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Brain stent offers alternative to shunt for fixing potentially blinding vein narrowing

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A team of interventional neuroradiologists and neurosurgeons at Johns Hopkins reports wide success with a new procedure to treat pseudotumor cerebri, a rare but potentially blinding condition marked by excessive pressure inside the skull, caused by a dangerous narrowing of a vein located at the base of the brain.

The Johns Hopkins team's latest study, to be published in the *Journal of Neuro-Ophthalmology* online March 14, is believed to be the first to show how directly lowering pressure inside the vein alleviates the condition and improves vision.

The study report on 12 patients describes the team's novel use of intravascular ultrasound imaging to delicately thread an expandable metal stent, roughly two inches long and attached to a catheter, through an opening in the groin, all the way to the main blood vessels in the neck and shoulders draining fluid from the brain. The goal is to precisely position the stent across the narrowed portion of the vein, called the transverse sinus, where it expands, allowing blood to drain more freely and relieving fluid pressure in the brain. The vein narrowing, or stenosis, is considered to be the leading cause of pseudotumor cerebri. The condition earned its name, which translates to "false" tumor of the brain, because in the era before detailed brain imaging became available, surgeons used to operate on people with similar optic nerve swelling, suspecting brain tumors, yet they would find none.

Because the transverse sinus drains cerebrospinal fluid, constriction of

the vessel causes fluid backup and raises intracranial pressure, which, if left untreated, can lead to permanent damage to the optic nerve, blurred vision and eventual blindness. Traditional treatments include drugs that reduce pressure and the surgical placement of a straw-like shunt between the lower spine and brain, or between the lower spine and stomach cavity to drain excess.

According to lead study investigator and Johns Hopkins interventional neuroradiologist Martin Radvany, M.D., the new, minimally invasive procedure, known as transverse sinus stenting, takes about two hours to perform, and could serve as a long-term, if not permanent fix to what he says is a vexing and growing problem, seen mostly in obese, premenopausal women between the ages of 18 and 40.

Radvany, an assistant professor at the Johns Hopkins University School of Medicine, says the new stent procedure has the potential to supplant the current standard of care using shunts, which come with risk of infection, including meningitis, and concerns about too little or too much drainage, and the need for shunt replacements every few years.

"Our study results, if validated in more patients, give us more than an alternative to shunts and bypassing the consequences of pseudotumor cerebri," says study co-investigator and interventional neuroradiologist Philippe Gailloud, M.D. "Our latest research helps us get to the root of the problem so that we can stop and possibly prevent the vein from narrowing in the first place," says Gailloud, who is also director of interventional neuroradiology at Johns Hopkins.

Study results showed that 10 of 12 patients treated had a complete and lasting recovery, with internal vein and spinal pressure readings returning to normal, loss of most symptoms, if not total reversal, and CT scan images showing no return of narrowing in the transverse sinus, nor any worsening optic nerve damage.

Pressure readings taken from inside the narrowed vein immediately before and after transverse sinus stenting showed initial differences between the healthy and constricted areas of the vein as high as 28 millimeters of mercury. These pressure gradients dropped back to and remained at 2 millimeters of mercury or even equalized with normal vein pressure at 0 millimeters of mercury.

Swelling in the eye went down to normal and never returned in 11 out of 12 patients treated. Extensive vision testing, including line-reading tests for visual acuity, distinguishing colors, and peripheral vision, found that eight patients' vision returned to normal. All had an immediate disappearance of headaches and hearing noises inside their heads.

One patient's symptoms later came back and deteriorated after treatment, and another's condition failed to show lasting signs of improvement, but did not get any worse. Both ended up having surgical shunts placed.

Gailloud, an associate professor at the Johns Hopkins University School of Medicine, points out that pseudotumor cerebri is one of many kinds of idiopathic intracranial hypertension, whose origins are unknown. But, he says, the immediate healthy rebound in vein pressure due to the stenting proves that the narrowing of the transverse sinus vein was not due to forces outside the vein, but instrinsic to it. "Now that we know more about what is actually happening, we can orient our research towards finding out what causes the transverse sinus to narrow."

In the study, 11 women and one man were offered transverse sinus stenting as an alternative to surgical shunting, but only after weight loss, dieting, and drug therapy with acetazolamide, or Diamox, a medication that lowers pressure in the brain, had failed to alleviate their symptoms. Participants came from the mid-Atlantic states, and ranged in age from 21 to 55; all were obese, with an average body mass index of 32.6.

All were treated at The Johns Hopkins Hospital between January 2008 and June 2011, and had spinal fluid pressure tests, angiograms and CT scans to confirm that pseudotumor cerebri and vein narrowing were causing their symptoms, the most serious of which is blurred vision, a sign of permanent nerve damage. Most study participants, Gailloud says, experienced initial symptoms of severe headache, with over half reporting "a whooshing noise" inside their heads. All were monitored in the hospital for two days after IVUS, which only required general anesthetic, and were followed through periodic checkups for between one year and as long as four-and-a-half years.

Senior study investigator and neurologist Abhay Moghekar, M.D., says the team will continue to monitor patients who choose transverse sinus stenting instead of surgical shunting, and to compare their progress over several more years.

Moghekar, an assistant professor and director of the Center for Cerebrospinal Fluid Disorders at Johns Hopkins, says having alternative and permanent therapies for pseudotumor cerebri is increasingly important, as the condition appears to be tied to obesity, now an epidemic in the United States. He says that a decade ago, he saw less than two dozen cases per year, but now sees well over a hundred.

"Pseudotumor cerebri is a serious condition, in which the prospect of going blind is very real," says Moghekar. "Until now, we had little evidence of why surgical shunting or IVUS stenting worked," he adds, pointing out that further studies can use pressure gradients to distinguish between people who respond best to one procedure or the other.

Provided by Johns Hopkins University School of Medicine

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