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Doctor of Physical Therapy Program Case Reports

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2018

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# Prognosis for Development of Early Motor Milestones in a Pediatric Patient with Myelomeningocele: A Case Report

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## Abstract

**Background:** Myelomeningocele, a type of spina bifida, is a prevalent birth defect that affects numerous individuals around the world. Significant research has been conducted to examine the influence of this condition on adolescents and adults, but limited evidence is available on how spina bifida influences development in childhood. **Purpose/Case Description:** The purpose of this report is to describe the development of motor milestones in a 27-month old female with L4-5 myelomeningocele to improve the current understanding of the expected prognosis for development of early motor milestones in children with spina bifida, specifically the diagnosis of lower lumbar myelomeningocele. Although the patient was progressing in her achievement of motor milestones, she was significantly delayed compared to her peers. **Outcomes:** The primary outcomes utilized in this case were tests and measures, largely qualitative, analyzing the patient's functional mobility. Additional outcome measures that could have been incorporated include the Bayley Scale of Infant and Toddler Development and the PEDI-CAT. **Discussion:** The patient with L4-5 myelomeningocele was achieving motor skills consistent with those around 10-12 months of age although her chronological age at the end of the nine-week time period was 29 months. The patient was steadily progressing in the development of new motor milestones over this time and, although behind peers without spina bifida, is expected to continue making progress. This case contributes to the current understanding of expected prognosis for motor skill development in children to assist physical therapists and other providers to establish an appropriate prognosis for timing of motor skill development in pediatric patients with myelomeningocele.

**Keywords:** Myelomeningocele; pediatrics; developmental milestones; physical therapy; rehabilitation

## Background

Myelomeningocele, a neural tube defect (NTD) commonly referred to as spina bifida, is a congenital birth defect affecting individuals from infancy through adulthood. Current estimates suggest that NTDs occur in 1 of every 2000 births in the United States and, in total, there are approximately 166,000 individuals living with spina bifida in the United States.<sup>6,11</sup> In addition, the rates of spina bifida in many countries throughout the world are higher than these numbers demonstrating the prevalence in the United States.<sup>6</sup>

The three most well-known types of spina bifida are spina bifida occulta, meningocele, and myelomeningocele.<sup>3</sup> Spina bifida occulta is the most common and mild form of this condition and it occurs when one or more vertebrae are incomplete, meaning they do not fully close during development. The spinal cord and nerves are not usually affected in spina bifida occulta and rarely is this type associated with physical disabilities. The second type of spina bifida is meningocele, the rarest type of NTD. This type of spina bifida occurs when the meninges and cerebrospinal fluid protrude through a cleft in the vertebrae, often resulting in minimal physical disability.<sup>3,13</sup> Myelomeningocele is the third and most severe type of spina bifida. This condition involves the meninges as well as the spinal cord protruding through a cleft in the vertebrae. With this involvement of the spinal cord, physical disability is extremely common in myelomeningocele and it is also associated with brain damage due to the pull of the cord on the hindbrain. This pull often causes a blockage in the flow of cerebrospinal fluid in the brain and results in hydrocephalus. Thus, myelomeningocele can lead to both physical impairments and learning disabilities.<sup>13</sup>

Although significant research has been conducted to examine the influence of spina bifida on individuals through adolescence and adulthood, limited investigation has focused on the influence that this condition has in early childhood. Additionally, research into the influence of this condition is complicated by the “multiple sources of phenotypic variability” including differences in lesion level, the presence of hydrocephalus and whether a shunt has been placed, and differences in socioeconomic status.<sup>6,8</sup> More specifically, little research has been aimed at analyzing the timing and achievement of motor milestones in early childhood and how this influences the prognosis of these individuals.

Based on the limited amount of research investigating the timing that early motor milestones are achieved by infants and toddlers with myelomeningocele, it is difficult for clinicians to monitor and assess the progress of these individuals they are treating. This case serves as a contribution toward the current, yet limited understanding of what clinicians such as physical therapists may expect for growth in motor skill development of children with myelomeningocele in infancy and toddlerhood. The purpose of this report is to describe the development of motor milestones in a 27-month old female with L4-5 myelomeningocele to improve the current understanding of the expected prognosis for development of early motor milestones in children with spina bifida, specifically the diagnosis of lower lumbar myelomeningocele.

## Case Description

The individual discussed in this case is a 27-month old female with L4-5 myelomeningocele who presented to physical therapy for an evaluation to assess current progress toward motor milestones and provide recommendations for an appropriate plan of care. At the initial evaluation, the patient’s mother shared the following questions and concerns: whether the patient should be getting outpatient physical therapy services, what to expect regarding her daughter’s progress toward motor milestones such as independent standing and gait, and the potential need for appropriate assistive devices and equipment. This patient was then seen for physical therapy treatments in the nine weeks following this initial evaluation. She was selected for this case report to describe the progress that she made in gross motor milestones to assist with gaining a better understanding of the expected prognosis for early motor development in children with spina bifida and how this varies by specific diagnosis.

As mentioned, the patient in this case has a medical history including L4-5 myelomeningocele that was repaired shortly after birth. In addition, she has a diagnosis of hydrocephalus for which she has a ventriculoperitoneal shunt. She was evaluated at age 26 months for possible right hip dysplasia,

but the acetabular index was found to be within normal limits. Additional diagnoses that the patient has include a speech and language delay, monocular estropia of the left eye, and a neurogenic bowel and bladder for which she has clean intermittent catheterization every four hours. Regarding her social history, the patient lives at home with both of her parents and two older siblings, and she is cared for by her mother during the day while her father is at work. She communicates primarily through facial expressions and does not use verbal communication to interact with the therapist. During this evaluation, it was also noted that the patient requires additional time to process and respond to requests made by the therapist.

At the time of the initial evaluation, the patient was currently receiving physical therapy services through the Area Education Agency (AEA) at a frequency of two times per month. In therapy, she is working on standing balance, cruising, and walking with a reverse walker. She also receives speech therapy through the AEA. In terms of orthotics, the patient has supra-malleolar orthotics (SMOs) on both feet and would soon be getting bilateral ankle foot orthoses.

The short-term goals of the patient's mom and therapist were for the patient to be able to complete sit to/from stand transfers with minimum assistance, maintain standing balance for at least 60 seconds with minimum support, and be able to cruise along furniture with unilateral upper extremity support. All of these goals were set for three to four weeks and they focused primarily on helping the patient be able to improve her functional mobility and engage with the environment to a greater extent. The long-term goals of this patient's family were to be able to complete sit to/from stand transfers independently, ascend/descend ramps and curbs with the use of a reverse walker, maintain standing balance for at least 60 seconds independently and while engaged in an upper extremity task, and be able to ambulate at least 15 feet without the use of any assistance other than her bilateral AFOs. These goals were set for 8 to 12 weeks and also focused largely on promoting safety and greater independence for the patient to complete functional mobility tasks.

Based on the limited amount of research regarding the timing of early motor milestone achievement in children with spina bifida, establishing appropriate goals for the patient based on her specific diagnosis was a challenge for the physical therapist. First, the limited amount of research available notes that there is significant variability in establishing a prognosis for individuals with spina bifida as the specifics of the condition and co-morbidities strongly influence outcomes. Two of the greatest variations in establishing an appropriate prognosis are which lesion level has been diagnosed and whether shunting was necessary for hydrocephalus.<sup>9</sup> In Rachel Thompson and colleagues' article regarding long-term functional outcomes in individuals with spina bifida, the authors found that functional outcomes are associated more strongly with the neurological level of injury than they are with potential hip complications - another problem frequently associated with spina bifida - in adults who have myelomeningocele.<sup>14</sup> This demonstrates that functional outcomes are indeed tied strongly to neurological level, such as lesion level of the myelomeningocele, but this article focused solely on individuals over the age of 18 years. Thus, at the time of this initial evaluation, establishing a prognosis for when the patient with L4-5 myelomeningocele should be expected to achieve the motor milestones of her peers was made difficult by the limited research available for this age range and the significant amount of variability within this diagnosis itself. As a result, the therapist must rely heavily on past clinical experience rather than the current literature to establish an appropriate prognosis for the individual.

The plan of care for this patient emphasized starting outpatient physical therapy to address the above goals that were established based on the hopes of the patient's family. The recommended frequency and duration of treatment were one to two therapy visits per week for eight to ten weeks beginning at the current date. The plan for discharge was that the episode of care will be complete when the patient has met all of the stated goals and no new goals are identified, or when she no longer requires skilled therapy services to progress toward these goals. Additionally, the patient and family were given recommendations to continue using the reverse walker and orthotics at home. The last part of the plan of care included patient and family education on what would take place during physical

therapy treatment sessions, namely functional mobility training including gait, lower extremity strengthening, balance training, and management of the patient's orthotics.

### **Clinical Examination**

At initial evaluation, the patient was evaluated using a variety of tests and measures to establish a baseline for determining progress over time in her achievement of motor milestones. The first assessments completed were on pain, range of motion, and strength. In addition, the patient's alignment and posture, muscle tone, and balance/postural control were assessed. Lastly, her overall functional mobility was evaluated by the therapist.

The first component of the objective examination looked at pain. The patient's mother was asked if she has noticed any concerns of her child having pain as the patient herself was not able to verbally express her pain level due to her age. The therapist also observed for any physical signs of pain that the patient may be experiencing such as facial expression, vocalizations, or compensatory movement strategies related to pain. With both of these assessments, no signs or symptoms of pain were noted.

Range of motion was then assessed in the patient, focusing primarily on passive range of motion completed by the therapist and active range of motion assessed through observation due to the patient's age. Her passive range of motion in her upper extremities was found to be within functional limits bilaterally. Regarding lower extremity range of motion, the patient demonstrated passive range of motion within normal limits for all hip motions, knee flexion and extension, and ankle dorsiflexion. Her plantarflexion was 20 degrees bilaterally with the interphalangeal joints of the toes noted to rest in flexion but able to passively correct to neutral extension. With the passive range of motion assessment, the patient was noted to have decreased tone throughout her lower extremities.

Due to the patient's age of 27 months, formal strength testing was not conducted and this assessment was performed primarily through observation of the patient's functional mobility. Her upper extremity functional strength appeared to be within normal limits as she was able to weight-bear through both arms in the quadruped position and also reach overhead. Observation also demonstrated her ability to activate musculature for hip flexion, knee flexion and extension, and dorsiflexion in both lower extremities. The therapist also noted activation of great toe extension and ankle inversion on the right but not on the left. Although the patient is able to stand in an upright position when she has bilateral upper extremity support, she has difficulty maintaining activation and control of the knees in neutral extension due to quadriceps weakness and significantly limited activation of hip extensors.

The patient's overall alignment and posture were then assessed in a variety of positions. When the patient was non-weightbearing, the resting posture of her feet was in a dorsiflexed position with extension at the metatarsophalangeal joints and flexion at the interphalangeal joints. Her lower extremities rested in a position of hip flexion, abduction, and external rotation when in supine. In a supported standing position, she was noted to have external rotation at the hips with bilateral out-toeing. To assess the patient's balance and postural control, she was placed in a sitting position on the mat and was given unexpected, mild perturbations. In response to these perturbations, she demonstrated good anterior and lateral trunk righting responses to correct her sitting balance. At the time of the evaluation, she was also able to maintain balance in short sitting position on the therapist's leg without back support.

Regarding the patient's overall functional mobility, she has gross motor delay which has limited her achievement of motor milestones. She is able to transition from independent sitting into prone. She can also transition into sitting from prone as well as from a sidelying position. The patient can also transition into a modified quadruped position characterized by significant hip abduction bilaterally. Using environmental supports to pull with her upper extremities in addition to moderate assistance provided at her trunk, she is able to pull to a standing position. She can maintain this standing position without upper extremity support as long as she has moderated assistance from the therapist. In addition, she can cruise short distances of approximately five feet using both arms on a solid support surface while also resting her trunk against the surface and using a side-stepping pattern. At the date of her initial

evaluation, the patient was not able to ascend or descend ramps or curbs independently or with the use of a walker. With gait, she requires moderate assistance provided as trunk and lower extremity support due to weakness in her hip extensors and abductors as well as weakness in her feet and ankles. Based on the objective findings from this evaluation, the patient has a medical history significant for paraparesis - with her left side being weaker than her right - as well as gross motor delay. Due to weakness in lower extremity musculature, primarily the hip extensors and abductors and around her feet and ankles, she would benefit from outpatient therapy services to focus on targeted trunk and lower extremity strengthening to assist with motor skill acquisition. Additionally, these services would allow for monitoring and management of any changes to her current bracing and equipment needs.

### **Clinical Impression**

Due to the limited research specifically addressing expected prognosis for achievement of early motor milestones in children with L4-5 myelomeningocele and hydrocephalus with ventriculoperitoneal shunt placement, the clinician had to rely heavily on clinical judgement to establish an appropriate prognosis. Thus, clinical judgement was the primary factor utilized to establish an appropriate impression and prognostic indicators. First, it was noted that the patient is at risk for adverse change due to associated medical comorbidities related to the diagnosis of myelomeningocele and shunted hydrocephalus. Since she has motor innervation to the L4-5 level, the patient has a good possibility of being able to walk with modified independence using progressively less adaptive support. However, this will largely be influenced by her ability to improve strength in her lower extremity musculature as well as postural stabilization. A variety of factors can also influence this prognosis, including: the capacity for this patient to improve muscle strength in specific lower extremity muscles is unknown at this time as there may only be partial innervation; vision impairments can influence her progress toward improved balance; and cognitive processing can impact her rate of progress toward learning new motor activities. Positive prognostic indicators noted were that the patient has strong family support with a commitment to maximizing her function, and the patient is also followed by other medical specialists to help monitor her condition including visits with a developmental pediatrician, speech therapist, audiologist, occupational therapist, and physical therapist every 6 to 12 months. As mentioned, this impression and the prognostic indicators above were established largely based on clinical judgement as the research in this area of the timeline of early motor milestone achievement in children with spina bifida is limited at this time.

### **Clinical Interventions**

Throughout the episode of care for this patient, a variety of treatment interventions were implemented to optimize the individual's functional mobility, promote the development of early motor milestones, assist with management of equipment needs including orthotics, and serve as a resource to help the family meet their needs and goals. In Dr. Apkon and associates' article, they provide a description of the role physical therapists play in managing and treating individuals with spina bifida as well as a foundation for interventions. First, they note the importance of treating these individuals from an interdisciplinary approach:

A comprehensive team of providers is optimal in the functional management of children with [myelomeningocele] and consists of specialists in orthopedics, developmental pediatrics, rehabilitation medicine, orthotic management, and therapies, including physical, occupational, and sometimes, speech therapy.<sup>1</sup>

For the patient highlighted in this case study, she received evaluations from a neurodevelopmental team every 6 to 12 months for a comprehensive assessment of her development. This team included providers such as a developmental pediatrician, physical therapist, occupational therapist, and speech therapist as recommended by the authors above. However, this neurodevelopmental team also included other providers that were incorporated into these intermittent evaluations as needed, including

audiology, dentistry, and social work. Therefore, the patient was actively involved in receiving thorough assessments to manage her condition. Following each team member's evaluation, the team discussed appropriate diagnoses for the patient as well as a shared plan of care, such as which specialties the patient should continue to see for treatment.

The physical therapy treatments provided to the patient in this case focused largely on functional mobility. These treatment sessions took place two times per week for nine weeks and were then continued at a frequency of one time per week after this period. Each session was one hour and was typically split into the three following focus areas: transitions, balance, and gait. To work on transitions, strengthening exercises and repetition of functional movements were performed. Activities were set up to encourage the patient to complete transitions from supine and prone to/from independent sitting, transitions in and out of quadruped, and progressing through tall-kneeling and half-kneeling positions up into standing. To work on transitions to sitting, the patient was placed in supine/prone and encouraged to reach for an object of interest to encourage her to transition up to a tall kneeling position. Objects would also be placed at a distance from her to encourage her to crawl forward and reach them. Over the nine weeks, the patient progressed from crawling in a modified quadruped position with significant hip abduction to crawling with hips closer to neutral alignment. The patient was also encouraged to complete sit to/from stands from progressively lower surfaces. This activity also started with allowing the patient to use bilateral upper extremity support on a stable surface to assist with the sit to/from stand, progressing to only having an unstable surface available for her upper extremities (such as a two-inch foam mat folded into a triangular shape and standing on edge), and eventually to no upper extremity support. Lastly, she was encouraged to complete transitions from supine/prone up through tall kneeling and half-kneeling to standing by being prompted to toss small plastic toys into a hoop standing two feet tall.

To work on balance, a variety of activities were incorporated that challenged the patient's core and lower extremity strength as well as her postural reactions. Balance activities were completed in standing by having the patient weight-shift and reach for objects to encourage unilateral upper extremity support rather than bilateral support. This was progressed to having the therapist hold a toy in open space and prompting the patient to hold onto and play with the toy which acted as an unstable surface. The plan to progress this activity was to have the patient hold objects while standing independently, but this was not yet achieved during the nine-week period covered in this case report. Balance activities were also completed by having the patient maintain a tall-kneeling position as well as half-kneeling positions on alternating lower extremities while engaged in upper extremity tasks. A similar progression of starting with a stable surface to an unstable surface to not allowing upper extremity support was utilized. Additionally, balance training and postural control were also addressed by having the patient straddle-sit on a peanut ball and maintain her balance while reaching for objects placed at different distances and heights. Assistance provided as trunk support to help maintain balance was progressively decreased over this episode of care as the patient improved in postural stability and balance.

In the treatment of this 27-month old female with L4-5 myelomeningocele, gait was a primary focus that also included the utilization of appropriate orthotics and trialing of assistive devices/technology that would best serve the patient. Table 1 below describes the various gait activities that were incorporated throughout the episode of care. In addition to the described gait training activities, orthotics and assistive technology and equipment were utilized. As mentioned, the patient presented to the initial evaluation with bilateral SMOs. Based on her level of functional mobility, the therapist determined that it would be appropriate for the patient to have bilateral AFOs to provide greater assistance with lower extremity control in weight-bearing positions. Two weeks after the initial evaluation, the patient had new rigid AFOs which she utilized bilaterally. At the initial evaluation, the patient required moderate support for standing as well as for transitioning into a standing position. Two weeks after she received her AFOs, the patient was able to maintain standing position and also transition into standing with minimum assistance.

**Table 1.** Gait Interventions in 27-Month Old Female with L4-5 Myelomeningocele.

Mode	Description
<b>Cruising along stable surface</b>	<ul style="list-style-type: none"> <li>- Patient was encouraged to cruise using upper extremity support on a solid, level surface (kitchen set).</li> <li>- From this surface, she was prompted to reach for items placed at different heights and in different locations along the kitchen set to challenge her ability to cruise, weight shift, and use unilateral upper extremity support rather than bilateral.</li> </ul>
<b>Cruising on variable surfaces with turns</b>	<ul style="list-style-type: none"> <li>- Cruising course was set up for the patient that included solid surfaces at varying heights (tables, chairs), unstable surfaces (a two-inch foam mat folded into a triangle and standing on edge), and gaps over which she was forced to reach between discontinuous surfaces while maintaining balance.</li> <li>- The course was set up to encourage her to make 90- and 180-degree turns while reaching, forcing unilateral upper extremity support rather than bilateral.</li> </ul>
<b>Pushing a shopping cart</b>	<ul style="list-style-type: none"> <li>- The patient pushed a shopping cart with four wheels (unstable) over carpet.</li> <li>- Encouraged to make turns to avoid props/obstacles.</li> </ul>
<b>Obstacle courses</b>	<ul style="list-style-type: none"> <li>- The patient was provided as much support as necessary (decreasing over time) through upper extremities via hand-held assist and/or support at her pelvis.</li> <li>- Course included rubber stepping stones of varying surface textures, challenging step length, weight shifts, and balance; 3"-wide balance beams that were raised 1" off of the ground and were 4' in length; and stairs, both 3" stairs and standard stairs which she was encouraged to ascend and descend with upper extremity support and as much assistance as needed provided at the pelvis.</li> </ul>

Over the nine-week period of bi-weekly treatments, various assistive devices for walking were also trialed. The patient practiced using a reverse walker in six of the sessions and a standard walker in three of them. With the standard walker, the patient made minimal progress and had a tendency to lean forward into the walker, requiring moderate assistance to prevent the walker from sliding too far anteriorly and to help her stay upright. With the reverse walker, the patient was able to progress from requiring moderate assistance to minimum assistance to help control the walker and maintain upright standing balance with gait. Her primary gait deviations with the reverse walker included lateral deviation and a tendency to lift her hands from the handles and reach forward as to assume a quadruped position for crawling. Regarding wheelchair considerations, the patient's mother relied on using a stroller for her and carrying her when necessary. Based on the patient's level of functional mobility, the therapist used clinical judgment on predicting her prognosis - in this case, ability to achieve locomotion with the use of assistive devices rather than a wheelchair - to make the decision not to suggest a wheelchair evaluation at this time. The current available research also notes that forearm crutches are typically used to facilitate ambulation in individuals with L4-5 myelomeningocele.<sup>1</sup> However, the clinician determined that the patient, at 27 months of age, does not yet have sufficient motor control and coordination to utilize forearm crutches in assisting with her gait. Therefore, for the equipment as well as assistive devices and technology for this patient, the episode of care for this case report included the use of bilateral AFOs and trialing of walkers with plans - shared with the family - to attempt forearm crutches when the patient is older and has further improved her strength.

**Outcomes**

In determining appropriate outcome measures for a 27-month old female with L4-5 myelomeningocele, the clinician relied heavily upon tests and measures analyzing the patient's functional mobility. Due to the patient's age, testing measures such as strength and active range of motion were primarily assessed through qualitative observation due to the patient's age and ability to follow specific commands. The individual's passive range of motion was assessed with the patient lying



in supine and with passive movement of each extremity. However, the primary outcome assessed at initial evaluation and compared at the end of this nine-week period was the patient's overall functional mobility, reported in terms of progress toward motor milestones. To determine the individual's progress toward these milestones, she was tested in supine, prone, sitting, quadruped, and standing both with and without support. Using the physical set-up of the environment as well as toys that interested the patient, the therapist assessed the patient in the following abilities: her primary means of mobility when transitioning in and out of supine/prone, transitioning in and out of independent sitting, army crawling or crawling in quadruped, pulling to stand to reach an object of interest, maintaining standing balance with and without upper extremity support, cruising, and walking independently.

In their article regarding the use of qualitative analysis of motor assessment, researchers Gajewska, Sobieska, and Moczko report the following comparison between utilizing a quantitative assessment versus a qualitative analysis when looking at motor development in infants:

Currently available discriminative motor tests specifically assess quantitative measures in comparison with peers. Most of these tests are reliable and validated; however, they only point out whether or not a child has performed the assessed activity. The qualitative assessment of motor performance verifies whether a specific activity is performed properly.<sup>7</sup>

Based on their research, these individuals established a formal assessment sheet for motor performance based on qualitative analysis of motor performance. They found that the qualitative assessment performed by both physical therapists and neurologists had high conformity and was a sensitive tool with good predictive value. Gajewska and colleagues add that "a detailed qualitative assessment allows us to predict the possible delay in motor development with greater accuracy."<sup>7</sup> Although this formal tool was not utilized for the evaluation of the individual of this case report, a qualitative approach to establishing a baseline level of motor performance was utilized.

When comparing the motor milestones of the 27-month old female with L4-5 myelomeningocele at her initial evaluation and then after nine weeks of physical therapy, there were notable improvements in her progress toward motor milestones. Table 2 below describes the specific changes made in the patient's functional mobility. One consideration to note is that her functional mobility status at initial evaluation and after the nine weeks of physical therapy included the use of orthotics. Originally, the patient utilized bilateral SMOs but, after the first two weeks, she was utilizing bilateral AFOs. These orthotics were worn during the assessment of the functional mobility tasks included in table. Mobility levels in supine and in prone remained relatively the same at both time points of this case report. With sitting, the patient made progress in that she was able to transition into and out of sitting from a variety of different positions and she significantly increased her efficiency with these transitions. In quadruped, she progressed by decreasing her excessive hip abduction closer to neutral and went from not moving in the modified quadruped position to moving distances of 10 to 20 feet consistently. In addition, her primary means of mobility shifted from scooting one to three feet to crawling in modified quadruped. Additionally, she progressed from completing pull to stands with moderate assistance to standby assistance, and her standing balance went from requiring moderate assistance to maintain standing position to being able to stand independently for two to five seconds. She made significant progress in her cruising/ambulation as well, as she no longer leaned against surfaces with her trunk, was cruising longer distances and with greater efficiency, could cruise along variable surfaces, and could maneuver gaps in the surfaces as well as reach 180 degrees to a different surface. Overall, the patient achieved a maximum level of being able to cruise up to three steps with unilateral upper extremity support and take one to two independent steps without losing her balance. Regarding gait training with the reverse walker as well as the pediatric-sized standard walker, the patient progressed slightly, improving from requiring maximum assistance to moderate assistance which was provided at her pelvis to help with weight-shifting and prevent anterior/posterior losses of balance.

**Table 2.** Qualitative Description of Motor Skill Development in a 27-Month Old Female with L4-5 Myelomeningocele over the Course of Nine Weeks in Physical Therapy.

<b>Position</b>	<b>Initial Evaluation</b>	<b>Following Nine Weeks of PT</b>
<b>Supine</b>	<ul style="list-style-type: none"> <li>- Able to roll left and right efficiently</li> <li>- Able to roll to prone efficiently</li> </ul>	<ul style="list-style-type: none"> <li>- Able to roll left and right efficiently</li> <li>- Able to roll to prone efficiently</li> </ul>
<b>Prone</b>	<ul style="list-style-type: none"> <li>- Able to roll left and right efficiently</li> <li>- Able to roll to supine efficiently</li> </ul>	<ul style="list-style-type: none"> <li>- Able to roll left and right efficiently</li> <li>- Able to roll to supine efficiently</li> </ul>
<b>Sitting</b>	<ul style="list-style-type: none"> <li>- Transitions into sitting from prone, progresses through a sidelying position; requires significant verbal cuing and extra time.</li> <li>- Transitions out of sitting by leaning anteriorly with her trunk, shifting weight anteriorly to push legs posteriorly, and moving into prone position.</li> <li>- Scoots distances of 1-3' intermittently, primary means of mobility.</li> <li>- Able to maintain independent sitting balance &gt;60 seconds.</li> </ul>	<ul style="list-style-type: none"> <li>- *Transitions in and out of independent sitting efficiently, from various positions, and without verbal cuing.</li> <li>- Able to maintain independent sitting balance &gt;60 seconds.</li> </ul>
<b>Quadruped</b>	<ul style="list-style-type: none"> <li>- From prone, patient can push off of ground through bilateral upper extremities and achieve elbow extension to neutral; she can also flex hips and knees to 90 degrees, but only achieves a modified quadruped position as hips remain abducted approximately 45 degrees bilaterally.</li> <li>- Pt does not crawl in this modified quadruped position.</li> </ul>	<ul style="list-style-type: none"> <li>- *Able to achieve modified quadruped position with hips abducted approximately 15 degrees bilaterally.</li> <li>- *Able to consistently crawl distances of 10-20' in modified quadruped position.</li> <li>- *Primary means of mobility.</li> </ul>
<b>Standing</b>	<ul style="list-style-type: none"> <li>- Can pull to stand using bilateral upper extremities on a stable surface and moderate assistance from therapist, provided at the patient's pelvis/trunk and proximal upper extremities.</li> <li>- Requires moderate support at trunk/pelvis to maintain hands-free standing balance.</li> </ul>	<ul style="list-style-type: none"> <li>- *Able to pull to stand with bilateral upper extremities on a solid surface with standby assistance from therapist; intermittent contact guard assist provided as tactile cuing to encourage her to progress from quadruped &gt; tall kneeling &gt; half kneeling &gt; standing.</li> <li>- *Able to stand independently for periods of 2-3 seconds on average and up to a maximum of 5 seconds.</li> </ul>
<b>Cruising/Walking</b>	<ul style="list-style-type: none"> <li>- Able to cruise along stable furniture using bilateral upper extremities, significant trunk lean against surface, and side-stepping; travels distances of 3-5 feet; does not cruise along surfaces of different height.</li> <li>- Requires moderate support at pelvis and occasionally at feet/ankles when ambulating with support of walker</li> </ul>	<ul style="list-style-type: none"> <li>- *Able to cruise along stable furniture using bilateral upper extremities and without leaning against the surface with her trunk; travels distances of 5-10' consistently and repeatedly.</li> <li>- *Able to cruise along surfaces of variable stability of distances up to 5'.</li> </ul>

	<p>(including reverse walker and pediatric-sized standard walker).                  - Requires maximum assistance when ambulating with the reverse walker and pediatric-sized standard walker.</p>	<ul style="list-style-type: none"> <li>- *Able to cruise along surfaces of variable height over distances of 5-10' consistently and repeatedly.</li> <li>- *Able to reach from one surface to another while cruising, both when there are 1' gaps between surfaces and when she is required to turn 180 degrees.</li> <li>- *Cruises with unilateral upper extremity support for up to 3 steps.</li> <li>- *Will take 1-2 independent steps before losing independent balance.</li> <li>- *Able to ambulate with the reverse walker and with the pediatric-sized standard walker with moderate assistance.</li> </ul>
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*\*Notes change/improvement from initial evaluation.*

The primary outcomes assessed with this patient focused on a qualitative assessment of her progress in motor milestones and functional mobility. Limited research has been conducted on the validity and reliability of a qualitative analysis of the patient's motor development at the two time points, but the clinician and patient/family were nonetheless able to monitor change in functional mobility and measure progress through using a qualitative description of her mobility in and from various positions. Although it was not used in this case, an alternative outcomes measure that could have been used with this patient was the Bayley Scale of Infant and Toddler Development. This scale, which measures development in children ages 1 to 42 months, assesses mental development, behavior, and motor development.<sup>5</sup> As stated in an article by Lomax-Bream and colleagues, "The Motor Scale evaluates skills requiring control of the gross and fine motor muscle groups. The scale does not differentiate between these two sets of skills, but assesses broad motor functioning."<sup>9</sup> The scores for this assessment are then reported as index scores that range from 50 to 150, with 100 being the average when comparing an individual's raw score to the norms for his or her age.<sup>9</sup> Overall, these scales are "known to have high reliability" with the motor scale having a correlation coefficient of 0.77 for test-retest reliability.<sup>5</sup>

A second alternative option that would have been appropriate for the patient of this case report is the Pediatric Evaluation of Disability Inventory - Computer Adaptive Test (PEDI-CAT), which could have been used at both the initial evaluation and at the end of the nine-week time period over which she received physical therapy. This assessment measures a child's ability in three primary areas: daily activities, mobility, and social/cognitive domains. This test can be used in a variety of settings and with a variety of diagnoses, making it appropriate for this 27-month old female with L4-5 myelomeningocele. In addition, the PEDI-CAT can be completed by parent report or based on the professional judgement of physical therapists or other clinicians. One of the applications noted for this test is for the "examination of improvement for an individual child after intervention," making it appropriate for this patient.<sup>12</sup> The test-retest reliability of the PEDI-CAT is also high for each of the four domains when completed by parents, with an intraclass correlation coefficient ranging from 0.96-0.99.<sup>4</sup> Thus, there were alternative options for standardized testing and outcome measures that were both appropriate for the patient's age and diagnosis, as well as reliable in assessing progress with motor development. However, the primary method utilized to monitor this patient's progress toward development of motor milestones over the nine-week timeframe discussed in this case report focused on a qualitative analysis of her functional mobility both in and from various positions.

## Discussion

In their 2014 article “Advances in the Care of Children with Spina Bifida,” Dr. Susan Apkon and colleagues provide a thorough analysis and explanation of the current state of understanding, treatments, outlook, and care for individuals with spina bifida. Similar to the information presented by other investigators, determining the lesion level is the primary factor involved in establishing a prognosis for the individual. As Dr. Apkon and colleagues write, “The motor level can be established based on active isolated movement, which is observed or determined through resisted manual muscle testing, which is typically reliable after age 5 years. Existing strength greatly affects the individual’s functional mobility and ambulation status.”<sup>1</sup> Again, due to the patient’s age in this case report, manual muscle testing was not formally completed and the therapist relied primarily on observation of functional mobility to assess strength. Based on their review of the current literature, Dr. Apkon and her colleagues predict that, with a lower lumbar lesion in myelomeningocele such as L4-5, the child should have hip abduction, knee flexion, ankle inversion and dorsiflexion, and toe extension.<sup>1</sup> As discussed above, the patient in this case study demonstrated the ability to activate hip flexion, knee flexion and extension, and dorsiflexion bilaterally; however, great toe extension and inversion were only activated on the right and were not observed on the left. Regarding functional mobility, the authors of this 2014 article predict household and community ambulation with the likely use of a wheelchair for long distances. In addition to a wheelchair, they predict that the child will likely need forearm crutches and ankle-foot orthoses.<sup>1</sup> The patient in this case presented with bilateral SMOs but was prescribed bilateral AFOs shortly after starting physical therapy. She did not have a wheelchair as she was primarily pushed around in a stroller by her mom or carried when out in the community. Regarding the use of forearm crutches, these were not yet attempted in therapy due to the patient’s limited balance in upright positions and coordination of her extremities, as indicated by her difficulty ambulating with walkers at this time. However, at the end of this nine-week period, it was determined that forearm crutches will be appropriate to try in the future once the patient has developed further motor skills and coordination.

When treating this patient with L4-5 myelomeningocele, a primary question of the patient’s family was what to expect in terms of their daughter making progress toward independent walking and motor milestones based on her diagnosis of gross motor delay compared to other children of 27-month age. In their article discussing neural tube defects such as spina bifida, Dr. Hope Northrup and Kelly Volcik note that lesion level is the most significant factor in determining if an individual with myelomeningocele will achieve independent ambulation. They report, “children with low-level lesions (low lumbar and sacral levels) are usually able to walk, although they may need the help of braces and/or crutches.”<sup>11</sup> The investigators also note that for lesions in the mid-lumbar region, more significant bracing and assistive devices are typically required and, if the lesion is in the upper lumbar region, children will often rely on wheelchairs as their primary means of mobility. Based on their review of the literature and update on the current understanding of spina bifida, they provide the following estimates regarding the likelihood of children walking based on the level of their myelomeningocele: “sacral, 100%; low lumbar, 95%; high lumbar, 30%; and thoracic, 33%.”<sup>11</sup> Based on this information, in combination with the information presented by Dr. Apkon and colleagues, the therapist can reasonably expect to include ambulation in the prognosis for this patient, yet it is also important to inform the family of the role that assistive devices will play in achieving gait.

As mentioned regarding orthotics and assistive devices, the patient in this case presented to therapy with bilateral SMOs but was not yet ambulating independently. Dr. Apkon and associates report that, “For children with midlumbar to low lumbar spina bifida, the use of a solid AFO has been shown to increase stability during walking, with increased stride length, increased speed, [ . . . ] and increased power at terminal stance.”<sup>1</sup> Orthotics for patients in toddlerhood are typically custom-molded and adjusted as needed, typically every three months and with full replacements when they no longer fit the child.<sup>1</sup> Although research has shown that children with L4-5 myelomeningocele may require a wheelchair, walker, or forearm crutches, the majority of research on assistive technology for individuals with spina bifida focuses on those who are older and past toddlerhood.<sup>1,8</sup> Dr. Kurt Johnson and colleagues conducted research on the use of assistive technology among 348 individuals with spina

bifida and determined that wheelchairs - including both electric and manual - were used by 57% of the participants, braces were used by 35%, and assistive devices for gait were used by 23%. Although this data is helpful in determining the likelihood that the patient in this case will utilize bracing and/or assistive technology in the future, this study was conducted among participants aged 13 to 27 and did not discuss those in childhood.<sup>8</sup>

Dr. Apkon and associates further state that the primary role of physical therapists in treating individuals with spina bifida varies widely depending on the patient's age and functional level, but it should always focus on achieving function such as independence in mobility.<sup>1</sup> According to an article by Laura Lomax-Bream and colleagues on influence of spina bifida on a child's development in infancy and toddlerhood, "a key motor milestone is the onset of self-generated locomotion (creeping, crawling, walking)."<sup>9</sup> The research conducted by these individuals found that achieving independent locomotion, regardless of the means, plays a large role in the development of motor, speech, and cognitive milestones.<sup>9</sup> Nicole Mueske and colleagues acknowledge that, in children with myelomeningocele who do ambulate, these children frequently demonstrate abnormalities in their gait pattern based on their lesion level and muscle function. As a result, they report that it is essential for providers such as physical therapists to identify gait abnormalities and - based on the cause of changes to their gait pattern - treat them in a way that will maximize the individual's functional mobility.<sup>10</sup> Thus, significant amounts of research report the importance of working on mobility when treating children with myelomeningocele.

When assessing this individual's progress toward the goals originally established by the child's parents and physical therapist, there were areas in which she achieved her goals and other areas where the goals were too aggressive. Again, this was in part related to the difficulty of establishing a timeline of motor milestone development and prognosis for individuals with L4-5 myelomeningocele over the first three years of life. One of the goals for this patient was to complete sit to/from stand transfers with minimum assistance in 3 to 4 weeks and independently in 8 to 12 weeks. At initial evaluation, the patient completed sit to/from stand transfers with moderate assistance provided at the pelvis and the knees. After three to four weeks, she continued to require moderate assistance; however, by week nine she was able to complete these transitions with contact guard assistance. Another goal was to maintain standing balance for at least 60 seconds, with a short-term goal of doing this with minimum assistance and a long-term goal of standing this long independently. She achieved her short-term goal of standing at least 60 seconds with minimum assistance after seven weeks of physical therapy and, at week nine, she was able to stand independently for a maximum of five seconds. Regarding the goal to cruise along furniture with unilateral upper extremity support, this goal was not achieved in the three- to four-week timeframe as planned, but she was able to complete this task for up to three steps by week nine. The goal of ambulating at least 15 feet without the use of any assistive device other than bilateral AFOs was not achieved during this timeframe, yet the patient was able to take one to two independent steps at the end of nine weeks. She did not achieve the goal of ascending/descending ramps and curbs with the use of a reverse walker during this window. Overall, qualitative analysis of her achievement toward these motor milestones suggests that the patient was able to achieve the majority of goals established by the therapist, but the goals were achieved after a longer period of time than anticipated. For example, although the short-term goals set for this patient were eventually achieved, it occurred later than the three to four weeks for which the goals were originally set. When considering goal setting, it is important to establish challenging goals to maximize the treatment sessions and outcomes, but one must also consider the overall delay in progress toward and achievement of these milestones as seen in this case.

According to the CDC, toddlers typically develop a variety of specific motor milestones during their second year of life. The physical skills that children often develop and exhibit during the age range of 24 to 36 months include: standing on tiptoes, kicking a ball, ascending and descending stairs with upper extremity support, initiation of running, throwing a ball overhand, and climbing onto and off of furniture.<sup>2</sup> These motor milestones are indicative of the functional skills that children at the same age as the patient in this case report typically achieve. As mentioned, the patient in this case with

myelomeningocele was able to perform the following motor skills at age 2 years and 5 months (after nine weeks of therapy): transition in and out of sitting independently and maintain independent sitting position; crawl 10 to 20 feet in modified quadruped, the patient's primary means of mobility; cruise with bilateral upper extremity support and without trunk lean against surface for distances of up to 10 feet; maintain independent standing for a maximum of five seconds; and take one to two independent steps. She was also able to ambulate up to 30 feet with the reverse walker and moderate assistance. However, she was not able to ascend or descend ramps or curbs and required at least moderate assistance with stairs and bilateral upper extremity support. In comparing these motor milestones with the CDC recommendations of typical timing of motor skill development, this patient has achieved milestones consistent with 9 months of age and was beginning to achieve those consistent with 12 months of age. At nine months, the CDC suggests that a child should be able to transition into sitting, sit without support, and crawl, all of which this patient was doing at the end of this nine-week time frame. At 12 months, they report that a child should be able to cruise, take a few steps without upper extremity support, and may stand alone.<sup>2</sup> This patient was cruising consistently, but she was only able to take one to two independent steps and maintain independent standing for a maximum of five seconds. Due to the patient's L4-5 myelomeningocele, she was therefore achieving motor milestones around 10 to 12 months of age although her current age was 29 months.

In their research on the influence of lesion level and shunt effects in motor development of children with spina bifida, Laura Lomax-Bream and colleagues report that children with spina bifida have decreased motor performance on the Bayley Motor Scale compared to age-matched peers without this condition. They further add that "having a shunt or a higher lesion level also predicted significantly lower levels of performance than having [spina bifida] without a shunt or having lower lesion levels, respectively."<sup>9</sup> In their research, they found that children without spina bifida had typically developed the majority of early motor milestones by age 36 months and, at that point, began to plateau in their growth or achievement of new milestones. Children with spina bifida, however, did not show a plateauing in motor skill development at age 36 months. The researchers propose that this is likely because many typically-developing children have already achieved the majority of early motor milestones at three years of age whereas those with spina bifida are still developing many of these skills such as independent locomotion.<sup>9</sup> As in the case of this patient with L4-5 myelomeningocele, she was significantly behind her peers in terms of motor milestone development as noted above. Although she was able to crawl approximately 20 feet in modified quadruped, this individual was on the verge of achieving independent locomotion for functional distances and was not yet ambulating more than one to two steps independently. Lomax-Bream and colleagues add that, although children with spina bifida continued to demonstrate development of new motor skills up through 36 months of age while those without spina bifida plateaued, the individuals without spina bifida did maintain significantly higher levels of skill performance on the Bayley Motor Scale throughout the first 36 months. Overall, children with low-level lesions continued to demonstrate growth in early motor skills throughout their first three years of life but they were significantly behind their peers in how early they were able to achieve these skills.<sup>9</sup>

## Conclusion

The purpose of this case report was to describe the development of motor milestones in a 27-month old female with L4-5 myelomeningocele in order to improve the current understanding of expected prognosis for early motor milestone development in children with spina bifida. More specifically, this case presented a patient with lower lumbar myelomeningocele who had also had a shunt placed. Based on the research described above and the case of this patient, it is evident that this individual was significantly behind her age-matched peers in terms of motor skill development. However, at the end of the nine-week time frame over which this case was assessed, the patient was on the verge of achieving independent locomotion for functional distances which has been shown to be a catalyst to further motor development.<sup>9</sup> Thus, when establishing a prognosis and goals for this patient going forward, the therapist can reasonably expect the patient to continue progressing in her

achievement of early motor skills through at least her first 36 months. Future research in this area - specifically to assess timing of achievement of motor skills over the first six years of life in those with spina bifida - would be beneficial to providers to establish an appropriate prognosis for patients and their families. In addition, it is essential to recognize the variability in lesions when conducting future research as the prognosis will likely vary significantly depending on level and severity of the lesion, directly impacting innervation for motor function. This case thus serves as a supplement to the limited amount of literature available to providers for establishing a prognosis for timing of early motor development in children with spina bifida.

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