What is Normal Pressure Hydrocephalus? Diagnosis and Treatment
Webcast
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Symptoms

Andrew Schorr:
Hello and welcome once again to Patient Power. I'm Andrew Schorr. Well, as we talk about conditions that affect older people, we have talked so many times about Alzheimer's and more broadly dementia, and we have talked about movement disorders like Parkinson's. But those are not the only conditions, and so it's so important to get an accurate diagnosis. Well, today we are visiting with a renowned neurologist at the Northwestern Memorial Hospital. He is also a clinical professor of neurology at Northwestern University's Feinberg School of Medicine and that's Dr. Allan Burke.

Dr. Burke, this condition that we are going to discuss is something that I am not familiar with. Help us understand it, normal pressure hydrocephalus. What are the symptoms of that, particularly in older people, and why could it be confused with some of these other conditions?

Dr. Burke:
The presentation of normal pressure hydrocephalus usually involves three cardinal features. First is a problem walking. It's called an apraxia, meaning really without action, but without a practiced action of walking, and it's as though the feet were glued to the floor. People just can't pick them up even though they have adequate leg strength to do so and can lift their legs and feet in bed without difficulty. Second there is urinary incontinence usually beginning with urinary urgency. And third there is cognitive impairment, dementia. Usually the first symptom the patient experiences is walking, difficulty walking, and that distinguishes it from Alzheimer's disease in which the presentation first involves memory loss and loss of insight into problems with thinking, and motor problems develop very late.

Andrew Schorr:
Now, if we talk about walking though would someone first think, well, that could be a movement disorder?

Dr. Burke:
It can be confused with Parkinsonian difficulties walking. Those patients also shuffle their feet and don't lift them, and as people get older they lose the ability to rotate the pelvis as they walk, which accounts for 50 percent of the length of each stride. So the steps become smaller. The French used to call it marché a petit pas, the walk of small steps, and it looks in part like an aging process. So it can be difficult to distinguish on the basis of just one item.
Effects on the Brain

Andrew Schorr:
Well, we'll come back to testing and how you decide which it is and someone getting an accurate diagnosis, but let's back up and have a little science class for a minute. What is normal pressure hydrocephalus? Tell us about the brain.

Dr. Burke:
The brain has a series of membranes around it in which it contains not only the brain and spinal cord, and that is one contiguous space, brain and spinal cord surrounded by these membranes, but the brain and spinal cord are bathed in a brain spinal cerebrospinal fluid called CSF, and there is a process through which the brain makes and reabsorbs CSF, and there is a disruption of the ability to do this on the surface of the brain in patients with normal pressure hydrocephalus. So they do not have increased pressure in the head, but there is increased fluid there so that the cavities through which the CSF travels, and they are called ventricles, increase in size even though the brain is not shrunken as much as one might expect for someone with these cavities enlarged.

Andrew Schorr:
So we think of this fluid now causing a problem. What causes that? Where does this come from? I know it's typically in people let's say over 60, so maybe it's part of aging. Help us understand the causes.

Dr. Burke:
There is a reabsorption of the brain spinal fluid, the CSF, on the surface of the brain. It's a little controversial about how it occurs. It can occur through structures called arachnoid villi. The arachnoid is one of the thin, as it sounds like, spidery layer of membrane which has the structures that would reabsorb spinal fluid. Or the CSF may be reabsorbed through the pulsatility of the brain. The brain actually expands as the heart contracts and then relaxes as the heart does and goes back to a smaller size. When it expands it opens some small capillaries and veins, and CSF may be reabsorbed there. Right now there is a little controversy. But whichever one is right, the process of reabsorption of the CSF is disrupted on the surface of the brain so that there is increase in the amount of spinal fluid present. It is being made at a normal rate, but the reabsorption of it pushing it back into the blood is decreased so that the water builds up, the CSF builds up in the head and spine and puts pressure on the brain, which the brain then can't tolerate because the cavities on the inside where the CSF starts are expanding, but the CSF can't be reabsorbed, and the volume is not kept normal.

Causes

Andrew Schorr:
So do we have any idea what causes this in people? Is it like you get a head injury or some prior disease or is it just you are getting older?
**Dr. Burke:**
There are two known identifiable causes, but for the vast majority of patients we don't know why this happens. And that is head trauma causing bleeding under that very thin, spidery membrane, the arachnoid, subarachnoid blood so there has to be not a little bump on the head, but a significant amount of head trauma, or a nontraumatic hemorrhage there such as would occur after an aneurysm bleeds. So this can be a late consequence of aneurysmal hemorrhage in the head, and that was actually how it was first described.

**Andrew Schorr:**
What about meningitis?

**Dr. Burke:**
Meningitis is another infection, will cause the meninges or the membranes around the brain, and from the inflammation and infection there can be scarring of the surface of the brain so the reabsorption of the CSF is impaired.

**Diagnosis**

**Andrew Schorr:**
All right. Now, let's go back to somebody out there listening and wondering do they have that problem walking, or in mom or dad. How is this usually discovered, and how do you rule out these other conditions and then know that you are working with this normal pressure hydrocephalus?

**Dr. Burke:**
In patients who have an identifiable predisposing cause such as subarachnoid hemorrhage or meningitis, that should be part of the history and it would tip off the physician early in the history taking as to what is causing the patient's trouble walking. For older patients more of a subacute process developing over weeks and usually months beginning with loss of ability to walk. The patient just seems to be walking more slowly. And then it can get to the point that the patient really can't get out of bed or can't stand up very easily without assistance. And then the other problems will start to develop shortly thereafter. Urinary urgency. The patient seems to have ten seconds between the time he or she thinks she has to urinate until he or she does. And then finally cognition just isn't as good. The patients usually have good recent memory, but in terms of putting big pictures together, figuring out what has to be done, they just are not as sharp as they once were.

**Andrew Schorr:**
So you get to the doctor and probably before long a neurologist, and whether through history or this constellation of symptoms you are starting to get to this diagnosis, but what tests are used to confirm that?
Dr. Burke:
In addition to a neurological exam and history, one usually proceeds to an MR of the brain which can be a little more sensitive than a CT which will show that the cavities in the brain, the ventricles containing the CSF, are enlarged more than one could explain by the degree the brain has shrunk for age. And there can be an additional measure performed of the stroke volume of the CSF in a connecting area between two of the ventricles, and that is a CSF stroke volume. As I mentioned, with each beat of the heart, the brain is going to be compressed as the pressure goes up in the head and the CSF is pushed from the earlier, the third ventricle, there are four, through a narrow channel called the aqueduct into the fourth, and one can focus the MR right in the aqueduct. And the CSF will flow forward with each beat of the heart, and an elevated CSF stroke volume, that is a great deal of CSF moving from the third to the fourth ventricle, indicates normal pressure hydrocephalus.

Andrew Schorr:
Dr. Burke, you are obviously an expert at this. Some people though may be seeing someone where it could be confused for something else. What are questions that a patient can ask to make sure that this is either being ruled out or being zeroed in on?

Dr. Burke:
If one thinks he has more than just memory problems, is to ask could this be normal pressure hydrocephalus, and a neurologist will then proceed to the next step. However most neurologists are going to order an imaging study for a patient who has problems thinking and walking. That would be almost automatic.

Available Treatments

Andrew Schorr:
All right. Let's carry it forward then, sir, and tell us what you can do to help this problem, and then we'll get to how do you decide whether to go ahead. So first of all what are the available treatments at Northwestern for this condition?

Dr. Burke:
The treatments are available universally, and the first one is part of the diagnostic test, and that is a spinal tap, a lumbar puncture with the removal of 40 cc, that's about one and one-third ounces of CSF. The CSF can be tested for other problems such as infection or cancer, but above all, removing that much spinal fluid should effect a rapid improvement in the patient's ability to walk. It should not be an ambiguous change. In other words I think all of us would like to think whatever we do for our patients is helping the patient, and patients want to feel that whatever the doctor is doing is helping. So a patient who says I may be a little better in my walking does not have an objective improvement. There should be a rapid improvement in the patient's ability to walk either by the time he leaves the office, but certainly by the next day. That should last for a few weeks. That is the best test for the definitive treatment for normal pressure hydrocephalus.
Should one still think that a patient has normal pressure hydrocephalus but has not responded to a lumbar puncture, a five-day lumbar puncture can be performed. It's called a lumbar drain. The neurosurgeon places a small needle in the spinal canal and collects spinal fluid, and now I am talking not just collecting one and a third ounces. I am talking about 10 to 20 ounces of spinal fluid over several days.

Andrew Schorr:
Wow.

Dr. Burke:
The patient must respond then to be diagnosed with normal pressure hydrocephalus. And the lumbar drain is more invasive than a spinal tap. The risk of meningitis from a single spinal tap is remote, beyond one in 10,000, beyond one in 50,000. With a lumbar drain because the needle is communicating with the outside world and the subarachnoid space, there is single digit risk of meningitis which could be serious or potentially fatal.

If that doesn't work, it is safe to say that surgery is not going to help that patient. If a lumbar puncture works, the patient would then have a recommendation from his neurologist to have surgery.

Andrew Schorr:
Okay. Take us through what the surgery is.

Dr. Burke:
The surgery is the placement of a permanent drain from the brain to the abdomen. And it is not a cosmetically disfiguring procedure. A small hole is drilled through the skull, and a catheter called a shunt is placed in the ventricle of the brain usually on the right side in one of the lateral ventricles. The first two ventricles aren't numbered. It's just right and left, and we usually place it on the right side because language is usually on the left side of the brain. If there is a complication, we do not want to impair a patient's ability to speak or understand speech. It is then run under the skin, behind the ear, down the neck, under the skin of the chest, and put into the abdominal cavity so that there is in effect a permanent spinal tap going on draining fluid from the brain into the abdomen correcting the problem of the inability of the brain to reabsorb its own CSF.

Andrew Schorr:
What are the risks and potential complications from this procedure?

Dr. Burke:
Here we are dealing with a risk somewhere between five and ten percent of complications which unfortunately can be serious and even fatal. First, putting in the catheter can lead to bleeding. One can strike a vessel inserting it. In addition as the pressure comes down, the brain shrinks, and some of the vessels bridging from the outer membranes around brain to the surface of the brain can tear, so
there can be serious even fatal hemorrhage. There can be two serious infections, and those are meningitis in the membranes around the brain and spine, or an abdominal infection, peritonitis in the cavity that lines the entire abdomen, and again that's a serious, potentially fatal infection.

Andrew Schorr:
Dr. Burke, so how do you decide when surgery would be recommended, offered to a patient? Is it based on age? Is it based on the response they had to the lumbar puncture? Take us through that thinking.

Dr. Burke:
I would not consider age alone a reason to eliminate surgery as a possibility. Certainly as all of us lose brain cells as we get older and the brains tend to shrink with increasing age, there would be a greater risk of bleeding from the reduction in pressure of a shunt. But if someone had shown marked improvement after a lumbar puncture at any age I would consider shunting if I thought the patient could return to a higher level of functioning and independence from the procedure. The patient must respond to a lumbar puncture or a lumbar drain dramatically as I mentioned.

And then I would recommend surgery having informed the patient or the family or both of the risks involved. In patients who have a history of subarachnoid hemorrhage, bleeding inside the membranes, or meningitis, the response rate is extremely high. The response rate is going to be 80 to 90 percent if not more. And those patients I would recommend it most strongly. Among the others I would say that the response rate is going to exceed 50 and probably approach 70 percent, and we are dealing with an otherwise untreatable condition which is going to leave someone bedridden, incontinent, and demented so that the other options, and there are no medications that will help this condition, are nil.

Andrew Schorr:
All right. So that leads to some questions about Northwestern. How experienced is your team with the procedures, all the way from lumbar puncture to neurosurgery approaches on the brain in doing this so that hopefully the risk of complications can be kept at bay?

Dr. Burke:
One of the neurosurgeons and I, Dr. Bernard Bendok, in the past year have followed about ten patients, eight of whom went to shunting without complications. He inserts a programmable shunt with a computerized magnet in it that he can adjust the rate and the degree of pressure at which it will drain fluid from the brain so that if a patient needs more drainage, he can adjust that in his clinic. If a patient needs less, he can adjust that. And of the ten we followed, eight went to surgery without complications.
Two of them did not. One, we chose not to do surgery because the gentleman never improved following lumbar puncture with removal of 40 cc of spinal fluid and then a lumbar drain. If one were to look at his case, he certainly had all the hallmarks of a normal pressure hydrocephalus.

The other patient was a younger patient with significant head trauma who despite having stroke volumes of CSF that were very high was getting better on her own. And I think that's critical. The clinical pattern the patient displays is most important. This patient was getting better. Her walking was getting better. She was taking up jogging. She was running five miles a day. She was not incontinent, and despite severe head trauma, her thinking was improving. So we have simply watched her and for reasons neither of us understands her CSF stroke volume fell from markedly elevated in a range which would suggest a shunt would help to normal. And we simply don't know why. I think it demonstrates that the brain is more complicated than we really understand.

**Andrew Schorr:**
Dr. Burke, in listening to you and discussing in the beginning how this condition can be confused sometimes with others, is it possible to have this condition, normal pressure hydrocephalus, but also have the beginnings of some of these other things like Alzheimer's?

**Dr. Burke:**
And that's a real problem. Yes. Absolutely. And the longer people live, the greater the potential for having more than one illness. When modern academic medicine started really in Baltimore with Dr. William Osler over a century ago, he used to talk about the unifying diagnosis, by which he meant if a patient had five symptoms, and a physician were giving that patient five different diagnoses, he had probably overlooked the one which would explain all five. I would point out that when Dr. Osler wrote that he was dealing with a country in which the median life expectancy was under the age of 50. So I think it's true with younger patients. I think particularly with older patients there can be multiple conditions, even multiple neurologic conditions going on.

**Research and Clinical Trials**

**Andrew Schorr:**
And that's exactly what I was wondering with the aging of the population. So you have research going on. Help us understand where you and other experts are hoping to take this. What's going on in clinical trials?

**Dr. Burke:**
The major issues that we have that are of interest, one, can we use the CSF stroke volume more? And two, there are ways of diagnosing the other conditions that might be helpful in differentiating normal pressure hydrocephalus from Alzheimer's disease. There is a PET scanning technique which is beginning to become commercially available, which shows amyloid plaque in the brain, but University of
Pittsburgh has been developing this over the past year or two, but I do not know whether it's commercially available yet. There is MR spectroscopy which may have ways to look for other conditions in the brain such as Parkinson's. And PET scanning again has a potential role there.

And I don't see why a patient with preexisting Alzheimer's disease or preexisting stroke could not develop normal pressure hydrocephalus later on, not as a consequence of the first disease, but coincident with it. And then I think the issue would be if the patient were severely demented to have an appropriate surrogate making the informed consent about what to do, and it would be the role of the physician to explain what improvement could be expected from shunting a patient.

**Andrew Schorr:**
Yeah. There are lots of questions as we have these multiple conditions, you have had past ones. So going forward, and certainly it's so troubling if a family listening to this, these symptoms are there. How would you recommend they get to whether your own department or get to somebody who is knowledgeable in this because obviously it's a very specialized condition?

**Dr. Burke:**
It is specialized, although I think it's one neurologists like to see. There are few conditions so dramatically responsive to treatment as normal pressure hydrocephalus, so that neurologists have been aware of it for 40 years, and I would say seeing a neurologist if there were questions would be the key. It went through a phase where we shunted people I think excessively along the idea that well, there is a small chance we can help them, we have nothing else we can treat. I think things are different now with treatments for many of the other disorders which can present similarly, and therefore it's important that we be more focused and truly discrete in how we approach the disease. I would think though this is one of the conditions which neurologists hold close to their hearts because it's so dramatically treatable.

**Andrew Schorr:**
Well, that is good news for patients and for family members, and then of course the decisions along the way. First, what are you dealing with? Get the accurate diagnosis, the tests, the different procedures that you say can make a dramatic difference and decide how far you have to go with it. For instance do you go to a surgery with a shunt.

Dr. Allan Burke, thank you so much for sharing your expertise in this condition. I know I have learned a lot, and I imagine that families that are concerned where this could be the diagnosis, they have learned a tremendous amount.
Of course for anyone listening, if you want to get more information or connect with Dr. Burke and the other experts at Northwestern, you can find it all on nmh.org, the website. And this is what we do every two weeks with Northwestern in the ihealth section of the website is connect you with leading experts like Dr. Allan Burke. Dr. Burke, thank you so much for being with us, sir.

**Dr. Burke:**
You're quite welcome. Thank you for having me.

**Andrew Schorr:**
Thank you. Well, this is Patient Power, and we like to say knowledge can be the best medicine of all, and I am sure it's helping people with this condition who are concerned about normal pressure hydrocephalus.

Thank you for joining us. We will be back in two weeks with connection to another Northwestern Memorial Hospital. You have been listening to Patient Power sponsored by Northwestern Memorial Hospital. Thanks for joining us.

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