Spinal Cord Injury in the Child and Young Adult

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Case vignette
Kyle, a male aged 4 years 6 months, suffered a spinal cord injury owing to an accidental gunshot wound 16 months ago. The locomotor training occurred five times per week for 76 sessions. His initial lower extremity motor score (4/50) and the time since the injury (16 months) both supported the presumption that Kyle was unlikely to recover his walking ability. He used a manual wheelchair permanently for mobility and had received the best current standards for therapy and equipment including a standing frame, wheeled stander, and reciprocating gait orthosis. Initially, locomotor training involved 1 hour of step and stand retraining on the treadmill, with approximately 30 minutes of each, followed by attempts to walk on the ground using the principles of locomotor training (see Table 28.1). The locomotor training used with Kyle had three components: (1) intense retraining of the neuromuscular system, which occurs on the treadmill with partial body weight support and manual facilitation by trainers, (2) transfer and assessment of walking skills on the ground (off the treadmill), and (3) daily life activities to guide choices supporting continued application of locomotor training principles, training, and skill progression outside of the clinic, termed community integration (Harkema et al 2011). From sessions 1 to 33, with the exception of successful thoracic extension, Kyle did not volitionally contribute to movement during locomotor training. From session 22, an emphasis was placed on augmentation of hip flexor and tibialis anterior cues while standing in a diagonal stride position during weight shift to facilitate a flexor response. In session 29, a right leg step without cuing was seen. This ‘spinal step’ occurred on the ground after a facilitated left leg step. It was characterized by synergistic hip, knee, and ankle flexion and Kyle was not aware it had occurred. In session 33, after being placed in a diagonal stance position, Kyle initiated and took seven independent (no cuing), consecutive steps on the ground using a postural control walker for balance. After this, walking ability progressed daily up to the final session (76) when he could initiate and sustain independent mobility with a reverse rolling walking. Several months later, after returning home, Kyle entered kindergarten as a full-time ambulator with a walker. Lower extremity motor scores
were unchanged from prelocomotor training (4/50) and did not reflect the significant change in capacity to generate steps, suggesting the significant contributions of the neural system below the level of the lesion to walking recovery in this child (Behrman et al 2008).

Introduction
There is little doubt that the central nervous system (CNS) is capable of significant plasticity. The evidence is clear that neural activity produced by experience can alter connections and have lasting effects. Although most extensively documented for learning and memory in adults (Buschkuehl et al 2012, Caroni et al 2012), the literature is rapidly expanding, showing that activity also can affect spinal circuitry (Edgerton and Roy 2009, Wolpaw 2010). Furthermore, plasticity is not a unique feature of the normal nervous system. Plasticity, in a number of systems, appears to correlate with, or has the potential to support, beneficial functional outcomes (Onifer et al 2011). Thus, the challenge for neuroscience and rehabilitation is to harness and drive plasticity toward end points that support desirable functional outcomes after spinal cord injury (SCI).

Neuroplasticity
Continually mounting evidence for therapeutically induced behavioral recovery after experimental SCIs in animal models provides a strong stimulus for the rapidly growing interest in, and push for, bench-to-bedside application for adults with SCIs (Kleitman 2004). Of particular interest are studies showing the effects of hind limb step training using a treadmill with animals that have anatomically complete spinal transections above the spinal centers innervating the hind limbs (Lovely et al 1986, Edgerton et al 2004, Ichiyama et al 2008, Courtine et al 2009, Rossignol and Frigon 2011, Rossignol et al 2011). Not only does stepping occur and improve in these animals with intensive training, but also improvements in functional performance are accompanied by changes at the circuitry (Courtine et al 2009) and molecular (Tillakaratne et al 2002, Wang et al 2006, 2009) levels. This work underlines the capacity of the spinal cord to produce basic stepping, independent of descending connections, and it shows that the specificity of the task appears to be key and intensive practice of non-task-specific activities may prevent desired task recovery and performance (Hodgson et al 1994, García-Alías G et al 2009). These findings provided the fundamental framework for locomotor training, which has become a standard of practice delivered to some adults with SCI across select sites throughout the Christopher and Dana Reeve Foundation NeuroRecovery Network (Harkema et al 2012a). Coupled with increasing evidence for training effects on neurotrophin expression (Gómez-Pinilla et al 2002), the possibility for promoting effective functional repair in the spinal cord with rehabilitation and other therapeutic modalities continues to gain more attention. Typically, advancements in the pediatric setting follow behind those made in research and clinical programs for adults. This is not surprising as the US SCI Model Systems are adult based, there are fewer children injured annually, and there are challenges associated with the inclusion of growing children in clinical trials.
Implications for rehabilitation

The emerging understanding of plasticity dramatically changes the way functional potential after SCI should be viewed and opens up opportunities for new rehabilitation paradigms. The current standard of care for rehabilitation of the patient with an SCI is based on the premise that the consequences of SCI (e.g. paralysis, weakness, sensory loss) may be recovered to some degree, but that compensatory strategies will be needed for mobility and function. Thus, patients with SCIs typically are taught and use compensatory movement strategies alone or in combination with adaptive devices to achieve some level of functional independence. Adaptive equipment used for upright postures and mobility has traditionally included static and passive standing frames and braces with walkers or crutches (see Chapter 22).

Although the intent of adaptive equipment is to enhance activity and function, it may be inconsistent with recovery-based approaches. The long-term effects of musculoskeletal inactivity and/or altered activity have an impact on more than just the nervous system. Immobility and a lack of active load bearing through the legs are correlated with multiple negative events including fractures, bone demineralization, osteopenia, and contractures (Giangregorio and McCartney 2006, Jiang et al 2006). Furthermore, the impact may be greater in children owing to ongoing skeletal growth and maturation processes (Vogel et al 2004, 2012, Parent et al 2011). Thus, one of the greatest challenges of rehabilitation, using a neurorecovery paradigm, is to determine how to best guide plasticity to maximize positive functional recovery and minimize dysfunction (Garraway et al 2011, Ferguson et al 2012a,b). The experiences that promote and drive changes in circuitry are key.

Based upon our knowledge of activity-dependent plasticity, the activity most engaged in (e.g. time, repetitions) will most strongly direct changes in, or maintenance of, synaptic connections. Thus, incorporating therapeutic elements into daily hours outside of therapy, in addition to during therapy, may more readily guide circuitry changes that will support greater neurorecovery. The critical threshold for tipping the balance toward change required for recovery or integration of a new skill is unknown and may even vary based upon the task. It is critical to appreciate the bidirectional relationship between plasticity and the desired activity or function. Activity drives plasticity and plasticity (circuitry modification) drives the behavior. This bidirectional relationship is well appreciated during nervous system development (Shatz and Stryker 1978, Yao and Dan 2005) and is critical for the continual shaping, refinement, and reinforcement of both the desired behavior and the underlying anatomical substrates.

Recovery-based rehabilitation is best designed when it promotes optimal CNS function in support of preserved, recovered, and new behaviors. The injured, remodeled, or essentially ‘new’ CNS after SCI (Edgerton et al 2004) is influenced not only by current experiences, but also by experiences before SCI. Although individuals injured as adults will have prior experience with all basic motor activities, a child may not. Dependent upon the child’s age at the time of his or her injury, their CNS may or may not have prior experience, for example, with independent sitting, crawling, standing, reaching, walking, or running. We do not yet understand how experience or lack of experience might be important in choice or design of therapeutic approaches. Preliminary data from our group show that a lack of prior walking experience does not preclude development of walking after SCI (see Fig. 28.1) (Howland et al 2010, 2011).
Sensory input is a critical component of any motor performance. A sensation may not only be the trigger for movement, but also provide feedback about ongoing movement so that movement features can be adjusted to achieve and perpetuate successful performance. The CNS circuitries that are necessary for many lower extremity reflexes, as well as basic stepping, are located within the spinal cord and may be activated by sensory cues via the peripheral nervous system. Studies assessing the H-reflex and basic stepping performance clearly show that, by using peripheral sensory cues on the lower extremities, basic reflex and stepping patterns can be modified in the neurally intact and the isolated caudal spinal cords (Nielsen et al 1993, Chen and Wolpaw 1995, Rossignol et al 1996, Thompson et al 2009). These studies, along with others (Lovely et al 1990, De Leon et al 1998), also show that the spinal cord is trainable and provide the basis for clinical approaches, which initially target activation of the spinal cord below the level of the injury using sensory cues which activate cutaneous and subcutaneous receptors, in particular the mechanoreceptors.

Based on studies in rats and cats (Rossignol et al 2011) and humans (Nadeau et al 2010), the caudal spinal cord contains a central pattern generator for locomotion. The extensive network of interneurons that comprise the central pattern generator responds to, and organizes its activities based upon, sensory information. It then interacts with motor neurons to produce leg movements associated with walking (Fig. 28.2).
Neurologic Recovery and Restorative Rehabilitation

Based upon our knowledge of activity-dependent plasticity and the neural control of walking (Roy et al 2012), the current activity-based locomotor training approach was developed for adults (Behrman and Harkema 2000, Behrman et al 2005, Harkema et al 2011). The key aspect of this training approach is its application of intense, walking-specific sensory input and practice on a treadmill using an adjustable body weight support system to promote normal movement patterns of the trunk, pelvis, arms, and legs. The treadmill facilitates intense walking practice. The repetition that can be achieved is critical in facilitating and establishing the circuitry required to support walking (Hebb 1949, Cooke and Bliss 2006). Equally important is the ability to adjust body weight support (Harkema et al 1997), alter treadmill speed (Beres-Jones and Harkema 2004), and enhance other sensory inputs through manual cues (Ferris et al 2004, Galvez et al 2007) provided by trainers. Repetitive training with constant reassessment of the patient’s neuromuscular capacity allows for daily adjustment of the sensory cues to continually challenge and promote the patient’s capacity. Immediately after training on the treadmill, the locomotor training approach includes evaluation and practice of walking, standing, and sitting activities on the ground on a daily basis (see Fig. 28.3). Locomotor training is

Fig. 28.2 After spinal cord injury owing to disruption of descending signals from the brain to the spinal cord to drive motor output, the intact sensory input from the periphery at the spinal cord level becomes even more critical for generating motor output. Sensory information from multiple sources (e.g. cutaneous, joint, muscle) enters the spinal cord through the dorsal roots to provide an ensemble of information about the state or activity of the individual. An interneuronal network (shown as white cells), which comprises the central pattern generator (CPG) for locomotion, lies within the gray matter of the spinal cord. This central pattern generator receives, integrates, interprets, and organizes this ensemble of sensory information into a ‘meaningful’ context. Based upon the interpreted context, information is sent forward to the motor neurons (MNs). The axons of these motor neurons form the ventral roots and then peripheral nerves, which innervate the muscles.
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guided by four training principles (Table 28.1), which emphasize plasticity and activation of neural circuitry, including the central pattern generator, below the level of an SCI to achieve the primary goal of walking activity. These principles (Behrman and Harkema 2000, Behrman et al 2005, Harkema et al 2011) are designed to provide the appropriate sensorimotor ensemble for walking on the treadmill and on the ground.

TABLE 28.1
The four principles of locomotor training

<table>
<thead>
<tr>
<th>Principle</th>
<th>Description</th>
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<tr>
<td>Maximize load bearing by the lower extremities, and minimize load bearing by the upper extremities</td>
<td>This principle is supported by reports of increased lower extremity electromyographic amplitudes as a result of increasing the weight through the legs in experimental animals and humans after SCI, as well as in able-bodied individuals during the stance phase of walking (Harkema et al 1997, Dietz et al 2002). An adjustable body weight support system allows modification of this important parameter as the neuromuscular system becomes capable of accepting more weight.</td>
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<tr>
<td>Optimize the sensory cues for walking</td>
<td>Training at normal walking speed, typically ≥1.0m/s (2.2mph), enhances stepping. This element of the sensory experience modulates electromyographic frequency and amplitude in individuals with complete and incomplete SCI, as well as able-bodied individuals (Beres-Jones and Harkema 2004, Lünenburger et al 2006).</td>
</tr>
<tr>
<td>Optimize the kinematics for the trunk, pelvis, and extremities</td>
<td>Appropriate load bearing and hip extension promote, during select epochs of the step cycle, critical afferent input from joint receptors (proprioception), muscle spindles and Golgi tendon organs (muscle length), and the cutaneous receptors (skin surface) during upright posture. In particular, these cues are triggers for the transition from stance to swing of the same limb and swing to stance of the contralateral limb (Harkema et al 1997, 2011). Manual cuing (stretch/pressure) of key muscle tendons at appropriate points during the step cycle are used to enhance normal afferent inputs and should be graded as the system response increases.</td>
</tr>
<tr>
<td>Maximize recovery strategies; minimize compensation strategies</td>
<td>To maximize recovery, strategies should promote receptor activation in spatial–temporal patterns similar to those used during normal movements. The goal is to foster more normal neural activity that supports a more normal sensorimotor experience. Promoting and approximating normal activity minimizes the use of compensatory strategies, particularly those that change task kinematics and decrease the motor output of the lower extremities, pelvis, and trunk (Visintin and Barbeau 1994). The choice of whether to use a device (e.g. orthosis or assistive device), the choice of device type, and how it is used all should be carefully made (see Fig. 28.3). For example, traditional walkers promote an anteriorly flexed posture, which prevents an upright trunk position and hip extension beyond neutral. These kinematic features diminish key sensory walking cues, including load bearing and maximal hip extension, important to normal walking. Thus, if using a walker, it is important to cue and train an upright posture to facilitate normal walking kinematics, as well as to maximize load bearing by the lower extremities. Alternatively, a postural control walker (positioned behind the patient with arm supports to each side) allows for a more upright posture.</td>
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Fig. 28.3 Three components of locomotor training. (a) Intense retraining in the treadmill environment with partial body weight support and manual facilitation by trainers. (b) Transfer and assessment of skills on the ground (off the treadmill) introducing postural control walker and overhead support for greater ease in progressing speed of walker and assisting with load bearing while initiating steps. (c) Application of locomotor training principles within the home and community to increase the intensity of training and advance practical skills (e.g. walking in the hardware store was motivating for the child and included environmental challenges including negotiation of puddles in the parking lot, displays, and other individuals in the store). Photographs printed with consent.

Locomotor training in clinical practice
Translation of locomotor training into the clinical setting began formally in the USA around 2005 with the formation of the Christopher and Dana Reeve Foundation NeuroRecovery Network through a cooperative agreement with the Centers for Disease Control (Harkema et al 2012a). Several elements supported and continue to support this process. These include (1) standardized training for any sites interested; (2) within the network, standardized training, standardized outcome measures, annual competency evaluations for staff, and ongoing program evaluation; (3) publication of a locomotor training manual (Harkema et al 2011); (4) publications of data acquired across the network to demonstrate standardization of procedures (Morrison et al 2012), evaluate established and newly developed outcome measures (Behrman et al 2012), determine outcomes (Buehner et al 2012, Forrest et al 2012, Harkema et al 2012b), and identify longitudinal patterns of recovery (Datta et al 2012, Lorenz et al 2012) within the network compared with existing literature (Harkema et al 2012c). The current translation is effective not only in providing a new cutting-edge neurorecovery-based approach, but also in establishing a reimbursement mechanism as a clinical treatment using standardized codes (e.g. neuromuscular re-education). Although children represent only 5% of the population with SCI, the structure and framework used with adults in the NeuroRecovery Network may potentially provide a basis for this approach with children.
**TABLE 28.2**
Comparison of locomotor training approach to compensation-based approaches for achieving mobility

<table>
<thead>
<tr>
<th>Feature</th>
<th>Locomotor training approach</th>
<th>Compensation-based approach</th>
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<tbody>
<tr>
<td>Expectation of recovery from physical</td>
<td>Expect neuromuscular recovery below the level of the lesion during acute and chronic periods</td>
<td>Expect neuromuscular recovery during ‘natural recovery’ period &lt;1y after spinal cord injury</td>
</tr>
<tr>
<td>rehabilitation</td>
<td>and physical rehabilitation to impact rate, magnitude, and trajectory of overall recovery</td>
<td>and rate diminishing with time with little expectation for physical rehabilitation to alter</td>
</tr>
<tr>
<td>View of spinal cord</td>
<td>Target substrate that is responsive, adaptable, trainable, and can alter motor output</td>
<td>recovery trajectory</td>
</tr>
<tr>
<td>Rehabilitation</td>
<td>Activate neuromuscular system below the level of the lesion through sensory cues followed</td>
<td>Focus on spared, voluntary control of muscles above and below the level of the lesion to</td>
</tr>
<tr>
<td></td>
<td>by integration of voluntary control. Incorporate neuromuscular system above and below the</td>
<td>compensate for weakness and paralysis</td>
</tr>
<tr>
<td></td>
<td>lesion</td>
<td></td>
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<tr>
<td>Functional goals</td>
<td>Pre-injury movement patterns (closest approximation) to achieve functional goals and</td>
<td>Alternative movement strategies, e.g. momentum, leverage, substitution to</td>
</tr>
<tr>
<td></td>
<td>independence</td>
<td>accomplish functional goals</td>
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<tr>
<td>Use of compensatory devices</td>
<td>Minimize or eliminate use of assistive devices and braces. Assistive devices introduced</td>
<td>Assistive devices, mobility aids, and braces are provided early in the rehabilitation</td>
</tr>
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<td></td>
<td>after training the neuromuscular system. How the devices are used is consistent with</td>
<td>process to achieve mobility as the end goal and are permanently integrated</td>
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<td></td>
<td>locomotor training principles. Two devices may be introduced to achieve two different goals,</td>
<td></td>
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<tr>
<td></td>
<td>e.g. endurance or independence</td>
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<tr>
<td>Sensory input</td>
<td>Focus on providing task-specific sensory experiences to retrain and activate the</td>
<td>Focus on achieving trunk/pelvic/limb alignment and stabilization via equipment, mobility</td>
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<tr>
<td></td>
<td>neuromuscular system and generate task-specific motor output</td>
<td>devices, bracing, or surgical intervention</td>
</tr>
<tr>
<td>Duration of training and type of sensory</td>
<td>Overall amount of time retraining the neuromuscular system increases as skills improve</td>
<td>Amount of time spent in wheelchair and/or using alternative mobility strategies, bracing,</td>
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<tr>
<td>experience</td>
<td>and targets consistency with normal (pre-injury) movement patterns. Training and promotion</td>
<td>and assistive devices provides repetitive sensory experience often inconsistent with</td>
</tr>
<tr>
<td></td>
<td>of locomotor training principles also occurs in the home and community with goals evolving</td>
<td>normal (pre-injury) movement patterns</td>
</tr>
<tr>
<td></td>
<td>relative to neuromuscular gains</td>
<td></td>
</tr>
<tr>
<td>Therapeutic effect</td>
<td>With training, expect therapeutic effect; thus, positive change in neuromuscular system</td>
<td>With rehabilitation, expect functional gains; however, no therapeutic effect (relatively</td>
</tr>
<tr>
<td></td>
<td>and functional gains via activation of the system below the level of the injury with</td>
<td>permanent change in motor capacity). If device is removed (e.g. brace), function is</td>
</tr>
<tr>
<td></td>
<td>locomotor training as well as above</td>
<td>diminished or lost</td>
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Priorities for research
Based on the existing literature, the broad research priorities for children with SCI should include (1) the development of pediatric-appropriate assessment tools across relevant domains that are sensitive for all SCI magnitudes and etiologies, (2) the use of common assessments across sites and studies for effective comparison of similar and different treatment approaches (locomotor training and others), (3) the assessment of activity-based approaches that are recovery focused, (4) the assessment of treatment effects across multiple systems, primary and secondary targets, and the domains of the International Classification of Functioning, Disability and Health, and (5) the assessment of immediate and long-term costs and benefits of treatment approaches.

Summary
Sufficient evidence from basic scientific research, developmental work, the adult spinal cord literature, and emerging work in children after SCI warrants a shift in our thinking regarding rehabilitation potential after pediatric-onset SCI. Research has shown (and continues to demonstrate) that the spinal cord is ‘smart’. It can receive, integrate, and respond to sensory input during the practice of specific tasks (Roy et al 2012). Furthermore, specific tasks can be designed to produce a desired motor output, such as stepping and/or postural control, and then adjusted during training to promote further learning and refinement of that task. In summary, spinal circuitry has the capacity to learn, change, and promote development or restoration of complex motor behavior. These findings have led to the development of the activity-based locomotor training intervention (Behrman and Harkema 2007, Harkema et al 2011). Although in some instances rehabilitation will continue to rely on the use of compensation to achieve functional goals, physical rehabilitation should also be used as an agent for neurologic change and restoration of function after pediatric SCI. Table 28.2 provides a comparison of approaches for mobility: activity-based and compensation including expectations for outcomes, assumptions concerning plasticity, and clinical decision making. This is a new era in rehabilitation for adults and children with SCI. New expectations and possibilities for improved outcomes based on recovery and development of motor skills with age-appropriate performance as the reference point should be pursued.

Disclosure
The contents of this chapter do not represent the views of the Department of Veterans Affairs or the US Government.

REFERENCES
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