

## Normal pressure hydrocephalus: a case report by a physician who is the patient

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**ABSTRACT** – This report describes the case of an elderly physician who endured a slowly progressive, ambulatory illness, which was erroneously diagnosed as Parkinson’s disease. After ten years of progressive illness the correct diagnosis of normal pressure hydrocephalus (NPH) was finally made, revealing itself, by accident, through incontinence and mild dementia. The patient–physician enjoyed an instantaneous remission induced by a large lumbar puncture (LP) sustained by a ventriculosystemic shunt. The patient-physician dedicated his renewed life to informing the medical profession about this dramatic syndrome, which he believes is more common and more reversible than generally thought. Although the patient had been virtually restored to normal, a series of complications typical of ventriculosystemic shunting (VSS) occurred, including significant hearing loss and subdural haematoma (SDH). The patient feels, however, that his clinical improvement far outweighs the complications and that every patient with NPH should have the opportunity to decide whether or not to have a VSS.

**KEY WORDS:** apraxia, cerebral atrophy, cerebrospinal fluid, intracranial pressure, normal pressure hydrocephalus, subdural haematoma, ventriculosystemic shunting

### The normal pressure hydrocephalus syndrome

Normal pressure hydrocephalus (NPH), a disease which was first described in 1965 by Hakim *et al.*<sup>1–3</sup> is characterised by a triad of symptoms – abnormal gait, incontinence and dementia – and by dilatation of the cerebral ventricles. It almost always occurs in patients who are over 65 years of age. The gait disturbance is usually the initial and predominant symptom. Patients with NPH shuffle and walk slowly with a broad-based, clumsy and irregular gait and take short steps of low step height. Their posture tends to resemble that of patients with Parkinson’s disease (PD), ie stooped with their necks craned forward. Despite the absence of a tremor or muscle rigidity, patients with NPH are often misdiagnosed as having PD. The incontinence is usually urinary, but may be

faecal as well. It often begins as polyuria or urgency rather than true incontinence. The dementia varies. It tends to be characterised by the loss of short-term memory, a lack of spontaneity, delayed responsiveness and apathy. The patients tend to be depressed and also occasionally exhibit hostility or paranoid behaviour.<sup>4</sup> Each of the symptoms tends to worsen gradually over a period of months or years and usually improves after ventriculosystemic shunting (VSS).

The ventriculomegaly, which is the *sine qua non* of the syndrome, can be demonstrated by computerised tomography (CT) or magnetic resonance imaging (MRI).<sup>5</sup> Although the entire ventricular system is enlarged, rounding of the frontal and temporal horns of the lateral ventricles is almost always present. The ventricular enlargement appears to be out of proportion to the sulcal dilatation, which is suggestive of cerebral atrophy, and which is also frequently seen in elderly patients.

Two types of NPH have been recognised – secondary (SNPH) and idiopathic (INPH).<sup>6</sup> Subarachnoid haemorrhage, brain surgery, cranial trauma, other types of intracranial bleeding or meningitis may lead to SHPH. Normal pressure hydrocephalus in patients without such precipitating disorders is defined as idiopathic. This type of NPH occurs in older patients with more severe symptoms, and is thought to be less responsive to VSS than those with SNPH.

The intracranial pressure in NPH is normal (<200 ml water<sup>7</sup>), but patients with slight elevations have been accepted as NPH, particularly when prolonged, nocturnal measurements are made.<sup>8</sup>

The pathogenesis of the NPH syndrome is not well understood, but it is hypothesised by Bateman that diminished resorption of cerebrospinal fluid (CSF) results in increments in the volume of CSF, in intracranial pressure (ICP) and in transvenular resistance in the region of the superior sagittal sinus, which result in decreased cerebral blood flow,<sup>9</sup> thus initiating dilatation of the cerebral ventricles. The increased pressure is exerted on the enlarged ventricular surface, which, in accord with Pascal’s principle, helps compensate for the increase in CSF volume and pressure, and permits minimal elevation of the intracranial pressure.<sup>2</sup> Holodny has reported several patients with NPH with sulcal dilatation, which

paradoxically decreases after successful VSS.<sup>10</sup> These observations indicate that sulcal dilation is not necessarily a sign of cerebral atrophy.

The diagnosis of NPH may be confirmed by the removal of a relatively large volume of CSF (50–60 ml), after which patients often exhibit prompt improvement.<sup>11</sup> Such a transient remission is usually predictive of a favourable response to the performance of a VSS. Such anastomoses, which are the standard treatment of NPH, divert CSF from the cerebral ventricles, to the peritoneal cavity, from which it is absorbed.

The large majority of patients with NPH who are shunted show impressive clinical improvement.<sup>11–13</sup> When crude indices of ventricular volume, such as Evan's index<sup>14</sup> are used, the ventricular volume does not always decrease after VSS.<sup>15</sup> When more sophisticated measurements, such as computerised nuclear magnetic resonance volumetric analysis, are used, a large majority of patients show a decrease in ventricular volume, although the decrease is often quite small.<sup>16</sup> Surprisingly, patients with sulcal dilatation, which is usually considered a contraindication to VSS, have often benefited from VSS.<sup>17</sup>

Shunt therapy is frequently followed by serious complications, such as subdural hematoma (SDH)<sup>18</sup> or infections<sup>19</sup> and shunt malfunctions, which require readjustment of the shunt valve pressure setting.

### Case history (patient's comments are italicised)

When his neurologic disorder began in early 1993 the patient was a healthy, 68-year-old squash- and bridge-playing physician who had recently retired from Yale University School of Medicine where he had studied, taught and practised hepatology for over 50 years. He had been actively seeing patients, teaching, lecturing and writing articles about his research in portal hypertension until his retirement. Initially, his chief complaint was slowness of walking, and later clumsiness, which gradually worsened until 2002, by which time he was unable to walk without using a walking frame or leaning on walls.

*I became aware that my gait was abnormal when a physician friend told me that I walked like a patient with PD.*

The clumsiness progressed until June 1997 when the patient consulted a senior neurologist at Yale who diagnosed an apraxia, an inability to walk normally in the absence of motor or sensory impairment. The patient's physical examination at the onset of his illness was normal except for mild hypertension (140/90 mmHg) and moderate obesity (height 5'9", weight 230 pounds). He had atrial fibrillation for which he had been anticoagulated for 20 years. Magnetic resonance imaging (MRI) showed 'prominence of the ventricles and sulci consistent with age-appropriate atrophy'. It was suggested that he might have had prior, silent 'cerebrovascular ischemic disease'. The patient was given a therapeutic trial of levodopa without clinical benefit. His hypertension was treated with beta-adrenergic blockade, which reduced the blood pressure to normal, where it has remained. A second senior neurologist on an independent examination concluded that the patient had PD, despite the absence of tremor or rigidity.

In 1998, the patient was seen by the first neurologist with no change in his neurologic examination or diagnosis. The patient, who played squash once a week, noticed that his gait had worsened, and that he was moving more slowly and less smoothly. He reluctantly stopped playing. By 1999 he began to spend winters in Florida where he had been appointed a consultant to the Liver Transplantation Division at the University of Miami (UM). He was referred by the Yale neurologist to the chairman of the Department of Neurology at the UM. The patient complained of stumbling, awkward gait and altered handwriting. The new consultant confirmed the PD-like gait and prescribed carbodopa, which had no effect. A second MRI showed 'slight enlargement of the lateral ventricles and sulci'. The new consultant accepted the referring physician's diagnosis.

*I believe that 'second opinions' are sometimes pro forma exercises in which senior consultants confirm the referring physician's opinion rather than introduce a new diagnosis in the absence of definitive new findings. The only significant difference in interpretations of the two MRIs was that the ventricles were described as being 'enlarged' rather than 'prominent'. The word hydrocephalus had not been used.*

Over the next two years the gait abnormality continued to deteriorate. The patient noted that it was sometimes difficult to initiate walking, and that his feet sometimes seemed stuck to the floor. In early 2002 the patient reported to the first Yale neurologist that he needed to urinate urgently and frequently. He was referred to a urologist who found no genitourinary abnormalities and introduced Detrol, which had no effect. In Florida in December the patient experienced several episodes of faecal incontinence for which he was referred to a gastroenterologist who thought they were related to 'old age and weakening of the anal sphincter'.

*The urinary symptoms represented the first clue to the diagnosis of NPH. Prior to the appearance of the urinary signs the patient had exhibited only a gait abnormality. Senile gaits are a common, diagnostically difficult problem in elderly patients.<sup>20</sup> Incontinence, together with difficulty walking comprise two of the three clinical symptoms of NPH, which should have been considered at that time by the neurologist, the urologist and the gastroenterologist.*

Almost five years after the Yale consultant had first examined the patient he saw the patient again. He found no significant changes from his initial neurologic exam except that the gait had worsened. The patient walked unsteadily, often leaning on walls or furniture for support. A third MRI in May 2002 showed 'diffuse enlargement of the ventricles and sulci consistent with volume loss'. Ventricular size had not changed.

*Neuroradiologists are reluctant to use the term 'hydrocephalus', which is more alarming than 'prominent or slightly enlarged ventricles'. It may induce physicians to undertake expensive diagnostic studies or shunt surgery in elderly people who they consider to be suboptimal surgical risks. Furthermore, NPH is a disease that is still emerging from doubts about its existence and reversibility.<sup>21</sup>*

After he had seen the MRI, the neurologist told the patient that 'cerebral atrophy' had advanced. He predicted that it would

probably progress until the patient was unable to walk at all, and that he would probably also lose cognitive function. Furthermore, he said that there was no effective treatment. The patient and his wife interpreted this progress report to mean that they should put their affairs in order.

*We were so depressed by this devastating prognosis that we cancelled our scheduled 50th wedding anniversary celebration and warned our family and friends of impending disaster.*

At about the same time it was discovered that the patient had type 2 diabetes mellitus, which both his parents and his three siblings had also had. The Director of the Diabetes Institute at UM became his attending physician. Despite his dismal prognosis the patient volunteered to participate in an investigation of a new drug for PD. The principal investigator, the fourth neurologist to see him, enrolled him in a clinical trial in which he was selected to receive an active experimental agent. The only effect of the drug was to induce somnolence.

Late in 2002 the patient's gait abruptly deteriorated until he lurched from place to place. He continued to drive his car because he felt that his driving seemed unimpaired.

*CM Fisher and others have noted that patients with NPH are able to move their legs normally while seated or reclining, but not while erect.<sup>22</sup> This bizarre phenomenon has been confirmed, but not explained.*

Walking, however, had become so slow and clumsy that the patient resigned his consultancy and he requested approval from his neurologist for a motorised wheelchair. The patient tried to continue as social director of his condominium, but was embarrassed that his colleagues might smell the odours of excrement, which he assumed surrounded him. The patient also realised that his bridge playing had deteriorated. He couldn't remember the number of trumps that had been played, an essential component of playing good bridge. One's inability to keep track of a number that varies from minute to minute may be the ultimate example of short-term memory loss.

*In frustration after playing particularly poorly one evening I stopped playing bridge. Whatever my illness was, it was destroying my life. I had given up squash and bridge, I had quit my job and I couldn't walk. I had reached the nadir of my life and felt that it could only get worse. For the first time I considered suicide. I decided to start accumulating sedatives and narcotics which might enable me to escape from this dreadful state quietly and with dignity. I was not even aware that my gait, incontinence and memory loss were all part of the same disease.*

The Yale neurologist did not approve the patient's request for an electric wheelchair. The patient showed the letter to his endocrinologist who was so incensed that he immediately referred the patient to a new, younger neurologist, who, he said '... will sign your application for the wheelchair and at the same time give you a "second opinion"'.

*The new consultant, the fifth, examined me and my MRIs and told me that I did not have PD, but that I had NPH. I had never heard of NPH. Ironically, the refusal of the wheelchair led directly to the correct diagnosis. I learned that NPH, unlike PD, is often reversible. He pointed out that a 'large' LP, of 50–60 ml, often*

*promptly induces a transient remission, which confirms the diagnosis of NPH, and indicates that the insertion of a VSS will probably be successful therapeutically. A neurosurgeon at Mount Sinai Medical Center in Miami Beach, FL, who had successfully shunted many patients with NPH was selected to perform the shunt.*

The neurosurgeon, who is one of relatively few board-qualified female neurosurgeons, concurred with the diagnosis of NPH. Five days after the patient's anticoagulation had been discontinued, she performed an LP.

*Although I had had difficulty climbing onto the trolley before the LP, once 60 ml had been drained I was able to walk. I am still amazed that such an instantaneous change could possibly have occurred. My spontaneity, my alertness and my sense of humour all seemed to have returned. It was an incredible experience.*

On 22 April 2003, the day of surgery, the remission induced by the LP had subsided. The shunting procedure was an anticlimax. The patient, who was known to fear hospitalisation, was an unhappy, uncooperative postoperative patient. When discharged from the hospital, however, he walked normally and his spontaneity, affect, mental acuity, and joie de vivre had all returned and have persisted since.

Within a few days of surgery the patient had abandoned his long-time career in hepatology, and dedicated his life to publicising NPH. This almost miraculous recovery has persisted for over four years, although not complication free. The patient's shunt pressure gradient was reduced several months after implantation following a recurrence of gait and urinary symptoms.

*On the following morning I realised that I had suffered significant hearing loss, which was confirmed by audiometry. This previously reported complication,<sup>23</sup> which is usually transient, appears to be permanent in my case.*

Seven months later, a routine computerised tomography scan revealed large, bilateral subdural haematomas (SDH) that were undergoing resolution. He could recall no trauma. His anticoagulation (for atrial fibrillation) was discontinued and his CSF pressure gradient was increased. The SDH, which had been completely asymptomatic, resolved over three months of decreased anticoagulation and of CSF pressure gradient.

*Several months later I recalled a minor fall in the bathroom that was clearly responsible for the SDH. I had underestimated the force involved in coup and contracoup.*

The long delay in making the diagnosis of NPH and the frequent difficulty differentiating NPH from other disorders such as PD, Alzheimer's, cerebral atrophy, etc are consequences of the relative rarity of NPH and the failure of family physicians, neurologists, urologists, gastroenterologists, rheumatologists and psychiatrists to consider NPH in patients who present with any one of the three components of NPH.

*In my view, the final decision about whether or not shunt therapy should be undertaken at any stage of NPH should be made by informed patients and their families rather than by their physicians.*

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