A *promise* to be your partner in pediatric care.

Services for Children with Neuromuscular Disabilities
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1. THE PROMISE TO CARE FOR EVERY CHILD AS IF THEY WERE OUR OWN

That’s the Nemours promise. As one of the premier pediatric health care systems in the nation, Nemours has made a promise to do whatever it takes to prevent and treat the most disabling childhood conditions. Every day at Nemours, the country’s top pediatric specialists, researchers, educators and caregivers are working together with our physician partners, schools and communities to fulfill that promise.

Established in 1936 as a legacy of Alfred I. duPont, Nemours cares for about a quarter of a million children annually in Delaware, Pennsylvania, New Jersey, Maryland and Florida, as well as children from across the nation and around the world. Ranked among the top children’s hospitals in the country, the Nemours/Alfred I. duPont Hospital for Children in Wilmington, Delaware offers intensive and acute inpatient and outpatient services in more than 30 disciplines. It is the only Level II Pediatric Trauma Center in Delaware. A progressive new Nemours children’s hospital and health campus is coming to Orlando, Florida in 2012. These facilities, along with outpatient clinics, prevention and health promotion initiatives, Nemours BrightStart! Dyslexia Initiative and numerous educational programs continue our founder’s legacy. Millions of children have survived catastrophic illness and thrived throughout childhood, thanks to the entire Nemours system of care.

2. SERVICES FOR CHILDREN WITH NEUROMUSCULAR DISABILITIES

Children with neuromuscular disabilities require the collaborative approach of a multidisciplinary team. At the duPont Hospital for Children, we bring together world-renowned specialists in orthopedics, neurology, neurosurgery and rehabilitation to determine the most effective way to help each child reach his or her maximum potential and enjoy the best possible quality of life.

We have been setting the standard for children’s orthopedic care since 1940, when the duPont Institute was established. For 40 years, it was devoted entirely to children’s orthopedics and led by renowned physicians in the developing field. Since then, the Hospital and its depth of expertise have continually expanded. We were ranked among the best children’s hospitals in the nation in the 2010-11 edition of the Best Children’s Hospitals, by *U.S. News & World Report*. The duPont Hospital for Children ranked sixth in orthopedics.

The team is further enhanced by specialists in neuro-developmental disabilities, medical imaging, physical, occupational and speech therapy, prosthetics and orthotics and primary care. Our family-centered, collaborative approach involves parents and caregivers in all aspects of evaluation and treatment. We can often set up appointments with multiple specialists for the same day. All members of the health care team can access medical information immediately through our electronic medical record system.
3. CEREBRAL PALSY PROGRAM

Cerebral palsy can range from mild to severe, and children with CP often need to see many specialists. The duPont Hospital for Children CP Program combines the expertise of neuro-developmental pediatricians and the neuro-orthopedic division of the Department of Orthopedics. An orthopedic surgeon specializing in CP sees children whose primary problems are musculoskeletal. A pediatric medical specialist evaluates children with specialized medical needs. When children need to be seen by both specialists, visits often can be scheduled for the same day to allow coordination of care.

We offer a unified program of medical, surgical and support services to meet the specific needs of children of all ages. The program includes:

- A team of orthopedic surgeons, neurologists, physician assistants, psychologists, physical therapists, occupational therapists and speech therapists
- An on-site clinical nurse specialist to coordinate services
- A social worker and a nutritionist for consultation and treatment as needed

The CP Program coordinates care and provides services through multiple specialty clinics established to meet the specific needs of the child with neurologic disabilities. Patients who can benefit from this expertise can be referred to the CP Program or directly to the specific service.

For information or referral to the CEREBRAL PALSY PROGRAM, call 302-651-5921.

A. Orthopedic Care

The orthopedic team generally starts to follow children in the CP Program between 18 and 24 months of age, with outpatient visits taking place every six to 12 months until the child reaches skeletal maturity. Through this care, we monitor and assess the child’s motor development, and provide evaluations and prescriptions for adaptive equipment such as walkers, wheelchairs and standers. We coordinate prescriptions for these devices with assessments done in school or outpatient therapy centers; if these are not available we will coordinate this through the duPont Hospital for Children Wheelchair Clinic or Equipment Evaluation Clinic.

We also evaluate children for orthotics and braces, confirming both fit and appropriate functional benefit. Most children need new orthotics every 12 months. Caregivers may use the orthotist of their choice. An orthotics shop also is available in the Hospital.

Additionally, we treat developing deformities in the growing child with cerebral palsy. We focus on treatments with documented outcomes, with the least invasive treatment to the child and family, and with the goal of maximizing the child’s long-term function and comfort. Specific areas of focus and our outcomes include:

**Hip Subluxation and Dislocation Prevention and Treatment**

Hip problems are common in children with CP. They have normal hips at birth, with problems generally occurring between ages 2 and 8 years and occasionally during the adolescent growth period. With proper orthopedic monitoring and management, all children can avoid having hip dislocations at skeletal maturity. Our goal is to prevent hip dislocation or to treat the hip before full dislocation occurs. Our program for hip management includes prevention, reconstruction and palliation.
SERVICES FOR CHILDREN WITH NEUROMUSCULAR DISABILITIES

PREVENTION

Prevention requires monitoring high-risk children—those who do not ambulate as the primary mode of movement (GMFCS IV and V)—with a physical exam every six months and X-rays every year from 18 months to 8 years of age. Open adductor lengthening is performed when the migration index on the X-ray is 30 percent or more. After age 8, X-rays are obtained every two years. For children who are primary ambulators (GMFCS I, II, III), an X-ray is obtained at age 2. If the physical exam continues with good adduction (greater than 30 degrees), then no further X-ray is needed unless there are concerns.

Results

- We monitored 129 hips in 65 children to skeletal maturity.
  - 90 percent of children who ambulated and developed hip subluxation were treated by a single soft-tissue surgery, and had good hips at maturity.
  - 60 percent of children who were not ambulating had good or fair hips at maturity.
  - Children whose hips failed muscle lengthening required bone reconstruction.1

Reference


HIP RECONSTRUCTION

Hip reconstruction is available for children over age 8 with hip subluxation whose hip disease was not successfully treated by muscle lengthening at a younger age, and those with dislocated hips without severe arthritis. Reconstruction involves an acetabular osteotomy, femoral varus shortening osteotomy and appropriate muscle lengthening. The varus osteotomy is always done bilaterally to maintain hip symmetry, to improve seating and standing and to avoid later contra-lateral dislocation. We encourage immediate mobilization postoperatively, with no cast immobilization.

Results

- We reviewed 51 children who had 49 subluxed and 21 dislocated hips reduced by reconstruction, who were followed for an average of 34 months.
- Two hips in two children redislocated and required a second surgery.
- Parents reported that 82 percent of children were pain-free, and 80 percent felt the procedure achieved their goals.1
- Follow-up at 10 years showed that 80 percent of these hip reconstructions still had a good outcome. The failure rate for children age 2 to 8 years is higher.2
- This reconstruction also is possible for teenagers who have reached skeletal maturity.
- Our results for 33 hips in 27 adolescents show a good outcome in 26 of the 33 hips.3
- Bilateral pelvic osteotomies may be performed in a single setting, with only a minimal increase in complications and less than twice the complication rate as unilateral surgery. We compared 29 children with bilateral pelvic osteotomies in one stage to 61 children who had unilateral pelvic but bilateral femoral osteotomies, and we found no difference in hospital stay or postoperative complications.4
- All children followed in our program since 1988 who have continued with routine follow-up and recommended surgery have reduced hips at maturity.
References

Palliation
Palliation is recommended for children who develop hip dislocation and severe degenerative arthritis to such a degree that they cannot be reconstructed. For these children, we try to decrease the movement and activity that causes the pain. Some will remain pain-free and need no management other than to restrict their activity to avoid pain-producing movement. For many children, however, pain becomes progressively worse with daily care such as toileting, bathing, dressing and transfers for further treatment. For children with ambulatory ability, we offer a standard total hip replacement that allows them to return to ambulation. For nonambulatory individuals, we offer a resection interposition arthroplasty.

Results
- We reviewed 14 hips in 11 patients who had an interposition arthroplasty with a shoulder prosthesis.
- 13 hips in 10 patients had pain relief relatively quickly and remained pain-free at average 16 months follow-up.\(^1\)
- Of 12 total hip replacements in the last 15 years, none have required revision.
- Two had immediate postoperative dislocation requiring cast immobilization.
- Castle resection arthroplasty is another option we can provide, but since pain relief is generally not seen for six to 12 months, we prefer interposition arthroplasty.

Reference

Spinal Deformity
Nonambulatory children with CP often develop scoliosis and/or kyphosis and lordosis. These spinal deformities occur primarily as the child moves from middle childhood (8-10 years) into adolescence, progressing rapidly at the adolescent growth spurt. There are no preventive or nonsurgical treatments that offer objective evidence of altering the progression of the scoliosis. While the use of a body brace (TLSO) does not alter the rate of progression of the spinal deformity, it can provide sitting comfort for some children. Our goal is to monitor the development of spinal deformity and use wheelchair adjustments and other seating support to allow as much growth as possible. Spinal fusion is performed when the scoliosis reaches approximately 90 degrees but before the stiffness increases to the point where the child needs both anterior and posterior surgery.
Results

- Although spinal fusion is a large operation with many potential complications, parent and caregiver outcomes are very good.

- Parents and direct caregivers feel the outcome of the spinal surgery is excellent in 96 percent of cases, with great improvement in seating comfort, appearance and general health.\(^1\)

- Our experience with scoliosis correction using the unit rod and spine fusion in more than 500 children has demonstrated that this can improve nearly every child’s quality of life.

- A review of 241 patients with average eight-year follow-up showed our complication rate for deep wound infection was 4.2 percent, and 4 percent for repeat surgery (almost all very minor procedures).

- We achieved 68 percent correction of the scoliosis and 71 percent correction of pelvic obliquity with no clinical loss of correction.\(^2\)

- The rate of pancreatitis as a complication was 40 percent (mostly requiring delayed feeding).\(^3\)

- Life expectancy after spine fusion is longer than for children who have not had a spine fusion (80 percent survival at 11 years compared with literature-reported 50 percent survival without spine fusion).\(^4\)

- There is no impact on gait function when the unit rod is used for fusion to the pelvis.\(^5\)

- Complication rates are most directly related to the severity of the neurologic disability.\(^6\)

References

1. Tsirikos AI, Chang WN, Dabney KW, Miller F. Comparison of Parents’ and 3 Caregivers’ Satisfaction After Spinal Fusion in Children.
2. Tsirikos AI, Lipton G, Chang WN, Dabney KW, Miller F. Surgical Correction of Scoliosis in Pediatric Patients With Cerebral Palsy Using the Unit Rod Instrumentation. SPINE 33(10):1133-1140 2008.
Gait Treatment

Our management of children with CP focuses on helping them reach the maximum function allowed by their neurologic ability. We encourage physical therapy, orthotics like the ankle foot orthotics (AFO), walking aids and botulinum toxin injections until the child reaches a physical development plateau, usually around age 6 to 7 years. About the time the child is ready for first grade, we consider surgery to improve deformities that limit the child’s function as determined by a full three-dimensional gait analysis. We focus on correcting all deformities in a single-event, multilevel surgical approach (SEMLS). Typically, the child’s function is then maximized and stays stable until the adolescent growth period; many children need a second surgical event during adolescence. We also consider spasticity management using intrathecal baclofen pumps as an option to improve gait function, especially in the 5- to 12-year-old age group.

Results

- We have focused on defining improved criteria for specific procedures, defining EMG patterns that respond better to rectus transfer,¹ and reviewing our long-term results of rectus transfer.
- We found excellent results in children with stiff knees in swing phase when they have better GMFM function.²
- We found excellent results from hamstring lengthening to help extend the knee at foot contact and in mid-stance, although there was an increase in anterior pelvic tilt.³
- In 59 knee capsulotomies in 35 children, our posterior knee capsulotomies showed knee flexion contracture improved from 26 to 17 degrees.
  - Seven knees had sciatic nerve palsy.
  - Three failed and required more surgery.⁴
- We are just completing a long-term outcome study of the function of young adults 25-35 years after our treatment of them as children. These young adults are continuing to maintain the function they had at the end of growth with very little drop in function.

References

Upper Extremity Function

In young children, we focus on therapy to encourage functional use of the hand. Splinting is only used to prevent deformity if it does not impede functional use. In our program of constrained use therapy, we apply a long arm cast that makes the good arm nonfunctional for four weeks. Through therapy, we encourage the child to use the involved arm. This program is used primarily for 5- to 12-year-old children with hemiplegia, whose involved arm appears to have much better physical function but tends to not be used functionally by the child. In the 8- to 14-year-olds, we also consider upper extremity reconstruction to improve either function or cosmesis. For children with severe quadriplegic pattern involvement and severe upper extremity contractures, we will consider surgical releases or fusions to improve comfort and custodial care.

Results

- Caregiver-perceived outcomes of shoulder releases were very positive relative to the goal of improving care.\(^1\)
- Elbow flexion, very common in younger children with hemiplegia, resolves as the children go through adolescence. Surgical release mildly improved this and was felt by families and children to be very positive.\(^2\)

References


For information or referral to the ORTHOPEDICS PROGRAM, call 302-651-5913.

B. Spasticity and Movement Disorders Program

The spasticity program is a multidisciplinary evaluation and follow-up program for spasticity or movement disorder management. The evaluation clinic is staffed by a physiatrist, orthopedist and physical therapist. We consider the goals of the family and the child with a strong focus on identifying functional goals that are achievable. Evaluation may include further diagnostic workup if there are concerns about etiology, medical treatment, botulinum toxin injection or intrathecal baclofen use. We also consider additional consultations with specialists in neurology, neurosurgery and psychiatry. Management of complex or progressive dystonia often requires intense combinations of therapeutic modalities requiring multispecialty input. New patients also may see a nurse practitioner, which may lead to separate appointments with other specialists on the movement disorders team. We may make immediate recommendations or we may bring the discussion to the monthly meeting of the entire team, which includes the primary team and experts in neurology, neurosurgery and psychiatry. A baclofen pump management follow-up clinic also is available for refills and follow-up problems. It is staffed by a pediatric physiatrist and nurse practitioner.

Results

- The caregiver-perceived outcomes of intrathecal baclofen therapy are very good; 80 percent feel it has achieved the goals that were set out.
- Complications include infections; our acute infection rate was 4 percent with 316 pump implants.
- Other frequent complications are constipation, which can be managed, catheter malfunction in 3 percent of implants and pump malfunction in 1 percent of implants.
- Of the families and caregivers questioned, 81 percent were happy with the treatment and 87 percent would recommend it to another family.¹

- We have carefully case-matched 25 children with baclofen pumps against 25 who did not have a pump and found there was no difference in the development of scoliosis. Therefore, we can conclude there is no impact on the development of scoliosis in children who have baclofen pumps implanted. It will neither prevent the development of scoliosis nor cause scoliosis to develop or progress more rapidly.²

- We also have carefully studied 62 patients who had a spine fusion and a baclofen pump and found no difference in the complication rate, compared to 103 children who did not have a spine fusion. There is no difference in implantation of the pump before, during or after spine fusion in our group of 62 implants with spine fusion.³

References


For information or referral to the SPASTICITY AND MOVEMENT DISORDERS PROGRAM, call 302-651-5921.

C. Gait Analysis Laboratory

Treatments for gait dysfunction have historically been prescribed based on visual observation of a child’s walking. Established in 1993, the duPont Hospital for Children Gait Analysis Laboratory provides state-of-the-art, computer-assisted motion analysis of the complex gait cycle. Detailed data can be gathered about the movement of various joints, patterns of muscle contractures, associated forces affecting the limbs and the energy demands of altered gait. The data are collected and analyzed by a team that includes a physical therapist, biomechanist and pediatric orthopedic surgeon. This information can accurately pinpoint cause and effect. The team uses this information to make recommendations that will provide better outcomes and improved functionality. It is especially useful when a new treatment plan is being developed or when a major change in treatment is being considered. Testing is determined by the patient’s age, level of functional independence, ability to cooperate for structured testing and treatment goals. Typical tests include:

- Video of the patient walking, shot from various angles
- Foot pressure analysis to measure pressure patterns and the interaction between the foot and the ground
- Kinematic evaluation using eight to 12 video cameras that are synchronized to record the child while he or she walks; measures the movement of the joints in the arms, trunk and legs, enabling us to determine what abnormal movements are present
- Kinetic assessment of the lateral or medial side of the foot, which uses force plates to measure the foot’s interaction with the ground and how the child generates power for walking
- Diagnostic electromyography using surface electrodes to capture muscle activity data
- Detailed physical exam that includes measurement of joint range of motion, strength, muscle tone and motor control
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- Metabolic analysis using a mask to measure volume and percentage of oxygen and carbon dioxide in the child’s expired air; combined with heart and respiratory rates, this information suggests the energy expenditure and overall efficiency of the child’s gait

- A specially developed balance assessment program that uses the kinematic information to assess the child’s balance as he or she walks; this can be further enhanced with our full Vestibular and Balance Assessment Program

About 40 clinical gait analysis labs exist in the U.S., with only a handful performing as many full gait analyses as the duPont Hospital for Children (approximately 400 each year).

For information or referral to the GAIT ANALYSIS LABORATORY, call 302-651-4248.

D. Wheelchair Clinic

The Wheelchair Clinic at the duPont Hospital for Children offers a team approach to the evaluation and recommendation of seating and mobility equipment for children from birth to age 18. The Wheelchair Clinic team includes a rehabilitation engineer, physical therapist, occupational therapist and rehab technician, and the wheelchair vendor. Families are invited to include anyone involved in their child’s care, especially physical and occupational therapists.

Our multidisciplinary team is up to date on the latest advances in seating and positioning technology. They are experts in helping find the solution that best fits the child’s needs. They strive to promote independence and better functioning within the home, school and community environments.

The Wheelchair Clinic team provides:

- A thorough evaluation of the child’s mobility and positioning needs
- Recommendations for the most appropriate equipment to meet those needs
- Access to a variety of manufacturers’ wheelchairs
- Evaluations for the feasibility and safety of power wheelchair use with a wide variety of controller options
- Collaboration with the child's orthopedic surgeon or pediatric physiatrist
- Documentation that will be provided to the wheelchair vendor and submitted to the child’s insurance company for authorization of the recommended equipment
- Assistance with insurance appeals, if necessary
- Fitting of the child’s wheelchair and seating system upon delivery to assure that the recommended equipment is providing the expected outcome

The Wheelchair Clinic also provides pressure mapping, custom molded seating, trials of various power seating systems and trials of alternative drive controls. We can make recommendations about supplemental assistive and adaptive technologies as well.

For information or referral to the WHEELCHAIR CLINIC, call 302-651-5850.
E. Adaptive Equipment

Bathing, toileting, feeding and dressing can be challenging for children with physical limitations. Occupational therapy can assist with evaluating and recommending equipment that will help children perform these tasks. Equipment evaluations for walkers and standers are typically performed by physical therapists as part of a child’s physical therapy program. We have many types of equipment available in order to properly match the adaptive equipment to the needs of the child.

For information or referral to PHYSICAL or OCCUPATIONAL THERAPY, call 302-651-4354.

F. Clinical Assistive Technology Services

The Clinical Assistive Technology Services (CATS) at the duPont Hospital for Children offer complete augmentative communication and computer access evaluations as well as individual therapy, consultation to community-based therapists and educators, and assistance with setup and customization of communication aids. These services are designed to help children utilize appropriate communication aids to supplement their natural speech and writing and to communicate and learn more effectively in their homes, schools and communities. Our multidisciplinary program includes speech-language pathology, occupational therapy and rehabilitation engineering services. This assures that we are able to consider each child’s unique communication and learning needs and abilities from all perspectives.

Teamwork is essential to the process of incorporating augmentative communication and computer-assisted learning and writing technology into a child’s life. We actively seek to build a strong collaboration between children, parents and family members, community-based therapists and teachers, and our clinic staff. Together we can facilitate the child’s most effective use of his or her skills and technologies in every setting. Consequently, we are able to offer both clinic-based services as well as on-site services with school and family teams.

Commercially available augmentative communication devices, assistive writing software and devices, and reading and learning technologies can be found for children of all ages, disabilities and cognitive levels, including:

- Young children, emerging communicators and children with physical, cognitive or sensory processing impairments who are just beginning to learn how to interact with and actively participate in their physical and social worlds
- Socially and pragmatically impaired communicators and children with acquired or developmental cognitive challenges
- Communicators who require flexible and generative language expression
- Children of all ages whose motor, sensory, processing and/or language disorders are accompanied by difficulties in achieving functional or age-appropriate writing, reading and literacy performance
As a complement to augmentative communication evaluation and assistive technology evaluation services, CATS offers follow-up services that include:

- Assistance with procuring funding for speech-generating devices (this includes providing additional documentation if families wish to appeal initial denials of coverage for devices through health insurance plans)
- Outpatient therapy in our clinic with individualized treatment plans and family education
- Assistance and instruction with setup and customization of a child’s speech-generating device or specialized computer software
- Consultation and in-service training programs for school and family teams in which we provide proven techniques for integrating augmentative communication and assistive technology into your child’s curriculum and learning experiences, functional activities, and day-to-day family care

For information or referral to CLINICAL ASSISTIVE TECHNOLOGY SERVICES, call 302-651-5850.

G. Physical Medicine and Rehabilitation

The rehabilitation program provides medical, nursing and therapeutic services for children and adolescents with a wide range of rehabilitation needs. Physicians, rehabilitation nurses, therapists, psychologists and other pediatric specialists deliver a comprehensive program designed to return the child to home and the community under the best possible circumstances. The goal of rehabilitation is to maximize the young person’s ability to be active, to communicate and interact with people, and to live as independently as possible.

The program offers a functional, comprehensive and family-centered approach to pediatric rehabilitation within a children’s hospital. Children and adolescents through age 17 receive coordinated, interdisciplinary care as inpatients on the rehabilitation unit, and in the comprehensive outpatient rehabilitation program.

The Comprehensive Outpatient Rehabilitation Program (CORP) is designed for children and adolescents who do not require the intensity of an inpatient rehabilitation program, but who are continuing to make progress toward specific functional rehabilitation goals. These children and adolescents live at home with their families while continuing a rehabilitation program at the Hospital. After discharge from an inpatient program, we maintain the child’s follow-up with the rehabilitation team through outpatient visits. During these visits, we evaluate the child’s progress and revise the individualized rehabilitation plan to meet his changing needs.

We offer individual levels of therapeutic outpatient programming Monday through Friday. We review each child’s progress, treatment plan, therapy schedule and goals in regularly scheduled team conferences. The family (and child, when appropriate) participates in the team conferences.

The minimum requirement for participation in this program is the need for two therapeutic services (physical, occupational or speech therapy). Additional supportive therapies may include psychology, academic therapy, cognitive remediation or aquatic therapy.

For information or referral to the INPATIENT or OUTPATIENT REHABILITATION PROGRAM, call 302-651-5605.
Children with physical disabilities, whether due to cerebral palsy, muscular dystrophy, spinal cord injury or another condition that affects their ability to walk and run, commonly develop low bone density over time due to partial or complete nonambulation. This is true even after correction of nutritional factors. Many will sustain fractures with minimal impact injuries, with 70 percent involving the lower extremities (legs, feet and ankles). In a sample of 117 children with moderate to severe CP, more than 25 percent of children over 10 years of age had already sustained a fracture. Of 186 patients with CP referred to the duPont Hospital for Children for a DXA, 53 percent had sustained at least one fracture, with 16 of these patients having had five or more fractures (with a maximum of 17).

In adults, bone density testing of the lumbar spine and hip is done to diagnose osteoporosis, to predict fracture risk and to monitor therapy. In pediatrics, however, no diagnostic criteria exist to define osteoporosis on the basis of bone mineral density (BMD); until recently we were not able to predict the fracture risk based on BMD. Furthermore, measuring BMD in children with contractures is technically difficult, or sometimes impossible. In addition, many of these children have metal in their hips, making measurement of the density of the hip not meaningful. We have developed a technique to measure the bone density of the lateral distal femur. The advantages are that it is accessible in nearly all patients with physical disability and is a common site for fracture. The scan time is short, and thus less susceptible to motion. Pediatric reference data for DXA measures of normal BMD in the distal femur were first published in 2002, and new norms were published in 2009. Data using the lateral distal femur have shown a strong correlation between fracture history and BMD Z-scores in the distal femur; 35 to 42 percent of those with BMD Z-scores less than minus 5 had fractures compared with 13 to 15 percent of those with BMD Z-scores greater than minus 1. There was a 6 to 15 percent increased risk of fracture with each 1.0 decrease in BMD Z-score.

Our bone metabolism program is staffed by a developmenta pediatrician, nurse practitioner and dietitian. The goal is to optimize bone health and prevent progression of osteoporosis where possible and to treat it when it has progressed to the point of causing pain or further disability via fractures. We routinely assess dietary intake of minerals and vitamins, check vitamin D levels and supplement minerals and vitamins to keep these at an optimum level. We encourage standing to develop bone strength, even in children who can’t walk. We obtain DXA measures of bone density in children at risk for osteoporosis, and based on this data assess their risk for fractures. When children develop nontraumatic fractures, we consider the use of bisphosphonates, which in young children usually means an intravenous bisphosphonate given as an infusion over four hours. In children who are over age 18, we consider the use of oral bisphosphonates, which are the same medications used by adults with postmenopausal osteoporosis.

Results

- Our staff participated in a prospective, randomized, placebo-controlled, double-blind study of IV Pamidronate given to six pairs of children with quadriplegic CP on three consecutive days every three to four months for a total of 15 doses. Eleven of 12 had previously had at least one nontraumatic fracture, and all had low bone density on DXA. The study showed that bone density improved dramatically in those who received Pamidronate. The greatest increase in BMD was seen in region one of the distal femur, ranging from 38 to 185 percent (average 89 percent + 21 percent) over 18 months. Z-score improvement also was greatest in region one of the femur, increasing from -4.0 + 0.6 to -1.8 + 1.0. During the 18-month period, the BMD of the placebo group (who received calcium plus vitamins) increased an average of 6 to 15 percent.

- We later published data that showed the bone density began to fall approximately six months after the end of treatment, and by two years was back to baseline, though without a recurrence of fractures.
More recently, we assessed the rate of fracture before and after a one-year course of Pamidronate in 25 children with quadriplegic CP, GMFCS level IV or V. All participants had previously experienced at least one nontraumatic fracture. Each received 15 doses of Pamidronate over an average of 13.6 months. Post-treatment observation ranged from one year to 10 years six months (mean 4 y 1.2 mo). The participants had experienced a total of 86 fractures before treatment began, occurring over 280.6 person-years, giving a fracture rate of 30.6 percent per year. During the post-treatment observation period, totalling 107.5 person-years, eight of the 25 children experienced a total of 14 fractures. This fracture rate of 13.0 percent per year is a statistically significant decrease ($p=0.02$).

References

For information or referral to the **Bone Density Program**, call 302-651-6040.

**I. Pediatric Hospitalist Medically Complex Comanagement Program**

Beginning in 2005, medically complex orthopedic patients undergoing elective surgery at the duPont Hospital for Children have been followed by pediatric hospitalists. These medically complex patients are identified preoperatively by one of the following criteria:

- They are taking multiple medications.
- They are being followed by multiple medical subspecialists.
- They are dependent on medical equipment.

Postoperatively, these patients are followed by the pediatric hospitalists, who round daily with the orthopedic service and coordinate medical care with the medical subspecialists, surgeons, nursing staff and families.

In a retrospective study of medically complex patients with neuromuscular scoliosis who underwent spinal surgery, comanagement by a pediatric hospitalist resulted in significantly improved clinical outcomes. Hospitalist-comanaged patients were hospitalized on average for three fewer days, they were less likely to need to return to the operating room or intensive care unit, were fed earlier and received fewer days of TPN and fewer laboratory and radiologic studies.
References


J. Medical Program for Children with Physical Disabilities

This program, staffed by a developmental pediatrician, nurse practitioner (who coordinates much of the care) and a dietitian, is designed to meet the medical and developmental needs of children with cerebral palsy and other physical disabilities who also have complex medical problems. Many of these children have more involved forms of cerebral palsy (GMFCS IV and V), as well as seizure disorders, swallowing difficulties, poor weight gain, recurrent pneumonias, low bone density, fragility fractures, constipation, gastroesophageal reflux, learning disabilities, attention deficit/hyperactivity disorder, sleep disorder, drooling and/or behavior problems. In this program, we assess and treat these problems and refer children to the appropriate pediatric subspecialist when necessary.

The medical team provides:

- A thorough evaluation of the child's nutritional status
- Referral for specific diagnostic tests to assess swallowing difficulties if clinically indicated
- Adjustment of the consistency of the diet and/or the quantity of nutrients; in addition, supplements or special formulas are recommended when indicated
- Assessment of bone density via DXA scan, as well as assessment of dietary intake of needed minerals and vitamins
- Treatment of excessive salivation with medication or referral for treatment by botulinum injections or surgery
- Assessment and treatment of constipation
- Assessment and treatment of sleep disorders
- Assessment of possible learning disabilities and/or AD/HD, with recommendations for appropriate school placement and/or medication treatment
- Referral to appropriate subspecialists, including gastroenterology, neurology, pulmonology, dental, surgery and others as needed
- Coordination of home care for medically complex patients
- Medical evaluation prior to complex surgery such as spinal fusion
- Postoperative care provided by hospitalists after complex surgery

For information or referral to the NEURO-DEVELOPMENTAL MEDICAL PROGRAM FOR CHILDREN WITH CEREBRAL PALSY, call 302-651-6040.
4. NEUROMUSCULAR PROGRAM

In the Neuromuscular Program, a multidisciplinary team directed by physicians specializing in neurology, orthopedic surgery and pulmonology evaluates and treats children with various forms of muscular dystrophy and other neuromuscular conditions. We begin with an initial evaluation including an appropriate diagnostic workup. We offer a coordinated clinic with the opportunity for our patients to see all three specialists, as well as other physician colleagues such as cardiologists if special needs exist. Social workers, nutritionists and physical therapists also see patients in the clinic. We can access the services of on-site orthotists and specialists in wheelchair seating, as bracing and mobility are essential to proper functioning. A nurse practitioner coordinates care and access to available services.

We treat many different neuromuscular conditions, most commonly Duchenne and Becker muscular dystrophies, limb girdle muscular dystrophies, and myotonic dystrophy. Other neuromuscular conditions we commonly treat include spinal muscular atrophy and Charcot Marie Tooth disease (an inherited neuropathy).

Many of these conditions are associated with weakness, muscle cramping, contractures, joint deformities and scoliosis. They are best managed in a multidisciplinary clinic where conservative measures such as bracing and therapy services can be utilized before surgery. Also, proper seating and adaptive equipment can help maintain function. Surgical intervention is often necessary given the progressive nature of these conditions. Surgical interventions include but are not limited to spinal fusion surgery for scoliosis and surgery to manage hip dysplasia, as well as correction of foot deformities.

A. Medical and Surgical Treatment for Muscular Dystrophies

DUCHENNE MUSCULAR DYSTROPHY

While there are no cures to date for these neuromuscular conditions, they are treatable. Management of Duchenne muscular dystrophy includes physical therapy and bracing to stretch the heelcords and limit contractures. Pulmonary and cardiac evaluations also are very important in managing these children, particularly as their disease progresses. They need ongoing follow-up and management of cardiomyopathy and respiratory insufficiency. Baseline pulmonary function tests, electrocardiograms and echocardiograms should begin at age 6 years and be offered routinely after that based on assessment and need.

Steroids can be used to medically manage Duchenne muscular dystrophy. Steroids most commonly used are prednisone or Deflazacort (a little harder to obtain as it is not available in this country). Most children begin steroids around 5 years of age. The steroids decrease muscle inflammation, preserving muscle function for a longer time. The goal is to maintain ambulation for two or three years beyond that without medication. Bone density is generally low in these children because of limited physical activity and less weight-bearing over time, and is lowered even further by the steroids. DXA scans should be done prior to starting steroids and repeated every one to two years after that for surveillance. Steroid use and low bone density make nutrition counseling integral for our patients. Meal intake and vitamin D levels are monitored and supplemented when needed.

Scoliosis commonly occurs with Duchenne muscular dystrophy. We evaluate boys in our clinic soon after diagnosis. Scoliosis progresses after boys stop ambulating, making monitoring essential. Orthopedic evaluations, including X-rays, occur every six months. We perform surgery when the curve reaches greater than 30 degrees. Considerations for surgery include comfortable seating and improved pulmonary function. Surgical techniques often involve posterior spinal fusion with unit rod generally T3 to sacropelvis, or T3 to L5, with allograft. These admissions generally require a day or two in the PICU until the patient is extubated and stabilizes. Patients usually remain in the Hospital for one week. In spite of the underlying weakness, respiratory and cardiac concerns, these patients manage this surgery quite well.
End of ambulation can be predicted with the combined value of the hip extensor and knee extensor lag of greater than 90 degrees and ankle equinus of greater than 15 degrees. Generally, this means that ambulation will cease in approximately six months. Some boys with Duchenne muscular dystrophy who are on the cusp of losing ambulation elect surgery in hope of prolonging ambulation. This surgery involves tendoachilles lengthening, anterior transfer of tibialis posterior tendon, illiotibial band release, and hamstring and rectus femoris lengthening. Postoperatively, patients are placed in long leg casts, walking shortly after surgery, and long leg braces or KAFOs (knee ankle foot orthoses) with drop locks. Surgery and bracing are most effective prior to the end of ambulation.

**SPINAL MUSCULAR ATROPHY**

Hip subluxations can occur in our neuromuscular patients, especially those with spinal muscular atrophy (SMA); those with type I and II are at higher risk as they are not ambulatory. We perform surgery if the hips are painful or function is limited by contractures. We also need to manage scoliosis in our SMA patients. Orthopedic evaluations that include X-rays occur every six months. We try physical therapy and standers that allow weight-bearing before we consider surgery. TLSO braces may slow the progression of the curve, but care must be taken to assure that breathing and pulmonary function are not adversely affected. Surgery is considered when the curve progresses with bracing, when the curve is 40-50 degrees, when the child no longer tolerates sitting for extended periods, or when a decline in upper extremity function is seen. Techniques for this surgery include growing rods, or posterior spinal fusion with unit rod from T3 to the sacropelvis.

**CHARCOT MARIE TOOTH DISEASE**

We evaluate patients with Charcot Marie Tooth (CMT) disease yearly or more frequently as needs arise. They often have X-rays to monitor foot deformities, scoliosis and hip dysplasia. The orthopedic surgeon will often request pre- and postoperative gait analysis through the Gait Laboratory to monitor the patient’s progress. Together, they will determine the best course of management for these patients, whether it’s therapy, orthotics, bracing or surgery. Hip dysplasias occur in 15 percent of CMT patients, with scoliosis occurring in 10 percent. We consider bracing before surgery for scoliosis treatment.

Many patients with Charcot Marie Tooth disease require surgical correction of foot deformities to improve gait and function as well as manage pain that occurs with these deformities. Surgery is varied. If the hindfoot is flexible, treatment is focused only on forefoot alignment, and calcaneal osteotomy is not necessary. If the midfoot cavus is flexible, plantar medial releases are done. If the midfoot cavus is fixed, plantar medial release and midfoot osteotomy are done. If the hindfoot is not flexible, hindfoot osteotomy is done. In children younger than 8 years of age, plantar fasciotomy and tibialis posterior tendon transfers can be done, provided the ankle dorsiflexors are greater than antigravity strength. In older children, surgical options include muscle transfer and anterior transfer of the tibialis posterior as well as osteotomies, either metatarsal, medial cuneiform, midfoot or calcaneal, and triple arthrodesis.

**B. Muscular Dystrophy and Genetics**

Duchenne muscular dystrophy is an X-linked recessive disorder, passed from mother to son. Sons of female carriers have a 50 percent chance of having the disease, with new mutations accounting for 30 percent of cases. The mutation results in an inability to produce dystrophin protein. This condition affects one in 3,500 males. Our neurologist leads the discussion about this condition’s genetics, course and prognosis. Genetic counselors are available for further discussions if needed. Families considering having other children are often encouraged to seek prenatal counseling and genetics evaluations from adult providers as well to fully understand the genetics of this condition and make informed decisions.
Spinal muscular atrophy is an autosomal recessive inheritance with incidence one in 6,000 to one in 10,000 live births. Carrier frequency is one in 40 to one in 60. Our neurologist describes the genetics of this condition when giving the diagnosis, along with treatment, management and prognosis. Genetic counselors are available for further discussions as needed. Just as in Duchenne muscular dystrophy, families wishing to have other children are encouraged to see the geneticist or obtain prenatal counseling for a full understanding of the condition.

Myotonic dystrophy in all three forms is caused by an abnormal triplet repeat. There is an apparent correlation between severity of illness, age at onset of symptoms and the length of the repeat expansion in myotonic dystrophy Type 1 (DM1) and congenital myotonic dystrophy. A worsening of symptoms in subsequent generations is common in DM1.

Charcot Marie Tooth disease is widely varied in its genetics. It is categorized by either demyelinating or axonal forms, then further classified by autosomal dominant and autosomal recessive forms. CMT1A is the most common, affecting 80 percent of CMT patients; it is demyelinating and autosomal dominant. CMT2 is next in incidence; it is autosomal dominant and axonal. CMTX is the next most common; it is X-linked dominant (absence of male to male transmission), and demyelinating.

For more information or referral to the NEUROMUSCULAR PROGRAM, call 302-651-5890.

5. SPINAL DYSFUNCTION PROGRAM

Our Spinal Dysfunction Program uses a team approach for the evaluation and treatment of children and adolescents with many different conditions that involve the spinal cord. The team includes specialists in neurosurgery, orthopedics, neurology, rehabilitation, physical therapy and social work. Conditions we treat include:

- Spinal cord injury
- Trauma or sports injuries to the spine
- Cervical spine disorders
- Sacral agenesis
- Transverse myelitis
- Spinal cord tumors
- Tethered cord syndrome
- Spina bifida
Evaluation and treatment of spinal cord disorders includes:

- Orthopedic evaluation and management
- Rehabilitation evaluation and management
- Nursing evaluation and management
- Urology evaluation and management
- Neurosurgery evaluation and management
- Physical therapy evaluation
- Bracing prescription, fitting and adjustment
- Wheelchair prescription, fitting and adjustment
- Radiological evaluations (MRI, CT scan, X-rays)
- Social work assessment
- Coordination of health care through collaboration with the child’s primary care physician, school and community
- Client advocacy
- Consultation to school and community programs

How we treat patients with hydrocephalus is one example of how children at the duPont Hospital for Children receive advanced care that results in better outcomes and fewer complications. Treatment for hydrocephalus has typically involved placement of a ventriculoperitoneal shunt. Now endoscopic third ventriculostomy (ETV) is becoming an increasingly common treatment. Surgeons create a small opening in the bottom of the third ventricle to allow fluid to exit the brain. This minimally invasive approach enables the large majority of children with spina bifida to avoid shunts and the difficult issues around shunt management and follow-up. Today’s options are even further expanded with the addition of a unique technique that combines ETV and choroid plexus cauterization (CPC).

For more information or referral to the SPINAL DYSFUNCTION PROGRAM, call 302-651-5007.

6. ARTHROGRYPOSIS PROGRAM

There are many variations of arthrogryposis, a condition of the joints and muscles that impairs a child’s ability to use his or her shoulders, wrists, fingers and lower extremities. They vary from mild to severe. In mild cases, only a few joints may be affected and the child may have nearly full range of motion, while in extreme cases nearly every joint is impacted, including those of the jaw and back. We treat arthrogryposis multiplex congenital/distal arthrogryposis and syndromes associated with arthrogryposis-like contractures.

At the duPont Hospital for Children, our team includes specialists in orthotics and prosthetics, medical imaging, physical and occupational therapy and rehabilitation to provide personalized treatment for children with this condition. From the early evaluation stages to the creation of a long-term rehabilitation plan, we work closely with families to determine the best course of care for the child. Services include clinical evaluation, diagnostic testing, genetic consultation, muscle biopsy, surgical intervention, splinting, therapeutic and rehabilitation services, home exercise program development, aquatic therapy and the Wilmington Robotic EXoskeleton (WREX).

For information or referral to the ARTHROGRYPOSIS PROGRAM, call 302-651-5007.
7. CHOOSE NEMOURS FOR YOUR PATIENTS

Nemours is one of the nation’s leading pediatric health systems, dedicated to advancing the highest standards in children’s health. We’ve made a promise to do whatever it takes to prevent and treat even the most disabling childhood conditions—a promise of specialty medical care, advanced hospitalization, applied research and advocacy integrated with health information, prevention and a continuous process of teaching and learning.

We invite you to learn more about the services for children with neuromuscular disabilities available at the duPont Hospital for Children. You may contact any of our programs directly, or call our CP Program Coordinator, who can assist you with information and referrals to any of the services in our system.

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<tbody>
<tr>
<td>Arthrogryposis Program</td>
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<tr>
<td>Wheelchair Clinic</td>
<td>302-651-5850</td>
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“It is a great honor that so many families dealing with neurologic disabilities trust us to be involved in their child’s care. With this trust, we strive hard to maximize the child’s function through treatment that minimizes interruptions to the family and child’s normal interactions and maturation.”

— Freeman Miller, MD, Orthopedic Surgeon, Director of the Cerebral Palsy Program and Medical Director of the Gait Analysis Laboratory, the Nemours/Alfred I. duPont Hospital for Children
Our founder came from one of America’s wealthiest families, but his family name could not protect his childhood from illness and tragedy. He was orphaned at age 13, and few knew that he was blind in one eye. He also suffered serious hearing loss, eventually becoming deaf.

Overcoming adversity, Alfred I. duPont became an innovator and inventor, always looking for ways to make the world better.

Today, that vision is fulfilled through the duPont Hospital for Children and the Nemours health system. Named among the best children’s hospitals by U.S. News & World Report for 2010-11 and one of the 25 best children’s hospitals by Parents magazine in 2009, the duPont Hospital for Children provides all the specialties of pediatric medicine, surgery and dentistry in a warm, family-centered environment. We are academically affiliated with Thomas Jefferson University and the University of Delaware, and offer outpatient pediatric specialty care in Wilmington, Delaware, Egg Harbor Township and Voorhees, New Jersey, and Lancaster, Newtown Square and Philadelphia, Pennsylvania. The duPont Hospital for Children is the only Level II Pediatric Trauma Center in Delaware.

With locations in Delaware, Pennsylvania, New Jersey and Florida, Nemours has grown to become one of the nation’s largest integrated pediatric health systems. With the addition of our new, state-of-the-art children’s hospital in Orlando, Florida in 2012, we provide hospital- and clinic-based specialty care, primary care, prevention and health information services, and research and medical education programs designed to improve the lives of children and families throughout the Delaware Valley and Florida—now and for generations to come. That is the promise of Nemours.
The Nemours/duPont Hospital for Children is easily accessible from Delaware, Pennsylvania, New Jersey and Maryland. Parking is free, and free valet parking is available at the inpatient entrance (near the playground) and visitor’s lobby (behind the Hospital).

**From the North (Route 202)**

Take Route 202 South. You will see the AstraZeneca campus on the right. Make a right at the light onto Powder Mill Road (Route 141). Driving under a walkway overpass, stay on this road and it will become Children’s Drive and take you through the Hospital gates.

**From the South (I-95)**

From I-95 North, take Exit 8 – Concord Pike/Route 202 N exit. Stay to the left. Once on Route 202, get in the far left lane. Turn left at the third light onto Powder Mill Road (Route 141). Driving under a walkway overpass, stay on this road and it will become Children’s Drive and take you through the hospital gates.

**From the West**

Take Route 141 North (Barley Mill Road), crossing the Tyler McConnell Bridge. Turn left at the duPont Experimental Station staying on Route 141 North (Powder Mill Road). Travel a mile and a half, making a right at the second light onto Children’s Drive. The Hospital is straight ahead through the gates.

**Directions for Rockland Center I, II, and Ronald McDonald House**

Follow the directions above. On Children’s Drive, facing the Hospital’s stone gates, turn left onto Rockland Road. Rockland I is the first office building on your left. Rockland II is the second building on your left. A parking garage is located between I and II.

The Ronald McDonald House is the large yellow building past Rockland II.

**RMHDE.org/302-656-4847**
To learn more about the services for children with neuromuscular disabilities available at the duPont Hospital for Children, please call the CP Program Coordinator at 302-651-5921.

For appointments with other Nemours specialists or more information about other Nemours programs, please call 800-416-4441.

Learn more at Nemours.org.