The Physical Medicine and Rehabilitation Division (PM&R) at Barrow Neurological Institute at Phoenix Children’s Hospital is dedicated to maximizing the functional recovery of children with neurological disorders. The primary goals are to reintegrate children back into school system and community as quickly as possible.

Ewa Brandys, MD, division chief of PM&R, received funding from the Leadership Circle to purchase the RTI 600, a robotic walking device with functional electric stimulation manufactured by Restorative Therapies, Inc. Many children with neurological disorders lose their ability to walk due to severe muscle weakness and impaired motor control, resulting in a sedentary lifestyle. This sophisticated, cutting-edge equipment presents the opportunity for children to reach their highest recovery potential in a shorter period of time and fulfill their dreams of walking. The device can be used on children as young as 3 years of age.

“Our goal is to deliver the best patient- and family-centered care to each child who comes to us.”
Neuromuscular Disorders: A Clinical Overview

Neuromuscular disorders present in one in every 1,000 live births. They include myopathy, muscular dystrophy, neurojunction disorders, and inflammatory muscle disorders. Neuromuscular disorders occur when nerve cells that control muscle function become unhealthy or die, causing a communication breakdown between the nervous and muscular systems. Eventually, the breakdown in communication causes the muscles to progressively weaken and deteriorate. Common symptoms include muscle twitching, cramps, aches, pains, and problems with joint and movement functionality.

Neuromuscular disorders often present with progressive weakness affecting the muscles of the trunk, arms, and legs. This weakness impacts mobility, resulting in the need for adaptive equipment such as wheelchairs, Hoyer lifts, toileting and shower chairs and, in some cases, special beds. Weakness and decreased mobility often lead to problems involving the bones of the lower extremities and spine. The most severe diagnoses impact the upper extremities, which makes it difficult to perform activities of daily living, such as eating and dressing independently. Measures to prevent these problems can be started early, but orthopedic surgery is often required.

The impact of neuromuscular disorders beyond the muscular system, often having a significant effect on the muscles of the heart and respiratory system. Muscles of the upper airways and respiratory system can weaken because of neuromuscular diseases in the same way other muscles of the body are weakened. Respiratory failure in neuromuscular patients occurs from three main mechanisms: aspiration secondary to oropharyngeal fatigue of respiratory muscles; or weak cough. Respiratory failure is especially prevalent in patients with myasthenia gravis and Guillain-Barre syndrome. Ongoing assessments with cardiology and pulmonology are important parts of a comprehensive care plan.

In addition to respiratory problems, many patients experience endocrine and gastrointestinal issues. Consultation with an endocrinologist is needed to address possible thyroid and growth-hormone irregularities. Gastroenterology consultation is also necessary to address feeding, swallowing and intestinal problems that often accompany this patient population.

Neuropsychological health is also impacted. The physiological processes, which cause the physical symptoms of neuromuscular diseases, can also affect behavior, learning and information processing. Pharmaceutical intervention may be required to treat behavioral issues. Neuropsychological testing can identify learning difficulties which can be addressed with educational adaptations.

Diagnosis of a chronic, progressively debilitating disease has an overwhelming impact on the overall mental health of children. Support services, including counseling from psychologists and Child Life specialists, can help ease the mental and emotional burden. Parents, siblings and other family members can also struggle with diagnosis. Learning a child has a progressive, lifealtering disease can be devastating. In addition coping with the diagnosis and the multiple medical interventions, families must make adjustments to their day-to-day routines, such as school and work schedules, coordinating sibling care and extracurricular activities.

From the Director

Each New Year offers hope for the year to come. It is our mission to improve the health and quality of life of children with neuromuscular disorders through the promise of state-of-the-art care, advanced research and professional and community education. As we embark on 2013, we intend to offer much more than just hope to children living with debilitating neuromuscular and neurological disorders.

In this newsletter we will highlight the innovative, state-of-the-art developments in medical management and clinical care that are improving the health and quality of life for patients with neuromuscular disorders at Barrow Neurological Institute at Phoenix Children’s Hospital.

The Neuromuscular Program offers hope to children and families with neuromuscular disorders as they are managed by a multidisciplinary team of specialists to provide state-of-the-art care for the treatment and management of these complex disorders. The program follows clinical pathways and algorithms to coordinate and centralize the subspecialty care provided to patients that has become the standard of practice at Barrow at Phoenix Children’s.

The life expectancy for children with neuromuscular disorders has increased due to the advancement of pharmaceutical interventions and rehabilitative treatment options. The Neuromuscular Program shares the latest evidence-based protocols to create individually tailored treatment plans that optimize patient outcomes.

Dr. Ewa Brandys is also optimizing patient outcomes by restoring ambulation in children who have lost their ability to walk by retraining the legs and spindled cord to walk again. The machine, which recently arrived at the hospital, will be part of our new inpatient and outpatient rehabilitation center, which will undergo a renovation that begins in the next few months. I look forward to sharing our progress and success with you along the way.

Improving the health and quality of life of children with neuromuscular disorders requires keeping up with the latest in science and clinical care. To that end, Barrow at Phoenix Children’s would like to invite you to the 16th Annual Children’s Neurosciences Symposium, being held from February 24 to February 27. The symposium will highlight the most recent advances and current issues in pediatric neuromuscs.

For more information on the symposium or to register, visit www.phoenixchildrens.com/cm2013.

This year is forecast to be an extraordinary one for Barrow Neurological Institute at Phoenix Children’s Hospital. It is my privilege to provide you with insight to the amazing care, science and knowledge being developed here.

I hope you find our newsletter helpful and informative. Please, feel free to contact me or the staff if you have any questions about improving our newsletter, other topics of interest, or improving our care of the children and service to you. Thank you very much.

I also encourage you to visit our website at www.phoenixchildrens.com/RNL to learn more about our programs and accomplishments or like us on Facebook.

The impact of neuromuscular disorders reaches beyond the muscular system, often having a significant effect on the muscles of the heart and respiratory system.

NEUROMUSCULAR DISORDERS

Muscular Dystrophies

- Myotonic dystrophy
- Duchenne muscular dystrophy
- Becker muscular dystrophy
- Limb-Girdle muscular dystrophy
- Congenital muscular dystrophy
- Oculopharyngeal muscular dystrophy

Motor Neuro Diseases

- Infantile progressive spinal muscular atrophy
- Intermediate spinal muscular atrophy
- Juvenile spinal muscular atrophy

Inflammatory Myopathies

- Polymyositis
- Dermatomyositis
- Inclusion-body myositis

Diseases of the Neuromuscular Junction

- Myasthenia gravis
- Lambert-Eaton (myasthenic syndrome)
- Congenital myasthenic syndrome

Diseases of Peripheral Nerve

- Charcot-Marie Tooth disease
- Friedreich’s ataxia

Metabolic Diseases of the Muscle

- Chronic inflammatory demyelinating polyneuropathy
- Guillain-Barre
The Neuromuscular Program at Barrow

The Neuromuscular Program at Barrow Neurological Institute at Phoenix Children’s Hospital is directed by Saunder Bernes, MD, and is a designated Muscular Dystrophy Clinic site by the Muscular Dystrophy Association. The program strives to provide consistent, coordinated, multidisciplinary collaborative care for children with neuromuscular disorders, a model that fosters positive outcomes.

The divisions of Neurology, Physical Medicine and Rehabilitation (PM&R), Orthopedics, Pulmonology, Cardiology, Endocrinology, Gastroenterology, and Palliative Care join together to create a multidisciplinary medical team. Support services from dedicated, specially trained health care professionals, such as nurses, social workers, and physical therapists, complete the comprehensive neuromuscular team.

“...The face of neuromuscular disorders is changing and we are changing with it. Pharmaceutical interventions and advanced treatment options are prolonging ambulation and increasing life expectancy. They are providing patients with hope for improved quality of life,” Saunder Bernes, MD.

Diagnosis and Services

Dr. Bernes is the program’s medical director and primary neurologist responsible for diagnosing specific neuromuscular conditions. The diagnostic workup includes a detailed patient and family history, neurological exam, diagnostic laboratory and genetic testing, and diagnostic workup includes a detailed patient and family history, presenting symptoms, diagnosis, and clinical findings. The patient is also responsible for diagnosing specific neuromuscular conditions.

After the initial assessment and diagnostic screenings are complete a comprehensive, coordinated care plan is developed. Referrals to the appropriate pediatric specialties are ordered based on the individual’s presenting symptoms, diagnosis, and clinical findings. The patient is also presented with the opportunity to register with the Muscular Dystrophy Association, if appropriate.

Clinical Team

Medical Director/Neurology
Saunder M. Bernes, MD
Nurse Manager/Coordinator
Nancy B. Quay, MS, RN, CNRN
Physiatry/Physical Medicine
Eva Brandtys, DO
Orthopaedics
Lee Segal, MD
Pulmonology
James Woodward, MD
Aparna Rao, MD
Kristen Meliska, BS, RN
Palliative Care
Tressia Shaw, MD
Endocrinology
Grazyna Pietros-Sobczak, MD
Cardiology
Erik Ellsworth, MD
ANCILLARY SUPPORT
Social Work
Child Life
Rehabilitation
Physical Therapy
Occupational Therapy
Speech Therapy
Gastroenterology
Neuropsychology
Psychology
Psychiatry

Who to Refer:

Children exhibiting the following symptoms are candidates for referral:
• Elevated CPK
• Progressive muscle weakness
• Progressive deterioration of motor skills
• Chronic muscle cramps
• Family history of a neuromuscular disorder

How to Refer:

By Phone: (602) 933-0970
By Fax: (602) 933-0068

Leadership

Saunder Bernes, MD
Pediatric Neurologist
Medical Director,
Neuromuscular Program

Eva Brandtys, MD
Division Chief,
Physical Medicine and Rehabilitation

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Diagnosis and Services

Dr. Bernes is the program’s medical director and primary neurologist responsible for diagnosing specific neuromuscular conditions. The diagnostic workup includes a detailed patient and family history, neurological exam, diagnostic laboratory and genetic testing, and surgical muscle biopsy. When available, nationally recognized, evidence-based practice guidelines are used to make the diagnosis.

After the initial assessment and diagnostic screenings are complete a comprehensive, coordinated care plan is developed. Referrals to the appropriate pediatric specialties are ordered based on the individual’s presenting symptoms, diagnosis, and clinical findings. The patient is also presented with the opportunity to register with the Muscular Dystrophy Association, if appropriate.

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Pediatric Neurologist
Medical Director,
Neuromuscular Program

Eva Brandtys, MD
Division Chief,
Physical Medicine and Rehabilitation

Many robotic devices for gait training are commercially available and cleared by the FDA. They improve the efficacy of therapies and patient satisfaction. In 2011, the RTI 600 was cleared by the FDA and is the first to combine a robotic motorized device with computerized functional electrical stimulation (FES).

RTI 600 consists of motorized footplates and can provide vibration and perturbation during standing. The patient is secured in a harness on the electrically powered lift. This allows support for an adjustable amount of body weight and is called partial body weight supported stepping or standing.

The machine’s SAGE smart functional electric stimulation system can power 10 muscle groups to produce walking movements by true muscle contractions. Electric current is delivered to muscles through adhesive electrodes in sequential pattern and uses the child’s own muscles’ activation to enable standing and walking exercise. FES has been studied in children and adults and has shown reverse muscle atrophy, improved circulation, increased range of motion, and reduced muscle spasms.

Many neurological disorders including stroke, cerebral palsy, brain injury, tumor, spinal cord injury, transverse myelitis, spina bifida; and multiple sclerosis have devastating effects on a child’s mobility. The sedentary lifestyle in children can be significant, not only affecting quality of life and self-esteem, but their overall health. Children who cannot walk or exercise frequently experience life-long complications such as muscle atrophy, joint deformities; brittle bones leading to fractures; poor cardiovascular endurance; obesity; premature cardiac disease; skin breaks; and diabetes. Restoration of function and neurological recovery occurs through neural plasticity, which can help children avoid these unfortunate consequences and live active lives.

Neural plasticity is activity dependent and occurs when a person is practicing the specific skills that need to be restored. Intense and frequent repetition of the activity is needed so motor learning can occur and strength is restored. Achieving this level of intensity is rarely possible with traditional rehabilitation approach. Traditional gait training requires prolonged time and assistance of one to three therapists per patient. These traditional therapy sessions usually can only accomplish between 10 and 200 steps and require significant physical effort of the treating therapists and patients. Gait patterns of these children are usually abnormal due to weakness and impaired coordination, so the motor learning of walking is not optimal.

Based on scientific evidence, more intensive and frequent practice produces better outcomes than traditional therapies. This high level of exercise intensity is achieved with use of sophisticated robotic equipment designed to replicate and support natural functional movements and allows patients to practice hundreds of steps during a single therapy session. Using mechanical devices also prevents physical exhaustion and injuries in therapists. Robotic devices provide motor precision of functional training and stimulate robust positive neuroplasticity.
Patient Story: 100% for Daniel
Family Copes with HH Diagnosis, Finds Hope and Healing at Barrow

Colds, coughs, sneezes and the flu are part of any cold weather season. In December of 2010, Daniel Mendez,was suffering from a viral infection when he began vomiting. Daniel’s mom, Nancy, brought him to the hospital for evaluation. While being treated doctors noted that Daniel had elevated liver enzymes and recommended that they follow up with their pediatrician upon discharge.

Suspected of having an enlarged liver (Hepatomegaly). Daniel’s pediatrician referred him to a pediatric gastroenterologist. After a repeat lab analysis, Daniel was given the clinical diagnosis of Duchenne Muscular Dystrophy. The diagnosis was given on March 11, 2011, a day Nancy will remember: “After getting the diagnosis we were devastated and lost with a diagnosis that we did not expect in the beginning of July, scheduling our appointment with Dr. Bernes. That was the day everything began to organize for us.”

Daniel was then referred to Saunder Bernes, MD and the Neuromuscular Clinic at Barrow Neurological Institute at Phoenix Children’s Hospital. He received a telephone call from Nancy Quay with referral to see a cardiologist, pulmonologist, physical therapist and orthopedic doctors.

One of the key components to understanding neuromuscular diseases is genetic testing. Acquiring the appropriate testing allows the team to create a more comprehensive, holistic and individualized plan of care for the patient. “Genetic confirmation is the standard of care. Identifying the exact deletion or duplication can predict prognosis, direct therapy and predict carrier status. Unfortunately, insurance companies are often reluctant to cover the testing needed. It is critical that we secure funding for genetic testing for our patients. This noninvasive method of diagnosis should be mandatory,” Dr. Bernes. Luckily for Daniel, they were able to have the necessary testing and develop a treatment plan specific to his needs. The goal of most treatment plans is to prolong the use, strength and functionality of the muscles and bones. Many patients, including Daniel, begin a steroid regimen as part of their treatment plan.

“Daniel started taking a steroid in May 2012 and although this drug has many side effects, right now it is giving him more strength. He is stable and happy.”

The care provided in the Neuromuscular Clinic gives families hope for the future. “Our lives changed completely and our future is uncertain. Although sometimes the sadness is very large, we have hope, waiting for a better treatment and cure for Duchenne Muscular Dystrophy.”

For more information and to register www.phoenixchildrens.com/cns2013

Upcoming Events

2013 Neuroscience Symposium
February 24 to 27, 2013
The Ritz-Carlton, Biltmore, Phoenix, AZ
The symposium will feature nationally prominent faculty, presenting topics that highlight the most recent advances and current issues in pediatric neurosciences and is broken into four half-day sessions:
• Developmental Pediatrics /Autism Spectrum
• Neurological Sports Medicine
• Fundamentals of Selected Neurological Disorders
• Nursing and Allied Health for the “Neuro” Child
• Brain Surgery for the PCP

Recent Advances in Pediatric Epilepsy
April 20, 2013
Phoenix Children’s Hospital, Melvin L. Cohen Conference Center
12:30 registration; 1-5 p.m. conference
Audience: Consumers, school nurses, public

Awards and Accolades

U.S. News & World Report Recognition
• The Hospital’s divisions of Neurosurgery and Neurology are ranked 40 nationally, in the 2012-2013 U.S. News & World Report Best Children’s Hospitals list.
• Seven physicians from Barrow at Phoenix Children’s were named to the U.S. News & World Report Top Doctor List. Congratulations Drs. Adelson, Bernes, Blitz, Haynes, Jarrar, Kerrigan, and Kottlow.
• Dr. Adelson is ranked in the Top 1 percent nationally for an Neurosurgery/Pediatric Neurosurgery.

Welcome to New Faculty and Staff

Barrow at Phoenix Children’s is excited to welcome Beth Trevino, MD and Laura Davis, MS, CRA to our institute.

Dr. Trevino joins the Developmental Pediatrics Team and has over five years experience in developmental and behavioral pediatric diagnosis and management. She is fluent in Spanish and has a special interest in medically underserved populations.

Laura Davis joins the Research Department as the Senior Grants and Contracts Administrator with over 10 years’ experience in research and administration. Ms. Davis looks forward to serving the Barrow at Phoenix Children’s Neuroscience community and supporting the growth of research funding.

Connect with us
Want to keep up with the latest news and events? Visit us on
www.facebook.com/BarrowAtPhoenixChildrens or www.phoenixchildrens.com/BarrowEvents

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Recently, through the generous support of two individual donors, Barrow Neurological Institute at Phoenix Children’s Hospital was able to open a new pediatric brain tumor lab in collaboration with Translational Genomics Research Institute to develop new strategies in the fight against pediatric brain cancer.

This investment in our Institute helps us develop new paradigms for clinical care and cures for these children. You can express your support to the Institute in ways that complement your own personal interests as you help us evolve and continue to set new standards in pediatric neurological care.

**WHAT PHILANTHROPY SUPPORTS:**

- Development/enhancement of existing and/or new clinical programs
- Funding cutting-edge programs in clinical, translational and laboratory/experimental research
- Community and professional educational initiatives
- Institute’s infrastructure supporting the Biorepository and Bioinformatics and Data Center.

**WAYS TO GIVE:**

- Make a memorial or honor gift
- Name an Institute’s Fund
- Contribute to the Director’s Fund
- Provide funds for Endowed Chairs for research and program leadership

For more information call Bonnie Morgan at (602) 546-2607 or visit [www.phoenixchildrens.com/DonateBarrow](http://www.phoenixchildrens.com/DonateBarrow)

Sign up for our e-newsletter at [www.phoenixchildrens.com/neuronews](http://www.phoenixchildrens.com/neuronews) or scan this code with your smartphone.