FIRST WITH IMPROVED CARDIAC DEVICE

A Connecticut Children's cardiologist recently became the first physician in the state to implant a next-generation heart monitor in a pediatric patient. Dr. Shailendra Upadhyay implanted the Medtronic Reveal LINQ Insertable Cardiac Monitor late last year in an 11-year-old boy who had complained of a racing heart.

“This new device is much smaller than previous versions—about half the size of a triple-A battery—but has the same capabilities as the larger ones” Upadhyay says. “It’s also MRI-compatible and so small that it’s barely visible under the patient’s skin.”

While earlier models required a sizable incision and the creation of a pocket, this unit comes in a pre-loaded syringe, so the physician simply inserts it under the skin with only a small incision, and no pocket is necessary. “This makes recovery and healing much quicker and results in less pain” Upadhyay says.

The device enables physicians to monitor the patient’s heart rhythm remotely using a cell phone and an adapter. The device can remain implanted and send signals for up to three years. “It’s a very good tool for monitoring patients who can’t express arrhythmia symptoms,” Upadhyay says. “And, in the case of patients who are passing out, it helps us determine whether abnormal heart rhythms are the culprit.”

Dr. Upadhyay is also the director of the Connecticut Adult Congenital Heart Service and an assistant professor at the UConn School of Medicine. He may be reached at 860.545.9400 or supadhyay@connecticutchildrens.org.

LENGTHENING LIMBS FROM THE INSIDE

A new device approved by the Food and Drug Administration early last year is revolutionizing limb lengthening in pediatric patients, and Connecticut Children’s orthopaedic surgeon Dr. Kristan Pierz is one of only a handful of Connecticut physicians certified in its use.

The PRECICE Intramedullary Limb Lengthening System consists of an adjustable nail that is placed within the tibia or femur and gradually lengthened using remote control technology.

“It’s remarkably different from external fixators,” Pierz says. “External fixators work, but they’re unsightly and painful, have a high infection risk and are a lot of work to keep clean.” By contrast, “The PRECICE device is all within the bone. No one can see it, the infection risk is less, maintenance is nothing once the incision heals, and it’s much more comfortable. Patients ‘cohabitate’ with the device.”

The PRECICE nail is a tube within a tube. Inside are gears that slide the tubes apart. There is a magnet inside the nail, and the patient has the countermagnet.

“When you put the countermagnet on top of the leg, the magnet inside the nail tries to push away, causing the gears to turn,” Pierz explains.

For proper healing, the bone can be lengthened by only 1 millimeter a day. As Pierz points out, “For every centimeter, that’s 10 days of lengthening. Then it has to heal for about twice as long as it took to lengthen.” The device extends to a maximum of 8 centimeters. It is removed at the end of the process.

Pierz acquired formal certification in the PRECICE system by participating in a training program presented by deformity expert Dr. Dror Paley. She has been using the device since early 2014.

When dealing with a pediatric population, Pierz notes, it’s important not only to know what the current limb-length difference is, but determine what the difference will be when the child grows to adulthood. For differences that will be small, other methods may be preferable. The PRECICE device is just one option.

Dr. Pierz is also an assistant professor of orthopaedic surgery at the UConn School of Medicine. She may be reached at 860.545.9100 or kpierz@connecticutchildrens.org.
PRESENTATION
A previously healthy 14-year-old female was referred to Connecticut Children’s emergency department (ED) by her primary care provider with one week of chest pain that worsened with inspiration and radiated to her right shoulder, and difficulty breathing while playing soccer. She had no fevers, shortness of breath, cough, hemoptysis, leg pain, nausea, vomiting, diarrhea, headaches or dysuria. One day prior, she had complained of sudden onset of binocular diplopia that worsened when looking to the right. One week prior, at the onset of her pain, she had been worked up with a chest CT angiography (CTA) at an outside hospital ED. The diplopia brought her to her primary care physician, who recommended a brain MRI, which was interpreted as normal. She had no recent illness or medical conditions, and was taking no medications. She was a 10th grade student who played soccer and lived with her parents. She owned a cat. She had not traveled recently.

The patient appeared well, but exam was notable for no right eye lateral abduction. Pupils were equally reactive to light, left extracocular motion was intact, and there was no proptosis, lid lag, scleral injection or icterus. She had lymphadenopathy. Heart and lungs were clear. She had some mild right anterior shoulder pain, but full range of motion of all extremities and no edema. She was alert and oriented, with a constant right cranial nerve six (CN VI) palsy. The remainder of CNs were intact. Strength was 5/5 bilaterally. She had no sensation deficit. She showed normal gait, heel-to-toe, and finger-to-nose.

DIAGNOSIS AND TREATMENT
Besides the CN VI palsy, the patient had no other focal neurological deficit. Her mild intermittent right shoulder pain with deep inspiration was relieved by oral nonsteroidal anti-inflammatory or ice pack.

She was evaluated by pediatric subspecialties including pulmonary, rheumatology, neurology, infectious disease, and hem/onc. She had normal complete blood count, urinalysis, complement (C3, C4), angiotensin converting enzyme (ACE), anti-neutrophil cytoplasmic antibody (ANCA), prothrombin time (PT) and partial thromboplastin time (PTT) and immunoglobulins. Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) were mildly elevated: 23mm/hr (lab range 0-20mm/hr) and 2.7mg/L (0-0.49mg/dL), respectively.

Negative interferon-gamma release assay, PPD, HIV, fungal cultures, Aspergillus, and Bartonella henselae. EBV IgG antibody positive while rest of the panel was negative, consistent with previous infection/exposure. Bone survey for occult malignancy/metastasis was negative. PFTs were normal.

Chest CTA confirmed multiple peripheral lung nodules including a right middle lobe subpleural nodule, later accessed for biopsy (Figure 1). Brain MRI and MRA showed no masses or vascular abnormalities.

Tissue biopsy from the subpleural nodule confirmed necrotizing granulomas and vasculitis with epithelioid granulomas in zones of necrosis along a lymphangitic distribution. Focal pleuritic granulomatous changes were in areas adjacent to subpleural granulomatous nodules. Special stains for acid fast bacilli and fungi showed no microorganisms such as tuberculosis and fungi. In the absence of infectious etiology, findings were consistent with necrotizing sarcoid granulomatosis (NSG).

DISCUSSION
NSG is a rare systemic disease first described in 1973. Prior studies have shown that extrarespiratory findings are more common in NSG. Inflammatory markers are mildly elevated, whereas ANCA and ACE are negative and normal respectively1. Most common pulmonary findings include solitary or multiple lung nodules and pulmonary infiltrates, while pulmonary function tests are normal and hilar lymphadenopathy is uncommon1. CN VII is most commonly involved in sarcoidosis and neurosarcoidosis2,3.

CN VI palsy, as in this case, has not been described in cases of sarcoidosis or NSG. Due to its overall benign nature, NSG has shown a good prognosis in children, many of whom have spontaneous remission or excellent response to treatment (often to corticosteroids)4.

After pathologic diagnosis, the patient was pulsed with one gram intravenous methylprednisolone daily for three days with complete resolution of right CN VI palsy and restoration of right eye lateral abduction. She was discharged home on oral prednisone. At two-week follow-up, she had no further respiratory or visual complaints. She remained on oral prednisone 30mg once daily (1/2mg/kg/day) for one month with further follow-up and continued resolution of symptoms. She went back to school and resumed playing soccer.

This case highlights the importance of awareness of NSG as a diagnostic entity in the pediatric population and also describes a rare presentation of this disorder in the form of CN VI palsy.

REFERENCES:

Figure 1. Chest CTA showed a right middle lobe subpleural nodule.

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.
**ANTIMICROBIAL STEWARDSHIP: WHAT EVERY PROVIDER CAN DO**

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

In my role as co-director of Antimicrobial Stewardship at Connecticut Children’s, I work to ensure that antibiotics are used appropriately. **That means the right drug in the right dose for the right duration.** The literature has shown us that up to half of all antibiotic usage is “inappropriate”—typically meaning that the coverage is too broad, or the duration is too long, or sometimes that an antibiotic isn’t needed at all. Faced with rising rates of antibiotic-resistant bacteria, and the fact that most new antibiotics consist of “me-too” drugs rather than novel classes, practicing prescribers are put in a quandary. Do we treat more aggressively to ensure we don’t get burned, or treat less aggressively to promote less future resistance?

The truth is, sometimes you need a bit of both. Sometimes the most appropriate antibiotic is one with broader coverage, and sometimes you treat for a longer period to prevent recurrence. How is this done without the help of a full antimicrobial stewardship program in your office?

A small but growing number of clinical guidelines are available to help any physician manage common ailments like respiratory infections, pneumonia, urinary infections, etc. Pick one, and make a change in your practice. You may be surprised how little outcomes actually change. In general, it is very appropriate and safe to utilize the “lowest” drug possible for empiric outpatient therapy. What about handouts for patients and families? Many parents or patients are seeking a solution—any solution—and if antibiotics aren’t it, then what can you tell the family to do?

All my efforts in the hospital are just the tip of the iceberg. As a profession, we all have a part to play in ensuring we can treat infections in the future. The CDC’s Get Smart campaign has tools and materials available online (http://www.cdc.gov/getsmart/specific-groups/hcp/outpatient.html). Furthermore, Connecticut Children’s has an approved MOC Part 4 module available on antibiotic usage, as if further incentive was needed to do the right thing! For information on the MOC module, call Eminet Abebe Gurganus at 860.837.5712.

Please direct questions and comments to our dedicated stewardship email: CCMCASP@connecticutchildrens.org. ■

**Dr. Bennett is medical director, Infectious Diseases & Immunology, and co-director of the Antimicrobial Stewardship Program. He is also assistant professor of pediatrics and adjunct assistant professor of pharmacy at the UConn School of Medicine. He can be reached at 860.545.9490 or nbennett01@connecticutchildrens.org.**

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**PREVENTING AND TREATING OVERUSE INJURIES**

With spring at hand, community pediatricians are likely to see an increase in young athletes with problems such as tendinitis, ligament sprains and avulsion fractures. These are most likely the result of overuse, says Dr. Carl Nissen, an orthopaedic surgeon and sports medicine specialist with Connecticut Children’s Elite Sports Medicine.

Just as the medical community’s understanding of concussion has grown in recent years, Nissen says, “We now know that overuse injuries are much more prevalent than we previously understood. Those injuries were probably always there, and they saw us, but we didn’t see them.”

Overuse injuries often occur in the spring, when student athletes who may have been snowboarding or playing basketball during the winter try to jump right into a throwing sport such as baseball or a pivot, run and cut sport such as lacrosse, without adequate preparation. “They’re not ready, and they do more than the body is capable of dealing with,” Nissen says. “Especially in the case of a growing skeleton, this will often lead to an overuse injury.”

Another culprit is playing the same sport year-round. “This is a recipe for overuse,” Nissen says. He stresses the importance of taking a break from one sport and playing another one for a while instead. “Do some other physical activity, but don’t do the same motion over and over again.”

When a patient presents with an overuse injury, Nissen recommends that primary care providers prescribe “active rest” in order to heal the injury. “As a pediatrician, you need to react to the injury, but also realize that this is a person dying to play his or her sport. The art of medicine is to find something that allows the individual to be physically engaged with friends in the sport, but not do further harm to themselves.” The provider may suggest, for example, that the athlete swing a bat, but not throw a ball; or do two weeks of cross-training before gradually progressing back to the sport. “You need an active plan, not a passive one,” he says.

Primary care providers should offer anticipatory guidance when the opportunity arises, explaining to patients the importance of getting into shape before plunging full-bore into the sport. “I tell patients they can’t go from zero to 60 in a day,” Nissen says. “If you push your car like that, it will respond and not be injured. But if you do this to your body, you’re asking for it to break. You need to spend time getting in shape first.”

With active rest, tendinitis should show signs of getting better within a week. “If someone doesn’t get better, there may be a more serious problem,” Nissen says. “That’s when the patient should be referred to Elite Sports Medicine.” ■

**Dr. Carl Nissen is also a professor of orthopaedic surgery at the UConn School of Medicine. He can be reached at 860-284-0220 or cnissen@connecticutchildrens.org.**
Sometimes it is difficult to know who the most appropriate pediatric specialist is to care for breast conditions in children. Pediatric surgeons are uniquely trained to care for the full spectrum of both gynecologic and breast conditions that can occur in children and adolescents. This article will review the most common pediatric breast and breast bud conditions, and provide practical guidelines for evaluation and referral.

EVALUATION OF BREAST MASSES
The overwhelming majority of pediatric and adolescent breast masses are benign, and their differential diagnosis is age dependent. When diagnostic uncertainty exists, referral to a pediatric surgeon can streamline evaluation and avoid unnecessary imaging. Ultrasound is the first line of imaging for breast masses if imaging is warranted, and readily distinguishes between solid and cystic lesions. Mammography and magnetic resonance imaging are rarely necessary. Fine needle aspiration and core needle biopsy may be used in special circumstances, but not without input from a pediatric surgeon.

NIPPLE DISCHARGE
Nipple discharge is an uncommon problem in children, but when present can be alarming for patients and their parents. Discharge is usually pus, cyst contents or blood. Serous brown/green fluid suggests cyst contents and is usually self-limited. Bloody discharge is usually the result of ductal ectasia in children. Galactorrhea is milk-colored nipple discharge unrelated to pregnancy, and prolactinoma is the most common cause in children.

MALE GYNECOMASTIA
Gynecomastia is the benign proliferation of glandular tissue of the male breast, and up to two-thirds of boys will develop gynecomastia during puberty. The highest prevalence occurs at 13 to 14 years, and the condition generally resolves within one year (~85 percent). When gynecomastia fails to resolve over time, subcutaneous mastectomy using a periareolar incision can achieve excellent cosmetic results in boys who are not obese.

BENIGN BREAST LESIONS
Breast masses occurring in prepubertal girls are almost always benign and self-limited. The most common cause is asynchronous thelarche. Premature thelarche should be distinguished from normal puberty, which usually commences after age 8.

Less commonly seen benign breast lesions in adolescents include mammary duct ectasia and cysts of Montgomery. Fibroadenomas are the most common pathologic breast masses seen by pediatric surgeons, and are usually small (1-2 cm), painless, well-circumscribed and mobile. Juvenile fibroadenomas typically affect younger adolescents around the time of puberty, and have the same propensity to regress.

Fibroadenomas can be diagnosed clinically (+/- ultrasound), and managed with close follow-up and reassurance since some will regress over time. The decision to proceed with excisional biopsy should be based upon family anxiety, family history of breast cancer, size greater than 5 cm (giant fibromadenoma) and interval growth.

MALIGNANT BREAST LESIONS
Fortunately the incidence of malignant breast tumors in children and adolescents is extremely low. Intraductal papilloma and phyllodes tumors are typically benign, but can be malignant.

Approximately 10 percent of phyllodes tumors occur in females younger than 20 years of age, and typically present as a rapidly enlarging breast mass (> 6 cm). Malignant breast lesions in children are classified as: primary malignancies, metastatic cancer and secondary malignancies. Virtually all children with malignant lesions have a palpable breast mass. If diagnostic uncertainty exists, referral to a pediatric surgeon should be considered.

PEDIATRIC BREAST DISEASE: PEARLS AND PITFALLS
1. Unilateral breast bud development is common, and errant biopsy of a normal breast bud can lead to disfigurement.

2. Pubertal gynecomastia in boys usually resolves completely within one year of onset.

3. Soft tissue infections in close proximity to the breast bud should be evaluated promptly and treated aggressively to avoid damage to the breast bud.

4. Bilateral gynecomastia occurring before age 8 (i.e., precocious puberty) should prompt endocrine evaluation to determine a specific cause (adrenal, ovarian and hypothalamic).

5. Supernumerary nipples (polythelia) may develop anywhere along the milk line (i.e., axilla to pubis) and occur in about 5 percent of children.

CONSULTS/REFERRALS
We will gladly facilitate referrals for breast problems and are always available for consultation. Contact the on-call pediatric surgeon at 860.545.9620. Contact Dr. Brendan Campbell is a pediatric surgeon with expertise in pediatric gynecology and breast disease. He is the surgeon champion for the Pediatric National Surgical Quality Improvement Program and the medical director of the Trauma Program at Connecticut Children’s. Reach him at bcampbell@connecticutchildrens.org or 860.545.9659.
Rapid Response to Airway Emergencies
Groundbreaking Initiative Has Saved Lives and Improved Patient Care.

Connecticut Children’s Critical Airway Response Team, which was created in mid-2012, has had approximately 50 activations so far, and all patients have survived their airway crises, according to pediatric otolaryngologist Dr. Nicole Murray, who spearheaded the airway rapid response initiative. The initiative’s success, Murray says, “is due to great collaboration among all parties involved.”

When an airway emergency code is called, responders come from anesthesia, otolaryngology, critical care and surgery. They bring with them a specially equipped cart stocked with advanced surgical, laryngoscopy and bronchoscopy equipment, a trach tray and other essential items. The program began with one cart. A second one was added for use in the NICU, thanks to a grant from the Will Rogers Foundation.

While the operating room is still the first choice for handling complex airway problems, Murray says, “Having this equipment gives us the ability to respond if we don’t have time to get to the OR. It doesn’t replace the OR, but makes us much more confident in our abilities to respond elsewhere if need be.”

The equipment is sometimes used for non-emergent situations, as well. It offers the advantage of having a video screen that allows everyone present to see what the clinician is seeing through the scope. “This adds a lot to patient care, because we can educate parents, nurses, medical students and other members of the team who might not otherwise see the airway pathology we’re evaluating when doing a non-emergent airway,” Murray says.

Murray chaired a symposium on rapid airway response at a recent annual meeting of the American Society for Pediatric Otolaryngology. Other presenters included representatives from children’s hospitals in Cincinnati, Philadelphia and Seattle. Prior to the meeting, the four hospitals had discovered that they’d been working independently to develop emergency airway response programs and that the systems they developed shared several key components.

As for Connecticut Children’s program, Murray says, “I’m really proud of it. It’s a great answer to a common problem.”

Dr. Murray is associate professor of otolaryngology at the UConn School of Medicine. She may be contacted at 860.545.9650 or lmurray@connecticutchildrens.org.

Welcome Aboard
We’re pleased to announce another fine addition to our medical staff.

Christine Matarese, DO
Neurology
- Child neurologist, Nemours Children’s Hospital, Orlando, Florida
- 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

Schedule a “Lunch & Learn”
Specialists from Connecticut Children’s are available to visit your practice to discuss clinical topics. Lunch is provided for physicians, APRNs and PAs. To schedule a talk, contact Trish Masse at tmasse@connecticutchildrens.org. A new talk has been added on pain management, sedation services, alternative therapies and relaxation techniques.

Webinars Offered
Connecticut Children’s will present several webinars during 2015, including:

**Evaluation of the Neck Mass in Infants and Children**
March 18
Dr. Scott Schoen, pediatric otolaryngologist, presents.

**Bariatric Surgery**
April 22
Dr. Christine Finck, chief of the Division of Pediatric Surgery and head of Connecticut Children’s Pediatric Obesity Center explains options and procedures.

**New Comprehensive Lyme Disease Program**
Late May date TBA
Dr. Lawrence Zemel, head of the Division of Rheumatology, discusses this new program.

In addition, four previously presented webinars are archived and available for viewing.

**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.
TRANSITIONS

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

Theresa Hendricksen, RN, who was executive vice president and chief operating officer, has accepted the position of chief nursing officer at Children’s Hospital of Michigan in Detroit.

Dean Rapoza has returned to the position of president of Connecticut Children’s Specialty Group, enabling Dr. Jeffrey Thomson to once again give his full attention to heading the Division of Orthopaedics.

Martha Schall, who served as president of the Connecticut Children’s Foundation is now the new deputy secretary at Yale University.

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

April 2, 2015
Allergy Update 2015

MAY 5, 2015
Social Media and Health Risks

ANDRULONIS CHILD MENTAL HEALTH EVENING LECTURE SERIES

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.