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Relationships Among Classifications of Impairment and Measures of Ambulatory Function For Children With Spina Bifida

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Abstract

Purpose—To examine associations between neurosegmental and functional level classifications in children with Spina Bifida, and determine which classification best reflects daily walking activity.

Materials and Methods—A prospective correlational study was conducted. Children with Spina Bifida were given ratings for lesion level [X-ray and International Myelodysplasia Study Group (IMSG) level determined by muscle strength] and functional level [Hoffer ambulatory level and Dias functional classification of myelomeningocele (FCM), Functional Mobility Scale (FMS)]. Daily walking activity was measured with a StepWatch monitor. Data were analyzed using Spearman rank correlation.

Results—Sixty-one children were included, [56% male, average age 9.8 (SD 2.7) years]. The neurosegmental level classifications, X-ray lesion level and IMSG level showed little to no correlation with each other ($r = 0.17$). Among functional classifications, the Dias FCM correlated strongly with the FMS ($r = 0.80 - 0.87$). Correlations with steps per day were moderate to good for the Dias FCM and the FMS ($r = 0.53 - 0.62$), fair for IMSG level ($r = 0.45$), and little to none for X-ray lesion level ($r = 0.03$).

Conclusions—The Dias FCM is comprehensive, including elements of neurosegmental level and function, and correlates well with walking activity. We recommend its use for classifying function in patients with Spina Bifida.

Keywords

Spina Bifida; classification; ambulation; functional level; neurosegmental level

Introduction:

Spina Bifida is a congenital malformation in which the neural tube fails to close, manifesting as a defect in the posterior elements of the vertebrae. Myelomeningocele is the most

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severe and common form of Spina Bifida. Failure of neural tube closure results in bowel, bladder, motor and sensory deficits below the level of the lesion, which can lead to functional limitations secondary to underlying paralysis.[1, 2] Comorbidities including obesity, cognitive impairment, orthopaedic deformity, and decreased motivation to ambulate also limit function.[3]

Classifying degree of impairment and function is important in this patient population. Categorizing function aids patients, families, and clinicians in anticipating life-long medical and treatment needs, potential response to treatments and prognosis for independence. Various classification systems have been used in the Spina Bifida population. These systems have primarily been focused on the body structure and function level of the World Health Organization's International Classification of Function (ICF) model,[4] such as anatomic lesion level determined radiographically, and lesion level based on muscle strength patterns. Classifications at the activity level of the ICF have also been used, such as rating ambulatory function, brace and assistive device use.

Level of impairment in children with Spina Bifida is often described by neurologic lesion level, as determined by prenatal ultrasound or spinal radiographs. Level determined by prenatal ultrasound has shown good to excellent correlation ($r=0.79$, $p=0.01$) with motor level and ambulatory function achieved by age 3–4 years.[5, 6] Lesion level as determined by x-ray agrees moderately with functional level when x-ray levels are grouped as thoracic, lumbar or sacral ($k=0.523$, concordance 74%), but only weakly when levels are defined more precisely ($k=0.273$, concordance 39%).[7] Gross motor function can change over time as a result of growth and increased body weight. Radiographic lesion level is fixed, and may not reflect function as well later in life.

Muscle strength is used to assign a neurosegmental impairment level based on recommendations by The International Myelodysplasia Study Group (IMSG).[8, 9] Muscle strength and strength patterns were shown to be predictive of later ambulatory status when categorized grossly (nonambulators, partial ambulators, community ambulators). Muscle groups contributing to variation in ambulatory level are the iliopsoas, gluteus maximus and medius, quadriceps and anterior tibialis.[10, 11]

Ambulatory function is most often classified using the scale reported by Hoffer, et al., which has four general categories: community ambulation, household ambulation, non-functional ambulation, and non-ambulatory.[12] Dias and Swaroop reported a classification system that incorporates both impairment of body structure and function (neurosegmental lesion level) and activity (need for bracing and/or assistive devices for walking).[13] It is a more comprehensive classification system than neurosegmental level alone, but is not currently in wide use.

Though traditionally used for patients with cerebral palsy, the Functional Mobility Scale (FMS) has more recently been applied to patients with Spina Bifida.[14, 15] The FMS rates mobility over three distinct distances representing mobility in the home, school, and community, based on patient and/or family report of devices used or assistance needed at each distance.[16] The FMS has been proven valid and reliable, and also responsive to

change with intervention in children with cerebral palsy.[17, 18, 19] FMS at all distances has been shown to correlate well ($r > 0.83$) with time spent in an upright position as opposed to supine in children with cerebral palsy.[20] No studies have investigated such relationships in patients with Spina Bifida.

The use of multiple classification systems of function in patients with Spina Bifida is problematic. Inconsistent classification of patients may lead to variability in service provision and quality of patient care among health care centers or geographic regions. Consistent use of clinically meaningful classifications would greatly enhance the quality of research in Spina Bifida. To date, there is no information in the literature regarding which of the various classifications is the best indicator of true ambulatory activity for these children. The purposes of this study were to examine associations between classifications of neurosegmental level and functional level, and to determine which classification system is the best indicator of daily mobility as measured with an activity monitor in pediatric patients with Spina Bifida.

Materials and Methods:

Children were enrolled in a 3-year prospective study of relationships between bone properties and walking ability conducted at a pediatric tertiary care hospital. Participants were recruited from the Spina Bifida Clinic at the center. Eligibility criteria included diagnosis of Spina Bifida, aged 6–16 years. All study procedures were approved by the hospital's institutional review board, and all participants and their parents or guardians gave written informed consent and assent as appropriate. For the current correlational study, participants able to walk at a community, household or non-functional level on the Hoffer scale who had complete activity monitor data were included, and data from the first year of the 3-year study were used. As part of the 3-year prospective study, one of three experienced physical therapists (PTs) with established reliability performed manual muscle testing to determine each patient's IMSG neurosegmental level. In cases of asymmetric involvement, the rating for the weaker side was used. The PT also assigned a Hoffer ambulatory level and a Dias functional classification of myelomeningocele (FCM) level to each patient. Descriptions of the classification systems are given in Table 1. The PT assigned FMS ratings based on patient and parent report of the participants' mobility in the home (5 meter distance), school (50 meter distance) and community (500 meter distance).[16] A single investigator (NM) assigned radiographic lesion level based on assessment of the earliest available spinal x-ray. Walking activity was assessed using the Stepwatch activity monitor, which was worn at the ankle of the participants' strongest limb during waking hours for one week. Average steps per day were tabulated for each participant. These variables were included in the current correlational analysis.

Statistical Analysis

Relationships among classifications of neurosegmental lesion level, ambulatory functional level, and average number of steps per day were analyzed using Spearman rank correlation. Strength of correlation was estimated using published general guidelines.[21] Correlations

were interpreted as little to none ($r = 0.00$ – 0.25), fair ($r = 0.25$ – 0.50), moderate to good ($r = 0.50$ – 0.75), and good to excellent ($r =$ above 0.75).

Results:

Sixty-one children were included in the current study, 34 males (56%) and 27 females (44%). Average age of participants at enrollment was 9.8 years (standard deviation (SD) 2.7, range 6 – 15). Forty-seven participants (77%) had myelomeningocele and 14 participants (23%) had lipomyelomeningocele. Thirty-seven participants (61%) had ventriculoperitoneal shunts, 26/37 (70%) of whom had undergone at least 1 revision (range: 1–9 revisions). Sixteen participants (26%) had undergone prior tethered cord release surgery. Twenty-two participants (36%) had undergone previous orthopedic surgery to the lower limbs.

Levels of neurosegmental impairment based on IMMSG and radiographs, Hoffer ambulatory levels and Dias FCM functional classifications are outlined in table 1. Participants took an average of 2687 steps/day (SD 2946). Average steps per day are stratified by classification system level in table 2.

The two classifications of neurosegmental level (IMMSG and vertebral lesion level) showed little to no relationship to each other. (table 3)

Among the functional classifications (Hoffer, Dias FCM, and FMS at all distances), the strongest relationships were seen between the Dias FCM classification and FMS at all distances (excellent correlation). The Hoffer classification showed fair to moderate relationships with the FMS at all distances and the Dias FCM classification. (table 3)

Between neurosegmental level and functional classifications, the IMMSG showed an excellent correlation with the Dias FCM, good correlation with the FMS at all distances and fair correlation with the Hoffer classification. X-ray lesion level showed fair to no correlation with all functional classifications.

The Dias FCM functional classification and FMS at all distances showed the strongest correlation with actual steps taken per day. IMMSG level and the Hoffer classification showed fair correlation with steps per day. Vertebral radiographic lesion level showed little to no correlation with average steps per day. (table 3)

Discussion:

In a previous study, our center reported baseline data on walking activity performance using the StepWatch activity monitor in ambulatory children with spina bifida. The study showed that children with sacral and low lumbar level lesions (based on IMMSG rating) function similarly to their typically developing peers, whereas those with mid-high lumbar lesions were less active. [22] The current study expands on the previous work by examining relationships among various classifications of neurosegmental and functional level, and their associations with actual ambulatory activity performance.

Lesion levels based on X-ray and muscle strength (IMSG) did not correlate well with each other, a finding which is contrary to the reports of Coniglio et al. and Kollias, et al. This disagreement is likely due to differences in participant characteristics and the means of determining muscle innervation or strength. The earlier studies assessed neurosegmental level at birth or at 3–4 years of age, and neither specifically described the method of doing so. In contrast, the current study included older participants and evaluated muscle strength using a standardized method. X-ray lesion level is a fixed factor (bony deformity), whereas muscle strength can vary with age, growth, and medical complications such as spinal cord tethering. Muscle strength profile can also change after orthopedic surgery. For instance, the anterior tibialis tendon is sometimes transferred to the calcaneus to correct foot deformity, which changes the muscle strength profile and worsens the IMSG level rating. We did not exclude such patients, since it was not always possible to know which patients had such procedures in the past.

Classifications of function (Hoffer, Dias FCM, FMS) all correlate well with each other and with actual steps taken per day. Use of the Dias FCM classification system, based on bracing and assistive devices, may be preferable to the Hoffer classification, as it better stratifies higher functional levels and correlated better with function as measured with the FMS. The Hoffer classification definition of community ambulation is very broad, and 78% of children in the current study fit the definition for this category. These participants were split among 3 categories using the Dias FCM classification system: low lumbar, high sacral and low sacral. The low lumbar Dias FCM group took less than 1/3 of the average number of steps per day taken by the Hoffer community ambulatory group, suggesting the Dias FCM classification may be more sensitive as an indicator of ambulatory ability in children with Spina Bifida.

Neurosegmental level as measured by the IMSG showed moderate correlation with ambulatory function based on both the Hoffer and Dias FCM scales, similar to the findings of Tita, et al. for adults with myelomeningocele.[23] However, neurosegmental level as determined by X-ray was only weakly correlated with steps per day and FMS scores. These findings suggest X-ray lesion level and IMSG neurosegmental levels should not be used in isolation to classify patient function and should definitely not be used interchangeably. They also illustrate the problems associated with classifying patients solely on the basis of body function/structure. Inclusion of a functional classification is recommended, as outlined in the ICF. The Dias FCM functional classification system is comprehensive, including elements of both neurosegmental level and function in terms of devices and assistance needed to walk. Because of this and the fact that it correlates well with ambulatory activity level, we recommend its use for classifying function in patients with Spina Bifida.

FMS rating (primarily at household and school distances) is a good indicator of activity level as measured by the StepWatch monitor. However, it is not a perfect proxy for actual activity performed. Correlations between the FMS at all distances and steps per day in the current study were lower ($r < 0.62$) than that seen between FMS and time spent in an upright position in children with CP ($r = 0.83$).[20] We do not know which monitor is a better indicator of actual steps taken (the UpTimer used in the study by Pirpiris, et al., or the StepWatch used in the current study). The FMS is assessed by report rather than direct observation. Because

of these factors, we feel the FMS should not be used in isolation as an activity measure, but should be used in conjunction with an activity monitor in research investigations.

There were several limitations of the present study. First, because of relatively small numbers and inclusion of patients in a single geographic area, participants may not be representative of the general Spina Bifida population. Second, patients with prior orthopaedic or neurosurgical procedures were included, which could have impacted muscle strength and IMSG classification and its associations with other variables. These patients were included to provide better representation of the clinical population. Lastly, we did not control for other factors which may impact overall balance and physical activity performance such as socio-economic status, access to therapy, parks and recreation, cognitive ability, presence of a VP shunt or other medical comorbidities.

In conclusion, classifications of ambulatory function in children with Spina Bifida are better indicators of actual activity performance than anatomic lesion level or neurosegmental level. The Dias FCM functional classification is more comprehensive than the Hoffer scale, correlates better with function, and may be preferable for use in the ambulatory spina bifida population. The FMS is an acceptable proxy for steps taken per day, but activity monitoring should accompany the FMS for research investigations. Future research should assess how neurosegmental impairments and activity levels change over time as children transition into adulthood.

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Implications for Rehabilitation:

- Functional classifications correlate better with daily walking activity than neurosegmental level classifications for patients with Spina Bifida.
- The Dias FCM includes neurosegmental and functional level elements, correlates well with daily activity, and is recommended for use in classifying Spina Bifida patients.
- Combined use of the FMS and activity monitoring is recommended for research and clinical assessment.

Table 1:

Classifications of neurologic and functional levels.

IMSG [5, 6]		
Neurologic Level		Muscles with 3/5 strength (each level includes all muscles from higher levels)
High lumbar	L1	Hip flexors (iliopsoas)
	L2	Hip adductors
Mid lumbar	L3	Knee extensors (quadriceps)
Low lumbar	L4	Knee flexors (medial hamstrings)
		Ankle dorsiflexors
		Ankle inverters (tibialis anterior & posterior)
	L5	Hip abductors
		Ankle evertors (peroneals)
Sacral	S1–S3	Hip extensors (gluteus maximus)
		Ankle plantarflexors (gastrocnemius, soleus)
Hoffer ambulatory level [9]		
Ambulatory Level	Description	
Community ambulator	Walk indoors and outdoors for most of their activities and may need crutches or braces, or both. Use wheelchair only for long trips out of the community.	
Household ambulator	Walk only indoors and with apparatus. Able to get in and out of chair and bed with little if any assistance. May use wheelchair for some indoor activities at home and school, and for all activities in the community.	
Non-functional ambulator	Walk in therapy session at home, in school, or in the hospital. Use wheelchair to get from place to place and to satisfy all needs for transportation.	
Non-ambulator	Wheelchair-bound but usually can transfer from chair to bed.	
Dias functional classification of myelomeningocele [10]		
Functional Level	Braces	Assistive Devices
Mid Lumbar and above	RGO or HKAFO	Walker or Crutches
Low Lumbar	AFO or KAFO with free knee joint	Forearm Crutches (or any other external support)
High Sacral	AFO	None
Low Sacral	UCBL or SMO in some cases	None

IMSG – International Myelodysplasia Study Group; RGO – reciprocating gait orthosis; HKAFO – hip knee ankle foot orthosis; AFO – ankle foot orthosis; KAFO – knee ankle foot orthosis; UCBL – University of California Berkeley Laboratory; SMO – supramalleolar orthosis

Table 2.

Participant characteristics (N=61)

	Classification, N (%)	Mean (SD) [minimum - maximum steps per day] [24]
IMSG	Thoracic – 1 (1)	1176
	High lumbar – 1 (1)	251
	Mid-lumbar – 28 (46)	1723 (1952) [range: 7–7932]
	Low lumbar – 10 (16)	3571 (3820) [range: 338–12252]
	High sacral – 15 (25)	3260 (3135) [range: 796–11716]
	Low sacral – 6 (10)	4938 (3778) [range: 1230–11882]
Hoffer	Non-functional ambulators – 7 (12)	1086 (1505) [range: 119–4400]
	Community ambulators – 54 (88)	2895 (3030) [range: 7–12252]
Dias FCM	Mid-lumbar and above – 7 (12)	1275 (1472) [range: 251–4400]
	Low lumbar – 16 (26)	803 (511) [range: 7–1602]
	High sacral – 24 (39)	3299 (2911) [range: 1010–12252]
	Low sacral – 14 (23)	4498 (3752) [range: 796–11882]
X-ray lesion level *	L2 – 4 (5.9)	1933 (1751) [range: 346–4400]
	L3 – 13 (20.6)	2186 (1950) [range: 251–6113]
	L4 – 28 (41.2)	3108 (3369) [range: 119–11882]
	L5 – 12 (17.6)	1956 (2005) [range: 7–6340]
	S1 – 3 (2.9)	5510 (5932) [range: 1088–12252]

* N=60

Table 3.

Correlation among classification systems and average steps per day.

	Steps per day	X-ray	IMSG	Dias FCM	Hoffer	FMS 5	FMS 50	FMS 500
Steps per day	1.0							
X-ray	0.03	1.0						
IMSG	0.45 ^a	0.17	1.0					
Dias FCM	0.58 ^a	0.29 ^a	0.79 ^a	1.0				
Hoffer	0.34 ^a	0.29 ^a	0.41 ^a	0.51 ^a	1.0			
FMS 5	0.62 ^a	0.24	0.66 ^a	0.85 ^a	0.49 ^a	1.0		
FMS 50	0.61 ^a	0.25 ^a	0.67 ^a	0.87 ^a	0.51 ^a	0.93 ^a	1.0	
FMS 500	0.53 ^a	0.27 ^a	0.63 ^a	0.80 ^a	0.47 ^a	0.80 ^a	0.90 ^a	1.0

^a $p < 0.05$ based on Spearman rank correlation.