

## Normal Pressure Hydrocephalus; Outcome after Ventriculoperitoneal Shunt

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### ABSTRACT

**Background:** Normal pressure hydrocephalus (NPH) is a treatable cause of dementia. Hakim and Adams described for the first time the symptoms and signs of NPH which include ataxia, progressive memory loss and urinary incontinence. In properly diagnosed cases, ventriculoperitoneal (VP) shunt leads to good outcome.

**Material and Methods:** This study comprised of 40 patients with NPH. The duration of study was 5 years from January 2009 to December 2013. The duration of illness before presentation ranged from one year to 6 years. The patients were diagnosed clinically and CT scan brain was done in all patients (MRI in some patients). All patients underwent Folstein Mini-Mental State Examination. In cases of possible NPH, drainage lumbar puncture (LP) was done and their response was noted. In 35 patients whose families gave consent for surgery, VP shunt was done while families of 5 patients refused surgery. The patients who underwent VP shunt were followed up, which ranged from one to 5 years.

**Results:** The age of the patients was between 52 and 70 years. The majority of patients (25) were between 61-70 years. Out of the 35 patients, 26 (75%) were male while 9 (25%) were female. The initial symptom in all these patients was gait ataxia and the duration of illness ranged from one year to 6 years.

**Conclusion:** Normal pressure hydrocephalus (NPH) is a treatable cause of dementia. In properly diagnosed patients, ventriculoperitoneal shunt gives good results.

**Key words:** Normal pressure hydrocephalus, dementia, ventriculoperitoneal shunt.

**Abbreviations:** NPH = Normal pressure hydrocephalus. VP = Ventriculoperitoneal. iNPH = Idiopathic Normal pressure hydrocephalus. CSF = Cerebrospinal fluid.

### INTRODUCTION

Normal pressure hydrocephalus (NPH) accounts for up to 6% cases of dementia. Hakim and Adams for the first time described the symptoms and signs of NPH which include ataxia, progressive memory loss and urinary incontinence. NPH can be classified as primary or idiopathic (iNPH) and secondary. In primary or idiopathic NPH, no definitive cause can be identified while secondary NPH has some underlying cause, like subarachnoid hemorrhage, head injury or meningitis. The mainstay of treatment of NPH is ventriculoperitoneal (VP) shunt. As it is difficult to differentiate iNPH from other forms of dementia, the key to successful treatment is its accurate diagnosis.<sup>1-4</sup>

### MATERIAL AND METHODS

Forty patients with NPH were included in the study. The duration of illness ranged from one year to 6 years. In all patients, the initial symptom was ataxia. The patients were diagnosed clinically and CT scan brain was done in all patients (MRI in some patients). All patients underwent Folstein Mini-Mental State Examination. All patients were given medical treatment (L-dopa) for gait ataxia and showed partial response. All patients were offered VP shunt but families of 5 patients refused surgery. In cases of possible NPH, therapeutic lumbar tap was done. In 35 patients whose families gave consent for surgery, VP shunt was done. These patients were followed for improvement, which ranged from one to 5 years.

**RESULTS**

The age of the patients was between 52 and 70 years. Thirty patients (75%) were between 61-70 years. Out of 40 patients, 30 (75%) were male while 10 (25%) were female. The initial symptom in these patients was ataxia and the duration of illness ranged from one year to 6 years. In the majority of patients, the duration of illness was over 2 years while functional status was markedly affected at the time of presentation (tables 1 – 3). As far as CT scan findings are concerned, most of the patients (85%) had ventriculomegaly along with atrophic changes and infarcts while only 15% had isolated ventricular dilatation (table 4). In patients with borderline features, therapeutic lumbar tap was done and it clearly helped (in over two-third patients) in

**Table 1: Duration of Dementia.**

Duration	No. of Patients	Percentage
3 months	4	10
6 months	2	5
1 year	4	10
2 years	16	40
2.5 years	8	20
Not present	6	15

**Table 2: Urinary Incontinence.**

Duration	No. of Patients	Percentage
6 months	8	20
1 year	8	20
1.5 year	18	45
Not present	6	15

**Table 3: Functional Status.**

Functional Status	No. of Patients	Percentage
Ambulatory	10	25
Ambulatory with walker	2	5
Ambulatory with 2 persons	10	25
Wheel chair – bound	12	30
Bed – bound	6	15

**Table 4: CT Scan Findings.**

CT Scan Findings	No. of Patients	Percentage
Ventricular dilatation with minimal cortical atrophy	22	55
Ventricular dilatation with minimal cortical atrophy with infarcts	12	30
Ventricular dilatation without cortical atrophy	6	15

**Table 5: Response to Therapeutic Tap (20 Patients).**

No. of Patients	Percentage	Response
4	20	Significant response
12	60	Partial response
4	20	No response

**Table 6: Outcome after VP Shunt (35 Patients).**

No. of Patients	Percentage	Outcome
21	60	Significant response
3	8.6	Partial response
3	8.6	Mild response
3	8.6	No significant response
5	14.2	Not known

decision making for shunt surgery (table 5). In patients who underwent VP shunt (35), 60% showed significant improvement while another 8% showed partial response (table 6). As far as shunt complications are concerned, one patient (3%) developed large, bilateral, acute subdural hematomas in early postoperative period, was operated for these but did not recover and died. One patient (3%) developed shunt infection and the shunt had to be removed. There were no cases of shunt obstruction / revision.

**DISCUSSION**

Normal pressure hydrocephalus (NPH) is a cause of dementia which can be treated. It accounts for up to 6% cases of dementia. Hakim and Adams described the symptoms and signs of NPH for the first time in

1964. These include ataxia, progressive memory loss and urinary incontinence.<sup>1-4</sup>

NPH can be classified as primary or idiopathic (iNPH) and secondary. In primary or idiopathic NPH, no definitive cause can be identified while secondary NPH has some underlying cause, like subarachnoid hemorrhage, head injury or meningitis. In idiopathic NPH, the cerebrospinal fluid (CSF) pressure is normal while it is moderately raised in cases of secondary NPH. Secondary hydrocephalus can present at any age while iNPH usually presents in the 6<sup>th</sup> or 7<sup>th</sup> decade of life. The mainstay of treatment of NPH is ventriculoperitoneal (VP) shunt. As it is difficult to differentiate iNPH from other forms of dementia, the key to successful treatment is its accurate diagnosis. Moreover, the disturbance of gait in NPH contributes to the morbidity associated with frequent falls in these patients.<sup>1-5</sup>

In a study by Tarnaris et al., all patients had ataxia along with dementia with/without urinary incontinence. CT or MRI was done in all patients which showed communicating hydrocephalus and an Evans index (maximum frontal horn width divided by the transverse inner diameter of the skull) more than 0.3. In another study, gait disturbance was present in all patients, dementia in 50.6%, and urinary incontinence in 69.8% patients. 90% of these patients improved after VP shunt (“responders”), while 10% did not (“non-responders”). Among the patients who responded to VP shunt, 42.5% had the complete triad of progressive dementia, ataxia and urinary incontinence while 57.5% had an incomplete triad; 25% had gait disturbance alone, 36% had gait disturbance and urinary incontinence, and 9% showed gait disturbance and dementia.<sup>4,6,9,10</sup>

In our study, all patients presented with ataxia while dementia and incontinence was not present in some patients (15% each) (table 1, 2).

Kim et al, conducted a large cine MRI study and noted that patients with NPH had a net flow of CSF from fourth ventricle into the third (opposite to that found in healthy people). Moreover, this “retrograde” net flow reversed to normal “anterograde” flow after VP shunt. This observation has important implications for the understanding of fluid dynamics of hydrocephalus and the effect of CSF shunting. It means that in hydrocephalus, there is transependymal absorption of CSF into the brain. These findings have been confirmed by other studies as well.<sup>5</sup>

The diagnosis of NPH demands careful consideration in the elderly, as it is a reversible cause of dementia if treated early. 70 – 80% of patients with typical clinical picture and positive CT findings would im-

rove significantly with VP shunt.<sup>7,8</sup>

In 2004, the Japanese Society of Normal – Pressure Hydrocephalus proposed clinical guidelines for diagnosis and treatment of idiopathic NPH. According to these guidelines, patients who have one or more symptoms of NPH with ventricular dilatation are defined as having “possible idiopathic NPH”, regardless of coexisting pathological features of Alzheimer’s disease. Those patients whose spinal tap test is positive, are classified as having “probable NPH”; and in those whose symptoms improve after CSF shunting, the disease is classified as “definite idiopathic NPH”. The most sensitive test in predicting a favorable outcome is external lumbar drainage of CSF and assessment of the clinical improvement of the patient. Furthermore, narrow CSF spaces in the high convexity on the coronal view of the MRI is the most important finding for suspected idiopathic NPH, even if the patient has atrophic hippocampus.<sup>1,5,9,10</sup>

Grading system for NPH established by the Research Committee on Intractable Hydrocephalus of the Ministry of Health and Welfare of Japan:<sup>10</sup>

Grade	Definition
<b>Gait Disturbance</b>	
0	Normal.
1	Unsteady but independent walking possible.
2	Unsteady but independent walking possible with 1 cane.
3	Unable to walk without 2 canes or a walker frame.
4	Unable to walk even with assistance.
<b>Dementia</b>	
0	Normal.
1	No apparent dementia, testing required to demonstrate deficit.
2	not disoriented but attention-disturbed, socially dependent, but independent at home.
3	Disoriented, partially dependent at home.
4	Disoriented, totally dependent.
<b>Urinary incontinence</b>	
0	Absent.
1	Absent but with increased frequency or urgency.
2	Sometimes, less than several times a week.
3	Frequent, at least once a day.
4	Total incontinence.

It is difficult to manage iNPH because establishing the diagnosis can be difficult and shunt surgery can have some complications. Patients who meet the criteria for probable NPH, a high – volume tap test (“30/30 tap test”) is performed. It is found to be useful in 70%

of cases. 30 ml CSF is removed and gait is evaluated at approximately 30 minutes. Tap test has a positive predictive value of 73 – 100% but sensitivity is low (26-61%). It helps to decide for VP shunt and avoids the more invasive tests of external lumbar drainage or intraventricular pressure monitoring.<sup>1,2,11-14</sup>

In our study, the majority of patients were between 60 – 70 years and 75% patients were male while in another study, the median age of patients was 76 years, while 57% patients were male.<sup>1,2,11</sup>

The differential diagnosis of NPH includes all pre-senile and senile dementias, post-traumatic cerebral atrophy, and cerebral arteriosclerosis. The definitive diagnosis of NPH is a bit difficult. Evidence of slowly progressive ventricular enlargement on serial CT/MRI scans is diagnostic. In some instances, hydrocephalus and cerebral atrophy coexist. Various protocols are followed to reach the diagnosis of NPH, a temporary improvement in symptoms following spinal CSF drainage is probably the most reliable indication for CSF shunting.<sup>12</sup>

Potential complications of VP shunting for NPH include subdural hematomas or hygroma, shunt infection, intracerebral hemorrhage, seizures, shunt obstruction or disconnection. The most likely symptom to improve with shunting is incontinence, then gait disturbance, and lastly dementia.<sup>1</sup>

Finally, we feel that NPH is not so uncommon in our population. Even more important is the fact that there is significant delay in diagnosis of these patients, as 70% of our patients had very poor functional status at presentation to the neurophysician (table 3). It ultimately leads to a delay in definitive treatment (VP shunt), and unsatisfactory/poor outcome. So an awareness campaign is required to educate the general physicians so that these patients can benefit from timely diagnosis and required treatment.<sup>14</sup>

## CONCLUSION

Normal pressure hydrocephalus (NPH) is a treatable cause of dementia. In properly diagnosed patients, ventriculoperitoneal shunt gives good results.

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