Hip Status and Long-term Functional Outcomes in Spina Bifida

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Background: Nearly 50% of individuals with myelomeningocele will develop a dislocated hip by skeletal maturity. The purpose of this study was to determine the influence of hip status on functional outcomes in a cohort of adult patients with myelomeningocele.

Methods: Patients with a diagnosis of myelomeningocele >18 years were prospectively enrolled over a 12-month period. Neurological level of involvement was obtained from chart review and interview. Clinical examination included hip range of motion and leg-length discrepancy. Reimer's migration index was calculated from a current anteroposterior pelvic radiograph. All subjects completed the VR-12 and the NIH PROMIS outcomes measures for pain interference and physical function. The χ^2, the Pearson correlation coefficients, and linear regression models were applied to evaluate the influence of hip status on functional outcomes.

Results: In total, 31 patients (average age 31, range 19 to 49) were included. Eight thoracic, 9 lumbar, and 14 sacral level patients participated. Twenty had bilaterally located hips, 5 had a unilateral subluxation or dislocation, and 6 had bilaterally subluxated or dislocated hips. In univariate analysis, patients with bilaterally located hips performed better in lower extremity function than those with unilateral subluxation/dislocation (36.7 vs. 26.0; $P=0.03$) but worse in pain interference than those with bilateral subluxation/dislocation (52.0 vs. 43.3; $P=0.03$). After controlling for neurological level, there was no statistically significant difference in the VR-12 mental ($P=0.32$) or physical component summary ($P=0.32$) scores, nor in the PROMIS lower extremity function ($P=0.26$) or pain interference scores ($P=0.33$) between groups. Decreased extension and abduction were indirectly correlated with VR-12 mental component scores ($P=0.0038$, 0.0032). Leg-length discrepancy was not associated with any outcome measure.

Conclusions: Long-term outcomes are not associated with hip status in adult patients with myelomeningocele. Functional outcomes are more closely correlated with neurological level and hip range of motion. These results suggest efforts to keep myelomeningocele hips reduced are likely without functional benefit and should be avoided in favor of maintaining motion with contracture release as needed.

Level of Evidence: Level III.

Key Words: myelomeningocele, myelodysplasia, neuromuscular hip, spina bifida

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Nearly 30% to 50% of patients with myelomeningocele will develop a dislocated hip as a result of muscular imbalance and subsequent acetabular and/or femoral dysplasia.[1,2] Previously, hip reduction by closed or open means was recommended for all children with a dislocated hip or a hip at risk[3] to avoid pain, decrease the risk of developing a leg-length discrepancy (LLD) and improve gait parameters. However, more recent research has revealed that there are no functional benefits gained by reducing either unilateral or bilateral dislocated hips in patients with myelomeningocele.[4–9] Rather, functional status, including ambulatory status and sitting ability/balance, is more closely associated with neurological level of involvement rather than status of hip reduction.[6,7,10] Further, most investigators agree that pain and psychological well-being are not affected by hip status (reduced vs. dislocated).

Despite this, some investigators continue to argue for surgical management of dislocated hips in patients with low-level myelomeningocele, as these patients are more likely to remain ambulatory into adulthood and theoretically more likely to suffer any negative consequences as a result of a LLD secondary to hip dislocation.[10,11] But there remains no long-term functional evidence to support this practice. As such, the purpose of this study was to determine the influence of hip status on functional outcomes in a cohort of adult patients with myelomeningocele.

METHODS

Patients were recruited for this institutional review board-approved prospective cohort study over a 12-month period (January to December, 2016) at a tertiary care

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rehabilitation hospital in an urban setting. All patients seen in the adult myelomeningocele continuity clinic were asked to participate. This included all functional neurological levels from thoracic to sacral regardless of ambulatory or hip stability status (history of unilateral or bilateral hip dislocation, including those that had been reduced by closed or open methods). All enrolled patients had a diagnosis of myelomeningocele, not including isolated related but dissimilar diagnoses (tethered cord, Arnold Chiari, diastametomyelia) and were 18 years of age and above. Patients were excluded for the use of any medications that could affect gait, balance, or muscular strength. Subjects were additionally excluded for any additional diagnosis (aside from myelomeningocele) that could affect gait, balance, or muscular strength.

All patients were evaluated by 1 of 2 treating orthopaedic surgeons (V.T.S. or L.D.). Neurological level of involvement, functional mobility scale, and history of hip surgery were obtained from chart review and patient interview. Clinical examination included hip range of motion and LLD. Reimer's migration percentage was calculated from a current anteroposterior pelvic radiograph.[12] All migration percentages were calculated by the same researcher (V.T.S.).

Each subject completed the VR-12 and the NIH PROMIS outcomes measures for pain interference and physical function. VR-12 is a generic instrument used to measure health-related quality of life. Along with the SF-12, it is the gold standard for all health-related quality of life outcome measures.[13] The effectiveness of the VR-12 in estimating health status and disease burden has been shown in several publications spanning multiple disease systems.[14] PROMIS has not been specifically validated for use in all neuromuscular diseases, but it has shown clinical validity among multiple chronic disease groups.[15,16]

Patients were compared in terms of outcomes by current hip status—bilaterally located hips, bilateral subluxation or dislocation, and unilateral subluxation or dislocation, defined by migration percentage, regardless of history of previous hip preservation surgery. A located hip was defined as a migration percentage <30%; a subluxated hip was defined as a migration percentage between 30% and 99%; and a dislocated hip was defined as a migration percentage of 100%.

TABLE 1. Neurological Level of Involvement

	N (%)		
	Thoracic Level	Lumbar Level	Sacral Level
Group 1	2 (10)	4 (20)	14 (70)
Group 2	3 (60)	2 (40)	0
Group 3	3 (50)	3 (50)	0

Functional outcome scores were compared between cohorts before and after controlling for neurological level of involvement and hip range of motion. The χ^2, the Pearson correlation coefficients, and linear regression models were applied to evaluate the influence of hip status on outcomes.

RESULTS

Over a 12-month period, 32 patients were recruited from the adult myelomeningocele health maintenance clinic. One patient was excluded for lack of complete information, leaving 31 patients for inclusion. There was near even representation of men and women—55% of the included subjects were female (n = 17) and 45% were male (n = 14). Included subjects were 31 years old on average (range, 19 to 49). Of those that were included, there were 8 thoracic, 9 lumbar, and 14 sacral level patients. Twenty had bilaterally located hips (group 1), 5 had a unilateral subluxation or dislocation (group 2), and 6 had bilaterally subluxated or dislocated hips (group 3) (Table 1 and Fig. 1).

In univariate analysis, we compared each group against the other in terms of MCS12 (psychological component score for the VR-12), PCS12 (physical component score for the VR-12), and the lower extremity function (physical score) and pain interference (psychological score) PROMIS measures. Patients with bilaterally located hips performed better in PROMIS lower extremity function scores than those with unilateral subluxation/dislocation (36.7 vs. 26.0; $P = 0.03$) but performed worse in PROMIS pain interference than those with bilateral subluxation/dislocation (52.0 vs. 43.3; $P = 0.03$). There were no significant differences found between any group in terms of the VR-12 outcomes measures in univariate analysis (Fig. 2). Further, there was no direct correlation

FIGURE 1. Radiographic representation of groups 1 to 3. A, Representative x-ray from a patient in group 1 with bilateral located hips. B, Representative x-ray from a patient in group 2 with a unilateral hip dislocation. C, Reprentative x-ray from a patient in group 3 with bilateral dislocated hips and pseudoacetabulum.

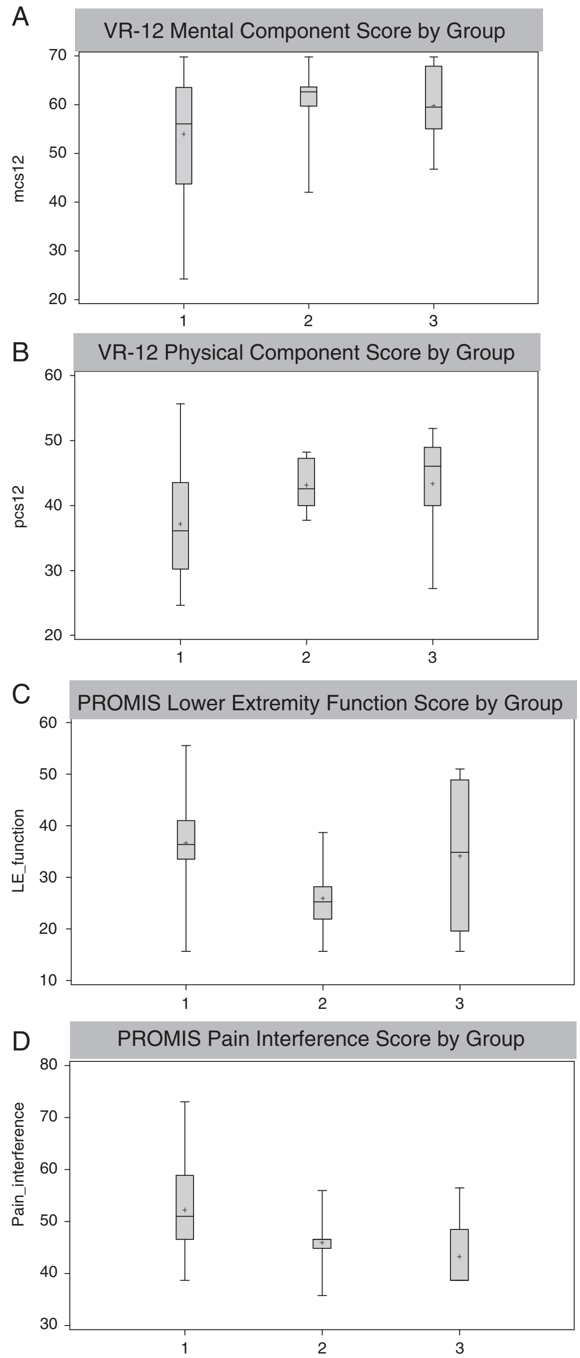

FIGURE 2. Outcomes measures. A, Correlation between VR-12 mental component score and hip status. B, Correlation between VR-12 physical component score and hip status. C, Correlation between PROMIS lower extremity function score and hip status. D, Correlation between PROMIS pain interference score and hip status. + indicates the mean, line is the median.

between hip migration percentage and any of the tested outcomes measures (Table 2).

Four patients had a history of previous hip preservation surgery in 6 hips. This includes 1 patient from group 1 who had a unilateral proximal femoral osteotomy at age 6. Of the 2 patients included from group 2, one underwent bilateral open reductions at age 4 and the other had a unilateral proximal femoral osteotomy at an unknown age. Of note, the hip that was subluxated at the time of inclusion was the operative side. The patient from group 3 had bilateral open reductions with a unilateral derotational osteotomy at age 1 and 2 years, respectively. All 4 of these patients tended to have lower PROMIS lower extremity function scores and higher pain interference scores than those who had not had previous hip surgery, but neither trend reached clinical or statistical significance. Two additional patients, one from group 2 and one from group 3, had histories of hip flexor releases, both of which had lower PROMIS pain interference scores compared with the remainder of the group.

After controlling for neurological level, there was no statistically significant difference in the VR-12 mental ($P=0.32$) or physical component summary ($P=0.32$) scores, nor was there any difference in the PROMIS lower extremity function ($P=0.26$) or pain interference scores ($P=0.33$) between groups.

After controlling for range of motion, there were no significant differences found between any group in terms of either VR-12 outcome scores nor in PROMIS lower extremity function scores. However, there was a statistically significant increase in PROMIS pain interference scores between groups 1 and 3 (mean difference 6.47; $P=0.0325$), although this did not rise to the level of minimal clinically important difference (mean difference <10).

Further investigation into range of motion revealed that when considering all included patients, decreased extension and abduction were both inversely correlated with VR-12 mental component scores ($P=0.0038$, 0.0032), but there was no correlation between any range of motion parameter and any other outcomes scores. There was no demonstrable correlation between LLD with any of the 4 outcomes measures.

DISCUSSION

Although the pendulum has swung against aggressive surgical management for hips at risk in children with myelomeningocele in the United States, there remain contemporaneous advocates for hip reduction in this population.[17] Variability in treatment protocols likely stems from a lack of published long-term patient-reported health-related quality of life outcomes in regard to hip status. In their place, surrogates for health-related quality of life outcomes measures—hip range of motion, ambulatory status, and radiographic outcomes[6,8,17,18]—have been used as justification both for and against surgical intervention for hips at risk in the myelomeningocele population. However, if we are to provide meaningful patient care for this pediatric population, we must interrogate and be guided by long-term quality of life measures. This statement is especially true considering the fact that the most commonly cited surrogate outcome measure—ambulatory status—is most closely linked to neuro-

TABLE 2. Correlation: Migration Percentage and Outcomes

	VR-12 Mental Component	VR-12 Physical Component	PROMIS Lower Extremity Function	PROMIS Pain Interference	Right Migration Percentage	Left Migration Percentage
Pearson correlation coefficients*						
VR-12 mental component	—	−0.04979	−0.13451	−0.27407	0.0779	0.24893
VR-12 physical component	−0.04979	—	0.21956	−0.61396	0.42243	0.35394
PROMIS lower extremity function	−0.13451	0.21956	—	−0.15061	−0.1036	0.0007
PROMIS pain interference	−0.27407	−0.61396	−0.15061	—	−0.44116	−0.48058
Right migration percentage	0.0779	0.42243	−0.1036	−0.44116	—	—
Left migration percentage	0.24893	0.35394	0.0007	−0.48058	—	—

*Values approaching 1 signify a high correlation.

logical level of involvement above all other factors[6,7,10,19] and not to hip status.

In this light, we sought to quantify the effect of hip status on both patient function/mobility and psychological well-being in adult patients with spina bifida. In this cohort of adult patients with myelomeningocele, long-term outcomes were not associated with hip status. Functional outcomes were more closely correlated with neurological level of involvement and hip range of motion, findings that are consistent with previously published reports.[4,6]

In fact, the discordant outcomes noted in univariate analysis were likely reflective of neurological level of involvement and hip range of motion rather than radiographic hip status. There were no significant differences in any outcome measure after controlling for neurological level and a clinically negligible difference in PROMIS pain interference scores between group 1 (bilateral located hips) and group 3 (bilateral subluxated or dislocated) when controlling for hip range of motion. Interestingly, on average, those with bilateral dislocations/subluxations and those with unilateral dislocations/subluxations reported less pain interference than those with bilateral located hips. Conversely, decreased range of motion was inversely correlated with the VR-12 mental component score.

There was also no correlation noted between LLD and any patient-reported outcome. Although LLD has previously been utilized as a surrogate measure for overall outcomes, these results suggest that LLD is irrelevant in terms of psychological well-being and physical function and pain. Further, while LLD is typically reported as an outcome negatively associated specifically with unilateral dislocations,[10] our adult self-reported outcomes do not reflect this presumed correlation between LLD and worsening quality of life. As such, the validity of any previous conclusions based on this surrogate measure should be called into question.

Although this study is the first to report on long-term functional outcomes in this population, it is limited primarily by its small size. Given that not every patient with myelomeningocele will have active orthopaedic concerns yearly, many patients do not continue regular long-term follow-up through an orthopaedic continuity clinic. However, we do believe that the included cohort was representative of the population as a whole as it spanned a broad age-range, had a well-balanced sex mix and included all neurological levels of involvement. Given the small cohort size, we were unable to compare outcomes between those patients who had not had previous hip surgery and those who have had previous surgery. Only 4 patients had a history of previous hip preservation surgery on 6 hips, 2 of which were subluxated and 2 of which were dislocated at the time of study. Interestingly, patients who had a previous hip surgery tended to have lower PROMIS lower extremity function scores and higher pain interference scores than those who had not had a previous hip surgery, but neither trend reached clinical or statistical significance. Resultantly, we compared all patient outcomes based only on current hip status regardless of surgical history. However, history of previous surgery—both successful and unsuccessful—may be a confounding factor. We did not explicitly compare treated to untreated hips within or between cohorts.

In addition, given the limited cohort size, we were unable to validate the PROMIS pain interference scores or physical function scores for specific use in this patient population. However, all outcomes measures had been previously validated in multiple adult populations with chronic diseases. Given that we had similar findings in 2 different outcomes measures for both physical function and psychological burden, we believe these findings to be reflective of true long-term outcomes in this patient population.

Future research is needed to validate PROMIS outcomes measures specifically in this population and to correlate long-term ambulatory parameters—energy consumption, speed, kinematics, kinetics—with both hip status and these long-term patient-reported outcomes measures.

Despite limitations inherent to a small comparative cohort study, these results suggest efforts to keep myelomeningocele hips reduced are likely without functional benefit and should be avoided. However, given the correlation between hip range of motion and patient-reported outcomes, efforts should be made to maintain functional motion with contracture release as needed.

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