





**STRETCHING**

**SPLINTING**

**ELECTRICAL STIMULATION**

**BRACING**

**DIAZEPAM**

**PUMP IMPLANTATION**



While visiting her father in South Carolina earlier this year, Karen Hewlett experienced one of the most frightening situations a mother can face: her 2-year-old son, Kyle, disappeared. After a frantic search, Kyle's grandfather found the boy in a nearby pond, jumped in and pulled him out. The grandfather quickly administered CPR and the family called 911—but the preschooler's medical ordeal was far from over. • Kyle was rushed to a hospital, where his muscles went into painful spasms. His back arched like an inverted "C" in a contortion so extreme that his toes nearly touched his head at times. His small hands clinched into tight fists, his ankles and feet contracted, his heart raced and his blood pressure rose. • Kyle received beta blockers to lower his blood pressure, narcotics to ease his pain and oral baclofen—a muscle relaxer and antispasticity agent—to relax his muscles. The medications, however, failed to control Kyle's spastic movements. His parents helplessly watched the painful episodes.

Children who have suffered damage to the part of the brain that controls voluntary movement or to the nerve fibers that travel from the brain down to the spinal cord commonly experience such spasms.

"If you have some sort of lesion of the central nervous system you experience spasticity and weakness," said Maurice Sholas, M.D., Ph.D., Medical Director of the Children's Healthcare of Atlanta Rehabilitation Services.

The most common causes of such lesions are cerebral palsy and brain trauma from severe head injuries, Dr. Sholas said. Other causes include near-drowning or suffocation, tumors, spinal cord injury, stroke, neurodegenerative illness and multiple sclerosis.

Whatever the cause, spasticity and weakness can lead

to a decrease in fine muscle control, said Dr. Sholas. In mild cases, nothing more than an awkward gait may result, particularly when a child runs. But more often, spasticity produces debilitating conditions. "Spastic muscles do not grow as quickly as nonspastic muscles, so it can lead to a contraction that causes the bones to be twisted or torqued in ways they should not be," he said.

This, in turn, can cause joints to become unstable, which can lead to joint degeneration and dislocation. Furthermore, children with spasticity often are extremely thin and inadequately nourished because of the number of calories the body expends to keep muscles in

spasm. Spasticity also is painful and increases a child's risk of falls. "Spasticity cannot only cause significant problems for mobility but it also can cause problems with care and comfort," said Dr. Sholas. "It is extremely difficult, for example, to open a child's legs to change their diaper if they are very spastic."

### Multiple treatment options exist

Throughout the continuum of pediatric spasticity, there are multiple treatments available. At Children's, for example, the response generally involves a multidisciplinary approach, consisting of medications, surgery and physical and occupational therapy. Here's a closer look at each step:

#### Medications

■ **Oral medications**—If spasticity is widespread, a variety of oral medications may be helpful. In addition to acting systemically, oral medications are noninvasive and reversible. For children whose symptoms fluctuate, dosages can be adjusted to accommodate changes. The primary drawback is the risk of systemic side effects, said Dr. Sholas.

For 16-year-old Christopher Saunders Lee of Douglasville, Ga., the side effects of oral baclofen prescribed to ease the spasticity resulting from cerebral palsy were not worth the minimal help they provided.

"He had a hard time focusing mentally," said his mother, Olgarita Lee. While oral baclofen is often

helpful for more widespread spasticity, for Christopher, whose problems were confined mainly to his lower body, a baclofen pump turned out to be a better option.

Oral medications commonly used for spasticity include:

- Baclofen (Lioseral<sup>®</sup>)
- Diazepam (Valium<sup>®</sup>)
- Dantrolene sodium (Dantrium<sup>®</sup>)
- Tizanidine (Zanaflex<sup>®</sup>)

■ **Injected Medications**—If a specific area of the body is affected, injected medications provide a more targeted treatment with less risk of side effects. "If someone has a specific muscle that is a problem, then you want a focal type intervention, so I would use those injectable-type medications because they are injected specifically into the muscles that are problematic," said Dr. Sholas. "If you just use injected medicine, it is focal and it is reversible when the medicine wears off."

Injected agents commonly used for spasticity include:

- Phenol
- Botulinum neurotoxin Type A (BOTOX<sup>®</sup>)
- Baclofen (Lioseral<sup>®</sup>)

#### Surgery

Many children benefit from orthopaedic surgery to correct the effects of spastic muscles. For Kyle, heel cord release stopped the contracture in his feet and ankles. Treating the source of spasticity, however, involves one of two neurosurgical procedures: the implantation of a pump to deliver medication directly to the spinal fluid or selective dorsal rhizotomy.

■ **Pump implantation**—For a reversible treatment, surgeons may implant a pump underneath the skin of the abdominal wall to deliver medication through a catheter directly into the spinal fluid to ease spasticity. The catheter's placement will depend on the area being treated.



#### IMPACT OF SPASTICITY

- Pain
- Contracture
- Functional limitations
  - Upper limb—hygiene and dressing
  - Lower limb—hygiene, transfers, seating, walking
- Fatigue
- Nutritional compromise
- Poor self-image due to abnormal limb posture
- Increased risk of falls
- Prolonged immobility
  - Skin breakdown
  - Infection



On April 28, Dr. Boydston implanted a pump and by May 12 Kyle's spasticity had stopped. His muscles were relaxed enough that he was able to sit in a wheelchair for the first time since his accident, according to his mother, Karen.

In Christopher's case the pump delivered medication directly to his legs, which were most severely affected.

"The benefits of pumps are that they are programmable and reversible," said William R. Boydston, M.D., Ph.D., Chief of the Children's Myelodysplasia Clinic and Neurosurgeon at Pediatric Neurosurgery Associates at Children's. "You can increase the dose using a magnetic programmer that you hold over the skin. You can program the pump to deliver any dose you want.

"If a child needs more medication after school—their muscles are tighter after school than in the morning—you can program it so it delivers more medicine in the afternoon hours than the morning hours," he said.

One downside of the pump is that it must be refilled every three months or so. Even so, refilling the pump is a simple procedure that involves minimal discomfort for the child.

For the Hewlett family, who brought Kyle to Children's in hopes of relieving his spasticity, an implantable pump that delivers baclofen to Kyle's spinal fluid proved to be the best solution. On April 28, Dr. Boydston implanted the pump and by May 12 the spasticity had stopped. His muscles were relaxed enough that he was able to sit in a wheelchair for the first time since his accident, said his mother, Karen.

#### SPASTICITY MANAGEMENT GOALS

- Decrease tone
- Increase range of motion
- Enhance fit of orthoses
- Decrease contracture
- Postpone orthopaedic surgery
- Decrease pain or spasm
- Enhance appearance
- Facilitate rehabilitation
- Improve positioning
  - Improve functioning
  - Activities of daily living
  - Mobility
  - Sleep
  - Dressing
- Decrease caregiver burden

■ **Selective dorsal rhizotomy** – For children with predominantly lower extremity spasticity, doctors and families may choose selective dorsal rhizotomy, a procedure in which sensory nerves involved in spasticity are partially cut to stop the abnormal impulses that cause the muscles to jerk too vigorously or continuously.

To perform a selective dorsal rhizotomy, the surgeon divides each of the sensory nerves in the lower part of the spine, stimulates some of these nerve fibers to see how they conduct and then cuts nerves that do not conduct normally.

In a typical procedure, about 50 percent of the sensory nerve fibers are cut, said Dr. Boydston. "It does not leave children numb in their legs," he said. "They still have normal sensation, but selective dorsal rhizotomy interrupts that abnormal reflex arch and allows them to have their spasticity reduced."

Following the procedure, children must be in the hospital for up to four days and then have frequent physical therapy for a couple of months.

Four months after his incident, Kyle is laughing and smiling. His mother said she is seeing familiar facial expressions and signs of his personality before the accident. Best of all, the pain and spasms in his back have ended.



"Rhizotomy is good because you are not placing pumps, which have to be refilled, reprogrammed and eventually replaced but at the same time if you cut a nerve, it is not reversible," said Dr. Boydston.

### Physical and Occupational Therapy

Regardless of the medical or surgical treatment chosen, children treated for spasticity will almost always require extensive physical and occupational therapy to maintain muscle and soft tissue length, improve body symmetry and facilitate functional activity. Interventions provided by physical and occupational therapists include:

- Ambulation and gait training
- Exercise (stretching, weight bearing)
- Activities of daily living training
- Modalities (heat, ice, ultrasound, electrical stimulation)
- Positioning (splinting, casting, bracing, seating)
- Caregiver and patient education

### Spasticity Clinics

Two days a month, Children's offers a spasticity clinic where children have the opportunity to be evaluated by physiatrists, surgeons, physical therapists and social workers to determine the best course of treatment for the problem.

Many families come to the clinic not realizing all of the options available to help their child.

"A lot of people do not necessarily think there is anything you can do with spasticity. But in truth, spasticity is something we can manage," said Dr. Sholas.

The treatment used and expectations of treatment results depend on the severity of the problem.

"There may be kids with very mild spasticity in their calves so they bounce a little bit when they walk and we can fix that completely," he said. "But there are other kids who are catastrophically disabled through spasticity and we can make them better and more functional," he said.

New treatments offer hope and help to many children. "We must offer a prognosis and a treatment appropriate to each child's case. We focus on what the child wishes to accomplish. Some kids are walking and they want to walk better or faster or with more energy," said Dr. Sholas. "Children who are not walking may wish to be more comfortable and make it easier for their caregivers to help them. Kids with fairly mild spasticity may want that last little bit of help so they can maintain their independence."

Once Dr. Sholas learns from patients and their parents what they want to accomplish, he works with them to design a treatment plan to meet their goals.

Today, the Hewlett and Lee families are encouraged as their children positively respond to their treatments.

Olgarita Lee said it is probably too early to know how much the baclofen pump will help Christopher, but she has noticed that when she opens his hands they do not immediately close back into fists the way they used to. And recently, for the first time in years, she saw him walking with both feet flat. "It will take a while before we really know," she said. "I hope that he will be running by next summer."

Four months after his incident, Kyle is laughing and smiling. His mother said she is seeing familiar facial expressions and signs of his personality before the accident. Best of all, the pain and spasms in his back have ended. Although she knows Kyle still faces a long road of rehabilitation, she is hopeful. "We have our little boy back." ©

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