

PhysicianUpdate

NEWS FOR PHYSICIANS FROM JOHNS HOPKINS MEDICINE

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More Than Nerve-Zapping for TN

A living hell. The suicide disease. Clinicians with even a passing knowledge of trigeminal neuralgia know that, for most untreated patients, life as they know it stops when the facial agony begins. The repeated episodes, the intense pain—dubbed lancinating and shock-like—“are broadly debilitating,” says **Michael Lim**, a neurosurgeon with the Trigeminal Neuralgia Center at Johns Hopkins.

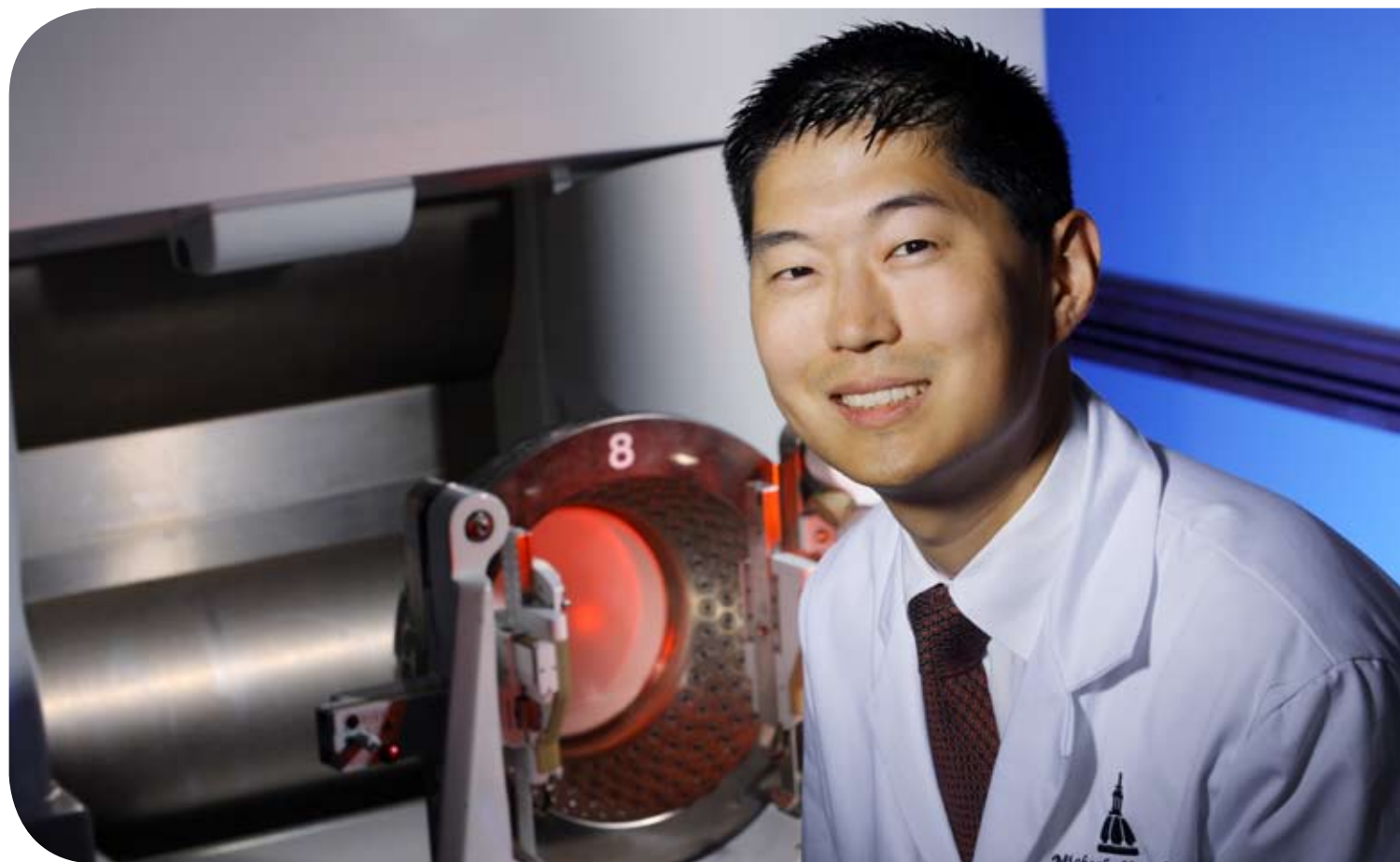
Therapies for trigeminal neuralgia are equally broad—a result of its persistence and the need to treat mostly older patients who suffer a variety of comorbidities. So the center offers percutaneous approaches, using glycerol, for example, to quiet an irritable trigeminal nerve.

And either Lim or neurosurgeon Ben Carson perform gold-standard microvascular surgery to relieve pressure on it.

But it’s the newest treatment at Hopkins, the gamma knife, that’s come into its own. The advantages of gamma knife radiosurgery (GKR), which concentrates ionizing radiation at a trigeminal nerve root target, are clear. “It’s minimally invasive,” says Lim. “If you’re working the day before you came in, you can work the day after. You can have dinner that night with your family.” Complications are low, and the most common one—localized numbness—can itself be a relief, patients say.

GKR is ideal for those who are frail or anti-surgery. “Also,” Lim adds, “it gets around the hazard of surgery for patients on coumadin or other anticoagulant ther-

“It gets around the hazard of surgery for patients on coumadin or other anticoagulant therapy.”



Neurosurgeon Michael Lim has made the gamma knife a specialty.

apy.” About 30 center patients have the procedure each year.

Yet it’s Lim’s experience with GKR and the way his earlier evidence-based research shapes his practice that’s put the therapy at Hopkins on a par with traditional decompression surgery for initial relief. “We’ve watched our successes go up,” he says, “and the two are comparable.

“It’s not simply zapping the nerve with radiation,” he adds. It’s an art to get the beam placed, its dosage and the length of nerve radiated just right. “Some studies show that the nearer you can safely go to the brainstem, the better the result.

In general, we aim 3 to 4 millimeters in front.” And how much nerve to treat? Lim’s research suggests 6-millimeter segments shorten the time to pain control.

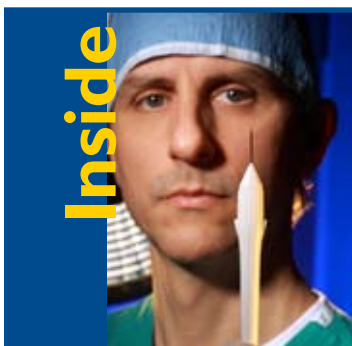
The initial workup also figures into the successes. The high-contrast, high-resolution MRI approach—FIESTA sequences—allow center clinicians to image brainstem arteries in all their tiny, nerve-encroaching detail. That not only sharpens trigeminal targeting but helps tell if radiosurgery’s appropriate.

The down side of gamma knife therapy is the relatively long time for pain relief to become complete—six to 10 weeks—as

radiated trigeminal axons and myelin shut down. “And though the treatment’s durable,” Lim says, “there’s still a 50 percent chance at least some pain will recur in five or 10 years.” Lim’s fine-tuning lessens that, but only so far, a fact that drives him to the lab. What’s ideal, he says, would be something that sensitizes the nerve to radiation—that and something to make normal tissue nearby less vulnerable.

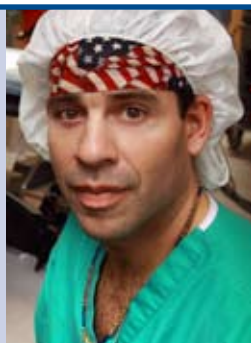
Meanwhile, Lim says, the center sticks to its motto: Leave no one in pain. “We offer all the therapies that are out and we really tailor them to the patient.”

☎ 410-955-9441 to refer a patient.



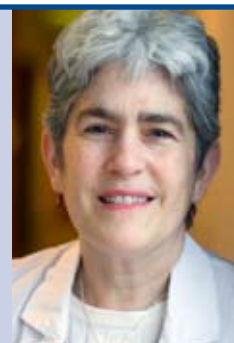
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Taking the Pressure Off Glaucoma

John Haddick took a certain pride in the eagle-eye vision he'd enjoyed for 24 years as a Navy pilot. He was even OK as his acuity softened from 20/15 to 20/40 during his later career with the Department of Defense. But as an advancing case of glaucoma began to darken his right eye's visual field on the eve of his 70th birthday last year, Haddick decided to make a move.

His ophthalmologist recommended one of the most common surgical solutions for adult glaucoma, a trabeculectomy, in which a hole is made in the sclera under a flap to release trapped intraocular fluid to the outer surface of the eye, relieving destructive pressure on the optic nerve fibers below.

Still, the operation has drawbacks. The ocular drainage system is permanently rerouted. The procedure often fails over time and puts the eye at permanent risk

“You ablate only the tissue in between the gap.”

of developing an infection, one that can be blinding. These details gave Haddick pause.

At Johns Hopkins' Wilmer Eye Institute, glaucoma expert **David Friedman** soon confirmed that other options are now available. In fact, Friedman says, about

one in five of the nation's 2.5 million glaucoma patients may qualify for a growing procedure that he calls “elegant.”

Named after the new technology that makes it possible, the Trabectome approach allows eye surgeons to restore fluid outflow to over-pressured eyes without the more radical restructuring posed by traditional trabeculectomies. The technique uses a fine probe with a curved tip that can precisely remove fine layers of clogged tissues that block aqueous flow.

“It all comes down to plumbing,” explains Friedman. With all glaucoma cases, he says, “we just need to make sure that either more fluid is leaving the eye or less is coming in.”

The Trabectome probe uses an electrical pulse that passes through a gap in the device's tip that spans less than 300 microns, nearly exactly the thickness of the trabecular meshwork, the area where aqueous flow is usually slowed. With the patient's afflicted eye numbed by anesthesia, the surgeon places the probe into the front part of the eye, just anterior to the iris, and slowly sweeps it through the affected layer of meshwork.

“You ablate only the tissue in between the gap,” explains Friedman. The unwanted material is then suctioned away by the probe, which has an aspiration port. After the probe is withdrawn, “you put one stitch in where you entered the eye,” he says.



David Friedman offers an alternative to the traditional trabeculectomy.

The Trabectome approach worked “perfectly” on Haddick, and Friedman has continued to extend the technology to dozens of new patients. He says about 70 percent will need no further surgery. “You can't perform the procedure on every-

body,” Friedman says, explaining that patients with angle closure glaucoma are not good candidates, along with those whose intraocular pressures must be brought to the lower range of the healthy scale.

☎ 410-955-6052 to refer a patient.

For Ataxia, No Longer Nowhere To Go

If someone published *Topics of Neurologists' Anxiety Dreams*, ataxia would probably be among the chapters. The loss of coordinated movement may not be so difficult to diagnose, but having to tell a patient that his ataxia is progressive and incurable is disquieting.

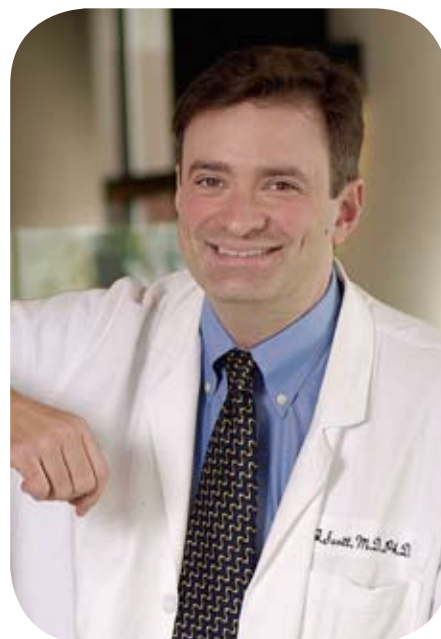
That's one reason that the Ataxia Center at Johns Hopkins opened last spring. Though the condition is rare, Hopkins neurologists have been seeing more than 350 people with ataxia or suspected ataxia each year. And access to the most current imaging and to sensitive bioassays, as well as to the hospital's immunologists, oncologists and geneticists, puts center clinicians in a good place to distinguish treatable ataxia from spontaneous or hereditary types. “We take unusual care in our testing,” says ataxia specialist **Joseph Savitt**,

“partly because of that.”

But it's after primary ataxia has been diagnosed that the center comes to its best use: to support patients as symptoms progress and to ensure the highest possible quality of life. Most have heard, *There's nothing we can do for you*. “They feel they have nowhere to go,” says Savitt, “and nothing to hope for.”

“Because we're not able to cure their ataxia, the interventions we can do, the strategies and the education we offer become extremely important,” explains clinic coordinator Katie McGuire. Patients certainly find benefit from gait-assistance devices, swallowing strategies, targeted speech therapies and medications for hypotension, bladder difficulties, muscle spasms or vision problems, for example.

“I've seen patients' lives vastly improve



with physical and speech therapy,” says Savitt, “often to a degree better than medication for some other disorders.” One man, for example, lived his life for ballroom dancing but his increasing unsteadiness cut that short. When aggressive physical therapy brought him back to the dance floor, “he was transformed.”

And not least, the center's one-stop experience is a plus. Seeing the neurologist, having speech, occupational and physical therapists on site, and having ready access, if needed, to a psychiatrist or ophthalmologist during a day's visit saves patients the stress of multiple returns.

☎ 410-955-4894 to refer a patient.

“People think there's nothing much you can do for ataxia patients,” says neurologist Joseph Savitt. “They're wrong.”

A Low-Impact Hysterectomy for Cervical Cancer

When Renee Burcin met with a surgical oncologist last spring, the diagnosis of a cancerous tumor in her cervix was still new and ominous. If she wanted a shot at ridding herself of cancer, the surgeon counseled, Burcin needed to undergo a radical hysterectomy. This was not a time for half-way measures, and the clock was ticking. Everything should come out, he said—the uterus, fallopian tubes, the ovaries.

Burcin certainly wanted to do whatever was needed to improve her prognosis. But the 46-year-old had also heard enough about radical hysterectomies, which normally must be performed via laparotomy, to know that she'd just as soon minimize the trauma.

Searching for an alternative to the open procedure, Burcin found her way to **Robert Bristow**, who directs Hopkins' Kelly Gynecologic Oncology Service. Bristow assessed Burcin's tumor and considered her eligible for radiation therapy, which could produce an equivalent cure rate to surgery but would also risk damaging the bowel and other surrounding pelvic tissues.

But Bristow, whose group has made

headway in adapting the newest robotic devices to minimize gynecologic surgeries, also told Burcin that her profile made her an ideal candidate for the less invasive approach to radical hysterectomy—her tumor was caught early, and her trim figure allowed easy access to the tissues along with an ability to endure the optimal head-down operating position. He would have to make only five dime-size slits in her abdomen for the entire operation.

Burcin opted for the four-hour procedure on May 28, in which Bristow operated from the controls of his department's robot stationed a few feet away from his patient. He selected and excised the troubled tissues within Burcin's abdomen—including the 3.5-centimeter tumor itself—removing them intact through the small incisions while another operator assisted from a hands-on position.

Burcin was discharged the next day, three days ahead of open-procedure patients, and returned to work in less than three weeks, half the traditional time.

Bristow says Burcin's not yet out of the woods on the cancer risk, but reports he removed 21 lymph glands in the procedure and they all came out clear.



Using the robot, Robert Bristow brings minimally invasive to a major operation.

This lower-impact approach to hysterectomy continues to grow in his practice here, says Bristow. His group has performed more than 80 of the procedures, all with good outcomes.

The best part of the new procedure, says Bristow, is that it's significantly

easier on the patient's quality of life. With hysterectomies currently at 650,000 per year and growing with the boomer generation, says Bristow, it's nice to have the robotic-assist technology to reduce the burden on patients. ☎ 410-502-4245 to refer a patient.

CARDIOLOGY

Pulmonary Valve Replacement: Getting It Right

Ask cardiologist **Jane Crosson** to name one of the biggest challenges facing adult congenital heart disease patients—and their physicians—and she doesn't skip a beat. "Timing," she says, "tops the list."

It's the sort of irony that persistently greets science and medicine: leaps in understanding and technology save lives, but also leave behind a new set of challenges.

In the case of adult patients with congenital heart conditions like tetralogy of Fallot and pulmonary stenosis, the conundrum centers on the optimal timing of pulmonary valve replacement.

Surgical valve repair of such congenital conditions at birth or shortly afterwards has translated to rising numbers of long-term survivors. Today, there are as many adults as children with congenital heart disease. Patients can go several decades—or a lifetime—without problems or symptoms.

But, often by their 20s or 30s, those patients begin experiencing the symptoms of right ventricular dilatation and dysfunction that include excess fatigue, shortness of breath and reduced exercise tolerance.



Adult congenital heart disease patients, says Jane Crosson, need a multidisciplinary approach.

"When that happens," says Crosson, "then they're looking at pulmonary valve replacement." There's some controversy surrounding the timing, though. Crosson says it's a balance between the fact that replacements don't last forever—stenosis and regurgitation are likely to occur within 10 years of replacement—and that waiting too long

may make it too late.

"Patients obviously don't like the prospect of multiple surgeries—and these are open-heart procedures," she says. But, delaying replacement may be akin to Russian roulette.

The key for adult congenital heart disease patients is multidisciplinary coordination that includes cardiology, imaging and cardiac surgery. Along with the leaps that have allowed these patients to survive into adulthood, other advances have been growing up as well, making it easier for physicians to advocate the right time for replacement.

"MRI is a good example of technology that is giving us a clearer view of the size and function of the pulmonary valve," says Crosson. "That has a great impact on decision making." And, Crosson says that though it's early and still in a study phase, percutaneous pulmonary valve replacement may soon be an alternative to open-heart surgery.

"Either way, patients will need lifelong cardiology follow-up," she says. "But, right now, traditional replacement outcomes remain very good and improve function."

☎ 410-614-0706 to refer a patient.

A Gentler Surgical Fix for Stubborn Pancreatitis

When it comes to tending the temperamental human pancreas, it seems, nothing is ever simple. Take the case of 27-year-old Sara Frank, a Virginia teacher who had been suffering upper abdominal pain and nausea since 2005. Her physicians went through a series of diagnostic efforts before tracking her symptoms to a form of pancreatitis stemming from a condition known as pancreas divisum. In people born with this anatomical variant, the pancreas is drained by two separate ducts instead of the usual one, and the resulting main drainage duct is prone to blockage.

Frank's physicians opened one duct with a stent, which relieved her symptoms for three months. Two more duct-clearing procedures followed, but both provided only short-term relief.

By last August, Frank was ready for a more lasting fix. The trouble, she'd learned, was that the traditionally favored surgical treatment of her form of chronic pancreatitis, the ambitious Whipple procedure, sounded too drastic for her condition. Not only does the Whipple entail a dramatic reduction in pancreatic tissue—removal of the entire pancreatic head, the gallbladder, the common bile duct and part of the duodenum—its completion brings a high risk of diabetes. Frank blanched at the prospect of a lifelong commitment to insulin shots.

When she visited with specialists at the Johns Hopkins Center for Pancreatitis to explore alternative remedies, she was pleased at the news of a more tailored option. She learned that a lesser-known operation called the Frey procedure had gained ground in Europe and that one of its most skilled U.S. practitioners is **Dana Andersen**, who heads up surgery at Johns Hopkins Bayview Medical Center.

The Frey approach, explains Andersen, drains the pancreas' primary duct while preserving much more of the organ than does the Whipple. Essentially, he says, he opens up the pancreatic head and "cores out the duct," which means relatively little of the organ is removed, but all of the gland is decompressed. The Whipple is the best answer to pancreatic cancer cases, says Andersen, but sufferers of pancreatitis "don't need a cancer operation."

The Frey procedure's reduced risk of diabetes—8 percent



Sanjay Jagannath and Dana Andersen.

compared to 25 percent—also sounded good to Frank. "I wanted them to take the least amount they could and still get the results I needed," she says.

The surgery unfolded during a five-hour procedure on Sept. 29. After some normal postsurgical pain and nausea, Frank was discharged one week later, several days sooner than she would have needed with the Whipple. Months later, she says she remains pain-free.

Andersen says the Frey approach has been shown to substantially relieve pancreatic pain in about 80 percent of patients, the same as with the Whipple procedure. It provides an extremely low risk of mortality, and only about one in 10 patients develop diabetes. He's very optimistic about Frank's long-term prognosis.

But Frank's case also spotlights the nuanced capability of Hopkins' Center for Pancreatitis, which has assembled a diverse team of gastroenterologists, radiologists, pathologists, surgeons, pain specialists and nutritionists who assess individual cases and carefully tailor a medical response. Gastroenterologist **Sanjay Jagannath**, who heads the center, says the group can handle just about any noncancerous disorder the pancreas throws at them.

Variably driven by a host of genetic, environmental and metabolic factors, pancreatitis has developed a reputation among clinicians as notoriously complex. It can be caused by gallstones, excessive use of alcohol, physical trauma, vascular disorders, pregnancy and even medications. The sheer complexities compel Jagannath to maintain lots of friends throughout the Hopkins science and medicine community.

"This is one of the few centers," he says, "where patients can see a full range of specialists all under one roof, and therapy can be tailored to the individual needs of the patient."

☎ 410-550-2821 to refer a patient.

Your Vital Links

Johns Hopkins Medicine offers the following links to physicians in the surrounding community. We also urge M.D.'s to use our Physician Liaison Service to offer suggestions and comments.

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410-502-2737 (Baltimore area and international calls) jhmcares@jhmi.edu

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