029
Feelings	0.0803	0.5811	0.0247	0.8649
Getting along	0.1445	0.3184	0.0655	0.6516
School	−0.2680	0.0596	−0.2078	0.1475
Total	−0.0531	0.7156	−0.1477	0.3060

* $p < 0.05$; HRQOL = health-related quality of life; BMI = body mass index.

the HRQOL of children who have MM in the L2 and above motor level group versus those in the L3-5 motor level group; and second, to determine if there is a correlation among age, BMI, and presence of a shunt on quality of life for these two motor level groups.

We note limitations to our study. First, although we plan to follow these children over time, this study was limited in that it was only a snapshot in time of their actual characteristics and not longitudinal in design. Second, we interviewed the parents rather than the children and reported only parental perceptions. Although self-report would be the ideal, we chose to have the parents complete the questionnaires so that we could access a better cross-section of participants at varied age, developmental, and cognitive levels. Even with guardian interviews, 22% of the guardians declined to complete the PODCI questionnaire during the clinic visit because of the length of the tool. Third, although the outcome tools were carefully chosen, there were important areas that they were not able to address such as the burden of care to families, socioeconomic status, number of shunt revisions, child cognitive levels, parental education level, and barriers to community participation. Fourth, if we had included youth who had sacral neurologic levels, we may have seen even higher levels of HRQOL for our clinic population of children with MM because they would likely have less medical complications and better functional health. We did not include this less involved population because we were attempting to keep our two groups more homogenous. Fifth, to look at obesity, BMI was calculated using standing or supine

height if the child was not able to stand. Hinderer et al. [12] suggest arm span should be used because the individuals often have a shorter stature. Although many of our subjects had elevated BMI and high BMI risk categories, this may be partly inflated by the fact that many of the subjects had short stature and broader trunks. A more accurate measure of health and cardiovascular risk than BMI would have been to determine truncal body fat with a body impedance assessment device or dual energy x-ray absorptiometry scan [12].

We found individuals with spinal lesions at or above L2 had lower scores on transfers and basic mobility, sports and physical function, and global function scores on the PODCI as well as health scores on the PedsQL^{TM} than those with L3-5 lesions. These findings are similar to those of some other studies [5, 9, 14, 18]. In general, children scoring in the low 80s or below on the PODCI typically function at a lower level than their normally developing peers [10]. In our population, the parents scored children in the low 80s and considerably below on the PODCI for all categories except upper extremity and physical function scores for both neurologic groups and in comfort scores for those children in the L2 and above group. This is in contrast to Leger [16] who reported that their sample of youth (age 15–25) with MM in the New England area had a high quality of life and participated in recreation and sports.

When we reviewed the correlation among demographics and HRQOL, we found an association with the presence of a shunt and lower HRQOL scores on the PODCI for transfers and basic mobility, sports and physical function, happiness, and global function. Many other studies reported a similar association [2, 3, 11, 12, 16, 28]. Davis et al. [9] looked at 158 adolescents and reported 84% of the individuals were shunted. They found the subjects had a 25% to 30% delay in autonomy skills, which was explained by cognitive ability more than the level of lesion [9]. We found age had little effect on HRQOL other than evidence of lower sports and physical function scores on the PODCI for those children who were older. Similarly, Young et al. [29] reported in a transitional study that children with MM have the best self-rated health, whereas adults with MM reported the worst self-rated health compared with those with cerebral palsy and traumatic brain injury. Holmbeck et al. [13] reported on children with MM in a younger age bracket (8–9 years) and noted that they had no difference between normally developing children for global self-worth, but did have difficulties with social immaturity, decreased social contacts apart from school, and were less physically active [13]. Although we found a high rate of obesity in our population, we were surprised that we found no association between BMI and HRQOL. Obesity may be the result of multiple complex issues and affects orthotic and wheelchair fit, ambulation abilities, endurance, transfers, and self-care. The presence of obesity and reduced activity levels in children and young adults with MM is concerning [1, 12, 23].

From the results of this study and our past clinical experience, we have enhanced our multidisciplinary team and developed the more focused aim of addressing community participation barriers and promoting optimum health and mobility so these children transition into independent young adults. Nutrition and dietary consults have been added for each clinic visit to help combat obesity and inactivity proactively. Recreation therapy and social work/care coordination are now providing referrals to sports and recreational activities as well as camps and spina bifida associations. We hope to create a pattern of lifetime social engagement and physical activity that is often lacking in current adults with MM. Since discovering the usefulness of the HRQOL tools, we have begun to use the PODCI and PedsQL^{TM} in other patient populations too. In the future, we would like to compare the results of our MM population with other disabled populations and expand on our current study to include a more longitudinal design with responses from children as well as guardians.

This study provides greater insight on activity level, functional health, and quality of life among children and adolescents with MM. Using two outcome tools that are not typically used during a clinical examination, we were able to provide a more comprehensive clinical picture of this population's strengths and deficits and areas that we need to address in the future. Young individuals with MM have multiple complex impairments, lower activity level, and decreased HRQOL, which only amplifies in the older MM population. Across the lifespan, clinicians need to assess the contribution of functional health impairments to activity limitations and develop programs to more fully meet their needs.

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