Interdisciplinary, Intensive, Activity-Based Treatment for Intrauterine Spinal Cord Infarct: A Case Report

Cara E. Felter, PT, DPT, MPH,[1] Erin E. Neuland, PT, DPT,[2] Shannon C. Iuculano, MS, OTR/L,[2] and Janet Dean, CRNP[2]

[1]Department of Physical Therapy and Rehabilitation Science, University of Maryland, School of Medicine, Baltimore, Maryland; [2]International Center for Spinal Cord Injury, Hugo W. Moser Research Institute at Kennedy Krieger Institute, Baltimore, Maryland

Intrauterine spinal cord infarcts (IUSCI) with resulting tetraplegia are extremely rare, and there is minimal evidence describing outcomes in this population. This case describes the functional progress of a 3-year-old girl born with IUSCI who participated in activity-based therapies (ABT). Children have developing nervous systems and are particularly suited to benefit from ABT. Over the course of treatment, the child in this case has demonstrated improvements in developmental milestone achievement including fine and gross motor skills and social/cognitive development. Intense, interdisciplinary ABT should be considered for the treatment of children with IUSCI. **Key words**: *activity-based therapy, case report, early mobility, intrauterine spinal cord infarct*

Forms of intrauterine or perinatal spinal cord injury (SCI) related to fetal position and birth trauma have been described extensively.[1-11] However, intrauterine spinal cord infarcts (IUSCI) with resulting tetraplegia are extremely rare,[12] and there is minimal evidence describing outcomes in this population, especially functional therapeutic outcomes.[13-18] Literature describing IUSCI is mainly limited to early medical intervention as most individuals affected do not survive early infancy.[18-21] Spinal cord infarcts are rare across the lifespan, accounting for only 0.3% of strokes.[22] Limited knowledge exists regarding long-term outcomes for individuals with spinal cord infarcts.[23-25]

Regardless of etiology, SCI critically impacts children's musculoskeletal and social development. Therapy focuses on reducing secondary complications including pressure ulcers, pain, bowel and bladder dysfunction, urinary tract infections (UTIs), contractures, scoliosis, spasticity, and depression. Currently, focus is shifting to include neuromuscular activities below the level of the lesion to capitalize on experience-dependent neuroplasticity.[26] This concept is embodied in activity-based therapies (ABT), which are "interventions that provide activation of the neuro-muscular system below the level of lesion with the goal of retraining the nervous system to recover a specific motor task."[27(p185)] ABT includes the principles of weight bearing (WB), functional electrical stimulation (FES), locomotor training (LT), massed practice (MP), and task-specific practice (TSP).[28] The term ABT has been used synonymously with activity-based rehabilitation. In children whose injuries occurred before or at birth, there would not be a baseline of motor function to rehabilitate. In such cases, ABT may be thought of as activity-based *habilitation* with the same goal of improving motor function. For children with SCI and other paralytic conditions, habilitative goals may include those for muscle use below the lesion including fine and gross motor skills,[29-34] social and cognitive function,[35,36] and improved physiologic function.[37-39] LT, FES, and early wheeled mobility may be among the interventions utilized. ABT can be performed in a unidisciplinary manner (ie, occupational therapy [OT] in isolation) or in an interdisciplinary manner (ie, OT, physical therapy [PT], aquatic therapy [AT], medicine, etc, working interactively toward patient-centered goals).

Corresponding author: Cara E. Felter, 100 Penn Street, Baltimore, MD 21201; phone: 410-706-7165; email: cfelter@som.umaryland.edu

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The patient may benefit from long-term gains in neurorecovery from ABT (ie, restored ambulation) while taking advantage of short-term strategies for independence (ie, learning to independently use a wheelchair). This case describes the functional progress of a 3-year-old girl born with IUSCI who participated in intensive, interdisciplinary ABT.

Presentation of the Case Report

The child was delivered by caesarean section at 35 weeks due to breech presentation and loss of fetal movement. She was born apneic without response to tactile stimulation. Her brain magnetic resonance imaging (MRI) was normal, and spine MRIs were consistent with SCI from the cervicomedullary junction to T2 with sparing at C3-C5. She was initially diagnosed with in utero transverse myelitis and received IV steroids and IVIg. At 1 month old, she was deemed medically stable and discharged to home; however, she was unable to perform the expected gross motor functions of a 1 month old. Following discharge, she presented for two brief hospitalizations for steroid-induced gastritis, failure to thrive, and UTIs. At 9 months, a follow-up MRI showed spinal cord volume loss from C4-T1 and she was evaluated in a neuroimmunology clinic. Based on prenatal history, MRIs, electromyelogram, nerve conduction study, and laboratory testing, her diagnosis was changed at that time to IUSCI.

At 11 months old, she could not perform any expected developmental milestones, so she began a 4-week course of inpatient ABT 6 days per week, 2 to 3 hours per day. Upon inpatient discharge, her caregivers had been trained in a home program to encourage developmental milestone achievement; however, she still required assistance with performing all developmental skills. At 12 months, she enrolled in outpatient therapy twice weekly for 4 months to promote motor skill acquisition and refine the home program. At 15 months, she transitioned to weekly clinic-based visits with an extensive home program for the rest of the week. Outpatient weekly, intensive interventions (5 hours per visit, including 2 hours of OT, 2 hours of PT, and 1 hour of AT) were complemented by in-home interventions carried out by the family, home-based therapists, and other caregivers. Activities practiced at nearly all visits and the relationship of those activities to the ABT principles are outlined in **Table 1**. A total of 146 OT visits, 120 PT visits,

Table 1. Interventions, duration used within an average session, and corresponding ABT principle(s)

		ABT principle reinforced				
Activity-based intervention	**Session duration**	**FES**	**WB**	**TSP**	**MP**	**LT**
Powered mobility training with a modified power wheelchair with joystick control	30-45 min			X	X	
TSCS during BWSTT (using Walkable pediatric harness/treadmill system)	30-45 min	X	X	X	X	X
TSCS during over-ground training (using Rifton Pacer Gait Trainer)	30-45 min	X	X	X	X	X
Body weight supported positioning using a harness to attain quadruped for developmental skill acquisition (ie, crawl) and FES to facilitate required UE, LE, and trunk muscles	20-30 min	X	X	X	X	X
Sensory input via weight bearing activities (ie, supported standing while teaching use of prone mobile stander) with FES for motor facilitation of LEs and trunk	30-45 min	X	X	X	X	
Sensory input via weight bearing activities (ie, sit to stand, supported standing, or quadruped in the aquatic environment)	30-40 min		X	X	X	
Functional mobility training of developmental skills with FES to facilitate UE, LE, and trunk muscles (ie, rolling and supine-to-sit trained on land and in aquatic environment)	30-40 min	X[a]	X	X	X	X

Note: BWSTT = body weight supported treadmill training; FES = functional electrical stimulation; LT = locomotor training; min = minutes; MP = massed practice; TSCS = trasnscutaneous spinal cord stimulation; TSP = task-specific practice; WB = weight bearing.
[a]Land only.

and 82 AT visits were completed between June 2015 and January 2018. Frequency and duration of the visits were based on available literature for similar conditions[29,33,35-44] as literature specific to IUCSI is very limited. The child's tolerance for therapy and the family's schedule also influenced the plan of care.

The child was routinely assessed using the Gross Motor Function Measure-88 (GMFM-88),[45] which is comprised of 88 gross motor skills that should be attained by 5 years old in typically developing children. The child's first assessment was performed upon inpatient admission in April 2015, and the most recent was conducted January 2018. She has shown steady progress on this measure, except for two instances of decline (see **Figure 1**). In one of those, the assessment was performed by a different evaluator; the other was after a 2-month break from therapy due to hospitalization for respiratory failure. Most notably, she has made progress with neck strength and control, rolling, sitting balance, and prone scooting. She has also demonstrated progress with transitions from sitting to supine but is limited due to upper extremity weakness.

The child has also been evaluated using the Physical Abilities and Mobility Scale (PAMS).[46] This scale is a 20-item measure created to complement the WeeFIM and is geared for children aged 2 years and up. Children are rated on a scale of 1 to 5 to determine recovery after injury, level of caregiver assistance, and tolerance to activities (ie, positioning, wheelchair use). It is primarily used in inpatient settings but is being developed for outpatient settings. The child is scored out of 100, with 0 being the most impaired and 100 being no impairments. Scoring allows for the use of compensatory movements to perform the tasks. Over 2 years, the child demonstrated improvements in the following domains: tolerance to positioning, head control, trunk control, rolling, transitioning supine/sit, and supported standing (see **Figure 2**). The PAMS was performed twice on inpatient and twice on outpatient. The large gap in time is due to the measure not initially being utilized in outpatient settings.

Clinically, she developed lower extremity (LE) sensory responses to tactile stimulation, volitional stepping, trunk and neck strength sufficient to support herself against gravity for up to 40 seconds, and upper extremity (UE) bilateral coordination for developmental play and fine motor skills. She achieved developmental milestones in functional

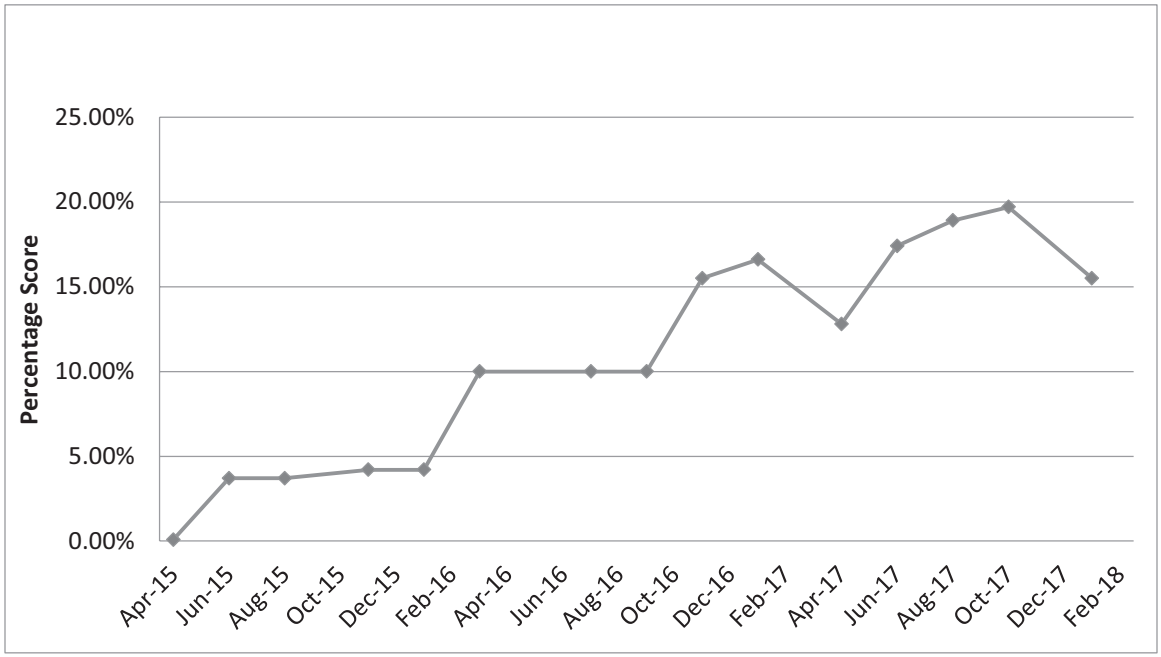

Figure 1. Gross Motor Function Measure-88.

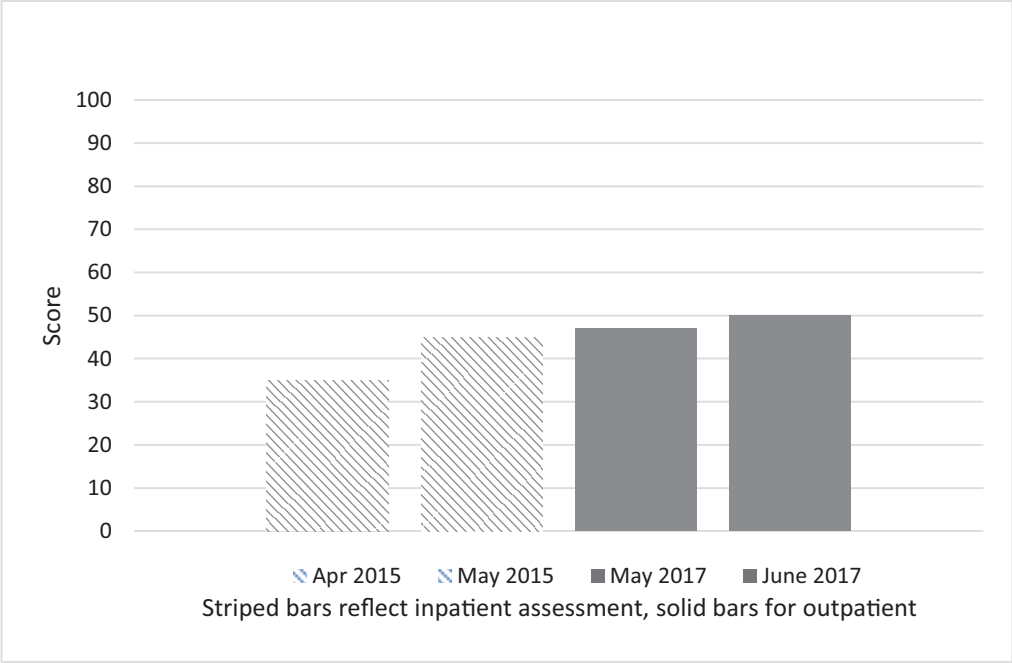

Figure 2. Physical Abilities and Mobility Scale.

mobility (ie, rolling supine to side-lying, sitting 5 minutes wearing a dynamic trunk orthosis [SPIO vest] with minimal assistance), social interactions (attending preschool with typically developing peers using her power wheelchair with supervision, playing hide-and-seek with friends), and UE function (self-feeding of finger foods and using a spoon with her dominant hand). She demonstrates spontaneous stepping with body weight supported treadmill training (BWSTT) combined with transcutaneous spinal cord stimulation (TSCS).[47] She began utilizing FES cycling and demonstrates reciprocal LE cycling activity. She has improved bladder sensation, which will be useful for continence as she can now recite the steps for intermittent catheterization (IC) and is beginning to work on opening packages for ICs. An additional goal of OT has been to assess, fabricate, and modify orthotics for her UEs to prevent contractures and increase function. While ABT promotes optimal neuromotor function through neuroplasticity, maintaining range of motion complements this goal and will allow for options like tendon transfer surgery should that be necessary to promote additional UE function in the future.

The child's parents and caregivers were trained in the implementation of activities (ie, prone mobile stander, FES, functional mobility, use of splints/orthoses) to carryover concepts from the weekly sessions into daily life at home, in her community, and at her preschool. The family demonstrated exemplary dedication to the program, averaging 10 hours per week of home program activities in addition to therapy visits.

Discussion

Evidence regarding IUSCI is very limited. Based on our review of the literature, we believe this may be the youngest reported case in which an ABT approach has been initiated. Children have developing nervous systems and are particularly suited to benefit from the technologies and advances utilized in an ABT paradigm.[48] Young children, especially infants, have shown better recovery than expected relative to the severity of their SCI, and this recovery can continue over many years due to increased neuroplasticity in the age range and, in part, due to their developmental processes.[49]

Infants have a very immature central nervous system that is undergoing reorganization and modification. This may mask the level of their injury due to the decreased myelination that slows signals from the brain to the spinal cord.[50] As their nervous system develops, we may find they have undamaged areas of the spinal cord that were masked by their immaturity. It has been determined that young children have excessive neuronal growth and connections. These connections will be naturally pruned from the central nervous system over time, in a nonjudgmental fashion.[51] This means we can take advantage of the excess neurons for the child's benefit.

We are also limited in our ability to fully assess children's level of function due to their limited ability to follow directions. Evaluations in this population are further impeded by a lack of appropriate outcome measures. For example, based on age, standardized OT assessments were not completed for this child due to the lack of available normative data. As for measuring gross motor skills, the GMFM-88 has only been validated for children with cerebral palsy and Down syndrome. The mechanism of this child's injury was not cerebral; however, it was acquired just before birth and impacts performance of the items tested with the GMFM-88. Therefore, we have found it useful to track her individual progress over time in the absence of comparable data. Specific developmental measures do not exist for children with IUSCI. Despite these challenges, clinical observations described above are sufficient to detail functional progress.

The child in this case has actively participated in ongoing therapy consistently for 3 years, demonstrating steady improvements in functional mobility and social interaction, as well as avoiding multiple hospitalizations which are usually seen in this population.[52,53] Over 3 years of ABT, she experienced only one hospitalization. This was due to respiratory syncytial virus and pneumonia and resulted in a 2-month interruption in ABT to allow her to recover. Upon return to ABT, it was noted that a significant decrease in trunk strength and endurance had occurred.

In this specific case, ABT did not *restore* function as IUSCI had impacted the child's baseline. Rather, the neurological and musculoskeletal system were trained to function as intended. While some compensatory aids were used, they were thoughtfully chosen to complement the ABT principles. For example, to participate in ABT the body must be free to move. However, this child could not achieve optimal trunk posture without support. A dynamic trunk orthosis was chosen over a rigid trunk orthosis to offer support while not completely restricting freedom of movement. Further, we acknowledged the need for short-term mobility and independence to promote social-cognitive development, so short-term goals were addressed (ie, use of wheeled mobility) while still incorporating ABT concepts such as TSP and MP.

This case supports the use of ABT to achieve constantly evolving therapeutic and functional goals. We believe that the gains made by the child are attributable to a combination of factors, not least of which is the consistent commitment of her family who regularly travelled to therapy sessions and performed the home program. This case provides a model to address therapy goals by coupling weekly intensive treatment with a home program to demonstrate meaningful progress for this child and her family. While IUSCIs are rare, the model implemented suggests possibilities for other children with early-onset neurological insult. Establishing a balanced, sustainable therapy program that focuses on improving function not only through compensation is vitally important to facilitate adaptive motor patterns in young children.

Conflicts of Interest

The authors report no conflicts of interest.

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