Reversible long-standing severe disability in idiopathic normal pressure hydrocephalus

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Idiopathic normal pressure hydrocephalus (iNPH) presents with the triad of impaired gait, impaired cognition, and urinary incontinence, with enlarged ventricles and normal CSF pressure.¹ Reports of CSF shunting and outcome after prolonged severe disability are few. We report a patient who responded dramatically to shunting despite severe motor and cognitive impairment and incontinence for more than a year.

Case report

A 71-year-old man with long-standing obsessive-compulsive disorder (OCD) developed hypomania with elevated mood, insomnia, and grandiosity at age 68. After responding to quetiapine, escitalopram and galantamine were added for OCD and “mixed dementia.” The latter diagnosis appears to have been based on functional decline and ischemic changes on brain CT, although this was not confirmed on subsequent MRI. One year later, he developed mania and improved with valproic acid and olanzapine, which replaced quetiapine. Walking became progressively slower with multiple falls. Despite discontinuation of olanzapine, he became nonambulatory. A levodopa trial produced visual hallucinations. Over the following year, he developed forgetfulness, apathy, disinhibition, multitasking difficulties, poor judgment, and double incontinence. He had hypertension, diabetes, hypercholesterolemia, and coronary artery disease. There was no family history of psychiatric disorders or dementia.

On admission to the hospital, he was alert and anxious. Insight was poor for cognitive problems but fair for motor disability. Medications included donepezil and valproic acid. Mini-Mental State Examination (MMSE) score was 5/30. Examination showed slowed saccadic eye movements, cogwheel rigidity, bradykinesia, upper limb postural tremors, and profound

Practical Implications

Idiopathic normal pressure hydrocephalus patients with severe and prolonged disability may benefit from shunting if there is an objective response to tap or drainage tests.

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Table 1 Physiotherapy and motor speed assessments

Physiotherapy				
Functional area/test	Pre tap	<24 h post tap	1 mo post shunt	5 mo post shunt
Sitting balance time	None	Up to 3 min	Up to 5 min	Unlimited
Transfer bed to chair	Mechanical lift or assist × 2	Assist × 2	Assist × 1	Independent
Ambulation	Unable	Unable	20 m assist × 2 with walker	Independent: 300–400 m with rollator, 5–10 m without
BBS	Not tested	Not tested	14/56	41/56
COVS	Not tested	Not tested	39/91	69/96
Motor speed				
Test	Pre tap	<24 h post tap	6 wk post shunt	
R thumb-index tapping	7.8 (range 7–11)	13 (range 10–20)	40+	
L thumb-index tapping	2.3 (range 1–3)	11 (range 11–11)	40+	
R foot alternate plantar flexion/dorsiflexion	11.6 (range 7–14)	15.4 (range 9–20)	12 (range 11–13), 2 trials	
L foot alternate plantar flexion/dorsiflexion	9.3 (range 7–11)	13.6 (range 6–18)	13 (range 13–13), 2 trials	

Abbreviations: BBS = Berg Balance Scale; COVS = Clinical Outcome Variable Scale.

Physiotherapy: Pre tap, he was unable to sit unsupported and required a mechanical lift or maximal assistance of 2 for transfers. On the first day post tap test, there were some gains in sitting balance and transfers. During the following 2 months post shunting, there was profound improvement in mobility that reached a plateau. Following adjustment of the shunt from 1.5 to 1, more improvement occurred.

Motor speed: For thumb-index tapping, the number of taps between his index finger and thumb was counted until no gap was visible (mean of 5 trials is provided). For the foot alternate plantar flexion/dorsiflexion, the patient flexed his toes up and down from rest up to the examiner's hand as fast as possible for 15 seconds (mean of 5 trials is provided). He showed some pre/post tap test gains in tapping speed. Five months post shunting, he declined to participate in testing.

lower limb stiffness. He could not stand or sit unsupported. Deep tendon reflexes were grade 2 bilaterally. Plantar responses were flexor. Valproic acid was discontinued to rule out drug-induced parkinsonism. The patient did not improve.

Diagnosis and investigations

Head CTs performed 2 years apart and brain MRI showed increasing diffuse ventricular enlargement, periventricular transependymal exudates, and mild cortical atrophy. Brain SPECT showed mildly reduced right temporal perfusion. Lumbar tap test was performed. Thirty mL of CSF was removed with difficulty in sitting position. Flow was so slow that pressure could not be measured reliably. Cell count, protein, and glucose were normal. Pre/post tap test mobility, motor speed, and neuropsychology assessments are shown in tables 1 and 2. He could not complete formal cognitive testing due to severe cognitive impairment. Within 24 hours after lumbar tap, he showed major improvement in sitting balance, with easier transfers and gradual improvement in motor speed over the next 6 weeks. Diagnosis of probable iNPH was considered.²

Shunting

Right frontal Medtronic Strata II valve ventriculoperitoneal shunt set at 1.5 was inserted. Frontal biopsy showed normal brain. Ten weeks post shunt, he required less assistance with gross

Table 2 Neuropsychology assessment

Neuropsychological test	Pre tap	10 wk post shunt
D-KEFS Trails 5 (Motor Speed)	Time discontinued (<1st percentile)	Time discontinued (<1st percentile)
Grooved pegboard (R) (motor dexterity)	Discontinued due to frustration after 30 s: 1 drop, no pegs placed	5 pegs in 66 s
Grooved pegboard (L) (motor dexterity)	Client did not want to attempt	5 pegs in 62 s
WAIS-IV Digit symbol	Discontinued due to confusion	1st percentile
RBANS Attention index	—	1st percentile
HVLT-R 1 Immediate recall (12-item list)	Discontinued due to frustration; 0 words on 2 trials	<1st percentile
RBANS Immediate memory index	—	2nd percentile
HVLT-R 1 Delayed recall	—	5th percentile
HVLT-R 1 Delayed recognition discrimination	—	16th percentile
RBANS Delayed memory index	—	1st percentile
RBANS Language index	—	<1st percentile
RBANS Visuospatial/constructional index	—	23rd percentile

Abbreviations: D-KEFS = Delis-Kaplan Executive Function System; HVLT-R 1 = Hopkins Verbal Learning Test-Revised form 1; WAIS-IV = Wechsler Adult Intelligence Scale, fourth edition; RBANS = Repeatable Battery for the Assessment of Neuropsychological Status.

There was no major improvement in the first 24 hours after tap. Five months post shunting, the patient declined to participate in testing.

motor function and ambulation. Cognitively he was testable but still severely impaired. Cognitive profile and behavioral observations suggested frontal-subcortical deficits, with relative sparing of information retention and visuospatial functioning. After shunt level was adjusted to 1, further motor and cognitive improvement occurred.

Five-month postoperative assessment showed mild hypertonia and normal muscle power, sensation, and coordination. He walked independently with a device and showed steady improvement in gait speed, step length, and distance (video at Neurology.org/cp). Incontinence resolved. MMSE score was 27/30. Behavioural Neurology Assessment-Short Form³ score was 78/114, whereas he was not testable preoperatively (cutoff for dementia is 82). Mild hypomanic symptoms emerged and improved with lithium.

DISCUSSION

Our patient with severe motor, sphincter, and cognitive impairment for more than a year showed clear improvement after a tap test and then marked improvement after ventriculoperitoneal shunt insertion. This confirmed definite iNPH.² Drug-induced parkinsonism was ruled out by absence of improvement following olanzapine and valproic acid discontinuation. Parkinson disease and Alzheimer disease are unlikely due to absence of levodopa response, normal brain biopsy, prominent response to shunting, and neuroimaging results. MRI did not support vascular dementia. Although mania has been described in 2 symptomatic NPH cases, postmeningitic⁴ and posttraumatic,⁵ this iNPH case started with hypomania.

Poca et al.⁶ reported predictive factors associated with unfavorable outcome in NPH: idiopathic form, prolonged duration (>12 months), dementia (MMSE < 24), cerebral

Video

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atrophy, incomplete clinical triad, absence of periventricular lucencies, and age over 64 years. However, they reported good shunt results in 12 patients with iNPH despite 4 poor prognostic factors in each. Our patient had 4 of these factors; compared with their patients, he had the lowest MMSE score and severely impaired gait and sphincter function. Nevertheless, outcome was excellent. A review of 44 published studies on treatment of NPH⁷ concluded that there is no reliable way to predict which patients will improve. iNPH patients with severe and prolonged disability may still benefit from shunting if there is an objective response to tap or drainage tests.

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