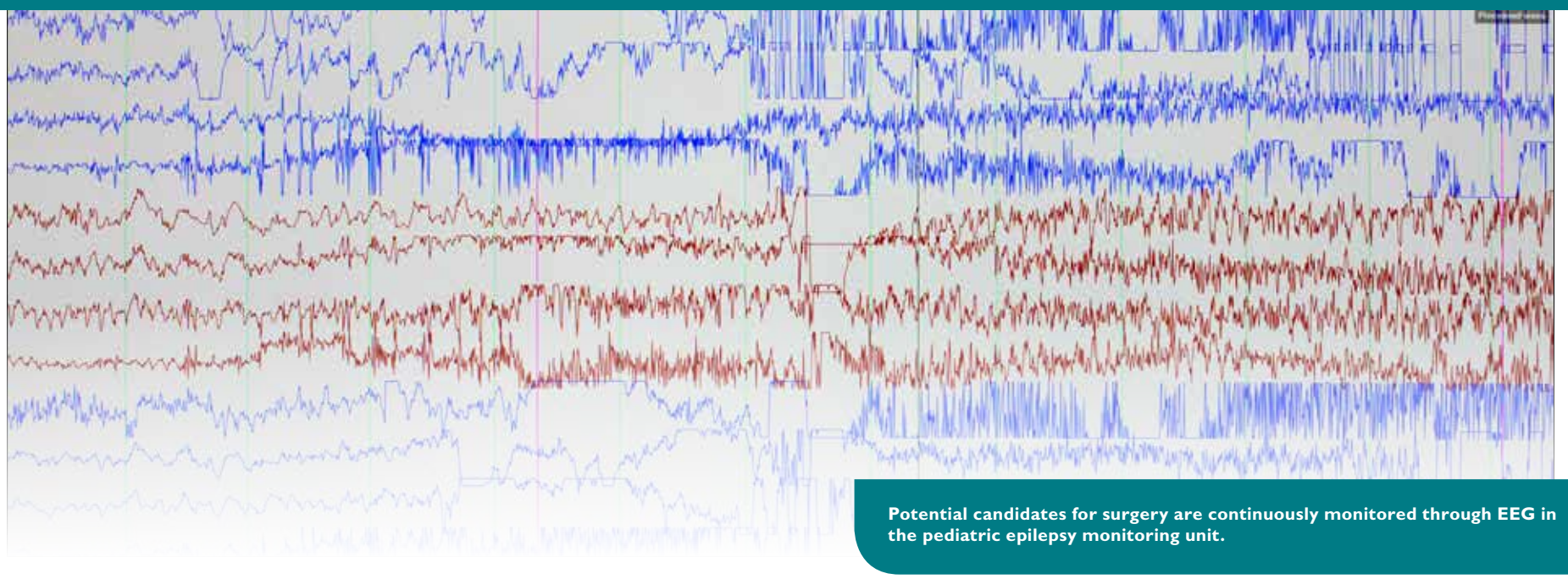
PEDIATRIC
NeuroNews

The latest on pediatric neurologic and neurosurgical care at the Johns Hopkins Children's Center

JOHNS HOPKINS CHILDREN'S CENTER

Winter 2017



Potential candidates for surgery are continuously monitored through EEG in the pediatric epilepsy monitoring unit.

Comprehensive Care for **Pediatric Epilepsy**

Of the estimated 150,000 people diagnosed with epilepsy each year in the United States, only two-thirds are effectively treated with medication. That leaves tens of thousands who will require different treatments to keep their seizures under control.

"We need other options," says neurologist **Sarah Kelley**, who directs Johns Hopkins' pediatric epilepsy monitoring unit. "That's why we have a comprehensive approach at the Johns Hopkins Epilepsy Center."

The center, which cares for patients of all ages from neonates through adults, is one of the oldest of its kind. Johns Hopkins began diagnosing and treating epilepsy as early as 1908. The center's pediatric practice got a boost in the 1970s, led by epileptologist John Freeman, an innovator who revived the use of the ketogenic diet and hemispherectomy, two treatments that can be highly effective for some patients but that had fallen out of favor decades ago.

Nowadays, the center includes a multidisciplinary team of experts who treat pediatric epilepsy, including pediatric epileptologists, neurosurgeons, neuroradiologists, dietitians, neuropsychologists and neuropsychiatrists, all working together to provide tailored care for each patient.

Every Tuesday, the team meets to discuss ongoing cases and patients with the most serious disease. Most of these patients, Kelley explains, start treatment with medication. About 20 epilepsy medications are currently available, and new ones periodically go on the market, she adds.

If patients fail on several different medications, dietary treatments may be an option. The Ketogenic Diet Center runs as part of the Johns Hopkins Epilepsy Center, providing consultations and overseeing treatment for patients interested in this high-fat, low-carbohydrate eating plan. In recent years, the center has overseen many patients on a modified Atkins diet, a less restrictive version. Studies have shown that dietary modification can ease symptoms for up to half of patients with refractory epilepsy, Kelley says.

Shenandoah "Dody" Robinson, left, and Sarah Kelly are among a multidisciplinary team of experts treating pediatric epilepsy. The group includes neuropsychiatrists, neuropsychologists, dietitians, neuroradiologists, neurosugreons, neurologists and pediatric epileptologists.

Surgery can also be a viable option that can either significantly ease or completely eliminate seizures for some patients, says pediatric neurosurgeon **Shenandoah "Dody" Robinson**. Potential candidates for surgery typically spend up to a week in the epilepsy monitoring unit. There, they're monitored continuously through video EEG to determine the frequency, severity and location of their seizures. A subset of patients will require additional monitoring through stereo-EEG or subdural electrodes.

Once monitoring shows that a patient is a surgical candidate, Robinson says, she and other neurosurgeons in the center offer a wealth of options based on each patient's condition. Resections that remove the seizure foci can be performed through open craniotomies or, in some cases, ablative procedures can be performed using minimally invasive techniques. Hemispherectomies and corpus callosotomies are effective for some patients. Neurostimulation, a technology that's recently grown in popularity for treating adult epilepsy, might also eventually be an option in children, Robinson says.

"It's hard to describe how disruptive epilepsy can be for children and families," Robinson says. "We aim to offer the entire range of treatment options here so families don't have to search for help." ■

To refer a patient, call: 410-955-4259

Alan Cohen and Carl Stafstrom

Colleagues Then and Now

In the late 1980s, Carl Stafstrom and Alan Cohen started their careers at New England Medical Center in Boston. Dr. Strafstrom was a resident there, and Dr. Cohen was an attending physician. Now, nearly three decades later, they've reunited at Johns Hopkins, with Dr. Stafstrom as the director of pediatric neurology and Dr. Cohen as the director of pediatric neurosurgery. They are dedicated to using their joint skills and leadership to provide the best care to patients with neurological diseases and disorders.

There are many areas where their work overlaps. For example, their teams collaborate closely when they care for patients with brain tumors: Neurology makes the diagnosis, and neurosurgery delivers a key component of the treatment. Similarly, they provide joint care for a number of other medical issues, including epilepsy, hydrocephalus, tethered spinal cords and birth malformations.

With 17 faculty members between their two departments, they're able to provide comprehensive care for the range of conditions that affect the nervous system. Neurosurgery recently added three of these faculty members to the roster, including Dr. Cohen and Shenandoah "Dody" Robinson, who came to Johns Hopkins in May of this year from Boston Children's Hospital, and Mari Groves, who did her residency and fellowship at Johns Hopkins as well as an additional fellowship in pediatric spinal deformity at Shriners Hospitals for Children in Philadelphia.

Both directors say they are thrilled to be working together again and leading their departments to provide the excellent care, training and research that form the pillars of Johns Hopkins' mission. ■

Michael Levy and Carlos Pardo discuss a patient case in the Transverse Myelitis Center—one of the few dedicated to treating the condition in the world.

Helping the **Youngest Patients with Transverse Myelitis**

Transverse myelitis (TM) symptoms in young patients frequently start the same way: complaints of back pain, which parents often don't take very seriously at first. But as the day passes, it becomes clearer that the problem is far worse than a simple muscle ache. Soon enough, symptoms progress and cause weakness in the legs or arms, and the patients can't urinate or move their bowels. By the time they arrive at the local emergency room, many can't walk.

No one is sure how many pediatric patients are diagnosed with this condition each year in the United States, says neurologist **Carlos Pardo**, who directs the Johns Hopkins Transverse Myelitis Center. Johns Hopkins is the region's top tertiary referral hospital for this condition, and the center is one of the few dedicated to treating TM in the world. Pardo and colleagues see dozens of cases in children each year.

Created in 1999, the center provides a comprehensive approach to diagnosing, treating and managing TM in patients of all ages, says Pardo's colleague **Michael Levy**, who specializes in treating neuromyelitis optica, a TM-related condition.

The first step when a lot of patients arrive at the center is to provide an accurate diagnosis, says Levy. Many conditions, such as spinal cord stroke or metabolic problems associated with mitochondrial disease, can masquerade as TM, which is characterized by inflammation in the spinal cord that damages the myelin sheaths on nerve fibers. In children, cases of acute flaccid paralysis linked to respiratory viruses are also of great concern, as patients present with similar symptoms to those of TM. To rule out other causes, the center has a close relationship with Johns Hopkins' Division of Neuroradiology, which provides diagnostic procedures, including spinal angiography, CT myelography, MRI and others, to gather clues as to the cause of patients' symptoms.

Because time is of the essence in treating TM—the longer inflammation persists, the more damage it does to the nerve fibers—some patients are treated with intravenous anti-inflammatory drugs even before doctors are certain of the diagnosis, Levy explains. Plasmapheresis is another common treatment. By exchanging plasma, Levy says, doctors can remove the offending immune factors from patients' bloodstreams and slow an ongoing attack. Although 60 percent of cases are idiopathic, the remainder are thought to be caused by autoimmune disease that may lead to future attacks.

Once they have inflammation under control, Pardo says, the center's job isn't finished. Over the past several years, he's worked to assemble a comprehensive team to care for patients as they recover, a process that can last months or years, or can be ongoing for patients with chronic disease. For example, pain specialists associated with the center are available to help ease the discomfort associated with this condition. Physical and occupational therapists provide aggressive rehabilitation to help return lost function. Ophthalmologists in Johns Hopkins' Wilmer Eye Institute help care for patients with ocular issues. And because depression is also associated with the condition, psychiatrists and psychologists also work with the center to help maintain patients' mental health.

After acute care ends, Pardo says, the center can continue to manage patients' care through regular checkups. Providers remain available by phone or email long after patients are discharged from the clinic.

"Like our adult patients, our goal for children is to get them back to as normal a life as possible," Pardo says. "We want them back to school, hanging out with friends, doing sports, whatever they enjoy. Our experts can help get them there." ■

To refer a patient, call: 410-502-7099

Treating a Spectrum of Neurosurgical Conditions: From Infancy to Adulthood

When pediatric neurosurgeon **Mari Groves** was receiving her fellowship training here four years ago, a friend's child began developing progressive weakness and other puzzling symptoms. Imaging showed that this toddler had an extensive thoracic tumor. Although some might have found such a tumor unresectable, remembers Groves, this patient underwent surgery at Johns Hopkins—a procedure so transformative that he's required little intervention since.

The case reaffirmed the reasons why she chose this specialty, she says. "With many adult conditions, there's little you can do to change the overall prognosis of the disease," she explains. "But with pediatrics, we often have the chance to change the course of a child's life."

After coming to Johns Hopkins for her residency in 2007, she spent the next eight years here receiving training through her pediatric neurosurgery fellowship, under the direction of former director of pediatric neurosurgery George Jallo. She received additional training in spine deformities at Shriners Hospitals for Children in Philadelphia, then joined the faculty of Johns Hopkins in January 2016.

"I want to bridge that divide and take care of these patients in adulthood as well."

—MARI GROVES

Groves cares for patients from infancy to adulthood. Several of the conditions she treats, such as myelomeningoceles and tethered cords, are typically diagnosed and treated very early in a child's life. Although some aspects of these conditions can linger into adulthood, Groves explains, many adult neurosurgeons are uncomfortable working with patients who have graduated from pediatric practice. "I want to bridge that divide and take care of these patients in adulthood as well," she says.

Groves treats a variety of conditions, including brain tumors, Chiari malformations, spinal cord tumors and spinal deformities—pathologies that are often extraordinarily challenging and complex. "Some of these patients have been told that nothing can be done," she says, "so they're particularly grateful when we're able to provide treatment."

Many of her cases require a team approach that involves a cadre of specialists in other disciplines. For example, Groves is part of the Johns Hopkins Greenberg Center for Skeletal Dysplasias, which aims to comprehensively treat this group of conditions with the help of experts in orthopaedic surgery, otolaryngology–head and neck surgery, pulmonology and other specialties.

For her pediatric tumor cases, Groves and colleagues from pediatric medical and radiation oncology work together to develop an individual treatment plan for each patient to optimize outcomes.

No matter what condition her patients have, her primary goal is to help patients and their families get back a sense of normalcy.

"Parents have faith and trust when they give the most important things in their lives to us," Groves says. "I want them to know that I understand where they are, and we'll do everything we can to help their children live full lives." ■

To refer a patient or for more information: 410-955-7337

Big Advances in Minimally Invasive Neurosurgery

The field of neurosurgery is practically unrecognizable compared with when it got its start at Johns Hopkins more than a century ago, says Chief of Pediatric Neurosurgery **Alan Cohen**. Antibiotics and corticosteroids are available now to reduce the risk of infection and damage to the brain. Preoperative imaging allows surgeons to craft a plan based on each patient's unique anatomy, feeding information to image guidance systems in real time during surgery. Intraoperative imaging can give surgeons an update on how successful a procedure might be before they even finish it.

"These are things that the founders probably never dreamed would exist," Cohen says.

One of these seemingly far-flung innovations has been a central focus of his career: Cohen's main clinical and research interest is minimally invasive techniques—those that access the brain and other neurological structures through keyhole openings, rather than the large, open incisions performed decades ago. He uses these techniques to treat a wide range of conditions, including brain and spinal cord tumors, craniosynostosis, hydrocephalus and arachnoid cysts. In the lab, he and his colleagues are working on developing new techniques and new tools that have the potential to transform this field.

Since joining Johns Hopkins from Boston Children's Hospital earlier this year, he's worked to set up a laboratory

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Big Advances in Minimally Invasive Neurosurgery *continued from page 3*

in Johns Hopkins' Carnegie Building, a structure that was erected around the time that neurosurgery was just gaining traction as a field. In his new lab, he's working with a host of researchers from other disciplines, including biomedical engineering and computer science, to innovatively solve current problems in neurosurgery. He's invited some of these researchers to directly witness surgery to point out some of the difficulties plaguing the field and invite ideas on how to fix them.

"By putting people from different disciplines together," he says, "we can benefit from the wisdom of the crowd."

That wisdom is already being put to good use. He and his colleagues are working now on improvements, such as endoscopic forceps that can control bleeding and a smart balloon that gently pushes tissues away, avoiding the need for manipulation that can lead to collateral damage in surgery.

Working with fellow surgeons, Cohen and his colleagues are also investigating new techniques that could eventually make tumors currently considered unresectable amenable to surgery. Through the use of cadavers and models made through three-dimensional printing, the researchers can assess the feasibility of these techniques and perfect them before testing them in patients.

Additionally, Cohen and his colleagues are using three-dimensional models to practice conventional procedures in complex cases using a patient's own anatomy prior to procedures. "It's like a golfer taking a practice swing," Cohen says. "This 'operation before the operation' allows us to plan out the best route to structures without compromising a patient's safety."

With the help of colleagues across Johns Hopkins, Cohen is hoping to revolutionize minimally invasive neurosurgery. "I feel lucky to be part of the Johns Hopkins community," he says. "With collegial talent in different specialties, we can cross boundaries to accomplish things that otherwise wouldn't happen." ■

To refer a patient, call: 410-955-7337

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