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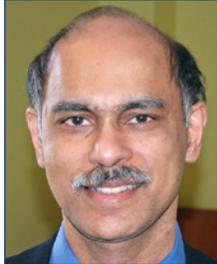
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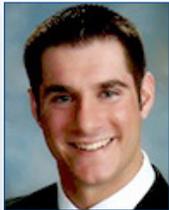
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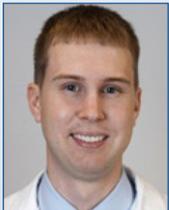
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Neurorehabilitation: achieving recovery after neural injuries

JON A. MUKAND, MD, PhD
GUEST EDITOR

A variety of neurological conditions can lead to disability, pain, and a decline in quality of life. As the American population ages, the prevalence of neurological disability will increase due to conditions such as stroke and Alzheimer's disease. Recent data show that about 800,000 strokes occur annually in the United States and someone dies of a stroke every four minutes.¹ Stroke survivors have to contend with outcomes ranging from mild impairments to total dependence. In the younger population, there is an epidemic of concussion related to sports. Every year about 182,000 football players sustain at least one concussion, primarily in youth (99,000) and high school (80,000) programs, or about 1 in 30 youth players and 1 in 14 high school players. Brain damage at this early stage of life has adverse effects for a long time.²

My perspective on neurological disability is that of a rehabilitation medicine specialist and medical director of the Southern New England Rehabilitation Center (SNERC) and the Sargent Rehabilitation Center (SRC). SNERC follows Medicare criteria for acute inpatient rehabilitation (medical stability, ability to tolerate three hours of therapy/day, goals and potential for progress, etc.); the center treats adults with neurological conditions including strokes, brain injuries, spinal cord injuries, Parkinson's disease, and multiple sclerosis. SRC is an outpatient facility that treats these conditions, especially stroke and brain injuries, as well as a variety of pediatric neurological problems (developmental delay, autism, learning disabilities, etc.).

This issue of the *Rhode Island Medical Journal* focuses on neurorehabilitation. In the first article, Marilyn Serra (a speech language pathologist by training and the president of SRC) and I discuss the pathophysiology of and rehabilitation after sports concussions. The second article is by Amanda Dragga, a speech language pathologist at SNERC, who describes treatments for speech, swallow, and cognitive problems after a stroke. As a clinician and faculty member

of the Orthopedic Surgery department at Brown University, I often collaborate with the orthopedic residents. Therefore, I'm pleased that some of these surgeons-in-training have contributed articles on orthopedic complications during neurorehabilitation. For instance, orthopedic surgeons can help with procedures such as tendon lengthening for spasticity when conservative measures are not working. In some cases, heterotopic ossification after a brain or spinal cord injury may require surgical intervention, so collaboration with orthopedic surgeons is important in neurorehabilitation. Back pain and radiculopathy are common conditions in the outpatient setting, but on occasion there can be concomitant conditions such as ruptured hamstring tendons, as described in a case report.

During the difficult process of neurorehabilitation, I admire my patients, their families, and their clinical team as they contend with challenging disabilities. Inpatient facilities such as the Southern New England Rehabilitation Center and outpatient centers such as Sargent Rehabilitation Center offer a therapeutic haven for people with disabilities who wish to improve their independence and quality of life.

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Guest Editor

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Concussions and Brain Injuries in Youth Sports

JON A. MUKAND, MD, PhD; MARILYN F. SERRA, MS, MEd, CCC-SLP

After Zackery Lystedt's brain injury, "he couldn't speak for nine months," his father said. Thirteen months later, "he could move his left arm a little; it took two years to get rid of the feeding tube and four years before he could move his right leg purposefully." Zackery had suffered a concussion during a high school football game in 2006, but "was twice returned to play and collapsed 60 seconds after the game was over."¹ The school district settled a lawsuit for \$14.6 million.²

About 1.6–3.8 million sports-related concussions occur every year,³ and a recent study found that "182,000 football players may sustain at least one concussion annually in youth (99,000), high school (79,640), and NCAA football programs (3,905)," or about 1 in 30 youth players and 1 in 14 high school players.⁴ Symptoms may appear mild but the injury can lead to life-long problems with physical function, concentration, memory, behavior, and emotion. Tragically, among high-school football athletes there were eight fatalities directly related to brain injuries in the 2013 season.⁵

At present, all fifty states have sports concussion laws modeled after the 2009 law in Washington that resulted from Zackery Lystedt's tragedy. Every law includes three components: education about the nature and risk of concussion and head injury for coaches, athletes, and parents/guardians; removal from play after a suspected concussion; and return to play with the approval of a trained and licensed practitioner.⁶

Definition of Concussion

The 4th International Conference on Concussion in Sport in 2012 (ICCS) defined a concussion as a complex process induced by biomechanical trauma, with the following common clinical, pathologic, and biomechanical features:⁷

1. Concussion typically causes short-lived impairment of neurological function that resolves spontaneously, but symptoms and signs may evolve over minutes to hours.
2. Concussion may cause neuropathology, but the symptoms reflect functional rather than structural changes, so standard neuro-imaging is normal.
3. The graded set of clinical symptoms may or may not involve loss of consciousness. Symptoms typically resolve in a sequential manner, but may be prolonged in some cases.

Symptoms of a Concussion

Symptoms after a concussion can be somatic, cognitive, or emotional – in varying combinations. The injured athlete

may feel "dazed" or "stunned," and may experience headaches, nausea, vomiting, impaired balance, visual problems, photosensitivity, phonosensitivity, and fatigue. Cognitive problems include mental "fogginess," slow information processing, slow speech, slow reaction times, impaired concentration, amnesia, and memory deficits. There may be emotional changes such as lability, irritability, anxiety, and sadness. Sleep patterns may also be affected, due to insomnia or drowsiness.

Warning signs of a more severe injury – intracranial bleeding, edema and impending herniation – include a severe headache, altered mental status, slurred speech, vomiting, a skull fracture, or a focal neurologic deficit such as diplopia. In these situations, prompt emergency evaluation is necessary.

Pathophysiology of Concussion

Trauma displaces the brain within the skull; compresses neural tissue; accelerates, decelerates, and rotates the brain within the hard casing of the skull; and causes a coup as well as a contre-coup injury. Cortical pathways are disrupted, as seen on diffusion tensor tractography, especially with frontal lobe connections;⁸ damage to the brainstem's reticular activating pathways alters consciousness. Pathologic changes include neuronal swelling and axonal disruption. Biochemical abnormalities include a sterile inflammatory response and metabolic changes. Injury to the young brain may also be related to elasticity of the skull sutures and the presence of vulnerable unmyelinated fibers in white matter tracts. Diffuse axonal injury involves mechanical disruption of the axon's cytoskeleton and axonal transport as well as axonal swelling, proteolysis, disconnection, and reorganization. Disruption of neural membranes affects ion channels, leading to potassium efflux, the release of glutamate, higher energy (ATP and glucose) consumption, increased lactate, increased Na-K pump activity, suppressed nerve activity, decreased blood flow, a hypometabolic state, and eventual cell death. Mitochondrial dysfunction and demyelination are also involved in diffuse axonal injury.⁹

Immediate Evaluation of a Concussion

If there are any symptoms of a concussion, the ICCS guidelines are clear:⁷

- A. The player should be evaluated and treated by a physician or other licensed healthcare provider and a cervical spine injury should be excluded.

- B. If no healthcare provider is available, the player should be removed from the field and promptly sent to a physician.
- C. Once the first aid issues are addressed, the concussive injury should be assessed with the SCAT3 or similar tools.
- D. The player should be closely monitored for the initial few hours following injury.
- E. A player with diagnosed concussion should not return to play on the day of injury.

Assessments after a Concussion

The Standardized Assessment of Concussion (SAC) is an effective, 6-minute tool for assessing orientation, concentration, immediate memory, and delayed recall. Laypersons may use the SAC at athletic events to identify a concussion, and the test has a sensitivity up to 94% and specificity up to 91%.^{10,11} A modified version for use in emergency departments has also been developed, with the addition of a Graded Symptom Checklist (headache, nausea, vomiting, blurred vision, etc.) and Neurologic Screening (amnesia, strength, sensation, coordination).¹²

Concussion in Younger Athletes

Although most (80–90%) concussions resolve within 7–10 days, the recovery process can be longer and more complicated in children and adolescents.¹³ Furthermore, younger athletes have a higher risk of severe symptoms and cognitive decline.¹⁴ This age difference in recovery and prognosis is probably related to the ongoing development of a child's brain. The primary senses, motor skills, and language are well developed by age ten. Frontal lobe maturation, however, goes on during the teenage years and even into the early 20s; these brain functions include abstraction, reasoning, judgment, insight, and emotional control.⁹ Consequently, achieving optimal recovery is critical for helping students with concussions.

Due to the more complex recovery process in young athletes, they need protection when they are most vulnerable. Recurrent concussions are especially destructive to the brain and are more likely during the first ten days after a concussion or if the athlete has had a previous concussion.¹¹ We recommend waiting at least seven days until return to play, regardless of the nature of the injury, because the long-term risks far outweigh any short-term benefits of the sport.

A Model of Concussion Care

Sargent Rehabilitation Center has a Concussion Management Clinic with a team of rehabilitation professionals for evaluation and treatment of student athletes. After a medical evaluation by the athlete's primary physician, Sargent's team evaluates cognitive function (concentration, memory, executive function, etc.) as well as behavioral, emotional, and physical changes. With the community school team, a comprehensive survey of the student in the classroom and

extra-curricular activities is performed. Concussion management includes short-term rehabilitation, school preparedness, and prevention of another concussion as well as monitoring and management of re-emergent symptoms.

The concussion clinic offers baseline/pre-season Immediate Post-concussion Assessment and Cognitive Testing (ImPACT) to school systems (Figure 1). This online test of attention span, memory, non-verbal problem solving, and reaction time is also used for follow-up evaluations in case of a brain injury. Among athletes with suspected concussions, the test was 91.4% sensitive and 69.1% specific. Notably, with athletes who denied symptoms but had a suspected concussion, ImPACT testing yielded 94.6% sensitivity and 97.3% specificity.¹⁵ The NeuroCom SMART EquiTest CDP® is used by the clinic's therapists to assess and retrain balance mechanisms with visual biofeedback (Figure 2). This system uses a stable or unstable surface (with a dynamic force plate) in a static or dynamic visual environment. Computerized protocols such as the Sensory Organization Test (SOT) measure the ability to maintain equilibrium with changes in

Figure 1. Visual-perceptual testing with the ImPACT test

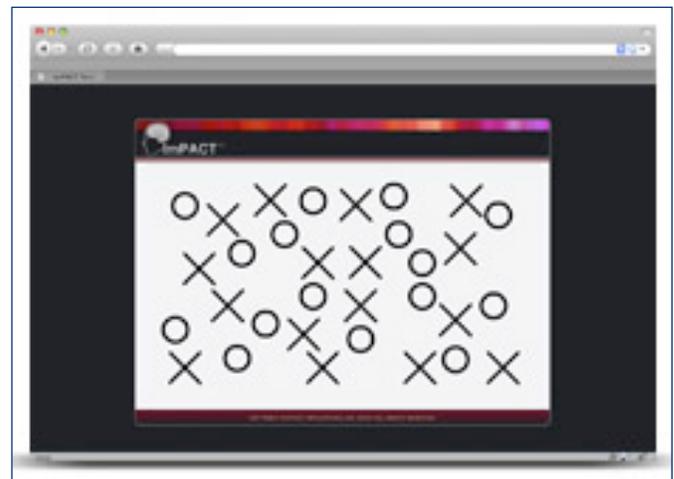


Figure 2. Balance testing with the NeuroCom SMART EquiTest CDP®



somatosensory or visual input or both. The SOT identifies concussions with a sensitivity of 48%–61% and specificity of 85%–90%.^{16, 11} Combining the ImPACT and Neurocom tests yields even better sensitivity and specificity for concussions, as the two assessments encompass cognitive, cerebellar, and visual brain pathways. The clinical program at the Concussion Management Clinic is described in the following case report.

Case Report

A sixteen-year-old male named “John” suffered a concussion due to a head butt in a soccer game. Two weeks later, he obtained physician clearance to return to school and the soccer field, but had decreased cognitive and physical functioning: an awkward gait and difficulty with comprehending texts. On the fourth day of his return, he fell and could not remember if he hit his head. Three weeks after the soccer injury, John fell down the stairs in his house and was diagnosed with a second concussion. Subsequently he had memory problems and severe headaches.

Unable to attend school, John was referred to Sargent Center and the intake committee determined that he was appropriate for the Concussion Management Clinic. His evaluations were scheduled in short sessions to accommodate his headaches and associated fatigue. John was provided educational, speech, and physical therapy three days a week. His progress was closely monitored by Sargent’s medical and nursing services. A coordinated plan of treatment and school management was developed by the concussion clinic and his school. The clinic staff also provided ongoing support to the family. After six weeks, John’s balance issues had resolved and the concussion clinic and school teams coordinated a return to school, three days a week for two hours each day.

The school was trained to reinforce the rehabilitation strategies for John to improve his memory, attention, processing speed, and balance. Over the next month his school attendance increased to five 2-hour days. The concussion clinic staff and the school continued working on John’s class instruction, learning strategies, and problem-solving. Four months after starting at the concussion clinic, he returned to school full-time. With educational accommodations, he successfully advanced to the next grade.

This case report describes a male soccer player, but girls are at especially high risk in this sport. A prospective study of female soccer players (ages 11-14) found 59 concussions during 43.7 thousand hours; a cumulative incidence of 13.0% per season; and a duration of concussion symptoms for an average of 9.4 days. About 30% of concussions were due to heading the ball. Longer recovery times occurred with the presence of light sensitivity, emotional lability, noise sensitivity, memory loss, nausea, and impaired concentration. Unfortunately, almost 60% of athletes kept playing in spite of their symptoms and only 44.1% sought medical attention.¹⁷ (These data raise important questions about the necessity, value, and risks of heading the ball in youth soccer.)

Rhode Island School and Youth Programs Concussion Act & Education

This law (2010, 2014) requires the Departments of Education and of Health to work with the Rhode Island Interscholastic League to educate coaches, teachers, school nurses, youth athletes, and parents/guardians about concussion and head injury (C & HI).¹⁸ To play sports after an injury, an information sheet must be signed by the athlete and the parent/guardian. All coaches, school nurses and volunteers must take a training course and an annual refresher on C & HI. Teachers and teachers’ aides are strongly encouraged to complete a training course. School districts are encouraged to arrange baseline neuropsychological testing. Parents/guardians should receive information about C & HI before the season and should acknowledge receipt of that material. Any “youth athlete who is suspected of sustaining a concussion or head injury in a practice or game shall be removed from competition.” The athlete “may not return to play until an evaluation by a licensed physician who may consult with an athletic trainer, all of whom shall be trained in the evaluation and management of concussions. The athlete must receive written clearance to return to play” from that licensed physician.

To help compliance with the RI Concussion Act, Sargent’s Regional Resource Center offers conferences and workshops on health care, education, policy, and the law. The target audience includes families, physicians, nurses, coaches, athletic directors and trainers, teachers, rehabilitation specialists, psychologists, counselors, social workers and school administrators. Education does reduce injuries. In a recent study of education about injury prevention, some coaches was not educated, another group received the Heads Up Football coaching program (HUF), and a third was educated about the HUF and also given the Pop Warner Football (PW) guidelines to restrict contact during practice. Among football players (ages 11 to 15), the concussions during practice were much lower in the HUF + PW group (0.14/1,000 athlete exposures (AEs)) compared to the non-educated cohort (0.79/1000 AEs).¹⁹

Education about injury prevention is especially important in youth football. These athletes are vulnerable to the second-impact syndrome, in which the patient with a brain injury has ongoing symptoms – and then has another head injury. Forensic studies suggest that the second trauma worsens the initial damage, which predisposes the brain to a more intense pathophysiologic response and leads to diffuse cerebral edema, brainstem herniation, and death.⁹

Conclusion

Sports offer many benefits to students, but their well-being and cognitive, emotional, and behavioral potential should never be compromised. Zackery Lystedt’s story is tragic, and there are thousands of other student athletes whose lives have been damaged by concussions to a less severe extent. Health care professionals have the responsibility to effectively and ethically manage the epidemic of youth sports concussions and brain injuries.²⁰

Suppliers

ImPACT Test: <https://www.impacttest.com/products/?The-ImPACT-Test-2>

Neurocom: http://www.natus.com/documents/015368A_SMART_EquiTest_EN-US_lo-res.pdf

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The Role of Speech-Language Pathologists in Stroke Rehabilitation

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INTRODUCTION

According to the American Heart Association's *2015 Heart Disease and Stroke Statistics Update*, stroke is the fourth leading cause of death in the United States (129,000 people per year) as well as a major cause of long term disability.¹ In addition to the physical impairments caused by stroke, many individuals may experience changes in their cognitive, communication, and swallowing abilities. A speech-language pathologist (SLP) is trained to evaluate and treat these types of disorders and is an integral part of the rehabilitation team in an acute inpatient rehabilitation facility (IRF). This article will provide a brief overview of these disorders and a description of the SLP's role in stroke rehabilitation.

COGNITION

"Cognition" refers to an individual's thinking skills, and includes the ability to concentrate on one or more tasks simultaneously, to recognize frequently encountered people and remember daily routines, to make decisions and solve problems, and to organize and carry out a sequence of steps to complete a task. Impairments in one or more of these areas can impact an individual's ability to safely and effectively perform activities of daily living.² A data collection study of the South London Stroke Register between the years of 1995 and 2010 suggested that the prevalence of cognitive impairment following stroke was around 22% at three months post-stroke and at annual follow up.³ It should be noted that for the purposes of this study, patients with severe aphasia, dysarthria, deafness, or visual impairment were excluded because they could not undergo formal cognitive testing; therefore, this figure may be a significant underrepresentation of the true prevalence of cognitive impairment secondary to stroke. Following a patient's admission to an IRF, the speech-language pathologist conducts an evaluation to identify the cognitive domains most severely affected by the stroke. This evaluation includes both informal testing and formal assessment measures such as the Cognitive Linguistic Quick Test (CLQT), the Assessment of Language-Related Functional Activities (ALFA), and the Brief Cognitive Assessment Tool (BCAT). The CLQT has tests such as clock drawing, generative naming of animals, and remembering details of a short narrative. In the ALFA, tests include simple math, understanding medicine labels, and transcribing

phone messages. Based on the patient's performance, the SLP then develops an individualized treatment plan, which may involve exercises to improve attention, memory, problem solving, executive functioning, and visuospatial skills. Treatment examples include using a memory log to improve recall of daily events; training in the use of environmental aids to assist with orientation; and using spaced retrieval training to improve the acquisition, retention and generalization of trained information and/or skills. In this particular method, individuals are trained to recall a specific target over gradually increasing time intervals. The SLP may also work in conjunction with the occupational or physical therapist to address the targeted cognitive skills in a functional context, such as preparing a meal or purchasing an item from the gift shop. These tasks are highly functional and require many cognitive skills including planning, organization, sequencing, divided attention, self-monitoring, problem solving, and memory.

APHASIA

A stroke on the left side of the brain often results in aphasia. This impairment in language may affect an individual's ability to speak, understand, read or write. Aphasia is present in 21%–38% of acute stroke patients.⁴ It is estimated that in the United States there are 80,000 new cases of aphasia each year and a total of one million people suffer from aphasia.⁵ Research has suggested that greater frequency and intensity of aphasia treatment leads to better recovery of language.⁶ Clinical practice guidelines suggest that individuals with stroke-induced aphasia should receive SLP treatment between two to eight hours a week, and treatment initiated early in the recovery process is more effective than when initiated later.⁷ Therefore, patients with stroke-induced aphasia may benefit from the more intense therapy schedule at an IRF, where individuals with aphasia receive a minimum of five hours of speech therapy per week. Depending on the type of aphasia, treatment may focus on word retrieval exercises, sentence formulation, following auditory or written directions, or training with alternative and augmentative communication aids. In some cases, patients with expressive aphasia may benefit from script training, which involves the rehearsal of specific responses to facilitate communication of basic wants and needs.

DYSPHAGIA

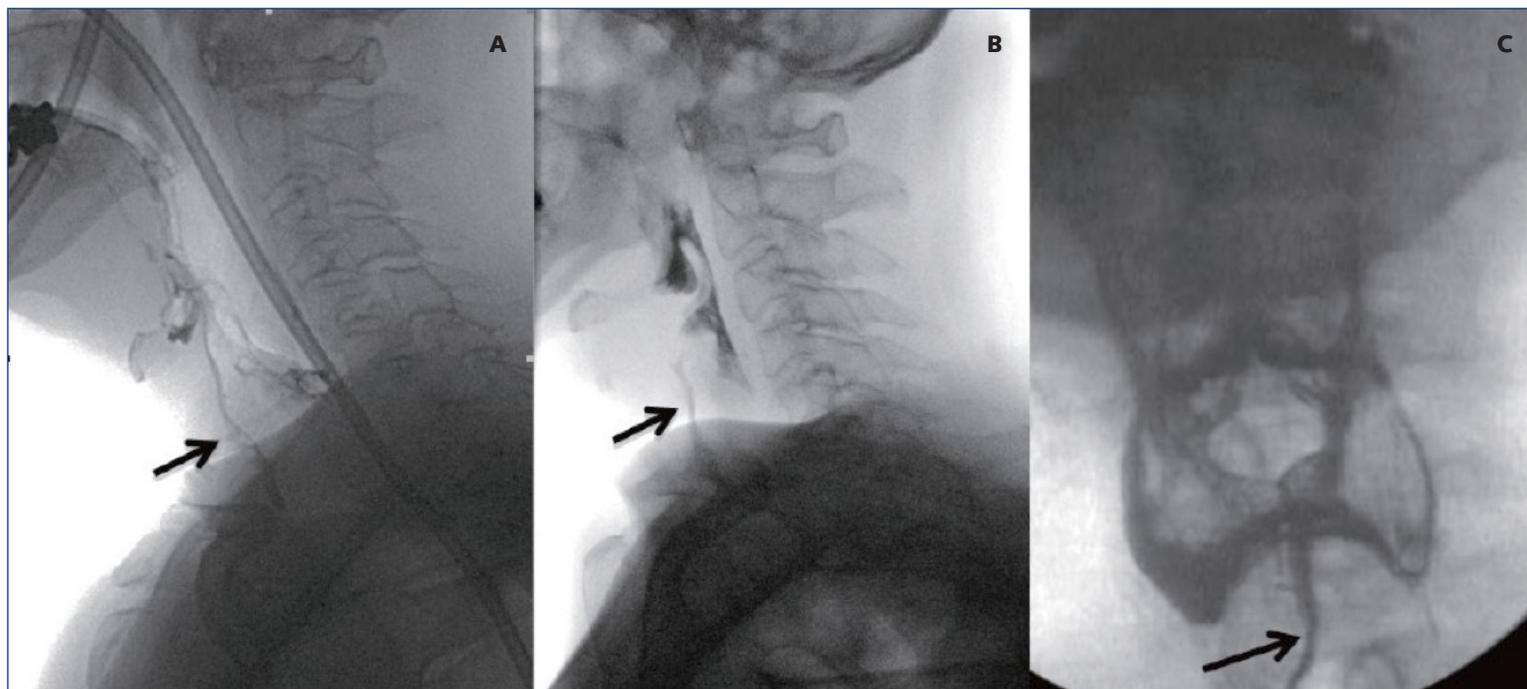
SLPs in the inpatient rehabilitation setting also evaluate and treat dysphagia. A person with dysphagia may have difficulty with the oral, pharyngeal, or esophageal phases of swallowing, and this can occur with liquids, solids or both. A 2009 study conducted by Falsetti et al. found that dysphagia occurred in more than one-third of consecutive patients admitted to a neurorehabilitation hospital following stroke; however, other studies have found a wide incidence, between 29% and 81%. Between 22% and 52% of individuals with dysphagia experience aspiration of material into the airway, and nearly half of aspirations in patients with stroke are silent. The presence of dysphagia has been linked to malnutrition, dehydration, pulmonary infections, prolonged hospital stays, and death.⁸ When a patient is suspected of having dysphagia, an SLP will conduct a clinical bedside assessment, but in some cases, that is not sufficient, especially if aspiration is silent. One of the greatest benefits of an IRF in a hospital setting is the on-site availability of a videofluoroscopic swallow study (VFSS). The VFSS is considered the “gold standard” of swallowing assessments and allows the SLP to objectively assess the patient’s swallowing function and to establish the safest and least restrictive diet textures (**Figure 1**). This minimizes the patient’s risk of aspiration and the associated complications. The VFSS is conducted in conjunction with a radiologist and allows the SLP to assess the oral, pharyngeal, and upper esophageal phases of the swallow mechanism with a variety of liquid

and solid textures, along with compensatory strategies that may improve the patient’s swallowing safety. For example, a patient with a delayed swallow trigger or reduced epiglottic deflection may benefit from the use of a “chin tuck” or “chin-down” posture while swallowing. This strategy widens the valleculae and places the epiglottis in closer proximity to the posterior pharyngeal wall. In some cases, the implementation of this strategy enables the patient to safely swallow thin liquids, avoiding the need to restrict the diet with thickened liquids. The VFSS allows direct visualization of how patients respond to food and liquids of various textures and to compensatory strategies. In contrast, empirical trials could lead to silent aspiration and to pneumonia. Once the cause of the dysphagia is established, the SLP develops a treatment plan which may consist of strengthening exercises for the oral, laryngeal, and pharyngeal musculature as well as compensatory strategy training.

CONCLUSION

After a stroke, patients often experience impairments of their cognitive, communication, and swallowing functions, which worsens their disabilities and quality of life. In acute inpatient rehabilitation facilities, speech-language pathologists offer advanced clinical techniques for the evaluation and treatment of these conditions as well as essential therapeutic time for the complex rehabilitation process after a stroke.

Figure 1. Three patients with aspiration at different points of swallowing: before (A), during (B), and after (C) elicitation of the pharyngeal swallow.



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Low back pain, radiculopathy, and bilateral proximal hamstring ruptures: a case report

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ABSTRACT

Low back pain (LBP) is a common complaint in the United States, with an incidence of 6.3%–15.4% and yearly recurrence in 54%–90% of patients.¹ Trends show more frequent diagnostic testing, opioid use, and surgical intervention as the incidence of LBP increases.² LBP is defined as pain at and near the lumbosacral region that can vary with physical activity and time.³

LBP is usually related to pathology of muscles, ligaments, spinal column joints, nerve roots, and the spinal cord. During the assessment of LBP, practitioners must also consider less common causes of pain in that region. For instance, patients with indolent or nighttime pain may have infectious or malignant processes. Referred pain from injuries to pelvic musculature or abdominal contents should be considered, especially following a traumatic event. One of these injuries, which can present as acute low back pain, is rupture of the proximal hamstring tendon. On rare occasion, concomitant LBP, radiculopathy, and hamstring injuries can occur. This diagnostic challenge is described in the following case.

KEYWORDS: Proximal hamstring rupture; low back pain; radiculopathy

CASE REPORT

A 46-year-old woman was seen for an initial rehabilitation medicine evaluation four years after slipping on a wet floor, catching herself, and not falling. She acutely developed sharp pain in her right hip, posterior thigh, and buttock. Her non-radiating pain was rated as 6/10 and worsened with movement and walking. Her relevant past medical and surgical history included back pain, hyperlipidemia, anxiety, depression, diverticulosis, and left shoulder rotator cuff surgery. She drank alcohol rarely and smoked one pack of cigarettes per day. Examination by an Emergency Department physician revealed pain with palpation at the buttock and posterior thigh. She was diagnosed with a muscle strain and myofascial pain and treated with ibuprofen, diazepam, and intramuscular ketorolac.

A week after the injury, she had persistent burning, stabbing pain in her right buttock, a positive straight leg raise at 10-20 degrees, and a mild limp. Her primary care physician

prescribed oral methylprednisolone and acetaminophen/propoxyphene. A magnetic resonance imaging (MRI) scan of her lumbar spine revealed mild degenerative changes of the disc spaces, but no significant narrowing of the spinal canal or neural foramina. There was hypertrophy of the facet joints at L4-L5, and a broad-based disc bulge at L4-L5 with mild narrowing of the neural foramen.

A month after her injury, she needed acetaminophen/propoxyphene three times a day. Her physician felt that her near-fall had worsened an asymptomatic spinal condition. Her physical therapy included trunk flexion and extension, spinal massage, stretching, ultrasound to the piriformis muscle, moist hot packs, and iontophoresis to the right hip. The pain initially decreased but persisted.

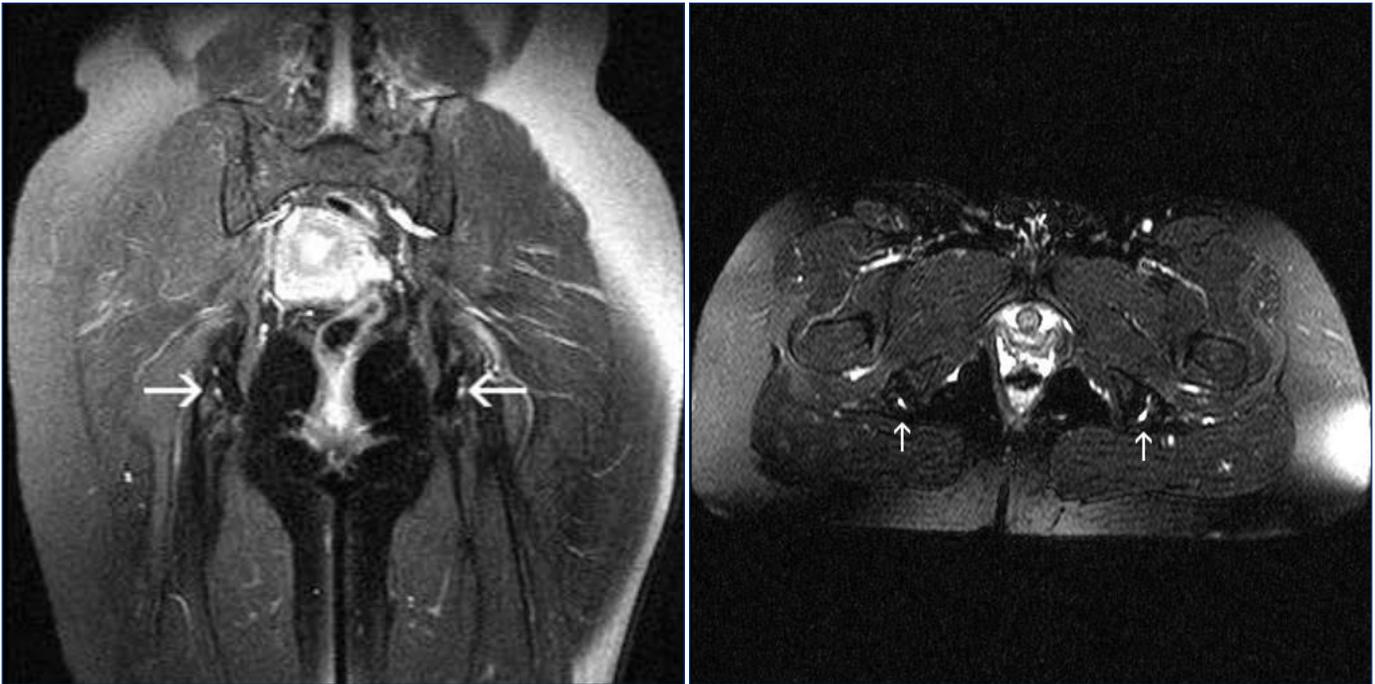
An orthopedic surgeon noted a short stride length on the right, tenderness at the posterior right greater trochanter and the sciatic nerve, and right hip pain with flexion of the lumbar spine. The surgeon felt that the disc bulge at the L4-L5 level and trochanteric bursitis were the likely cause of her radiculopathic symptoms; he injected the trochanteric bursa with methylprednisolone and bupivacaine, with some improvement in pain.

At follow-up with the surgeon six months after the near-fall, she still had tenderness in the ischial tuberosity and greater trochanter. Her symptoms worsened with adduction across the midline and hip flexion. An MRI of the pelvis revealed tendon ruptures: small fluid collections underlying the origin of the conjoined tendon of the hamstring tendon bilaterally, left greater than right (**Figure 1**). An EMG/NCV study revealed radiculopathies at the L4 and L5 nerve roots, with fibrillations at the L4 and L5 paraspinals, the right anterior tibialis, and the left peroneus longus muscle.

DISCUSSION

Acute hamstring injuries are commonly experienced by athletes.⁴ The semitendinosus, semimembranosus, and biceps femoris tendons originate on the ischial tuberosity and are at risk of injury with eccentric contractions during hip flexion and knee extension.⁵ Proximal hamstring ruptures represent 9% of all hamstring injuries.⁶ Ruptures occur in adults at the myotendinous junction; however, patients aged 16-25 years may sustain an avulsion fracture of the ischial apophysis.⁷ Hamstring ruptures may occur in elite or middle-aged recreational athletes.⁷ Injuries have been reported

Figure 1. MRI images showing bilateral proximal hamstring ruptures from the ischial tuberosity (arrowheads). The tears are the white signals noted by the arrows; a normal tendon would be gray/black.



during water-skiing, running, soccer, American football, ice hockey, dancing, tennis, wrestling, and bull-riding as well as during slip and falls.^{8,9} Timely evaluation of possible proximal hamstring injuries within 48 hours may avoid a delay in diagnosis.¹⁰

Patients with hamstring injuries complain of acute shooting pain in the posterior thigh. They may have a stiff-legged gait pattern in order to limit painful hip and knee flexion during ambulation.⁵ Physical examination often reveals tenderness over the ischial tuberosity as well as ecchymosis due to hematoma formation. Depending on body habitus, a palpable step-off may be present at the location of the tear; however, this is not a reliable sign of injury. The bowstring sign may be the best way to distinguish between complete and partial tears and was present in 23/23 patients with complete tears.⁹ It is present if there is no palpable tension in the distal hamstrings with the patient prone and the knee flexed to 90 degrees.

Neurological testing of the lower extremity is important, as chronic hamstring ruptures can present with sciatic neuralgia.⁸ Chronic injuries may also present with “hamstring syndrome,” or local posterior buttock pain over the ischial tuberosity. In one series of chronic hamstring injuries, 52/59 patients experienced relief of their symptoms following surgical release and nerve decompression.¹¹ Peroneal nerve function must also be assessed; injury to this nerve can result in foot-drop or weak ankle eversion.⁷

Radiographs will often be negative, although a small avulsion of the ischial tuberosity is possible. MRI is the gold standard and can distinguish complete versus partial

rupture, allowing for grading of the injury. Grade 1 injuries show only muscle edema on MRI with no architectural disruption of the muscle while Grade 2 and 3 represent partial and complete tears, respectively.⁵ Modifications to this grading system include sciatic nerve tethering and the degree of muscle retraction, with > 2 cm being a relative indication for surgery.¹² Ultrasound may be useful as a diagnostic tool but is operator-dependent. In one study, MRI diagnosed hamstring strain in 70% of patients while ultrasound correctly identified 75%.¹³ At six weeks, MRI identified 35.7% of patients with abnormalities compared to 22.2% for ultrasound. MRI may be superior to ultrasound for follow-up imaging but either appears acceptable for initial diagnosis.¹³

The treatment of proximal hamstring ruptures depends on the patient and expectations for future activities. Cohen *et al.* suggested a treatment algorithm based on MRI findings.⁵ Acute single tendon tears with retraction 1-2 cm tend to scar and adhere to the intact tendons; they are managed conservatively with relative rest for 6 weeks, with likely return to full strength.⁵ Tears of all three proximal hamstring tendons often result in significant retraction of ≥ 5 cm, and these injuries should be managed operatively, especially in high-level athletes.⁵ There is currently no consensus on the management of two-tendon proximal hamstring tears. Some recommend surgical treatment of two-tendon proximal hamstring ruptures with ≥ 2 cm of retraction in patients younger than 50 who are recreational athletes; these patients may have an injury to the third hamstring muscle at the musculotendinous junction that is not apparent on MRI.⁵ Failure to repair may result in chronic pain, weakness, and dysfunction.

Systematic reviews of outcomes after surgical repair of proximal hamstring rupture favored surgical repair in retracted, complete proximal hamstring tears but noted the paucity of higher level studies.^{14,15}

Conservative management of proximal hamstring ruptures consists of relative rest with modalities including ice, ultrasound, electrical stimulation, non-steroidal anti-inflammatory medications, and gentle stretching with progression to therapeutic exercise and gradual return to sports.⁵

Most cases of LBP will resolve with conservative therapy. LBP has been attributed to injury, disc herniation, stress, weather, and aging but may have a psychosomatic component.¹⁶ Nerve entrapment is over-diagnosed and leads to an overuse of surgical intervention.¹⁷ Neurological abnormalities in strength, sensation, and reflexes, especially with bowel or bladder dysfunction, require prompt surgical evaluation and treatment in order to avoid complications of cauda equina syndrome. In our patient, neurological and radiological abnormalities were accompanied by EMG findings of radiculopathy but she was safely treated in a conservative manner.

SUMMARY

Proximal hamstring ruptures can be a source of low back pain and disability for both young, athletic patients who sustain an injury during sports as well as older patients who sustain a fall. Treatment options range from conservative measures with gradual resumption of activity to surgical repair of the ruptured tendons. In our patient, the diagnosis of hamstring tendon injuries was complicated by low back pain and radiculopathy. Co-existent neurological and musculoskeletal conditions can create a diagnostic challenge, but vigilance for these rare situations leads to better diagnosis and treatment.

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Orthopaedic Management of Spasticity

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ABSTRACT

Spasticity is a common manifestation of many neurological conditions including multiple sclerosis, stroke, cerebral palsy, traumatic brain injury, and spinal cord injuries. Management of spasticity seeks to reduce its burden on patients and to limit secondary complications. Non-operative interventions including stretching/splinting, postural management, physical therapy/strengthening, anti-spasticity medications, and botulinum toxin injections may help patients with spasticity. Surgical management of these conditions, however, is often necessary to improve quality of life and prevent complications. Orthopaedic surgeons manage numerous sequelae of spasticity, including joint contractures, hip dislocations, scoliosis, and deformed extremities. When combined with the efforts of rehabilitation specialists, neurologists, and physical/occupational therapists, the orthopaedic management of spasticity can help patients maintain and regain function and independence as well as reduce the risk of long-term complications.

KEYWORDS: Spasticity, Orthopaedics, Multiple sclerosis, Stroke, Cerebral palsy, Traumatic brain injury, Spinal cord injuries (SCI)

INTRODUCTION

Spasticity is a common manifestation of many neurological conditions including multiple sclerosis (MS), stroke, cerebral palsy (CP), traumatic brain injury (TBI), and spinal cord injury (SCI).¹⁻⁴ Spasticity can lead to severe physical, psychological, and social impairments.⁴⁻⁶ While numerous non-operative treatments are available, surgical management is often necessary to improve quality of life and prevent complications.

EPIDEMIOLOGY

Spasticity affects up to one-third of all stroke survivors. Six months after stroke, as many as 50% of patients have developed contractures.^{5,7,8} Up to 90% of patients with MS experience spasticity and as many as one-third modify their activity as a result.^{5,6} Spasticity has been reported in 25% to 89% of patients with TBI and is a common feature in SCI (65% to 78% of patients) and CP (72% to 91%).⁹⁻¹⁴

DIAGNOSIS

“Spasticity” refers to “disordered sensorimotor control resulting from an upper motor neuron (UMN) lesion, presenting as intermittent or sustained involuntary activation of muscles.”^{1,2,4,6,15} Patients commonly exhibit increased tone, hyperreflexia, clonus, a Babinski sign, reduced velocity of movement, reduced motor control, weakness, and loss of dexterity.^{1,2,4,16} Increased tone manifests as a resistance to passive motion that is mediated by exaggerated spinal motor neuron responses to muscle stretch.²⁻⁴ Over time, sarcomeres in underutilized muscles are replaced by fat and connective tissue, resulting in contractures.² Upper extremity spasticity often presents with hypertonia in the shoulder adductors, elbow, wrist, and finger flexors; and forearm pronators.^{1,2} Lower extremity spasticity usually presents with high tone in the hip adductors, knee flexors, ankle plantarflexors and invertors, and great toe extensors.^{1,2,4}

NON-OPERATIVE MANAGEMENT

Stretching/Exercise/Posture

Passive stretching is a mainstay of spasticity treatment, as it decreases the excitability of motor neurons and maintains flexibility.² Exercise improves motor control, strength, and overall function in addition to helping trunk, pelvic, and shoulder girdle muscles to control distal movements.^{2,3} Exercise may not directly reduce spasticity, but it does not worsen hypertonia, as was previously thought.^{2,3,17} Additionally, weight bearing reduces spasticity, improves bone mineral density, enhances psychological health, and aids lung, bowel, and bladder function; all of these benefits are especially important for people with disabilities.^{2,3}

Medications

Table 1 summarizes the oral and injectable medications commonly used in the management of spasticity. Oral agents may unmask weakness and should be started at low doses and titrated up as needed.^{2,3} Targeted Botulinum toxin (Botox) injections cause selective weakening of spastic muscles, obviating the generalized weakness associated with oral agents.² Similarly, neurolysis with phenol injections can achieve targeted muscle weakening, but should be performed only with pure motor nerves to avoid the risk of chronic neuropathic pain.²⁻⁴

Table 1. Medications for the Treatment of Spasticity²

Table 1 illustrates many of the commonly used oral medications for the treatment of spasticity.

GABA: "Gamma-Aminobutyric acid." QHS: "At bedtime;" 1XD: "One time daily;" 2XD: "Two times daily;" 3XD: "Three times daily."

Drug Name	Administration	Mechanism	Dosage	Side Effects
Baclofen	Oral, Intrathecal	GABA agonist	Starting dose: 5mg 3XD. Increase 5-10mg weekly until desired effect. Max dose: 90-120mg/day	Weakness, drowsiness, dizziness, sexual dysfunction, urinary incontinence, reduction of seizure threshold, withdrawal
Benzodiazepines	Oral, Intravenous	Potentiates GABA system	Starting dose (clonazepam): 500µg QHS. Max dose: 1mg QHS	Drowsiness, dizziness, fatigue, respiratory depression, dependency, withdrawal, seizures, hypotension, tachycardia
Gabapentin	Oral	Stimulates GABA biosynthesis	Starting dose: 300mg 1XD day 1; 300mg 2XD day 2; 300mg 3XD day 3. Increase by 300mg every 2-3 days until desired effect. Max dose: 3600mg daily	Weight gain, gastro-intestinal disturbances, confusion, depression, hostility, sleep disturbance
Pregabalin	Oral	GABA agonist	Starting dose: 75mg 2XD. Max dose: 300mg 2XD	Weight gain, gastrointestinal disturbances, confusion, depression, hostility, sleep disturbance
Tizanidine	Oral	Central α-2 adrenergic system agonist	Starting dose: 2mg QHS. Increase by 2mg weekly as needed. Max dose: 36mg (divided into 3-4 daily doses)	Dry mouth, gastrointestinal disturbance, hypotension, acute hepatitis, withdrawal: hyperadrenergic syndrome
Dantrolene	Oral, Intravenous	Blocks calcium release from sarcoplasmic reticulum, blocking contraction of muscle cells	Starting dose: 25mg 1XD. Increase by 25mg per week as needed. Max dose: 100mg 3-4 times daily	Hepatotoxicity and rare fatalities (need regular liver function tests)

OPERATIVE MANAGEMENT

Intrathecal Baclofen

Baclofen inhibits the spinal cord's reflex arc, which reduces resting muscle tone. It can be delivered via intrathecal pumps in small doses of high concentrations that can be titrated to a desirable level of inhibition.^{2-4,16,18-21} Patients who rely on some tone to maintain posture will benefit from such a reduction without elimination of their spasticity.^{2,3,16} As with any implantable device, infection is a concern.^{16,22-24} Furthermore, errors in surgical implantation or catheter-related problems can cause baclofen overdose or withdrawal.^{4,16,25-27}

Selective Doral Rhizotomy

Selective dorsal rhizotomy (SDR) reduces afferent input to the spinal reflex arc, dampening the heightened response to muscle elongation seen in spasticity.^{2,16} SDR is a relatively permanent and cost-effective solution when compared to baclofen pumps, which require regular maintenance and refills.^{16,28} SDR in children reduces the need for future orthopaedic procedures.^{16,29} The procedure has also been shown to improve motion throughout the gait cycle.^{30,31} Because SDR affects all afferent signals at the dorsal root, decreased proprioception and sensory function may impair walking and standing.^{16,32}

ORTHOPAEDIC MANAGEMENT

Upper Extremity Management

Reconstructive surgery of the upper extremity in patients with spasticity can improve range of motion (ROM), strength, functional grasp, dexterity, two-point discrimination,

stereognosis, and limb positioning.³³⁻³⁷ Surgical options include tendon transfers, muscle/tendon lengthening, and joint stabilization.^{35,37,38} **Table 2** summarizes numerous reconstructive options for the management of common spastic problems of the upper extremity.

Forearm pronation deformity can be addressed by rerouting or releasing the pronator teres (PT) muscle.³⁸ Tenotomy is preferred in patients who are able to supinate and have

Table 2. Common Soft Tissue Reconstructive Procedures for Treatment of Spasticity of the Upper Extremity^a

Location	Deformity	Procedures
Elbow	Flexion	• Lengthening: Biceps, brachialis
Forearm	Pronation	• Releases: Pronator teres, pronator quadratus • Rerouting: Pronator teres
Wrist	Flexion +/- ulnar deviation	• Lengthening: FCR/FCU • Flexor pronator slides • Tendon Transfers: PT, BR, FCU, ECU, FCR to ECRB/ECRL
Fingers	Flexion	• Lengthening: FDS • Tendon Transfers: FCU/BR to EDC, FDS tenodesis, lateral band rerouting
Thumb	Thumb-in-palm	• Releases: Adductor pollicis, 1st DI • 1st web space Z-plasty • Lengthening: FPL • Tendon Transfers: FCR/PL/BR to APL, FCR/PL/BR to EPB, PL/BR to EPL • Rerouting: EPL

a: Adapted from Upper extremity Surgical Treatment of Cerebral Palsy. Van Heest et al., 1999

continuous PT spasticity.³⁸ Tendon transfer is favored for phasic PT contractions during supination.

A spastic flexor carpi ulnaris (FCU) can lead to wrist flexion and ulnar deviation, which impairs grasp and release. Patients with voluntary control of the FCU may benefit from transfer to the extensor carpi radialis brevis (ECRB), as described by Green.³⁹ Patients lacking finger extension may be best served with transfer to the finger extensors or combining the Green procedure with lengthening of the finger flexors.³⁷ Transfer of the FCU to the extensor carpi radialis longus (ECRL) can correct ulnar deviation. Additionally, transfers of the pronator teres, brachioradialis, or extensor carpi ulnaris (ECU); flexor carpi radialis (FCR) transfer to the ECRB; lengthening of the wrist flexors; or wrist fusion can be used to address the flexion deformity.³⁷

Adduction of the thumb due to a spastic adductor pollicis (AP) muscle characterizes thumb-in-palm deformities.³⁸ Spasticity of the flexor pollicis muscles, metacarpophalangeal joint instability, and interphalangeal joint flexion or hyperextension can also contribute to the deformity.³⁸ In patients with voluntary control of the extensor pollicis longus (EPL), a radial transfer of the tendon from the third to first dorsal compartment on the radial side of the thumb can help restore thumb extension when combined with a release of the AP.^{40,41} Associated first web space contractures can be addressed via z-plasty.³⁸ The deformity can also be addressed by tendon transfer to the thumb abductors and extensors and/or lengthening of thumb flexors.³⁸

Hip Abnormalities

Hip deformities such as coxa valga, femoral anteversion, and acetabular dysplasia are common in patients with spasticity.⁴² Up to 75% of patients with CP may experience hip subluxation, with more severely affected patients experiencing higher rates of subluxation and dislocation.⁴²⁻⁴⁵ Hip abnormalities should be identified early in at-risk patients to prevent long-term complications.⁴²

Radiographically, hip subluxation can be quantified by the Reimer migration index (RMI) and acetabular dysplasia can be evaluated by the acetabular index (AI), as illustrated in **Table 3**.⁴² Independent ambulators should obtain a baseline

Table 3. Radiographic Measurement of Hip Dysplasia⁴²

Table III illustrates two techniques for the radiographic evaluation of hip dysplasia. The Reimer migration index (RMI) is designed to evaluate subluxation of the femoral head while the acetabular index (AI) calculates dysplasia of the acetabulum. Increased AI suggests that a pelvic osteotomy may be necessary. AP: Anterior-Posterior.

Classification	Measurement	Normal Values	Abnormal Values
Reimer Migration Index	Calculates width of uncovered femoral head compared to whole head	< 30%	> 40%: Hip at risk > 60%: Dislocated hip
Acetabular Index	Measured on AP pelvis by calculating angle between acetabular roof and Hilgenreiner's line	> 5 yo: 20o < 5 yo: 25o	> 5 yo: > 20o < 5 yo: > 25o

AP pelvis film between age two and four, and should be followed clinically unless the exam suggests that further imaging is needed.⁴² Patients who ambulate with assistive devices or not at all and who have an RMI of less than 30% should obtain yearly radiographs until age eight, and then biannual films until skeletal maturity.⁴² Patients with an RMI of more than 30% require serial radiographs every six months.⁴²

Numerous soft tissue operations for spastic hips in skeletally immature patients can prevent or address deformity and/or dislocation.⁴² Adductor and iliopsoas lengthening or tenotomy have been shown to improve ROM, prevent dislocation, and reduce the need for bony reconstruction in children with hip muscle spasticity.^{42,46,47}

Bony procedures about the hip may address acetabular and femoral deformities in skeletally immature patients with promising results.^{48,49} Patients without substantial acetabular dysplasia benefit from proximal femoral osteotomy alone or combined with adductor/iliopsoas soft tissue procedures (**Figure 1**).⁴² Such osteotomies often produce varus angulation to address coxa valga and rotation to address femoral anteversion. Patients with an abnormal AI may also require pelvic osteotomy.⁴² Children under eight years of age with an RMI of 30%-60% may be treated operatively with adductor and iliopsoas lengthening/release; however, children over eight with an RMI greater than 40% and all children with an RMI greater than 60% should undergo a pelvic osteotomy combined with proximal femoral shortening/varus osteotomy and soft tissue releases.⁴²

Skeletally mature patients with spastic hip deformities are considerably more challenging to treat. Nevertheless, periacetabular osteotomies combined with varus and de-rotational proximal femoral osteotomies can be successful.^{50,51} In addition, proximal femoral resection procedures have been used to reduce pain and improve sitting in non-ambulatory adult patients with spastic hip deformities.^{52,53} Using the resected femoral head to cap the resection arthroplasty reduces the risk of postoperative heterotopic ossification⁵⁴ Alternatively, a valgus-producing proximal femoral osteotomy pointing the femoral head away from the acetabulum allows indirect load transferring, which prevents proximal migration of the femur seen with resection arthroplasty.⁵⁵⁻⁵⁷ Finally, some authors have demonstrated success with total joint arthroplasty in skeletally mature patients with spastic hip deformities.^{42,58,59}

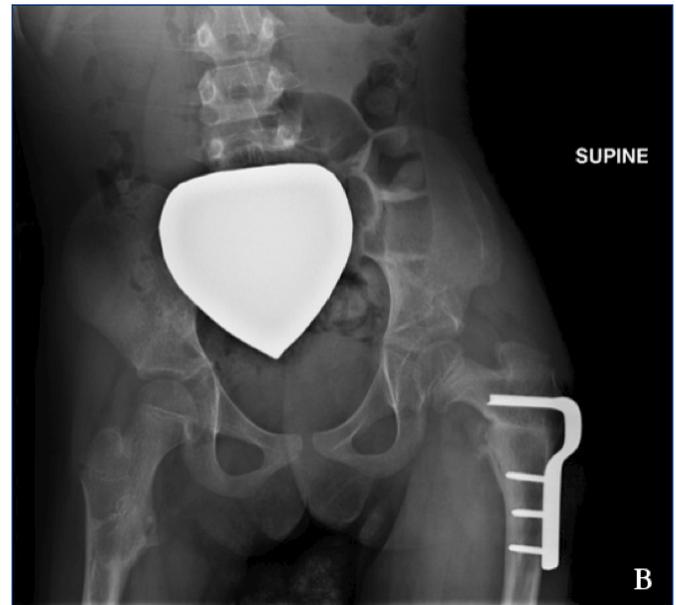
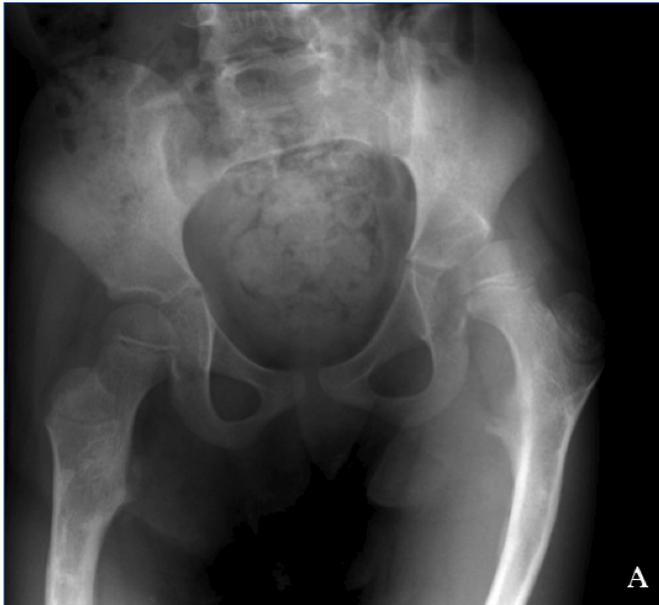
Foot Abnormalities

Foot abnormalities are common in patients with spastic conditions and can lead to pain and difficulty with ambulation, shoe wear, and bracing.⁴² In patients with CP, planovalgus and equinovarus deformities are most common.⁴² Surgical correction of such deformities is reserved for patients aged ten or older, while younger children can often be managed with orthotics.

The flexible planovalgus foot is first addressed

Figure 1. Spastic Hip

Figure 1A illustrates a child with cerebral palsy suffering from a hip dislocation on the left side secondary to spasticity. This child was successfully treated with a proximal femoral osteotomy. The post-operative radiograph is displayed in Figure 1B. *Radiographs courtesy of Dr. Jonathan Schiller.*



with lateral column lengthening through the calcaneus (in patients who ambulate without assistive devices) or via reduction of the talus and calcaneus and a subtalar fusion (in patients with more severe gait problems).^{42,60-62} The medial column can then be assessed; if there is residual forefoot supination a tibialis posterior advancement (in mild cases) or plantarflexion osteotomy of the first ray is performed.⁴² Particularly severe cases may require a talonavicular arthrodesis.^{42,63} Rigid planovalgus foot is treated with lateral column lengthening and triple arthrodesis.⁶⁴

The equinovarus foot is common in hemiplegic patients and may be treated with split tendon transfers in mild, flexible cases.^{42,65,66} Patients with forefoot/midfoot inversion may benefit from split anterior tibial tendon transfers while patients with hindfoot varus are treated with split posterior tibial tendon transfers (but some patients require both procedures).⁶⁵ In more severe but flexible cases, an additional wedge or sliding calcaneal osteotomy is used to address residual hindfoot varus.⁴² In cases of rigid equinovarus foot, triple arthrodesis is often required.⁶⁷

Both planovalgus and equinovarus deformities can be associated with and exacerbated by Achilles contractures related to gastrocnemius tone.⁴² Lengthening of the gastrosoleus complex may be necessary to achieve appropriate correction of these deformities; however, great caution must be observed in diplegic patients with a crouching gait, which may be worsened by plantarflexion weakness.^{42,68}

Spine Abnormalities

Spine deformity is common in patients with spasticity (Figure 2). Scoliosis has been reported in up to 77% of children with CP and is common in more severely affected patients.^{42,69-73}

Figure 2. Neuromuscular Scoliosis

Figure 2 illustrates a patient with cerebral palsy with severe neuromuscular scoliosis. *Radiograph courtesy of Dr. Jonathan Schiller.*



Ambulatory patients develop curves similar to that of idiopathic scoliosis patients; those with severe disease and/or tetraplegia develop a long C-shaped curve that involves the pelvis.⁴² Progression is more common in younger, skeletally-immature children; however, patients with large curves (>50°) may progress after skeletal maturity.^{42,71,72} Furthermore, ambulatory patients exhibit less curve progression compared to non-ambulatory patients.^{42,72} SDR, laminectomy, laminoplasty, and baclofen pump placement (controversial) have been linked to scoliosis development in patients with spasticity.^{42,74-79}

Progression of disease can lead to cosmetic deformity as well as functional, physiologic (respiratory and alimentary), and postural impairment.⁴² Ambulatory patients may be treated like patients with idiopathic curves.⁴² If patients have severe neuromuscular curves, the treatment focuses on improving spinal balance, posture, and sitting ability.^{42,80} Non-operative intervention, such as seating modification, bracing, and serial casting may delay future surgical intervention such as fusion.^{42,80} Caution must be observed in patients with chest wall deformities who may succumb to respiratory compromise with rigid bracing.⁴²

Insertion of growing rods in anticipation of definitive fusion is an option in children with significant growth potential.^{42,81,82} Non-ambulatory children may be best served with a primary T2 to pelvis fusion, which minimizes the incidence of junctional kyphosis and addresses pelvic obliquity, respectively.^{42,83} While most fusions are performed posteriorly, the presence of a rigid pelvis may justify additional anterior surgery or hip reconstruction.⁸⁴ Furthermore, anterior fusion may be indicated to avoid the crankshaft phenomenon in growing children.⁴² Lastly, while some ambulatory patients may avoid fusion to the pelvis, ambulation can be maintained after such a procedure.^{42,85}

CONCLUSION

Spasticity is a devastating manifestation of numerous common neurologic conditions. A multi-disciplinary approach, with non-operative and surgical options, is required to adequately treat patients suffering from this condition. Orthopaedic surgeons can play an important role in alleviating symptoms, preventing complications, and improving function in patients with spasticity.

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Heterotopic Ossification in Neurorehabilitation

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ABSTRACT

Neurogenic heterotopic ossification (NHO) involves deposition of bone in extraskeletal tissue in the setting of a neurological disorder, and its pathophysiology is incompletely understood. NHO can lead to significant disability and functional impairment. NHO initially manifests as pain and joint stiffness. Early diagnosis requires appropriate suspicion and imaging studies to detect the uncalcified collagen matrix that forms in the early stages of NHO. If diagnosis is made in the early phase of NHO, progression may be halted with bisphosphonates, indomethacin or radiation therapy. If NHO progresses to its final stages without intervention, it may restrict joints and render them dysfunctional. Surgical treatment of NHO may restore function, but complications may occur, and prophylaxis and aggressive rehabilitation are essential.

KEYWORDS: Heterotopic ossification, bisphosphonates, indomethacin, radiation therapy, rehabilitation

INTRODUCTION

Neurogenic heterotopic ossification (NHO) refers to the aberrant formation of bone in extraskeletal tissue in the context of a neurological condition.¹⁻⁴ NHO is one of the most common complications associated with traumatic spinal cord (SCI) and brain injuries (TBI), with an incidence of over 50% in SCI and 20% in TBI patients. NHO may also develop with other neurologic conditions including stroke, cerebral anoxia, Guillain-Barre syndrome, tumors and infections.⁵

Various risk factors have been associated with the development of NHO, including prolonged immobilization, muscle spasticity, and a long hospital length of stay.¹ NHO has a tendency to develop around larger joints, particularly the hip and knee; however, it may develop near any joint, including the shoulders, elbows, and spine.² The exact pathophysiology that causes NHO formation is not completely understood.⁶

Pathophysiology

NHO is thought to be caused by the induction of pluripotent mesenchymal stem cells by signaling factors that are present in patients with neurologic conditions or trauma. Recent evidence suggests that these mesenchymal cells originate from muscles and their differentiation is dependent on

soft tissue macrophages.⁷ The resulting osteogenic cells lead to aberrant bone formation in extraskeletal tissue.⁵ Many humoral factors have been implicated, particularly BMP-4 and substance P; however, none have been definitively proven to be the primary culprit in NHO.⁷⁻⁹

Several studies have investigated the role of peripheral nerves in NHO. Campos da Paz et al. suggested that altered proprioception could lead to tissue irregularities that predispose patients to NHO.¹⁰ Salisbury et al. demonstrated that sensory nerves are stimulated by BMP-2, an osteogenic factor that induces a neuroinflammatory response and leads to the proliferation and release of osteogenic cells. NHO can be a painful condition, but the etiology of the pain is unclear. Local factors such as substance P and neuropeptides released in local soft tissues are thought to stimulate peripheral nerves, leading to the perception of pain.¹¹

NHO formation occurs in three phases.^{12,13} First, immature NHO is deposited in extraskeletal tissue. This is primarily comprised of well-organized collagen fibers with minimal calcification. Second, an inflammatory process leads to the vascularization of this tissue, which allows osteogenic cells to further induce immature bone formation. During the final phase, remodeling and maturation occurs. This last phase is associated with mature bone and minimal activity on 99m-technetium bone scanning.

Diagnosis

Early NHO manifests as joint stiffness, skin erythema, swelling and pain. Without appropriate suspicion, the diagnosis of early NHO is elusive because the condition is not evident on radiographs or computed tomography scans.¹⁴ Patients with neurological disabilities are at high risk of deep venous thromboses (DVTs), and this condition should be excluded. Once critical diagnoses like septic joint, tumor, and deep venous thromboses are ruled out, a patient at high risk of developing NHO must have further studies to support the diagnosis.¹⁵

Early diagnosis of NHO is important because it allows for interventions that may stop its progression. Early diagnosis can be accomplished with 3-phase 99m-technetium bone scanning, MRI or ultrasonography. Bone scans may detect lesions suspicious for NHO in a high-risk patient; however, the specificity for making the diagnosis is low and further testing is required to prove its efficacy. Similar to bone scans, MRI is able to reliably detect NHO but the specificity is low.

Recently, it has been shown that ultrasound has proven to be a reliable method of diagnosing NHO in its early phases.¹⁴

Laboratory studies are not diagnostic for NHO; however, they may help identify the presence of NHO in its early inflammatory phase. Simultaneous elevations of phosphorus and alkaline phosphatase have been associated with NHO.¹⁶ Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) are typically elevated in the acute inflammatory phase of NHO and normalize once this phase is over.¹⁷

Treatment

Research studies of prophylaxis with bisphosphonates and indomethacin to prevent heterotopic ossification have shown similar efficacy.¹⁸⁻²⁰ These drugs target the early inflammatory phase of NHO. Generally, indomethacin is not tolerated as well as bisphosphonates, with adverse effects being reported in up to 31% of patients.²⁰ Although bisphosphonates are effective in NHO prophylaxis, they are

up to six times more costly.²⁰

In the early stages of NHO, radiation therapy has also been effective in halting the progression of NHO.¹² Unlike bisphosphonates and NSAIDs, which target the inflammatory cascade that activates osteogenic cells, radiation therapy inactivates the pluripotent osteoprogenitor cells that have been mobilized by the inflammatory cascade.²¹ In patients diagnosed with early NHO as well as those who had NHO resection and radiation therapy, progression or recurrence was prevented in over 70%.²²

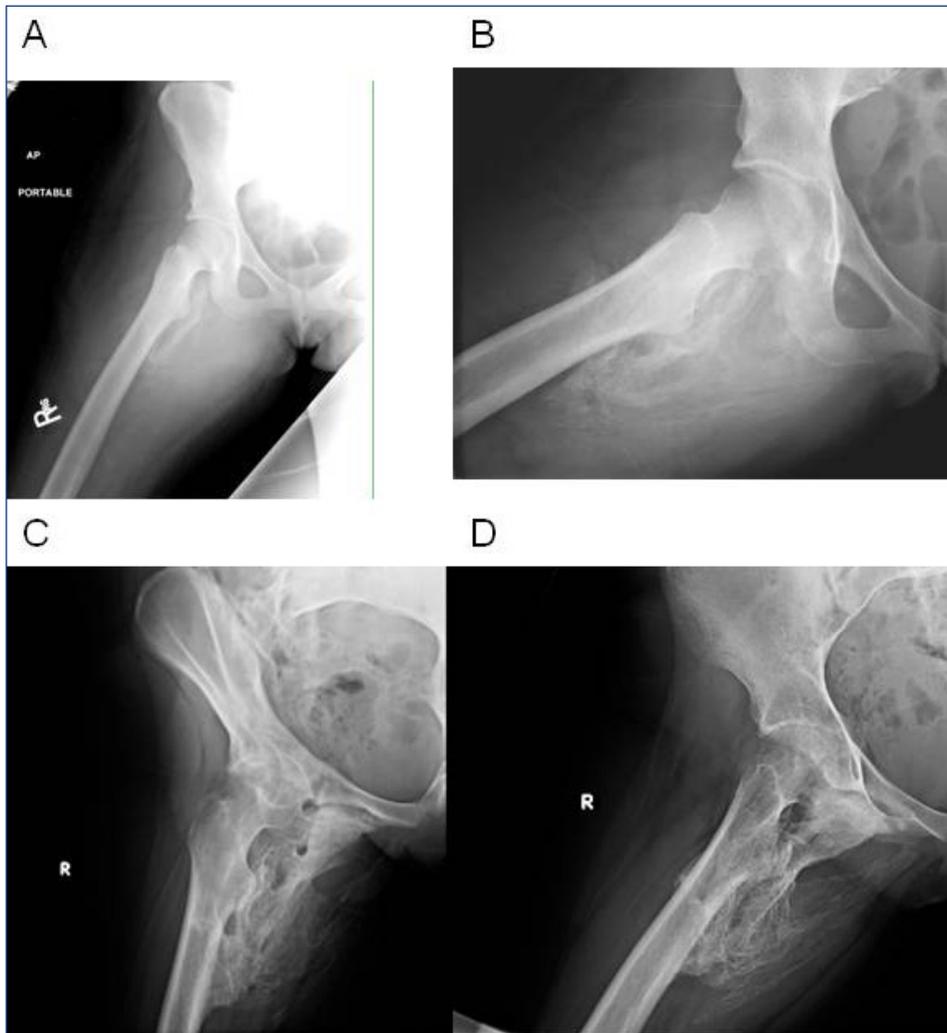
In the late stages of NHO, associated with functional impairment and pain, excess bone must be excised surgically (**figure 1**).²³ It has been reported that excision should not be performed until the growth and maturation phases have ended, which has been estimated to be over one year for NHO related to SCI and over 1.5 years for TBI.^{12,24}

However, despite confirmation of the end of the growth or maturation phases of NHO, recurrence rates of NHO have been reported in over one-third of patients who undergo excision.^{12,25}

Fortunately, surgical resection that is combined with prophylactic bisphosphonates and radiation therapy has significantly reduced the recurrence of NHO.²⁶ This suggests that the risks and benefits of waiting to excise NHO until the maturation phase must be weighed with the risk and benefits of earlier excision. Earlier excision may require less surgery if appropriate prophylaxis is used to prevent recurrence, as it may avoid the soft tissue contractures that often develop with NHO and the resulting immobilization.

Regardless of the phase of NHO or intervention utilized for prophylaxis or treatment, physical and occupational therapy have a significant role in the management of NHO. Physical therapy has not been proven to independently prevent the formation of NHO.¹² However, range-of-motion exercises are important in preserving joint function by preventing soft tissue contractures.¹² In the late stages of NHO, where function is impaired by a restricted joint, excision is generally combined with postoperative prophylaxis and a rehabilitation program to optimize function and prevent recurrence.²⁷

Figure 1. 19-year-old female with heterotopic ossification of the right femur following a brain aneurysm rupture. She had an angiogram with resultant hematoma formation. Images A and B were taken 1 month after the hematoma formation and images C and D were taken at 9 months.



CONCLUSION

NHO deposition of bone in extraskeletal tissue is a disabling condition that is common following SCI, TBI, and other neurological conditions. Early diagnosis requires appropriate suspicion and imaging studies. If the diagnosis of NHO is made in the early phase, progression can sometimes be halted with bisphosphonates, indomethacin or radiation therapy. If NHO progresses to its final stages without intervention, it could restrict joints and render them dysfunctional, ankylosed, and painful. In cases where excision is performed to restore function, prophylaxis and regimented rehabilitation are required to maintain function and improve outcomes.

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