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PhysicianUpdate

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This newsletter is published for the Johns Hopkins Clinical Practice Association by Johns Hopkins Medicine Marketing and Communications

Clinical Practice Association

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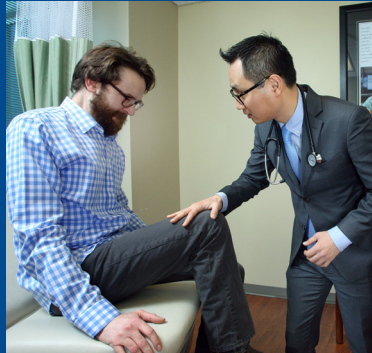
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PhysicianUpdate

NEWS FOR PHYSICIANS FROM JOHNS HOPKINS MEDICINE

FALL 2015

New Thinking on Myositis Rehab

When it comes to managing patients with myositis, says **Tae Chung**, fear of causing harm can be overstated. The physiatrist and director of the neuromuscular rehab team at Johns Hopkins explains that physicians often discourage exercise for patients with the rare inflammatory muscular disorder out of concern that it could raise creatine kinase (CPK) levels and further damage weakened muscles. That fear is understandable, says Chung, considering how little research has been conducted on these patients to demonstrate their threshold for physical therapy and which exercises might prove helpful.

But, says Chung, he and his colleagues have found a way to customize rehab for patients with myositis while prescribing medications to fight their autoimmunity. "These patients want to do something to strengthen their muscles," says Chung, "and we've developed rehab protocols at various stages of the disease."

The exercises—mostly focused on high-intensity resistance and core muscle strengthening—are based on studies using objective measures, notes Chung. Protocols are modeled after numerous studies performed in Sweden, which the Swedish team has shared with him. Since the neuromuscular clinic opened last fall, Chung has been documenting each patient's baseline strength and tracking blood values to build on this research.

Among those who appear to be benefiting is Matt Darnell. The 33-year-old engineer from Chicago knew something was amiss last summer when he had trouble doing his morning pushups, a habit he's sustained since his years in the Navy. "I felt an overall weakness," he says, "and was worried when it didn't improve."

Blood work showed elevated CPK, and an electromyography was consistent with irritable myopathy. This was followed by a biopsy. But, as is the case for many patients with myositis, which resembles many other neuromuscular diseases, his neurologist couldn't pinpoint a diagnosis. Meanwhile, Darnell was advised not to exercise.

An online search led Darnell to studies on myositis, many of which he found were authored by Johns Hopkins researchers. So he scheduled an appointment with Chung and flew to Baltimore. Chung suspected and confirmed that Darnell has immune-mediated necrotizing myopathy with HMGR antibody, a disease entity discovered in neurologist **Andrew Mammen's** lab, where Chung worked some years ago. It's a rare complication of statins. Darnell, however, had never taken these types of cholesterol-lowering drugs.

Despite the long day spent meeting with Chung, other specialists, and physical and occupational therapists at Johns Hopkins' Myositis Center, Darnell says he values the thorough team approach because "it puts different eyes on the same problem."

Chung was the first of Darnell's physicians to prescribe immunosuppressants. "We need to control the immune system," Chung told Darnell, "so the muscle is no longer inappropriately attacked." Darnell says he's tolerated the drugs well, and that they have lowered his CPK.

In between the monthly visits that have followed, Darnell has stayed in close touch with Chung and physical therapists **Albert "Fin" Mears** and **Ruben Pagkatipunan** and occupational therapist **Sarah Hess**.

Roughly 70 percent of the patients with myositis now under the team's care are from out of state or from overseas. And, because patients' local therapists are often uncomfortable about prescribing PT, says Mears, "we spend a lot of time educating patients, practicing exercise programs and conferring with their therapists." ■



Tae Chung tests Matt Darnell's reflexes on a follow-up visit for myositis care. Darnell says he wishes more patients with his condition were aware of the benefits of early physical activity.

Neuromuscular Rehab Clinic at a Glance

The Johns Hopkins Neuromuscular Rehab Clinic is among the first to focus on the diagnosis and rehabilitative management of neuromuscular disorders, including all known forms of myositis: dermatomyositis, inclusion body myositis, juvenile myositis and polymyositis. Directed by Tae Chung, the center combines the expertise of physical therapists, occupational therapists and speech-language pathology therapists. When needed, referrals are made to neurology, rheumatology and pulmonology. Treatments are individualized based on diagnosis and severity of disease, which are scientifically assessed. Each patient's baseline strength and blood values are tracked to build on neuromuscular research studies.

To make an appointment at the clinic, call 410-502-2272. International inquiries: +1-410-502-7683

ROUGHLY 70 PERCENT OF THE PATIENTS WITH MYOSITIS NOW UNDER THE TEAM'S CARE ARE FROM OUT OF STATE OR FROM OVERSEAS.



An Electrifying Approach to Epilepsy

About one-half of patients with epilepsy can control their seizures with medication, but when that doesn't prove successful, surgery to remove the seizure focus—the brain region triggering the episodes—can often render patients seizure-free. For some, however, surgery isn't a viable option. Those who have more than one seizure focus or more than one in locations where removal would lead to neurological deficits, for example, aren't candidates for the procedure.

In the past, these patients had few other options.

Now, neurologist **Gregory Bergey** and neurosurgeon **William Anderson** are offering a new treatment, responsive neurostimulation therapy, which recently gained U.S. Food and Drug Administration approval.

This therapy is delivered via a device consisting of a small computer and stimulator, about half the length of a dollar bill and the thickness of a finger. Anderson can implant the apparatus in a recess carved out in the skull. Extending from the central unit are two leads ending in electrodes, which are placed through holes in the dura into the brain regions near the seizure foci, as determined by previous intensive monitoring studies.

Once activated, Bergey can tune the closed-loop responsive stimulator to detect epileptiform electrical activity from the seizure focus. It then delivers a brief electrical pulse through the leads that can cut a seizure short within a couple of seconds. Patients don't feel the pulse, and many don't even feel the beginning of the seizure, which never evolves further.



Gregory Bergey, left, and William Anderson now offer responsive neurostimulation therapy to control epileptic seizures—a treatment that was in clinical trials at Johns Hopkins and other sites, and was recently approved by the FDA.

Clinical trials over the past several years, at Johns Hopkins and other sites, have shown that the majority of patients who receive this device have an early reduction in their seizures that increases over time. Up to 60 percent of patients eventually have the number of their seizures cut in half. About 10 percent of patients eventually become seizure-free.

Though the stimulator requires periodic battery changes—how often depends on how frequently patients' brain activity triggers a stimulating event—the neurostimulator requires no day-to-day maintenance or thought from patients. "It's not like taking a pill, where you'll have seizures if you forget," Bergey says. ■

Call: 443-287-1609
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A Revealing Biopsy for Sjogren's Syndrome

Sjogren's syndrome is one of medicine's most puzzling enigmas. Recognized as an autoimmune disorder, its constellation of symptoms can seem bafflingly unrelated, causing problems as diverse as multiple dental caries, chronic dry eye, painful intercourse and digestive problems. Though researchers have tied many of these issues to an attack on the body's exocrine glands, including salivary and lacrimal, its cause and exactly how it exerts its effects remain a mystery.

"What we don't understand is the root cause of the process," says Johns Hopkins otolaryngologist **Jean Kim**. "But what we do know is that this syndrome is excruciatingly debilitating for patients."

So much about Sjogren's still remains unknown that even diagnosing the disease can be a challenge. That's where Kim's expertise comes in. Through working with numerous patients in Johns Hopkins' Jerome L. Greene Sjogren's Syndrome Center, the only clinic in the world dedicated to patients with this condition, she's perfected a technique for biopsy of the minor salivary glands in the lower lip. The results from this biopsy provide the largest and most definitive clue available for separating Sjogren's from other conditions with related symptoms.

Thousands of minor salivary glands line the

mouth and upper airways, providing the mucus necessary for proper aerodigestive function. However, explains Kim, few physicians are trained or experienced in effectively and safely dissecting minor salivary glands from surrounding tissue. These glands are superficial and surrounded by a delicate lacework of nerves, she says, necessitating an approach as noninvasive as possible into the right layer of tissue, rather than a simple wedge resection that often doesn't produce any of the desired glands and can result in numbness of the lower lip.

Having standardized this approach at the Sjogren's Syndrome Center, Kim and her colleagues are able to move patients efficiently from diagnosis to therapy. She sees patients on Wednesdays in the clinic, working with other specialists to provide supportive therapy for the individual facets of this condition. Treating patients effectively often involves experts in rheumatology, neurology, ophthalmology, gynecology and numerous other areas.

In addition to treating patients, Kim also performs basic research on this condition, searching for Sjogren's root cause and clues that could provide better treatments. Some of the minor salivary glands from patients' biopsies also go toward this cause. Kim and her colleagues are currently



Jean Kim has developed a biopsy technique that offers the most definitive diagnosis of the enigmatic Sjogren's syndrome.

studying the genetic makeup of patients' glands and performing experiments to identify the functional differences between these and glands from healthy patients. Eventually, she says, their findings could provide new hope for patients plagued by this mysterious condition.

"It's not enough to say that we don't understand Sjogren's and that it's just going to be an enigma," Kim says. "The patients we see fuel the fire that keeps us moving toward better treatments and, eventually, a cure." ■

Call: 443-997-6467
International inquiries: +1-410-502-7683

The Planning Stage of Complex Fracture Repair

When **Erik Hasenboehler** sees a patient with a complex fracture involving 10 to 20 fracture segments in an area, his first thought is not about the exact approach he will use to repair the injury. He initially thinks about the optimum time he should wait before repairing the injury.

As the primary orthopaedic traumatologist at Johns Hopkins Bayview Medical Center, Hasenboehler specializes in complex fracture repair in the upper and lower extremities. Planning the proper surgical approach for such extensive injuries, he says, involves studying the fracture pattern, and that can take some time. But the main reason for waiting is to allow the body to relax. "The amount of soft-tissue swelling goes down, which decreases the morbidity and the infection risk after surgery," says Hasenboehler.

Complicated fracture repair and reconstruction involves contemplating not only the right time and approach, but also each particular incision, the specific implant and the potential need for more than one operation.

For example, when Hasenboehler plans to repair a pilon fracture in the tibia and fibula near the ankle, he waits between 14 and 20 days after the injury occurred. Then, he says, "open reduction through an incision allows us to visualize the fracture, reduce it with clamps and wires, and then put the pieces back together."

Typically, a pilon injury requires multiple approaches with several plates and screws at multiple levels. Each step is deliberately chosen because if it's not done perfectly, he says, the repair will cause more harm, and he will have to repeat the process.

For another complex fracture—a tibial plateau fracture involving the knee—Hasenboehler waits 10 to 14 days for the swelling to decrease. For the repair, he chooses from various possible



Timing is crucial when it comes to planning the proper surgical approach for extensive fractures, notes orthopaedic traumatologist Erik Hasenboehler.

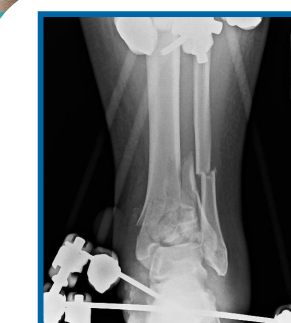
approaches to combine with an internal fixator for the tibia. The incisions are made on the inside and outside of the knee.

When an implant is required, Hasenboehler knows all the options. "There are so many available," he says, "but one is usually better than another for a particular type of injury."

If he doesn't want to use internal hardware because of the possibility of an infection, Hasenboehler chooses a multiplanar external fixator to definitively treat the injury, since this keeps all the hardware on the outside of the body.

Patients come to the orthopaedic traumatologist from hospitals across the region for complex fracture care and remain in his care until they are healed. The process usually takes six to nine months, or even longer if the patient needs multiple surgeries. ■

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Above, a highly comminuted pilon fracture has an external fixator applied. Below, after surgery, the fracture is repaired.



Endarterectomy in Patients Over 70

At 94, Doris Russell has a workout routine that puts to shame most people half her age. She swims laps three times a week and competes annually in the Maryland Senior Olympics and YMCA Nationals. But three years ago, Russell had a series of transient ischemic attacks. Because these mini-strokes are forerunners of full-blown ischemic stroke, they could not only derail Russell's workouts, but also precipitate devastating neurologic injury. So when the spry nonagenarian ended up in the office

of Johns Hopkins

vascular surgeon

Bruce Perler,

the decision to proceed with carotid endarterectomy was a no-brainer.

"Strokes are particularly debilitating in the elderly due to their

lower physiologic reserve, but also because the aging brain tends to have preexisting microvascular damage, so the effects of a stroke can be more extensive and clinically severe," Perler says.

Russell is among a growing number of people over 70 who undergo both therapeutic and preventive carotid endarterectomies. In fact, about 40 percent of the nearly 2,000 carotid patients Perler has operated on are in their 70s, 80s and 90s. Performing carotid surgery among elderly patients with symptomatic disease—marked by TIAs and notable carotid occlusion—is not controversial, Perler says.

"Between 30 and 40 percent of people who have a mini-stroke go on to have a full-blown stroke," Perler says. "Doing nothing poses a grave risk."

However, performing surgery for asymptomatic disease in this advanced-age group remains debatable because the risk-benefit ratio is less clear. Yet, Perler says, when done correctly, the procedure—gently peeling away the calcified atherosclerotic plaque from the arterial wall—is both safe and justified.

Older symptom-free patients with substantial carotid occlusion—80 to 90 percent or more—stand to benefit from pre-emptive surgery because the risk of

full-blown stroke looms large.

Tailoring intraoperative and postoperative care to the physiologic needs of elderly people can minimize risk, and so can careful patient selection, Perler says.

For example, Johns Hopkins anesthesiologists use minimal sedation, and the patient is awake promptly after surgery, which lasts about an hour. Most go home the following day. A careful preop exam and ultrasound can pinpoint the occlusions—usually at the carotid bifurcation—and ensure a smaller incision. And since carotid stenosis is fueled chiefly by atherosclerotic plaque, all patients should be on statins, Perler adds, because those who take them fare better overall. "If they are not already on a statin," he says, "we make sure they start taking one, ideally at least a month before surgery." ■

Information: 410-955-5165 (option 3)
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