Original Article

The need for an institution-specific normal pressure hydrocephalus management protocol

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ABSTRACT

Background: Despite the publication of international guidelines, the management of normal pressure hydrocephalus (NPH) varies due to clinician preference and varying clinical evidence. An audit was performed to review the current pathways used in clinical practice with the aim of formulating an institution-specific protocol to optimize and standardize care.

Methods: An internal audit was performed on the management of patients with NPH who presented to the Princess Alexandra Hospital, Brisbane between January 2016 and February 2019.

Results: Forty-one patients were included in the study. Lumbar puncture (LP) was the main diagnostic test used (63.4%). About 14.6% underwent lumbar drain (LD) insertion instead. About 12.2% did not undergo either LP or LD before definitive treatment. Only 60% of all patients underwent ventriculoperitoneal shunt insertion. Overall, five treatment pathways were noted. LP + VP shunt showed the greatest average improvement in Montreal Cognitive Assessment (MoCA) or Mini-Mental State Examination (MMSE) (+3.8 ± 3.18), followed by LD + VP shunt (+3.25 ± 3.52) and sole treatment with LP (+1.83 ± 1.18). Both pre and post intervention assessment of gait and cognition were only performed in 31% and 48.8% of patients, respectively. Four types of cognitive assessment were used (MOCA 46.4%, MMSE 46.4%, Rowland Universal Dementia Assessment Scale 3.6%, and Addenbrooke's Cognitive Examination-III 3.6%). MoCA showed greater cognition improvement (2.07) compared to MMSE (1.3) post intervention. There was no consistent objective gait assessment test used.

Conclusion: The multiple NPH treatment pathways, low rate of pre and post objective symptom assessment, and lack of standardized gait and cognitive assessment tests demonstrate a clear need for an institution-specific NPH management protocol to standardize diagnostic workup, definitive management, and allied health assessment.

Keywords: Diagnosis, Normal pressure hydrocephalus, Protocol, Treatment

INTRODUCTION

Normal pressure hydrocephalus (NPH) is characterized by a triad of gait disturbance, cognitive impairment, and urinary dysfunction. This is usually associated with ventriculomegaly and normal CSF pressure.[2,20] This condition was first described by Hankins and Adams in 1965, where diagnosis relied on the presence of all three clinical signs.[6,19,20]

NPH did not have published management guidelines until recent years.[3,10] The first guidelines were proposed in 2004 by Ishikawa.[5] These were last updated before this study in 2012 by Mori *et al*.[12] and then recently by Nakajima *et al*. in 2021[13] Clinical evidence is variable and limited,

leading to significant variation in management between individual clinicians and neurosurgical centers.[1,7] In general, diagnosis is based on a combination of clinical history, physical examination, brain imaging, and confirmatory tests such as a lumbar puncture (LP) or lumbar drain (LD).[9,16] The use of either LP or LD for initial diagnosis is variable.[4,9] Lumbar drainage has a higher sensitivity and specificity compared to LP. However, LP is less invasive, easier to perform and has a lower risk of complications.[9,10,12] Definitive diagnosis is demonstrated by a positive response to shunt surgery (e.g., ventriculoperitoneal shunt), which also serves as a definitive treatment.[6,10,19]

The effectiveness of a confirmatory test or definitive treatment is usually assessed by subjective and objective improvements in gait and cognitive impairment.

Gait disturbance can manifest as an ataxic wide-based gait or a short stepped, shuffling gait.[10,12,19,20] There is no accepted assessment test for gait disturbance from NPH. Some studies and guidelines have suggested subjective evaluation as well as the time-up-and-go (TUG) test or a short distance straight walking test.[6,8,12,13,19]

Cognitive impairment in NPH is characterized by frontosubcortical dysfunction. This manifests as psychomotor slowing as well as impaired attention, short-term memory, and executive function.[6,12,15,19] At present, there is no consensus on the most appropriate cognitive assessment tool for NPH.[12,18,19] The most recent guidelines by Nakajima et al.[13] suggest the Mini-Mental State Examination, the Wechsler Adult Intelligence Scale-III digit symbol coding and symbol search tasks, and the Frontal Assessment Battery. Various studies and neurosurgical centers each use a different assessment battery, which include Mini-Mental State Examination (MMSE), Montreal Cognitive Assessment (MoCA), FAB, trail making test A and B (TMT A and B), and many others.[6,11,14,17,21]

The purpose of this study is to review the diagnostic and management pathways of NPH within an Australian tertiary hospital and to formulate an institution-specific protocol to standardize care.

MATERIALS AND METHODS

This study was a retrospective analysis of the diagnostic and management pathways of patients with NPH who presented to the Princess Alexandra Hospital (Brisbane, Australia) between January 2016 and February 2019.

Patients were included if they had a confirmed diagnosis of NPH and were investigated or treated for NPH at PAH between January 1, 2016, and February 30, 2019.

Notes containing each patient's presenting symptoms, imaging reports, diagnostic method/confirmatory tests,

and definitive treatment were manually extracted from the hospital's electronic medical records system (iEMR) and the state-based electronic records system "The Viewer" through discharge summaries and correspondence from GPs and private specialists.

RESULTS

Forty-one patients underwent diagnosis and/or treatment for NPH at the Princess Alexandra Hospital between January 2016 and February 2019. The ages ranged from 55 to 91 with the average age being 73. The average age of NPH diagnosis was 70 years. The ratio of males-to-females was 29:16.

Overall, 63.4% had a LP as a part of their care, compared to 14.6% who had a lumbar drain. About 12.2% did not undergo either LP or LD before definitive treatment. About 57.7% of those who underwent LP went on to have ventriculoperitoneal shunt inserted. About 83% of those who underwent LD went on to have a ventriculoperitoneal shunt inserted. About 60% of all patients had a ventriculoperitoneal shunt inserted.

Overall, five main treatment pathways were noted: LP followed by VP shunt (36.6%); LP only (26.8%); VP shunt only (12.2%); LD followed by a VP shunt (12.2%); and LD only (2.4%). About 4.8% underwent other surgical interventions and another 4.8% patients refused treatment [Figure 1].

Four types of cognitive assessment were used: MMSE, MoCA, Rowland Universal Dementia Assessment Scale (RUDAS), and the Addenbrooke's Cognitive Examination-III [Figure 2].
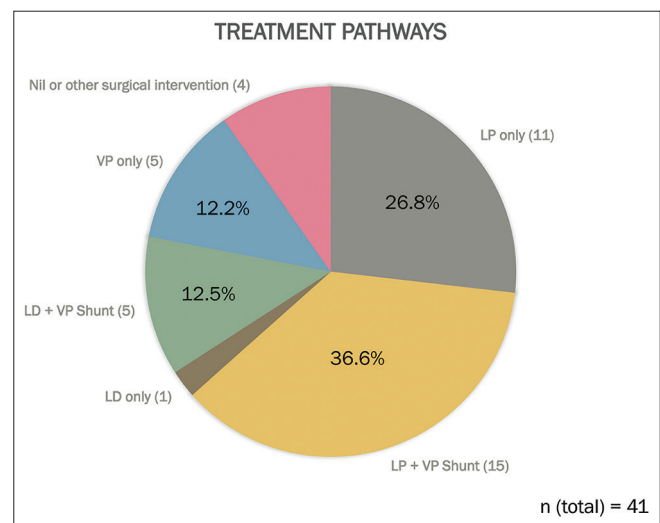
Figure 1: Distribution of treatment pathways, there were five treatment pathways noted: lumbar puncture (LP) only (26.8%), lumbar drain (LD) only (2.4%), ventriculoperitoneal shunt (VP) only (12.2%), LP + VP (36.6%), and LD + VP (12.5%).
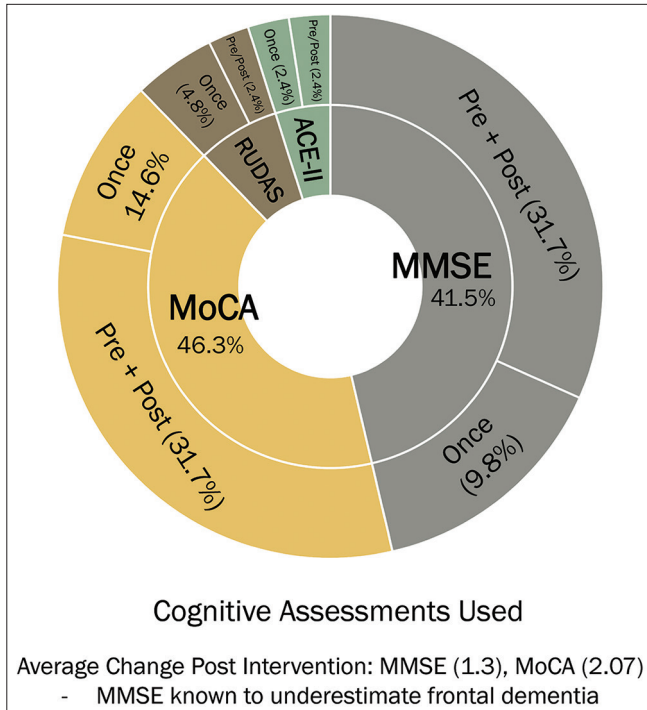
Figure 2: Distribution of cognitive assessments, there were four cognitive assessment tests used: Montreal Cognitive Assessment (MoCA, 46.3%), Mini-Mental Status Examination (MMSE, 41.5%), Rowland Universal Dementia Assessment Scale (RUDAS, 7.2%), and the Addenbrooke's Cognitive Examination III (ACE-III, 4.8%).

Despite 73.1% undergoing cognitive assessment at some point, however, only 48.8% were assessed both pre and post intervention. In those who underwent both pre and post intervention assessment, MOCA and MMSE were equally used (46.4%), while RUDAS and ACE-II were each used in 3.6% of patients.

Only 31% of patients underwent an objective gait assessment (e.g., TUG) despite 80% being assessed by a physiotherapist.

Cognitive and gait assessment were performed at variable times pre and post intervention and only when patients were admitted as an inpatient. These were not performed on outpatient follow-up.

Only 50% who underwent LP had their opening pressures recorded, while the average volume of CSF removed was 30 ml.

MoCA showed a greater average score change post intervention than MMSE with changes of 2.07 ± 1.38 and 1.30 ± 1.14 out of 30, respectively. The average change following LP was also +2.63 ± 2.09 (MoCA) and +1.2 ± 0.96 (MMSE), while the average change following a VP shunt was +1.55 ± 1.26 (MoCA) and +0.25 ± 1.68 (MMSE).

Lumbar drain insertion showed a greater increase in MMSE score (+2.5 ± 2.29) compared to LP (+1.2 ± 1.09).

Regarding treatment pathways, those who only underwent a LP showed an average improvement in cognitive assessment score (in either MoCA or MMSE) of +1.83 ± 1.18 out of 30; while having a lumbar drain followed by a VP shunt showed a change of +3.25 ± 3.52. Patients who underwent a LP followed by a VP shunt showed the greatest average cognitive score change of +3.8 ± 3.18. Cognitive assessment for patients who only underwent a VP shunt was not performed.

The average improvement in TUG was 4.77s ± 5.57 following an LP, 3.37s ± 9.38 following a lumbar drain, and 3.75s ± 4.56 following any intervention.

DISCUSSION

This study allowed for an audit of the clinically applied management pathways of NPH in an Australian tertiary hospital.

For diagnosis, LP was the main confirmatory test used (63.4%). This was likely due to several factors: more widespread skill competence among medical staff, shorter inpatient stays, and reduced risk of adverse effects. LP has a sensitivity of 58% and specificity of 75% and is the recommended CSF drainage test.[13] Insertion of a lumbar drain occurred in smaller proportion of patients (14.6%).

42.3% of those who underwent LP did not proceed with definitive management. This was likely due to inadequate improvement to justify surgical intervention or patient objection to surgery. A higher proportion of patients who had a lumbar drain inserted (83%) proceeded to insertion of a ventriculoperitoneal shunt compared to those who had a LP (57.7%). This could be attributed to greater volume of diverted CSF allowed by a lumbar drain (~10 ml/h) over 72 h compared a single reduction in CSF volume of 30 ml in LP.

12.2% of all patients did not undergo a confirmatory test and proceeded to ventriculoperitoneal shunt insertion. While the correlation of symptoms and radiological findings can provide strong diagnostic evidence for NPH, the foregoing of a confirmatory CSF drainage test can risk insertion of a VP shunt in a patient with no subsequent clinical improvement.

Five treatment pathways were likely due to varying clinical evidence and individual clinician preference. The most used pathway (36.6%) was diagnosis with a LP, followed by definitive management by insertion of a ventriculoperitoneal shunt. This is the pathway that is most recommended by the latest published guidelines by Nakajima *et al.*[13]

There was a low rate of pre and post intervention objective assessment of NPH symptoms by allied health staff. Only 48.8% underwent pre and post cognitive assessment, while only 31% underwent pre and post objective gait assessment. This was likely due to the lack of a management protocol
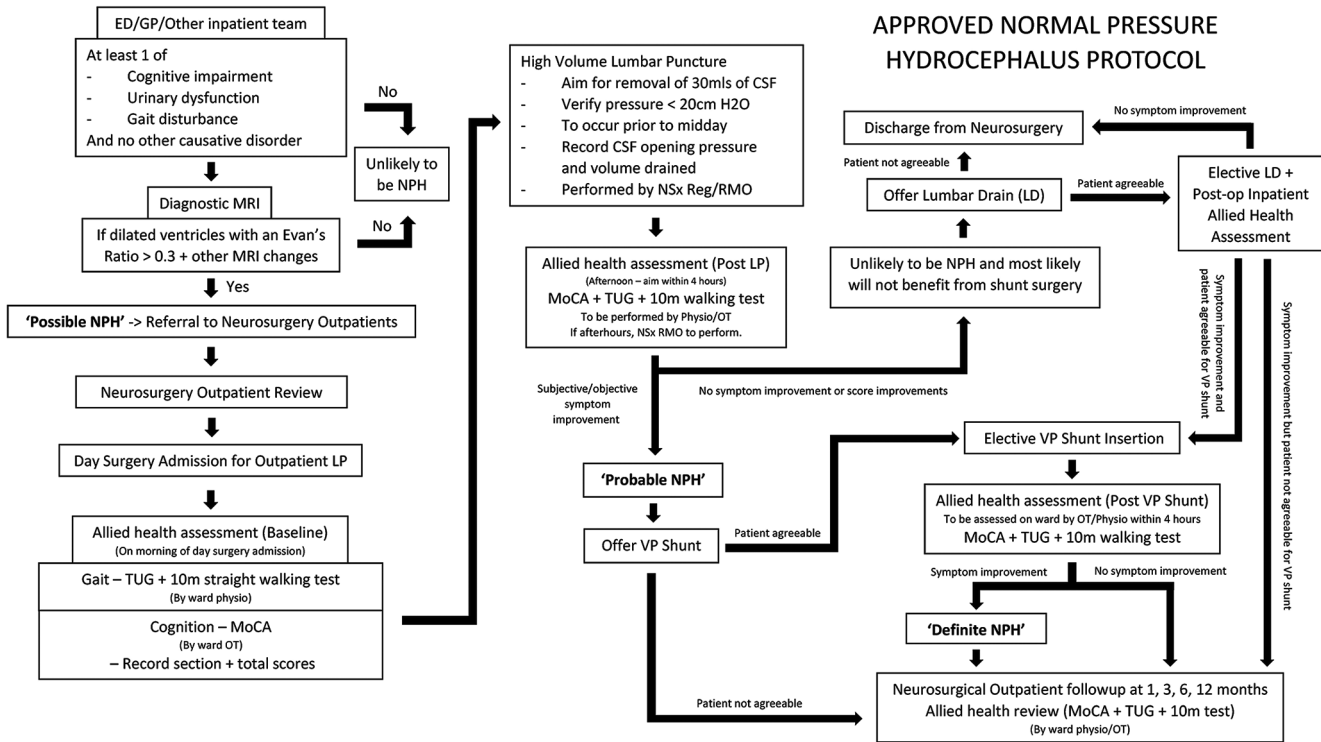
Figure 3: Approved normal pressure hydrocephalus management protocol.

mandating assessment pre and post intervention at our institution as well as inadequate allied health staffing to facilitate this service when requested by the medical team.

The clear need for an institution-specific NPH management protocol was shown by the multiple treatment pathways, the low rate of pre and post objective symptom assessment and the lack of standardized gait and cognitive assessment tests.

It must be stated that our study had a small patient population and low pre and post intervention symptom assessment rates. As such, our single institution study had very limited study power. This is demonstrated by our statistically nonsignificant findings. Nevertheless, our findings were consistent with known guidelines. Regarding cognitive assessment test, MOCA and MMSE were used equally. Our study suggested that MoCA was possibly more sensitive than MMSE, which is consistent with the previous evidence that MMSE underestimates frontal dementia when compared to other assessment tests.[15,20] There were insufficient data to include RUDAS and ACE-II in the comparison. The treatment pathway involving a LP followed by a ventriculoperitoneal shunt was suggested to be the most effective in improving cognition. This is consistent with many other studies and is the pathway suggested by the most current guidelines.[13] Interestingly, lumbar drain insertion was suggested to show greater improvement in MMSE score than LP. Regarding gait assessment, TUG was generally used when objective assessment was undertaken. There was insufficient and inconsistent assessment of gait to provide a comparison between tests; however, the literature and current guidelines suggest the use of a timed 10 m straight walking test and a TUG.[8,13]

In future studies with a multi-institution study with a larger study population, a proper analysis of treatment pathways and cognitive assessment tests would be possible.

Following consideration of the current published guidelines, local resources, and discussion with relevant allied health and medical staff, we created a hospital-specific NPH management protocol [Figure 3]. This was presented and approved at the PAH neurosurgery department meeting and implemented into clinical practice. Anecdotal evidence has shown that this has resulted in more standardized NPH management and improved patient outcomes; however, a follow-up study is in progress and will provide objective evidence.

CONCLUSION

In our hospital, there are multiple NPH treatment pathways, a low rate of pre and post objective symptom assessment and a lack of standardized gait and cognitive assessment tests. We created a hospital-specific NPH management protocol to standardize care at our institution. Given our findings, we suggest that other institutions also review their current practices to optimize and standardize NPH management.

With further research across multiple institutions and with a larger study population, statistically significant findings when comparing individual assessment and interventions are likely and an externally valid protocol could be created.

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