COMPREHENSIVE LYME DISEASE PROGRAM LAUNCHED
The only one of its kind in the country.

This spring, Connecticut Children's Division of Rheumatology introduced a Comprehensive Lyme Disease Program that is a unique resource for families and for referring providers with patients who either don't respond to treatment or who have complicated Lyme disease.

A Multidisciplinary Approach
Dr. Lawrence Zemel, chief of rheumatology, directs the program, which is staffed by a multidisciplinary team that includes pediatric subspecialists from neurology, infectious diseases, orthopaedics, cardiology, pain medicine and behavioral health. Patients see appropriate subspecialists, and the entire team meets monthly to discuss complicated patients. “This is the first time anyone has put together a comprehensive program like this,” says Zemel. “I have not heard of another program like it in the country.”

Zemel notes that primary care providers today typically recognize early Lyme disease, especially if the patient has a rash. The primary care physician will provide evidence-based treatment, and, most often, the patient will be successfully treated. “We generally don’t need to see those straightforward patients,” Zemel says. “However, if patients don’t respond to treatment or if there’s a question about diagnosis, we want to see those patients.”

Close Collaboration with Primary Care Providers
The Medical Center is taking the unusual step of promoting the program to the general public and inviting families to contact the program directly. Zemel says that's because there's an abundance of misinformation about Lyme disease available on the Internet and from advocacy groups and other sources. Because of this misinformation, families may bypass their own primary care provider.

A QUARTER-CENTURY OF INJURY PREVENTION
Children everywhere are safer because of Connecticut Children’s Injury Prevention Center.

When Connecticut Children's Injury Prevention Center (IPC) began in 1990, "We were unique in that the center was based in a department of pediatrics, focused solely on children, and had a mission that was statewide, broad and comprehensive," says IPC co-founder and longtime Director Garry Lapidus. "That is, we had four core activities: research, education and training, outreach and public policy advocacy."

Since then, the IPC has pursued its mission with passion and rigor and become a model for other programs across the United States. It's also made great strides in identifying underlying causes of injury, raising awareness of safety issues and shaping public policies that have achieved measurable gains. Continued on page 3
Diagnosing Lung Lesions: Cast a Wide Net

Dr. Sumith Madhavarapu, a resident at Connecticut Children’s, authored this issue’s case in collaboration with Dr. Anand Sekaran, chief of hospital medicine. The patient was referred to Connecticut Children’s by Dr. Ruth Loomis of Pediatric Associates in Bristol, Connecticut.

PRESENTATION
A 17-year-old female with Type 1 diabetes mellitus presented to Connecticut Children’s Medical Center with three weeks of cough, left-sided chest pain and gradually worsening dyspnea. A chest X-ray two weeks prior had revealed bilateral, multifocal opacities. Azithromycin was started with no improvement. When a repeat chest X-ray showed interval worsening, she was sent to Connecticut Children’s Emergency Department for further evaluation and treatment. Review of systems was positive for intermittent otalgia, congestion and myalgias. She had no fever, weight loss, joint pain or rash. There were no known TB exposures or recent long-distance travel.

In the ED, she was febrile to 38.8°C. She was well appearing and had no increased work of breathing. On exam, she had decreased breath sounds in the left lower lung. HEENT, heart, abdomen, musculoskeletal, lymph and skin exams were normal. Chest X-ray confirmed multifocal bilateral pulmonary nodules. CBC, LFTs and urine dip were normal. CRP was 3.8 (normal <0.5) and ESR was elevated at 68 (normal <21). Contrast-enhanced CT of the chest showed “multiple large pulmonary nodular opacities, some with central cavitation.”

DIAGNOSIS AND TREATMENT
She was admitted to the pediatric hospitalist service, and consults were obtained from infectious diseases, rheumatology and pulmonary. It was decided to avoid antibiotics until the diagnosis was better elucidated. CRP decreased to 2.6 but the ESR increased to 100. PPD and rapid HIV were negative. Immunoglobulin levels were normal. Multiple infectious studies were obtained, including blood culture, mycoplasma titer, sputum culture, and sputum for fungus, anaerobes and AFB. Rheumatoid factor was weakly positive at 37 (normal <14). C3 and C4 were normal. ANA was negative, and angiotensin converting enzyme was negative. Her p-ANCA was normal but c-ANCA was positive at >8 (normal <1). This result suggested a diagnosis of granulomatosis with polyangiitis (GPA), formerly known as Wegener’s granulomatosis. An additional antibody test confirmed that GPA was the most likely diagnosis: PR-3 ANCA was 22,118 (normal <20)! The family was presented with the option to obtain a confirmatory biopsy of one of the lung lesions, but elected to initiate treatment for the probable diagnosis of GPA.

In coordination with the rheumatology service, the patient received IV methylprednisolone, rituximab and oral prednisone. Three days after treatment was begun, her cough and chest pain resolved. A repeat chest X-ray two weeks later showed marked improvement (see images).

DISCUSSION
Granulomatosis with polyangiitis is a small- and medium-sized vessel vasculitis. Mean age at diagnosis is 80 years. In the pediatric population, incidence is estimated to be one case per million with a mean age of 15.4 years. It typically involves a triad of systems: sinus, pulmonary, and renal disease, though over time can involve other systems as well. Initial symptoms can include otalgia, persistent otitis, hearing loss, congestion, sinusitis or persistent cough. Renal involvement usually manifests as asymptomatic hematuria or proteinuria. Approximately 90 percent of patients will have nasal involvement. Glomerulonephritis is present in 20 percent at the time of diagnosis, but 80 percent will eventually develop renal disease within two years, even with active treatment.

There is a limited form of GPA isolated to the respiratory tract at time of diagnosis. This occurs in 25 percent of cases and typically happens in younger women. Up to 80 percent with this form will eventually develop glomerulonephritis.

Laboratory and radiographic studies can aid the diagnosis. The combination of a positive c-ANCA and PR3-ANCA provides a high positive predictive value for GPA. Diagnosis is best made via biopsy of viable, affected lung tissue, as biopsies from other affected sites are less likely to show granulomas.

Therapy is aimed at inducing remission, with continued maintenance treatment to prevent relapse. Cyclophosphamide was formerly the mainstay of treatment, but more recently rituximab has been shown to be as effective, with an improved side effect profile and a lower relapse rate. Induction therapy also involves a long course of steroids for six to nine months. Maintenance therapy follows for 12 to 24 months and may include rituximab, steroids, and sometimes methotrexate or azathioprine. Treatment can be initiated without biopsy if clinical suspicion is high and lab testing is highly suggestive.

When presented with pulmonary cavitary lesions in a pediatric patient, it is important to consider a broad differential, including infectious, rheumatologic, oncologic and congenital causes.

REFERENCES


Have you had an interesting case involving Connecticut Children’s?
Contact Medical News Managing Editor Dennis Crean, RN, at 860.837.6248 or dcrean@connecticutchildrens.org.
Comprehensive Lyme Disease Program Launched, continued from page 1

A Quarter-Century of Injury Prevention, continued from page 1

results in preventing injuries and fatalities among children and teens.

Shaping Public Policy
“Through the years, we’ve been fairly successful in taking the research and going to the Capitol to advocate for safety laws,” Lapidus says. The IPC was instrumental in Connecticut’s enacting laws governing child car safety seats, bicycle helmets, graduated driver’s licenses for teen drivers, and gun control. These measures, along with education and outreach efforts, have made a significant difference. Lapidus points out that, since 1990, injury death rates for children and teens have decreased by 50 percent in Connecticut. As a result of passing graduated driver’s license legislation, Lapidus says, “There has been a 20 to 40 percent reduction in teen car crashes in the state.”

Statistics such as these are reflected across the United States, because other organizations also began focusing on injury prevention. “We were right at the beginning of a broad movement around the country,” Lapidus says. In 1992, the IPC became the state chapter for a national campaign called SafeKids. It also became one of 40 sites across the country—mostly at children’s hospitals—for the Injury-Free Coalition for Kids. The IPC has played an important role in these efforts, and its research has contributed to the science of injury prevention.

A Growing Center
The IPC’s success is also reflected in how much it has grown over the years. When the IPC was launched in 1990, Lapidus and Dr. Leonard Banco were the only staff members, and they were part time. Today, the center has 12 staff members (eight full-time equivalents) and nine affiliated scientists. Founded with a $4,000 grant from the American Academy of Pediatrics, the center last year attracted more than $800,000 in external funding.

Connecticut Children’s pediatric surgeon Brendan Campbell says that Lapidus has been the catalyst for the IPC’s success. “One of Garry’s great qualities is that he’s a team-builder, and he always finds a way to do the right thing,” Campbell says. “He’s been a leader nationally in the field of injury prevention.” Campbell adds, “The Injury Prevention Center is central to the mission of Connecticut Children’s as a Pediatric Level I Trauma Center.”

Looking to the Future
One of the issues the IPC is focusing on now and will continue to work on going forward is domestic violence. The center’s staff members are engaged in research and educational activities aimed at reducing domestic violence, which Lapidus says is “devastating” to children and sets up a cycle of violence that continues through generations. “Babies are not born violent,” Lapidus says. “It’s a learned behavior. We want to change the culture of how people view domestic violence because it is so harmful to women, families, children and communities.”

Primary Care Doctor’s Role
Asked what referring providers can do to help prevent injuries, Lapidus says, “I urge our affiliated providers to find one or two injury prevention messages they can integrate into their clinical practice.” PCPs might, for example, focus on car seat safety, bike helmets, working smoke detectors or domestic violence. “If they pick one or two things and talk about them routinely, that would make an enormous difference.”

Garry Lapidus is an associate professor of pediatrics and public health at the University of Connecticut School of Medicine. He may be contacted at 860.837.5318 or glapidu@connecticutchildrens.org.

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Receive important news and information from Connecticut Children’s quickly and easily. Sign up for emails today by visiting connecticutchildrens.org/for health professionals/education/register for emails.

NEW OUTPATIENT RADIOLOGY HOURS
Connecticut Children’s Radiology Department now offers expanded hours for outpatient general X-rays. The new hours are Monday through Friday, 8 a.m. to 6:30 p.m., and Saturdays from 9 a.m. to noon.
PRESIDENT AND CEO ANNOUNCES RETIREMENT

This past spring, Martin J. Gavin informed Connecticut Children’s board of directors of his plan to retire as president and chief executive officer. Gavin, 65, will continue to lead the hospital until a successor is chosen and in place, but expects to step down prior to the close of the 2015 calendar year.

Connecticut Children’s board of directors has formed a search committee and begun a national search to identify a replacement for Gavin.

“I have been incredibly fortunate to have had the opportunity to serve as president and CEO of Connecticut Children’s. I’ve always said that I have the best job in the world; it’s truly been a gift,” said Gavin. “But after nine years, it’s time for me to move on and let a new generation of leadership continue to advance our mission of caring for children and families.”

POWASSAN VIRUS IN PERSPECTIVE

This spring, media reports of an emerging, “potentially deadly,” tick-borne infection known as the Powassan virus alarmed Connecticut residents. Connecticut Children’s pediatric infectious disease specialist Dr. Nicholas Bennett says that, despite the hype, it’s important to keep the disease in perspective.

“This is not a new virus or a new disease,” Bennett says. “It’s rare, and it’s very similar to other diseases pediatricians might see in the summer, such as West Nile virus or Eastern equine encephalitis.”

According to the Centers for Disease Control and Prevention (CDC), only 50 cases of the virus have been reported in the United States in the past 10 years. “Chances are that physicians will go their entire professional careers and never see a case,” Bennett says.

Powassan is one of scores of arboviruses. It can cause encephalitis or meningitis. Many people who contract the virus experience mild symptoms or none at all. Symptoms may include fever, headache, vomiting, weakness, change in mental status and seizures. In severe cases, hospitalization for supportive care may be necessary. The mortality rate is about 10 percent, versus the 30 percent mortality rate associated with Eastern equine encephalitis.

Studies have shown that the virus can be transmitted from an infected tick to a host in as little as 15 minutes. The incubation period ranges from a week to a month. Bennett encourages community practitioners to advise parents to use the same precautions with their children to prevent Powassan as they would to protect against Lyme disease. Those precautions, as outlined by the CDC, include avoiding brushy or wooded areas, wearing long pants tucked into socks, using a DEET-containing repellent, wearing specially treated clothing and checking the body for ticks after outdoor activities.

Dr. Nicholas Bennett is medical director of infectious diseases and immunology at Connecticut Children’s and an assistant professor of pediatrics at the University of Connecticut School of Medicine. He may be reached at 860.545.9490 or nbennett01@connecticutchildrens.org.

TRAUMA PROGRAM EARN S REVERIFICATION

Connecticut Children’s has been reverified as a Level I Pediatric Trauma Center by the American College of Surgeons (ACS) and the Connecticut Department of Public Health. This distinction makes Connecticut Children’s one of only 25 freestanding children’s hospitals nationwide that are Level I Pediatric Trauma Centers. The re-verification is for a three-year period ending in September 2017.

To earn re-verification, a hospital must demonstrate through an application and a rigorous site visit that it meets all of the quality metrics specified in the ACS manual, Resources for Optimal Care of the Injured Patient.

Connecticut Children’s first attained Level I status in 2008 as part of the Trauma Institute formed with Hartford Hospital. This collaborative model of trauma care continues, but the majority of pediatric patients are now brought directly to Connecticut Children’s. “Over the past several years, we’ve demonstrated that we can care for patients with multisystem injuries,” says Dr. Brendan Campbell, medical director of Connecticut Children’s Pediatric Trauma Program. “We’ve transitioned to having all injured pediatric patients come to Connecticut Children’s because we have the resources in place to care for the most critically injured children.”

Those resources include pediatric surgeons, neurosurgeons and orthopaedic surgeons; 24-hour coverage by specialists in pediatric emergency medicine, anesthesiology and intensive care; an on-site pediatric rehabilitation program; a massive transfusion protocol and more.

Campbell says that the re-verification “emphasizes our goal of providing the highest-quality care to injured children in Connecticut while continuing to be a national leader in trauma education and injury prevention research.”

Dr. Brendan Campbell is assistant professor of surgery and pediatrics at the University of Connecticut School of Medicine and surgeon champion for the National Surgery Quality Improvement Program at Connecticut Children’s. He may be contacted at bcampbell@connecticutchildrens.org or 860.545.9659.
LEARN ABOUT GENETICS IN PRIMARY CARE

Jackson Laboratory speakers to offer lectures.

Scientists from Jackson Laboratory’s Farmington-based genomic medicine institute, in collaboration with Connecticut Children’s, will present two talks on how pediatricians and family medicine specialists can incorporate genetics in their practices to enhance patient health.

A Grand Rounds lecture planned for Jan. 12, 2016, will focus on the role of family history-taking in assessing a child’s risk of common conditions such as cardiovascular disease, diabetes, atopy, intellectual disabilities and mental illness. Attendees will work through clinical cases to identify genetic risks and discuss the implications of family history for the child’s health.

Also in 2016 (date TBA), Jackson Labs will provide an evening lecture and workshop at the Pond House in West Hartford on genomic risk assessment.

Participants will practice collecting targeted family history information, identifying red flags and patterns and developing a management plan based on risk assessment.

For more information about these programs, contact Diane Mouradjian at 860.837.6264; dmouradjian@connecticutchildrens.org.

WEBINARS AVAILABLE ON DEMAND

Webinars previously presented by Connecticut Children’s subspecialists are archived and available for viewing. They include:

New Comprehensive Lyme Disease Program
Dr. Lawrence Zemel, head of the Division of Rheumatology, discusses this new program. Recorded May 2015.

Evaluation of the Neck Mass in Infants and Children
Dr. Scott Schoern, pediatric otolaryngologist, presents. Recorded March 2015.

How Fecal Transplantation is Changing Lives
Pediatric gastroenterologist Dr. Zev Davidovics explains how fecal transplantation transfers the good bacteria of a healthy donor to the patient experiencing symptoms. Recorded January 2015.

What You Need to Know About Ebola
Pediatric infectious disease specialist Dr. Nicholas Bennett addresses the actual risks related to Ebola from both a natural health and epidemiological point of view. Recorded November 2014.

Syncope in Children
Dr. Derek Obayashi, 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.

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

For more information, please see the For Health Care Professionals section of connecticutchildrens.org.

NEW NICU CHIEF NAMED

Dr. James E. Moore has been named chief of the Neonatal Intensive Care Unit at Connecticut Children’s and head of the Division of Neonatology in the Department of Pediatrics at the University of Connecticut School of Medicine. Dr. Moore comes to us from Children’s Medical Center of Dallas. He will assume his position at Connecticut Children’s in August 2015.

PHYSICIANS NAMED TO “TOP DOCS” LIST

Connecticut Magazine’s 2015 “Top Docs” list includes four of Connecticut Children’s many outstanding subspecialists. Those named to the list are Drs. Jeffrey Hyams, gastroenterology; Paul Kanev, neurosurgery; Joseph Newell, pediatrics, and Catherine Wiley, pediatrics. Congratulations!

FREE “LUNCH & LEARN” TALKS FOR YOUR PRACTICE

Would you like to learn more about clinical topics you may encounter in your practice? Connecticut Children’s can help. Our specialists will visit your office to present information on topics you choose and engage in informal discussions with you and your colleagues. The talks are free of charge. Lunch is provided for physicians, APRNs and PAs. To schedule a talk, contact Trish Masse at tmasse@connecticutchildrens.org or 860.837.6251.

WELCOME ABOARD

We’re delighted to announce this addition to our medical staff.

Dr. Christine Matarese
Neurology

• Child neurologist, Nemours Children’s Hospital
• Child neurologist, Child Neurology Center of Orlando
• Fellowship in clinical neurophysiology, Warren Alpert Medical School of Brown University
• Residency in child and adolescent neurology, Mayo Clinic
• Residency in pediatrics, Orlando Regional Healthcare
• Internship, Philadelphia College of Osteopathic Medicine and St. Joseph Medical Center

• DO, University of New England, College of Osteopathic Medicine
• BA, biology, and BA, psychology, Lehigh University

*Editor’s note: We regret that an incorrect photo appeared with Dr. Matarese’s announcement when it appeared in the spring 2015 issue of Medical News. The correct photo appears above.

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CONTINUING MEDICAL EDUCATION PROGRAMS

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

PEDiatric evening lecture series

Sept. 17, 2015
Hernias, Hydroceles & Undescended Testicles

Nov. 12, 2015
Common Neurosurgical Problems Seen in Primary Care

Feb. 11, 2016
More Than Meets the Eye: Common Problems in Pediatric Ophthalmology

April 7, 2016
The 5 E’s to an Exceptional Eczema Experience

ANDRULONIS CHILD MENTAL HEALTH EVENING LECTURE SERIES

Oct. 15, 2015
Psychiatry for the Primary Care Physician: A Toolkit for Pediatric Health Care Providers

Jan. 12, 2016
Addressing Postpartum Mood and Anxiety Disorders in Primary Care: From Screening to Triage

March 1, 2016
Assessment and Treatment of Autism and Transition to Adulthood

May 10, 2016
Gender Nonconformity and Dysphoria in Childhood & Adolescence: Clinical Issues for the Primary Care Pediatrician

To register or obtain more information, contact:

Diane Mouradjian – 860.837.6264, dmouradjian@connecticutchildrens.org
Deidre Palmer – 860.837.6281, dpalmer01@connecticutchildrens.org