

Hidden Pressure

Often misdiagnosed as Parkinson's, Alzheimer's, or the inevitabilities of aging, "normal pressure" hydrocephalus is a treatable condition. But first it has to be recognized.

BY MICHAEL SMOLINSKY

When Nancy Murray's balance and memory began deteriorating in her 40s, her doctor told her, "You're approaching 50. What do you expect? You can't be 20 for the rest of your life."

But Murray had been experiencing balance problems since her 20s. It wasn't until she totaled her car that this 55-year-old advertising manager from McLean, VA, was examined for adult normal pressure hydrocephalus (sometimes abbreviated NPH), a neurological condition that can cause problems with memory, bladder control, balance, and "gait" (one's manner of walking).

"I'm lucky to be alive," Murray says. "My balance had gotten so bad that I was leaning over to the right all the time. I drove off the right side of the road into an embankment."

One diagnostic challenge is that "the classic symptoms of adult normal pressure hydrocephalus—trouble with walking and balance, thinking and memory, and bladder control—are also the most common problems of aging," says Michael A. Williams, M.D., medical director of the Brain & Spine Institute at LifeBridge Health and director of the Adult Hydrocephalus

“The improvement can be substantial,” observes Dr. Williams. “Some people who were originally in wheelchairs will come in after shunt surgery with only a cane, or nothing.”

Center at Sinai Hospital, both in Baltimore, MD.

Bob Fowler's experience was similar to Murray's. The late Texas oilman dozed off on a drive to Houston and woke up in a roadside ditch. Fowler was misdiagnosed with Parkinson's and Alzheimer's for almost 10 years before being told he had adult normal pressure hydrocephalus and undergoing shunt surgery in 1999. He passed away earlier this year (from leukemia, which is unrelated to hydrocephalus), but not before raising awareness of this treatable disease.

The cognitive deficits of adult normal pressure hydrocephalus can resemble those of Alzheimer's disease, and the gait disturbances can resemble Parkinson's. Researchers at the Cleveland Clinic estimate that as many as 10 percent of people with dementia attributed to other disorders may actually have adult normal pressure hydrocephalus.

Compared to Fowler and Murray, 71-year-old John Howland was fortunate. This former submariner and director of the U.S. Navy's Deep Submergence System Division was only medicated for Parkinson's for two years before being evaluated for hydrocephalus.

“My wife was getting more and more impatient,” Howland remembers. “Eventually she said, ‘That's it, I'm calling Hopkins,’” referring to the Johns Hopkins Hydrocephalus Center in Baltimore, MD, which is where they first were evaluated by Dr. Williams.

“The lucky ones are able to be their own health advocates or have a loved one advocating for them,” says Dory Kranz, executive director of the Hydrocephalus Association in San Francisco, CA (hydroassoc.org). “They keep asking the same questions of different doctors until they find out what's wrong.” Kranz learned about the condition through the experience of her father-in-law, Dick Kranz. It took at least six years from the onset of Dick's gait problems to get a correct evaluation. (See box on page 26 for symptoms.)

WHAT IS HYDROCEPHALUS?

Once called “water on the brain,” hydrocephalus is an abnormal accumulation of cerebrospinal fluid within cavities in the brain called ventricles. This fluid, which is produced in the ventricles and circulates through them continuously, has a number of functions. It surrounds the brain and spinal cord and acts as a cushion against injury, contains nutrients needed for the normal function of the brain, and carries waste products away from surrounding tissues. Ev-

ery day the body makes roughly 8 oz. of it, and about the same amount is reabsorbed into the bloodstream.

Hydrocephalus is due to an imbalance between the amount of fluid that is produced and the absorption rate. As the fluid builds up, it causes the ventricles to enlarge and the pressure inside the head to increase, compressing surrounding brain tissue and leading to neurological complications. Adult normal pressure hydrocephalus occurs most often in people 60 and older and can result from bleeding in the ventricles, meningitis (inflammation often due to infection), head trauma, tumors or cysts, or for no known reason (“idiopathic”).

THE CHALLENGES OF DIAGNOSIS

“Some patients and families don't think of the symptoms of hydrocephalus as a problem,” Dr. Williams says. “They consider them a normal part of getting older.”

In Dick Kranz's case, embarrassment got in the way. “Dick and my mother-in-law conspired to keep the incontinence from us and the doctors,” Dory Kranz recalls.

Doctors often fail to recognize the condition as well, something that experts hope will change. “If any one of the symptoms is evident, normal pressure hydrocephalus should be considered and ruled out before moving on to a diagnosis of dementia or Alzheimer's disease,” says Anthony Marmarou, Ph.D., professor and vice chairman of the department of neurosurgery and director of research at the Medical College of Virginia in Richmond. “Too many people may be living with normal pressure hydrocephalus and suffering needlessly.”

Headaches, head fullness, and vision problems—common symptoms in other forms of hydrocephalus—may offer more obvious clues to abnormal pressure inside the brain. But in adult normal pressure hydrocephalus, these symptoms often never show up, and the pressure can remain hidden.

“Our studies have found that only about 15 percent of elderly idiopathic normal pressure hydrocephalus patients have headaches or head fullness. This is much less frequent than the young and middle-aged adult group, in which 55 percent have headaches or head fullness,” Dr. Williams says.

Dr. Williams examines computerized tomography (CT) or magnetic resonance imaging (MRI) scans to see if a patient's ventricles are larger than they should be for his or her age. “But because the ventricles get

“Normal” Pressure?

The name “normal pressure” came out of a 1965 medical paper describing cases of hydrocephalus where the symptoms occurred in the presence of supposedly normal cerebrospinal-fluid pressure. The paper was published before continuous pressure-recording techniques were available. We now know that “normal pressure” is a misnomer.

larger with age, you can't make a diagnosis on the basis of CT or MRI scan alone, except in rare circumstances," he says.

For Dr. Williams, a patient's response to cerebrospinal fluid drainage—done by lumbar puncture (spinal tap)—is the key to diagnosis and the best indicator of whether shunt surgery will be an effective treatment, and one worth the risks.

"The nice thing about cerebrospinal fluid drainage is that all the other variables—age, other disorders, overall health—stay the same," he notes. "A temporary spinal catheter is inserted in the back to drain the fluid. We look for a 'Wow!' response, where the patient feels substantially better, especially in terms of gait, and the physician can document the improvement. Ninety to 95 percent of the people who respond this way will have success with shunt surgery."

John Howland was one such patient. "I had a lot of trouble walking," he remembers. "My gait was awkward and my feet felt funny, like I was walking in mud. I would lose my balance on

uneven ground. Dr. Williams did a spinal tap and said, 'Let's see you walk down the hall and the stairs,' and I walked straight as an arrow, no problem."

TREATMENT

Hydrocephalus is most often treated by surgically inserting a shunt. "The shunt is the only effective treatment for hydrocephalus," says Dr. Williams, "with one exception: Between five and six percent of elderly patients have obstructive hydrocephalus, caused by tiny blockages in the passageways of the ventricles. An alternative treatment for these patients is endoscopic third ventriculostomy, which involves poking a hole in a membrane in the third ventricle to create a detour for the fluid."

There are three components to the shunt: a thin flexible tube that goes into the brain; the valve, which is a spring mechanism; and another tube that carries the fluid to the abdominal cavity or some other part of the body to drain.



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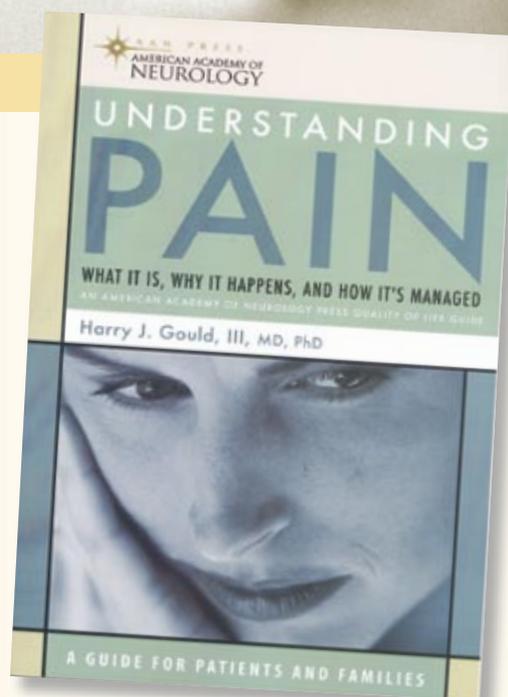
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The development of the adjustable shunt has made a big difference in shunt's success and safety. "You can change the tension on the spring, thereby changing the pressure that allows it to open," Dr. Williams says. "These adjustments take 10 or 15 seconds and can be made without further surgery. It's like adjusting medication dosage: You want enough spinal fluid to drain, but not too much."

A study conducted by Dr. Marmarou and his colleagues of 151 patients with normal pressure hydrocephalus (published in the *Journal of Neurosurgery* in 2005) found that two-thirds (100) improved with temporary drainage. That group then underwent surgery to implant an adjustable shunt, and 91 percent of them benefited from the implant.

In Dick Kranz's case, the surgery was almost immediately successful. However, he soon became a victim of a rare but serious complication: a stroke. Careful monitoring for this post-surgical problem can minimize or prevent serious consequences, Kranz says.

Other complications may include mechanical failure of the shunt, infections, blockages, the need to lengthen or replace the tube, and over-draining, which can result in a subdural hematoma (bleeding). "There is a gap between the brain and the skull that contains blood vessels," Dr. Williams explains. "If too much fluid is drained, the brain pulls away from the skull, which can cause bleeding."

According to the Idiopathic Normal Pressure Hydrocephalus Guidelines (published in the journal *Neurosurgery* in 2005), the reported incidence of subdural hematoma varies widely, from 2 percent to 17 percent, and the reported incidence of infection varies from 3 percent to 6 percent.

"The risks of the procedure include the risk of general anesthesia (very low), the risk of passing the shunt into the brain (very low), the risk of infection (low), and the delayed risk of subdural hematoma (low)," Dr. Williams points out. "But those risks have to be put into context. If you put a shunt into a patient who has very little chance of responding, then those risks are proportionally greater because they're not balanced by benefit."

A NEW LIFE

"The improvement can be substantial," observes Dr. Williams. "Some people who were in wheelchairs will come in after surgery with only a cane, or nothing. They join what some patients call the 'I put my wheelchair in the garage' club. This is really satisfying—you give people their lives back. They have improved bladder, mobility, and cognitive function. Many patients have more modest improvement, depending in part on whether they have other health problems contributing to their

Symptoms of Normal Pressure Hydrocephalus

- **GAIT DISTURBANCES** can range in severity from mild imbalance to the inability to stand or walk. Patients may have trouble picking up their feet, which makes climbing stairs difficult and often results in falls. They may also have difficulty turning around, rotating slowly with multiple small steps. Gait disturbance is usually the most pronounced symptom and the first to become apparent.
- **MILD DEMENTIA** shows up as a loss of interest in daily activities, forgetfulness, difficulty dealing with routine tasks, and short-term memory loss. These cognitive symptoms are usually less severe than full-blown dementia and are often overlooked. Sometimes cognitive changes can only be detected with extensive neuropsychological testing.
- **IMPAIRMENT IN BLADDER CONTROL** is usually characterized by urinary frequency and urgency in mild cases, and a complete loss of bladder control (incontinence) in more severe cases. Urinary frequency is the need to urinate more often than usual, sometimes as often as every one to two hours. "Urgency" is a strong sensation of the need to urinate. Sometimes the urge is strong that it can't be held back, resulting in incontinence. Some people never display signs of bladder problems.

Source: Hydrocephalus Association (hydroassoc.org)

symptoms, but it can be enough to make the difference between dependence and independence."

For Nancy Murray, the treatment has been life-changing. "The surgery was probably the best thing that's happened to me in about 30 years. I feel better than I did when I was 30 years old. And now, I still get up five years later and think, I feel so good today it's just unbelievable. My balance is better, my memory is better, my cognitive skills are better."

Left untreated, people with hydrocephalus do get worse with time, though the degree to which relief of pressure following shunt surgery can minimize or reverse damage to the brain is not well understood.

"Hydrocephalus can go on for a long time and still get better," Dr. Williams says. "The brain seems to tolerate it better than a number of other disorders, and this is something that people in the field are researching now." NN



For more information on hydrocephalus, please see **RESOURCE CENTRAL** on page 37.