Outcome of modern shunt therapy in patients with idiopathic normal pressure hydrocephalus 6 years postoperatively

Clinical article

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Object. Shunt therapy in idiopathic normal pressure hydrocephalus (iNPH) can improve symptoms in 84% of patients 1 year postoperatively. Therefore, implantation of a ventriculoperitoneal shunt (VPS) prevents or at least prolongs the time during which patients are not dependent on care from others because of gait disorder or dementia. In this study, the medium-term results of modern shunt therapy were evaluated.

Methods. The authors retrospectively studied outcome in iNPH patients in whom VPSs were implanted. At yearly follow-up examinations over at least 6 years, clinical symptoms were recorded according to the NPH recovery rate (NPH-RR): (preoperative Kiefer Scale [KS] score – postoperative KS score)/(preoperative KS score x 10). Time and cause of death over this period were also analyzed.

Results. Among the 147 patients treated for iNPH in the period between 1997 and 2006, 69 had died at the time of the authors’ survey. Sixty-one patients reached the 6-year examination. The median age was 64 years (range 33–83 years) at the time of VPS implantation and 73 years (range 41–91 years) at the time of this study. Six years after shunt implantation, the median KS score was significantly lower than the preoperative score (4.3 vs 8.1 points). Fifty-nine percent of 61 patients had an excellent outcome, 15% had satisfactory benefit, and 26% had unsatisfactory results 6 years postoperatively. Three of the 147 patients who underwent implantation of a VPS died of cerebral diseases 4–10 years postoperatively.

Conclusions. Implantation of a VPS is a safe procedure and can improve symptoms in 74% of patients with iNPH in the longer term.

(Please note: This article contains some figures that are displayed in color online but in black-and-white in the print edition.)

Key Words • iNPH • outcome • medium-term results • shunt • normal pressure hydrocephalus

Patients with idiopathic normal pressure hydrocephalus (iNPH) present with gait disorder, urinary incontinence, and cognitive decline. This complex, known as Hakim’s triad, can include headaches and vertigo, but enlargement of the ventricles on cerebral imaging is mandatory for the diagnosis. Over the years, the cognitive decline can evolve into true dementia. Those affected by iNPH are mostly older than 60 years, and in Norway there are approximately 5.5 new cases diagnosed per 100,000 persons per year. Currently, the estimated number of undetected cases of iNPH may be higher than expected. Approximately 9%–14% of nursing home inhabitants show symptoms of iNPH, and a further increase in cases can be expected given the aging demographic of the population. The economic cost of a comparable dementia disease, such as Alzheimer’s disease, is around €43,765 per patient per year. Thus, a considerable economic burden can be expected for iNPH as well.

Recently, the SVASONA (Shunt Valves plus shunt Assistant versus Shunt valves alone for controlling Overdrainage in idiopathic Normal pressure hydrocephalus in Adults) study showed that overdrainage complications can be avoided by implanting gravitational valves instead of conventional programmable valves with a number needed to treat of 3 to maintain therapeutic efficacy. Thus, gravitational valves have been proven to significantly increase patient safety and should be seen as the standard of care. After conducting an earlier MEDLINE search, some authors concluded that 24%–100% of patients with
shunted iNPH experience improvement postoperatively. Recent publications have shown that 84% of the patients who underwent shunt surgery for iNPH have good postoperative results and that dependency on nursing care can be reduced 1 year after treatment. The aim of this retrospective study was to evaluate the medium-term results of modern shunt therapy in patients surgically treated before 2007 in our neurosurgical department.

Methods

All patients included in this retrospective study had a standardized diagnostic pathway. Those with gait disorder, possible symptoms of iNPH, and enlargement of the ventricles on cerebral imaging (Evans Index ≥ 0.3) were diagnosed using an intrathecal infusion test and a CSF tap test. Patients with iNPH showed pathological elevated resistance ($R_{\text{out}} > 13$ mm Hg/[ml/min]) during the infusional test and an improvement of at least 20% in walking speed and step count after the CSF tap test. In cases of doubt—that is, when only one test had a positive result—lumbar drainage was performed for 72 hours to reveal late-onset improvement during gait testing. After VPS implantation, patients were prospectively scheduled for follow-up examinations at 3, 6, and 12 months postoperatively and yearly thereafter for at least 6 years. During follow-up exams, symptoms were scored using the Kiefer Scale (KS) for iNPH, which describes the main symptoms of gait disorder, cognitive decline, incontinence, headache, and vertigo by assigning 0–6 points according to symptom severity. Yearly follow-up exams documented clinical symptoms using the NPH recovery rate (NPH-RR), according to Meier et al. (preoperative KS score – postoperative KS score)/(preoperative KS score × 10). The NPH-RR defines excellent outcomes as 7–10 points. Good outcomes are represented by 5–6 points, satisfactory results by at least 2 points, and bad outcomes by less than 2 points. In addition, each patient was assigned a Comorbiditity Index (CMI) of 0–23 points to account for additional diseases, which influence the clinical presentation and outcome of patients with iNPH. During follow-up exams, valve pressure was adjusted to the individual needs of each patient. For deceased patients, time and cause of death were determined by interviewing their families, doctors, and proper authorities retrospectively.

Statistical evaluation was performed using Prism for Mac OS X (GraphPad) and Microsoft Excel for Windows (Microsoft Corp.). Unpaired samples were tested using the Mann-Whitney and Friedman tests, and the testing of more than 2 groups was performed using the Kruskal-Wallis test. The significance level was set at $p < 0.05$.

Results

Patients

In total, 147 patients suffering from iNPH were surgically treated in the Department of Neurosurgery of the Unfallkrankenhaus Berlin between 1997 and 2006. Fifty-three of these patients had already died by the time of this evaluation. After thoroughly researching, we confirmed the death of another 16 patients and loss to follow-up of 17 patients. Therefore, 61 patients were followed up for at least 6 years postoperatively. Twenty-five of these patients were female and 36 were male. The median age of the group was 64 years (range 33–83 years) at the time of VPS implantation and 73 years (range 41–91 years) at the time of this evaluation.

Twenty-eight patients primarily received a nonprogrammable gravitational valve (DualSwitch valve, Aesculap-Miethke). During a second operation, 7 of these valves were replaced with conventional programmable valves (Codman Medos programmable valve, Codman and Shurtleff, Johnson & Johnson) and 3 valves were replaced with a programmable gravitational valve (proGAV, Aesculap-Miethke). In 16 and 17 cases, respectively, conventional programmable valves and programmable gravitational valves were implanted primarily.

Fatalities

The cause of death among all 147 patients was known in few cases. Seven patients died of severe pneumonia. Beforehand, 2 of these patients were immobilized by a fracture of the lower extremity. Five patients suffered from malignant disease, and 5 other patients died of cardiovascular diseases and heart attacks. Another 3 patients had multiple organ failure due to liver dysfunction, 2 patients died of renal failure, 1 patient developed sepsis due to peritonitis, and 1 patient died due to acute paraplegia. Cerebral diseases caused the death of 3 patients. One of them suffered from an intracerebral hematoma 4 years postoperatively, and 2 patients had cerebral ischemia 9 and 10 years postoperatively. A direct association between these deaths and VPS implantation did not exist.

Midterm Success of Shunt Therapy

The median KS score was 4.3 points at 6 years after VPS implantation, which was significantly lower than the preoperative KS score of 8.1 points (p < 0.0001; Fig. 1). Compared with the 1-year postoperative KS score (3 points), an increase was observed over the following 5 years.

The KS score of the deceased patients was 7.3 points before death. The difference in this score compared with the preoperative KS score of 8.1 points among patients with a 6-year follow-up was not significant (p = 0.41). Figure 2 shows NPH-RRs over 6 years. In 59% of the cases the clinical outcome was excellent, in 15% the outcome was satisfactory, and in 26% the outcome did not improve at least 20% and was therefore described as poor. All in all, in 74% of the patients, a positive outcome was observed 6 years after VPS implantation.

Moreover, there was a statistically significant difference in the CMI between the patients with an excellent outcome and those with a poor outcome (2 vs 4 points, p = 0.001). In addition, the median age was 64 years in the patients with an excellent outcome, 72 years in those with a good outcome, 70 years in those with a satisfactory outcome, and 67 years in those with a poor outcome. These differences were not statistically significant.

Complications and Revision Surgeries

The overall complication rate was 13% (19 of 147 pa-
Midterm outcome of shunt therapy in patients with iNPH

Valve revision surgeries were necessary in 12 cases of valve dysfunction, overdrainage, and underdrainage (12 patients [8.2%]). The abdominal catheter was revised in 2 patients because of dislocation (2 patients [1.4%]). Chronic subdural hematomas led to another 2 operations (2 patients [1.4%]). The infection rate was 2% (3 patients) and shunts were explanted immediately in those patients.

Discussion

The primary aim of this study was to evaluate the 6-year outcome of a relatively large group of patients with iNPH who had a standardized diagnostic pathway and VPS surgery. Such midterm results for a single hydrocephalic entity have rarely been published. This study may provide valuable information about the prognosis of shunt therapy and thus may be helpful for individual clinical decisions. Nevertheless, the study has some clear limitations. Although all patients had the same diagnostic pathway, because of the long running time of the study, a selection bias due to a partial crew change cannot be excluded. Another limitation is the range of valve types used in the study.

These limitations aside, the acquisition of KS scores for each patient annually and the relatively high number of patients studied give this analysis relevant impact. In contrast, data about deceased patients and the causes of death were only recently surveyed; therefore, these results are incomplete and less conclusive.

Complications and Revision Surgeries

The overall complication rate of 13% in this study is comparable to recent international data (12%–15%) obtained from studies of shorter duration.26 This finding indicates that the majority of complications occur within the 1st year after shunt surgery. Most revision surgeries were performed to exchange the valve. In part this revision can be explained by the implantation of 28 nonprogrammable gravitational valves during the first operation. The revision rate tended to decrease with the use of programmable valves since a number of revision operations were replaced by simple reprogramming.

Fatalities

There were no directly postoperative deaths during the in-hospital stay after VPS implantation in the 147 patients. One patient suffered from intracerebral bleeding 4 years postoperatively, and cerebral ischemia was reported as the cause of death 2 and 10 years after VPS implantation in 2 cases. A latency of 2–10 years between VPS implantation and the ischemia or bleeding seems rather long to infer direct causality. The incidence of strokes in Germany, including transient ischemic attacks, is estimated to be around 250 cases per 100,000 persons per year.4 Approximately 80% of strokes are ischemic in nature, and 20% are caused by bleedings.7,8 In the present study, 3 of 147 patients suffered from a stroke, which is a significantly higher proportion. In addition to shunt therapy as a possible predisposing factor for stroke, the relatively high comorbidity of the study group compared with that in normal populations must be taken into account. Whether shunt therapy caused these pathologies cannot be answered definitively, but we must remember that shunt therapy can have both short-term and long-term risks, which emphasizes the need for strict diagnostics and indications for the surgical therapy.

The median CMI of all patients was 3 points, and the median patient age at the time of this study was 73 years. Given this age distribution and the comorbidity leading to a higher risk for cardiovascular disease, our patients can hardly be compared with the standard population in Germany.

Contracting pneumonia surely does not have a pathophysiological connection to shunt therapy per se, but it must be noted that being bedridden due to a gait disorder and dementia increases the risk of developing pulmonary infections. Therefore, the occurrence of pneumonia in this study cohort could hint at an ineffective therapy in affected patients.28

Midterm Success of Shunt Therapy

Our analysis showed positive results in 74% of the cases followed up 6 years postoperatively. In a recent search of the literature, we found that the midterm success of shunt therapy varies between 56% and 87% (Table...
The success of VPS implantation by the preoperative CMI, which significantly influences immobility and bedriddenness are imaginable explanations for the inability to reach our clinic for follow-up examinations. Unfortunately, it is much less likely that some of these patients were free of symptoms since at least a light progression of iNPH symptoms has to be expected for the majority of patients over time.

In summary, implantation of a VPS in patients diagnosed with iNPH has a low complication rate and ensures therapeutic success over several years postoperatively for more than half of the patients. Thorough diagnostics and patient selection seem to be crucial factors for the outcome prognosis after VPS. Careful clinical examination and invasive diagnostics remain the basis of the long-term success of shunt therapy in iNPH.

Conclusions

In summary, implantation of a VPS in patients diagnosed with iNPH has a low complication rate and ensures therapeutic success over several years postoperatively for more than half of the patients. Thorough diagnostics and patient selection seem to be crucial factors for the outcome prognosis after VPS. Careful clinical examination and invasive diagnostics remain the basis of the long-term success of shunt therapy in iNPH.

Disclosure

Dr. Lemcke received clinical or research support from B. Braun Aesculap/Miethke for the study described. Financial support was received from B. Braun Aesculap for previous studies to cover travel expenses and as remuneration for lecturing.

Author contributions to the study and manuscript preparation include the following. Acquisition of data: Gölz, Ruppert. Analysis and interpretation of data: Gözl. Drafting the article: Gözl, Lemcke. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Gözl. Study supervision: Meier.

References

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**TABLE 1: Medium-term success of shunt therapy with different VPS systems**

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Patients</th>
<th>Follow-Up Duration (mos)</th>
<th>% of Patients w/ Clinical Improvement</th>
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<tr>
<td>McGirt et al., 2005</td>
<td>179</td>
<td>24</td>
<td>75</td>
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<tr>
<td>Kiefer et al., 2002</td>
<td>91</td>
<td>26</td>
<td>85</td>
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<td>Zemack &amp; Romner, 2002</td>
<td>147</td>
<td>26.7</td>
<td>78.9</td>
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<td>Mori, 2001</td>
<td>120</td>
<td>36</td>
<td>73.3</td>
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<tr>
<td>Black, 1980</td>
<td>62</td>
<td>36.5</td>
<td>61</td>
</tr>
<tr>
<td>Meier &amp; Lemcke, 2006</td>
<td>117</td>
<td>62</td>
<td>60</td>
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<td>Kahlon et al., 2007</td>
<td>23</td>
<td>62</td>
<td>56</td>
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<tr>
<td>Gözl et al., 2013</td>
<td>61</td>
<td>72</td>
<td>62</td>
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<tr>
<td>Pujari et al., 2008</td>
<td>55</td>
<td>84</td>
<td>80–87</td>
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* Studies in boldface indicate follow-up times ≥ 6 years.


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