Amazing Recovery

A young patient has made a dramatic recovery from severe injuries, thanks to fast, comprehensive trauma care.

Madison Wilkinson is an active, happy honor student who rides horses, runs track and field and roller skates. But, not long ago, a traumatic injury threatened her life and her future. The fact that she is well today underscores the value of a trauma system that integrates care from the first 911 call through rehabilitation.

Madison was 12 years old and enjoying a summer boat ride with family when tragedy struck. A sharp turn of the boat caused her to fall into the water, where her head was struck by the boat’s propeller. She was underwater for approximately one minute before family members were able to pull her from the lake and call 911. A nurse who lived nearby administered first aid to control bleeding. Emergency medical responders rushed the unconscious child to the Backus Hospital Emergency Department, where she was found to have a skull fracture and brain extrusion. Physicians inserted an endotracheal tube and administered intravenous pain medication and antibiotics. Madison was transported by LifeStar to Connecticut Children’s, arriving there within two hours of her injury.

Trauma Team in Action

Connecticut Children’s trauma team was mobilized prior to the patient’s arrival, and an operating room was made ready. An immediate assessment in the Emergency Department found that Madison had fixed, dilated pupils and decorticate posture, suggesting a severe head injury. Laboratory tests and a CT scan were performed to determine the extent of blood loss, coagulopathy and presence of electrolyte abnormalities.

She was taken to the operating room directly from the CT scanner for treatment of her head injuries, including elevation of depressed skull fractures, partial frontal lobectomy, placement of a dural graft and insertion of an intracranial pressure monitor. As the neurosurgical procedures were being performed, the plastic surgery

Interstate Clinical Collaboration Launched

Connecticut Children’s Medical Center and Baystate Children’s Hospital have launched a new collaboration addressing potential ways to improve access to high-quality and high-value health care for children in Connecticut and western Massachusetts.

The two organizations, which both provide high-level inpatient pediatric and neonatal care as well as comprehensive outpatient services for children and adolescents, will work together to determine whether they can increase the availability, sophistication and coordination of pediatric services throughout the Connecticut River Valley, and collaborate with community pediatric providers to improve the overall health and wellness of children in the region. Both have been recognized by U.S. News and World Report as being among the top U.S. children’s hospitals.

Examples of areas where both organizations agree that a collaborative approach could improve access and quality of care include pediatric neurosurgery, ophthalmology, pulmonology and urology, all areas where current provider shortages can make getting care difficult for patients and families.

Level 1 Trauma Care

Beginning Jan. 1, 2015, all pediatric trauma patients will be transported directly to Connecticut Children’s for care.
A 17-year-old female presented to Nephrology for a history of edema and proteinuria. Prior to her presentation, she had developed a presumed anaphylactoid reaction with significant edema, lip swelling and difficulty breathing two days after starting bupropion for depression and anxiety. She was evaluated at an outside Emergency Department and received intramuscular prednisone and Benadryl. Two days following this, she again developed edema and was evaluated at the same Emergency Department. She received a prednisone taper over one week and hydrochlorothiazide 12.5 milligrams daily, with recommended follow-up with Nephrology. The patient’s medical history was relatively benign. She was a full-term infant without delivery complications. She was followed by a psychiatrist for depression and anxiety. Her only medication at the time was hydrochlorothiazide 12.5 milligrams daily. Apart from a distant relative with a renal transplant for unknown reason, her family history was benign for renal disease. Review of systems was positive for fluctuating edema. She denied any weight loss, fatigue, joint pain or skin lesions.

DIAGNOSIS AND TREATMENT

Initial serum laboratory workup revealed an essentially normal complete blood count (CBC), with a hemoglobin of 13.4 g/dL and a hematocrit of 40.5%. Platelets were normal at 450 Thou/μL. White blood count was mildly elevated at 12.4 Thou/μL. Electrolyte panel revealed a creatinine of 0.5 mg/dL, a calcium of 8.7 mg/dL and albumin of 2.3 g/dL. Other electrolytes were within normal limits. Complement C3 was 244 mg/dL, and complement C3 was 62 mg/dL. Anti-nuclear antibody screen was negative. Acute hepatitis panel was negative. Urine protein to creatinine ratio revealed nephrotic range proteinuria at greater than 7. Retroperitoneal ultrasound revealed two normal, symmetrically sized kidneys with normal echotexture. Given the patient’s age at presentation, the decision was made to perform a renal biopsy. Pathology revealed findings that were consistent with minimal change disease. Twenty glomeruli were sampled, none of which showed lesions of focal segmental glomerulosclerosis (FSGS). Immunofluorescence (IF) revealed trace mesangial staining for IgG, IgM, kappa and lambda. Shortly thereafter, the patient was started on prednisone 60 milligrams daily in conjunction with a low-salt diet. One week after starting steroids, the patient reported resolution of her edema and return of her energy. About four weeks into prednisone therapy, the patient had a rapid weight gain of 20 pounds, increased edema, decreased appetite and emesis. Given her significant symptomatology and lack of response to prednisone, it was decided to start her on steroid-sparing therapy with tacrolimus with a rapid taper of prednisone. A chest X-ray was performed due to difficulty breathing, and it was normal.

Shortly after starting tacrolimus, the patient was admitted to the hospital with complaints of weight gain, fatigue, low back pain, nausea and vomiting. During her evaluation, it was noted that a complete blood count showed a decreasing trend in all cell lineage. Uric acid was elevated at 9.8 mg/dL. Due to the concern for an oncologic process with hyperuricemia, the patient was treated with rasburicase acutely, and allopurinol. Upon further questioning, the patient reported additional complaints, including night sweats and pruritis. A positron emission tomography (PET) scan was performed and revealed areas of intensity in the neck, with mediastinal, chest, abdominal and sacral lymph nodes. Oncology was consulted, and a bone marrow biopsy and bone marrow aspirate were non-diagnostic. Sacral biopsy was also non-diagnostic. Given the patient’s recent course of high dose steroids, it was decided to continue her steroid taper and repeat biopsies two months following completion of taper.

PET scan was repeated, and it showed bulky mediastinal nodes and supravacular lymph nodes, in addition to rib and sacral uptake. A cervical lymph node biopsy was performed, which confirmed classic Hodgkin lymphoma (HL). Based on the recent scans, it was staged at IIB, with concern for IIIb. The patient had a port placement and was started on chemotherapy.

DISCUSSION

Minimal change disease (MCD) or idiopathic form is the most common cause of nephrotic syndrome (NS) in children. It consists of a triad of edema, proteinuria and hypoalbuminemia. The pathogenesis is postulated to be related to a T-lymphocyte disorder due to the initial observation of relapse occurring in association with measles; a state of depressed cell immunity. Several lymphokines may play a pathogenic role with the emergence of serum analysis from a large cohort of children with MCD. The idiopathic form is frequently restricted to younger ages between 2-6 years of age and typically is a remitting and relapsing course, likely to be steroid responsive with resolution by the third decade of life. When older children and adolescents present with NS, a wide range of etiologies must be considered given the atypical nature in presentation. One such category includes extrarenal malignancies. NS is an extremely well-described paraneoplastic manifestation, particularly in HL. The most common glomerulopathy by renal biopsy is MCD in patients with HL. Multiple large studies recently published regarding the association of NS and HL reveal an incidence of 0.4% and 0.6% in adult and pediatric patients respectively. The development of glomerulopathy in HL is likely related to abnormal cytokine release and thus implication of T-cell disregulation.

Clinical presentation of HL-associated MCD is no different from that of classic MCD. However, the diagnosis of HL should be considered when patients present with atypical features including older age and unresponsiveness or partial response to standard steroid treatment. The timing of NS varies with respect to HL, but the syndrome is noted after lymphoma in 42% of cases. Therefore, treatment with steroids in the atypical population is cautioned against as it can delay the diagnosis of HL as was the case in our patient.

Treatment of the underlying lymphoma almost always results in remission of NS. A cumulating amount of studies recently reveal 100% of patients treated with chemotherapy went into remission after successful treatment of HL, regardless of whether steroids were used pre-diagnosis of malignancy. In summary, MCD is a rather common and not devastating disease in the pediatric population. However, when children present at older ages, MCD is less likely, and HL is a rare but treatable etiology in this age group. Standard chemotherapy regimens are the first and best line of treatment in patients with MCD-associated HL.

REFERENCES:
Dr. Silva is also assistant professor of pediatrics at the University of Connecticut Health Center. She may be reached at csilva02@connecticutchildrens.org or 860.545.9395.
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The collaboration may extend beyond clinical care delivery as the organizations will also jointly explore the potential for expansion of a new pediatric accountable care organization (ACO) that is now being developed in western Massachusetts by Baycare Health Partners, Baystate’s affiliated physician-hospital organization. Among the goals of an expanded children’s ACO will be to improve the coordination of care among pediatric primary care providers, specialists and hospitals throughout the Connecticut River Valley.

SPOTTING MEASLES

By Nicholas Bennett, MA(Cantab), MBBChir, PhD, FAAP

I have seen a grand total of one case of confirmed measles in my life. It was in a young boy admitted to the hospital with pneumonia and a provisional diagnosis of scarlet fever. From an infectious disease perspective his story made no sense: The X-ray looked nothing like group A strep, and he was too young to see strep commonly anyway. I questioned how real this “pneumonia” was, until I saw him—tachypneic, hypoxic and lethargic in his mother’s arms. He looked up at me with bloodshot eyes ... wait ... bloodshot eyes?! Cough, coryza, conjunctivitis, rash, fever ... and he was a recent immigrant. Mom didn’t know what vaccines he’d had; only that “the last one was before he could walk.” That was all I needed to know. He had measles.

My generation of physicians has been spoiled. While some of us may remember having these diseases, or seeing cases in our friends as kids, during our training there has been a happy dearth of vaccine-preventable diseases to learn from. This makes diagnosing them that much more difficult. As vaccination rates drop in some areas, there is the possibility of imported cases spreading. The key is to be mindful. Measles mimics other diseases found between the Red Book covers, such as adenovirus, Kawasaki disease, leptospirosis, mycoplasma and scarlet fever, making pattern recognition difficult and confirmation bias a problem.

Measles is one of the most contagious diseases known, and while the majority will recover without incident, some (like my young friend) can be seriously ill with pneumonia or encephalitis. Subacute sclerosing panencephalitis is the rare, but untreatable and fatal neurologic complication of wild-type measles.

There is hope. The United Kingdom recently reported its highest-ever MMR uptake of 94.5 percent*. Maybe we can reverse the trend and follow its lead.

*Most recent figure for the U.S. is 91.9.

Dr. Bennett is medical director of the Division of Pediatric Infectious Diseases and Immunology and co-director of antimicrobial stewardship at Connecticut Children’s. He is also assistant professor of pediatrics at the University of Connecticut School of Medicine. He may be reached at nbennett01@connecticutchildrens.org or 860.545.9490.
**RESEARCH AIMS TO PREVENT ACL REINJURY**

**CAN COMPUTERIZED MOTION ANALYSIS BETTER PREDICT WHEN IT’S SAFE TO RETURN TO SPORT?**

Two Connecticut Children’s programs, Elite Sports Medicine and the Center for Motion Analysis, are collaborating on a study to see whether advanced technology can improve return-to-sport decisions for adolescents who have had anterior cruciate ligament reconstruction.

“The ultimate goal” says Dr. Matthew Milewski, the primary orthopaedic surgeon involved in the study, “is to more safely return these athletes to sports and, hopefully, minimize reinjury rates.”

Literature shows that risk of injury to both the injured and contralateral side of the knee after ACL reconstruction is as high as 25 percent, and patients who have already sustained one ACL injury are 15 times more likely than control subjects to have another. “Although there may be multiple causes of reinjury, premature return to activity is one possibility,” says Milewski. He and his colleagues hope their work will result in more objective criteria for returning athletes to play.

The researchers are using computerized, three-dimensional motion analysis to study kinematics and kinetics in subjects as they run, hop and perform other motions associated with participation in sports. Ideally, subjects would show bilateral symmetry during these activities, says Sylvia Ounpuu, MSc, director of research at the Center for Motion Analysis. However, she notes, “We have found significant asymmetries in kinematics and kinetics in these patients. These asymmetries, which are not visible to the naked eye, may contribute to increased reinjury rates.” Preliminary review of data suggests that at least half of the patients have a greater than 10 percent difference in parameters such as peak knee flexion during landing, peak knee extensor torque and peak knee power absorption. The motion data show that some patients shift their trunks differently to lessen lower extremity loads.

The team expects to have preliminary results and a better understanding of asymmetries in January 2015. In addition to helping guide return-to-play decisions, the study may aid in developing targeted rehabilitation protocols.

Dr. Milewski is also assistant professor of orthopaedic surgery at the University of Connecticut School of Medicine. He may be reached at 860.284.0220 or mmilewski@connecticutchildrens.org. Ms. Ounpuu, who is also an associate professor at the University of Connecticut School of Medicine, may be reached at 860.282.0202 or sounpuu@connecticutchildrens.org.

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**EXPERT SURGICAL CARE FOR HEART DEFECTS**

**CONNECTICUT CHILDREN’S TREATS ALL CONGENITAL HEART DEFECTS AND ACHIEVES OUTCOMES SUPERIOR TO THE NATIONAL AVERAGE**

Anyone, from newborn to adult, who needs surgical correction of a congenital heart defect can have the procedure performed at Connecticut Children’s, confident that the care provided is among the best in the nation. “No one has to leave Connecticut for this type of surgery,” says Harris Leopold, MD, division head of cardiology at Connecticut Children’s.

Pediatric cardiovascular surgeons Dr. Paul Kirshbom and Dr. Mohsen Karimi, who serve as chief and associate chief, respectively, of cardiovascular surgery, are experienced, highly skilled surgeons who work with a multidisciplinary team dedicated to caring exclusively for children and adults with congenital heart problems.

“Our treatment runs from very simple defects to very complex single ventricle physiology,” says Karimi. For example, this fall, Kirshbom performed two Norwood procedures for patients with hypoplastic left heart syndrome. Both patients did well and went home shortly after surgery.

The cardiac surgery program is a combined program with Yale-New Haven Children’s Hospital. Its formal name is Northeast Pediatric Specialists. The two surgeons divide their time evenly between the hospitals and are available 24/7.

Connecticut Children’s contributes to a national database operated by the Society of Thoracic Surgeons. Kirshbom says the data show that “Connecticut Children’s survival rate for congenital heart surgery is better than the national average and better than expected, based upon the complexity of our patients. Our total mortality and risk-adjusted mortality are significantly better than the national average.” The data are for 200 cases annually, evenly divided between the two hospitals.

Another distinguishing feature of Connecticut Children’s cardiac surgery services is the use of practices that minimize blood loss and, therefore, reduce or eliminate the need for transfusions. “Conservation of blood has become a priority for us during cardiac surgery because of donor blood shortages, risks associated with use of allogeneic blood products and the costs of these products. Further, transfusions expose patients to a variety of potential cellular and humoral antigens, pose risks of disease transmission and immunomodulation, and may alone represent proinflammatory stimuli in the perioperative period. Our strategies to reduce bleeding and transfusion requirements include recognizing preoperative risk factors, developing transfusion practices, and conserving red blood cells using sophisticated operative equipment we have at Connecticut Children’s.”

Dr. Kirshbom is chair and Dr. Karimi is associate chair of pediatric cardiothoracic surgery at the University of Connecticut School of Medicine and Yale School of Medicine. Dr. Kirshbom may be reached at pkirshbom@connecticutchildrens.org. Dr. Karimi may be contacted at mkarimi@connecticutchildrens.org.
WELCOME ABOARD!
We’re pleased to announce these new additions to our medical staff.

Samriti Dogra, MD
Nephrology
- Fellowship in pediatric nephrology, Children’s Hospital at Montefiore
- Residency in pediatrics, Children’s Hospital at Montefiore
- MD, University of Maryland School of Medicine
- BA, biology & society, Cornell University

Miriam Harel, MD
Urology
- Fellowship in pediatric urology, Connecticut Children’s Medical Center
- Residency in urology, SUNY Downstate Medical Center
- MD, SUNY Downstate Medical Center
- BA, psychology, CUNY Brooklyn College

Kalyani Raghavan, MBBS, MD, DCH
Pain and Palliative Medicine
- Attending physician, Pediatric Sedation Service, Children’s Hospital of Michigan
- Residency, University of Michigan, Ann Arbor, Michigan
- MD, Maharaja Sayajirao University and Government Medical College (India)
- DCH, Maharaja Sayajirao University and Government Medical College
- MBBS, Maharaja Sayajirao University and Government Medical College

Heather Tory, MD
Rheumatology and Quality
- Fellowship in pediatric rheumatology, Boston Children’s Hospital
- Residency in pediatrics, Yale-New Haven Children’s Hospital
- MD, Tufts University School of Medicine
- BA, neuroscience, Middlebury College

More Additions To Our Community
This fall, Connecticut Children’s welcomed seven new fellows, 20 new residents and three chief residents. For information about these talented individuals, visit WWW.CONNECTICUTCHILDRENS.ORG.

WEBINARS AVAILABLE
Two webinars presented earlier by Connecticut Children’s subspecialists are archived and available for viewing. See the For Health Care Professionals section of connecticutchildrens.org.

Syncope in Children
Derek Obayashi, MD, pediatric cardiologist, helps physicians better understand the evaluation and treatment of children experiencing syncope as it relates to pre-existing, and possibly undiagnosed, heart conditions. Recorded October 2014.

Posterior Urethral Valves: Prenatal and Postnatal Evaluation and Management
Eric Nelson, MD, a pediatric urologist, helps physicians better understand the prenatal and postnatal care for boys with posterior urethral valves. Recorded July 2014.

Treatment and Diagnosis of Idiopathic Scoliosis
Jeffrey Thomson, MD, director, orthopaedic surgery, helps primary care physicians better diagnose and treat patients with scoliosis. Recorded May 2014.

Obtain MOC Credits
Connecticut Children’s offers quality improvement projects approved by the American Board of Pediatrics for meeting maintenance of certification (MOC) requirements. Connecticut Children’s program is unique; it focuses on bridging quality improvement initiatives from our organization to the practicing pediatric community. Physicians who participate in Connecticut Children’s Portfolio Projects and meet ABP completion requirements will receive Performance in Practice (Part 4) MOC credits. For more information on Connecticut Children’s MOC Portfolio Program, please visit cme.connecticutchildrens.org or contact MOC Program Manager Eminet Feyissa at efeyissa@connecticutchildrens.org or 860.837.5712.

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New Offerings and Locations
Departments of Diabetes & Endocrinology and Neurology – Have relocated to 505 Farmington Ave., Farmington. Will continue to see patients in Hartford.

Department of Nephrology – Now seeing patients on Friday mornings at 399 Farmington Ave., Farmington.

Division of Orthopaedics – Now offers Saturday office hours from 8:30 to 11:30 a.m. at Connecticut Children’s Medical Center in Hartford for urgent, routine and walk-in visits.

Elite Sports Medicine – Now offers a same-day sports medicine clinic at 399 Farmington Ave., Farmington. Call 860.284.0220.

Watch for dates and topics of webinars to be presented in 2015.
TRANSITIONS

Several members of Connecticut Children’s medical staff have embarked on new paths.

Elizabeth Estrada, MD, Endocrinology, has relocated to Chapel Hill, North Carolina.

Robert Greenstein, MD, Developmental Pediatrics/Genetics, has retired. He is succeeded by Thyde Dumont-Mathieu, MD.

Cristian Marek Ionita, MD, Neurology, has relocated to Seattle Children’s Hospital.

Francisco Sylvester, MD, Digestive Diseases, has relocated to Chapel Hill, North Carolina.

CONTINUING MEDICAL EDUCATION PROGRAMS

All programs are held at the Pond House Café, 1555 Asylum Ave., West Hartford, Conn., and begin at 5:30 p.m. with registration and buffet dinner.

PEDIATRIC EVENING LECTURE SERIES

Feb. 12, 2015
Nephrology Update

April 2, 2015
Allergy Update 2015

ANDRULONIS CHILD MENTAL HEALTH EVENING LECTURE SERIES

Jan. 15, 2015
Anxiety Disorders

March 10, 2015
Cannabis Harmfulness to Youth Wellness: The Emperor's New Policies

May 5, 2015
Social Media and Health Risks

To register or obtain more information, contact:
Diane Mouradjian - 860.837.6264, dmouradjian@connecticutchildrens.org
or Deidre Palmer - 860.837.6281, dpalmer01@connecticutchildrens.org.