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Clinical outcomes after ventriculoatrial shunting for idiopathic normal pressure hydrocephalus

Ann Liu, Eric W. Sankey, Ignacio Jusué-Torres, Mira A. Patel, Benjamin D. Elder*, C. Rory Goodwin, Jamie Hoffberger, Jennifer Lu, Daniele Rigamonti

Department of Neurosurgery, The Johns Hopkins University School of Medicine, Baltimore, MD, United States

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ABSTRACT

Introduction: Idiopathic normal pressure hydrocephalus (iNPH) is a neurological disorder that classically presents with a triad of progressive gait impairment, urinary incontinence, and cognitive deterioration. Treatment predominantly involves ventriculoperitoneal (VP) shunting, but one alternative is ventriculoatrial (VA) shunting. This study sought to describe and evaluate the clinical outcomes of patients with iNPH primarily treated with VA shunting.

Materials and Methods: A retrospective review of patients with iNPH who were treated with VA shunting at a single institution, from 2003 to 2013, was performed.

Results: 58 patients with iNPH underwent primary VA shunting at a median age of 74 (IQR: 70–80) years. The most common comorbidities included hypertension ($n = 39$, 67%) and diabetes mellitus ($n = 11$, 19%). Median duration of symptoms prior to VA shunting was 24 (IQR: 12–36) months. All patients had gait impairment, 52 (90%) had cognitive decline, and 43 (74%) had urinary incontinence. Forty-three (74%) patients had all three symptoms. At a median last follow-up of 16 (IQR: 7–26) months, median iNPH score improved from 6 to 3 ($p < 0.0001$), mini mental status exam (MMSE) tended to increase from 26 to 29 ($p = 0.082$), timed up-and-go (TUG) improved from 18 to 13 s ($p < 0.0001$), and Tinetti score improved from 19 to 25 ($p < 0.0001$) after VA shunting. 78% of patients had improvement in at least one of their symptoms with 66% of patients having improvement in gait, 53% having improvement in their cognition, and 52% having improved urinary incontinence. A total of 21 patients (36%) had improvement in all 3 symptoms.

Conclusions: There were significant improvements in functional outcomes as evaluated via the iNPH score, TUG, and Tinetti score, while improvement in MMSE trended toward significance. Patients also had improvement of clinical symptoms related to gait, urinary function and cognition. These results suggest that VA shunting can be an effective primary treatment alternative to VP shunting for iNPH.

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1. Introduction

Normal pressure hydrocephalus (NPH) is a neurological disorder commonly seen in the elderly that classically presents with a triad of progressive gait impairment, urinary incontinence, and cognitive deterioration [1,2], associated with ventriculomegaly. Idiopathic NPH (iNPH) is diagnosed when no etiologies, such as traumatic brain injury, bleeding, tumor or infection, are identifiable [3].

Treatment for iNPH involves CSF diversion, most commonly via ventriculoperitoneal (VP) shunting [4]. One alternative to VP shunting is lumboperitoneal (LP) shunting, recently shown to be an effective treatment option for iNPH in the open-label randomized trial, SINPHONI-2 [5]. Ventriculoatrial (VA) shunting, which uses the cardiac atrium as the distal location of the shunt catheter, is another potential alternative to VP shunting in patients with previous abdominal surgery, a history of peritonitis, morbid obesity, or VP shunt failure [6]. In comparison to VP shunting, VA shunting provides a consistent low-pressure outlet as well as intraoperative radiographic confirmation of placement location [7]. Given the increased rates of obesity among the general population, these characteristics may be increasingly useful; however, VA shunting is still rarely used as a first-line treatment for iNPH due to surgeon

* Corresponding author at: Department of Neurosurgery The Johns Hopkins Hospital 1800 Orleans St. Room 6007 Baltimore, MD 21287, United States. Fax: +1 410 955 9126.
E-mail address: belder4@jhmi.edu (B.D. Elder).

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technical preferences and bias, as well as concerns of cardiopulmonary and renal complications.

Although outcomes of general CSF shunting in iNPH have been examined [4,8–13], the global functional outcomes of VA shunting in iNPH, specifically, have not been reported previously. Thus, the objective of this study was to examine the clinical outcomes of patients with iNPH who were treated with VA shunting.

2. Materials and methods

2.1. Patient selection

Following Institutional Review Board (NA_00044584) approval, the records of all patients with iNPH who underwent primary VA shunting at our institution from 2003 to 2013 were retrospectively reviewed. Inclusion criteria included age greater than 21 years, clinical symptoms suggestive of hydrocephalus, no previous treatment, and an Evans Index greater than 0.3. Using preoperative magnetic resonance imaging (MRI), non-obstructive hydrocephalus with normal morphology of the third ventricle was confirmed. In addition, all patients received a lumbar puncture (LP) prior to treatment and had a preoperative opening pressure < 25 cm H_2O, supporting the diagnosis of iNPH. Patients with a known cause for hydrocephalus such as trauma, tumor, infection, or bleeding were excluded. Patients that were previously treated for iNPH with VP shunting or ETV were also excluded. All patients were treated by the senior author with percutaneous placement of the VA shunt under preoperative radiographic guidance and ultrasonographic monitoring [6]. In the senior author's practice, earlier in the series, VA shunts were preferentially placed if there were challenges to VP shunting such as abdominal pathology, infections, or obesity. However, recently, VA shunts have become the senior author's initial procedure of choice if there is favorable venous anatomy on intraoperative ultrasound, due to reduced operative time.

Demographic factors including gender, age, and comorbidities were reviewed. Clinical information included prior treatment, preoperative symptoms and duration, postoperative symptoms, complications, and length of follow-up. Preoperative and postoperative functional status were described using the iNPH grading scale (from 0 to 12) [14], timed up and go (TUG), Tinetti score (out of 28 points), and Mini Mental State Examination (MMSE, out of 30 points). For iNPH score and TUG, lower values suggest better outcomes. For both Tinetti score and MMSE, higher values reflect better status. Data from the last follow-up was used to identify improvements in clinical symptoms.

2.2. Statistical analysis

Descriptive statistics regarding patient demographics, clinical presentation and function, follow-up, and postoperative function were calculated with GraphPad Prism 5.0 (GraphPad, La Jolla, CA, USA). Categorical variables were described by frequency (%) and analyzed with paired McNemar chi-square test. Continuous variables were calculated as median (IQR) and analyzed with the paired Wilcoxon signed rank test. Box-and-whisker plots displayed medians with minimum and maximum values. Statistical significance was defined as $p < 0.05$.

3. Results

From 2003 to 2013, 58 patients underwent primary VA shunting for the treatment of iNPH, and met the study's inclusion criteria. Patient demographics are presented in Table 1. Median age at treatment was 74 (IQR: 70–80) years. All but one patient were Caucasian and 33 (57%) patients were male. Common comorbidities included hypertension ($n = 39$, 67%), diabetes mellitus ($n = 11$, 19%), prior transient ischemic attack ($n = 8$, 14%), prior coronary bypass ($n = 8$, 14%) or stenting ($n = 7$, 12%), and heart arrhythmia ($n = 7$, 10%).

Table 1
Patient demographics ($n = 58$).

Patient characteristics	Patients ($n = 58$)
Male, *n* (%)	33 (57%)
Caucasian, *n* (%)	57 (98%)
Age at VA shunt, years (IQR)	74 (70–80)
Comorbidities, *n* (%)	
Hypertension	39 (67%)
Diabetes	11 (19%)
Cardiac bypass	8 (14%)
Transient ischemic attack	8 (14%)
Arrhythmia	7 (12%)
Coronary stent	6 (10%)
Cerebral infarct	5 (9%)
Myocardial infarction	5 (9%)
Heart failure	2 (3%)
ICA stenosis	2 (3%)
Valvular disease	2 (3%)
Aortofemoral bypass	1 (2%)

Abbreviations: IQR: interquartile range; VA: ventriculoatrial; ETV: endoscopic third ventriculostomy; VP: ventriculoperitoneal; ICA: internal carotid artery.

Clinical characteristics are presented in Table 2. The median duration of symptoms prior to VA shunting was 24 (IQR: 12–36) months. All patients had gait impairment, 52 (90%) had cognitive decline, and 43 (74%) had urinary incontinence. Forty-three patients (74%) presented with the complete Hakim's triad. Other symptoms included dizziness ($n = 7$, 12%), headache ($n = 6$, 10%), and subjective visual complaints ($n = 2$, 3%). Using the iNPH grading scale, preoperative median cognitive impairment grade was 2 (IQR: 2–2), gait disturbance grade was 3 (IQR: 2–3), and urinary disturbance grade was 2 (IQR: 1–3). Total median iNPH score at baseline was 6 (IQR: 5–8), median baseline TUG was 18 (IQR: 14–25) seconds, and median baseline Tinetti score was 19 (IQR: 14–22). Median baseline cognitive score, as evaluated by the MMSE, was 26 (IQR: 24–28). Median Evans Index was 0.35 (IQR: 0.33–0.38).

Two patients experienced postoperative subdural hematomas due to overdrainage, requiring surgical evacuation and shunt revision. One patient developed bilateral subdural hygromas necessitating surgical shunt revision with insertion of an anti-siphon device (ASD) two years after VA shunt placement. Three other patients had evidence of overdrainage without subdural hematoma/hygroma, which was corrected by shunt reprogramming in two patients and by surgical shunt revision in one patient. Other postoperative complications included: shunt obstruction ($n = 3$), CSF infection ($n = 1$), and malpositioned distal catheter ($n = 1$). No intraoperative or postoperative cardiopulmonary complications occurred as a result of distal catheter placement within the atrium. No cases of shunt nephritis were observed.

Median time to last follow-up was 16 (IQR: 7–26) months. At last follow-up, median cognitive impairment grade using the iNPH grading scale was 1 (IQR: 0–2), gait disturbance grade was 2 (IQR: 1–3), and urinary disturbance grade was 1 (IQR: 0–2). Paired analysis showed that the total median iNPH score at last follow-up improved to 3 (IQR: 2–5) ($p < 0.0001$). The median TUG improved to 13 (IQR: 9–15) seconds ($p < 0.0001$), and median Tinetti score improved to 25 (IQR: 22–27) ($p < 0.0001$). Median cognitive score, as evaluated by the MMSE, improved to 29 (IQR: 25–30) ($p = 0.082$). Comparisons of preoperative and postoperative values are observed in Fig. 1. Although the majority of patients continued to have some degree of gait problems ($n = 47$, 81%), cognitive issues ($n = 32$, 55%) and urinary incontinence ($n = 28$, 48%), 78% of patients had improvement in at least one of their symptoms with 66% of patients having improvement in gait, 53% having improvement in