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Course of disease in patients with idiopathic normal pressure hydrocephalus (iNPH): a follow-up study 3, 4 and 5 years following shunt implantation

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Abstract
Background In spite of recent advances in the diagnosis and treatment of iNPH, favorable outcomes following CSF diversion continue to be limited by complications, both valve dependent and valve independent, as well as by a reduction, over time, in the response to shunting.
Materials and methods Between September 1997 and December 2006, 148 patients underwent ventriculo-peritoneal shunt surgery in our department. All patients underwent the implantation of gravitational valves. These patients were followed-up 3, 6 and 12 months after surgery and then at annual intervals.
Findings The mean age of the 94 men and 54 women in our study was 68 years. The perioperative mortality was 0.7% (one patient died from a pulmonary embolism). A further 23 patients died during the follow-up period from causes unrelated to iNPH or the surgery. This study reports on groups of patients followed-up for 2 years (n=92), 3 years (n=62), 4 years (n=38) and 5 years (n=21) postoperatively. Valve independent complications occurred postoperatively in 6% of patients (n=10). Of these, five patients (3% of the total) had an infection and catheter displacement was recorded in a further five. Valve dependent complications occurred in 24 patients (16%), with overdrainage found in seven patients (5%) and underdrainage apparent in 17 (11%). Responder rates were 79% at 2 years, 79% at 3 years, 64% at 4 years and 60% at 5 years. The optimal valve opening pressure in programmable valves with a gravitational unit was between 30 and 70 mmHg.
Conclusions Sixty percent of patients with iNPH who underwent a ventriculo-peritoneal shunt using a gravitational valve continue to benefit from surgery 5 years postoperatively.

Keywords Idiopathic normal pressure hydrocephalus · Gravitational valve · Programmable Medos-Codman-Valve · Miethke proGAV · Outcome · Gravitational valve-shunt operation · Clinical outcome study

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Introduction

The successful treatment of normal pressure hydrocephalus continues to represent a challenge to practitioners. Optimal long term outcomes are reached via a lengthy path which begins with the appropriate choice of surgical indication. The selection of individually tailored methods for controlling internal CSF drainage is the next step and the journey ends with careful postoperative management over many years. The aim of this paper is to prospectively investigate outcomes following surgical treatment of idiopathic normal pressure hydrocephalus (iNPH) using gravitational valves (valves in which the opening pressure is posture-dependent) 3, 4 and 5 years after implantation.

Materials and methods

Patients, 148, diagnosed with iNPH at a major acute hospital (Unfallkrankenhaus Berlin) between September 1997 and December 2006 were enrolled into this prospective clinical outcomes study. Treatment took the form of surgical implantation of a gravitational ventriculo-peritoneal shunt. The course of their disease was recorded postopera-

tively using the Kiefer score [7], initially at 3, 6 and 12 months following implantation and thereafter at yearly intervals. All patients underwent a computed tomography scan of the head (head CT) before surgery, 4 days post-surgery and regularly during the follow-up period to assess the Evan's index [8].

The same diagnostic pathway was used with all patients. To fulfil the requirements for further assessment of possible iNPH, a patient had to demonstrate gait ataxia during an initial clinical examination as well as at least one additional symptom of the Hakim triad. An enlarged ventricular system (as evidenced neuroradiologically with an Evan's index of >0.3) was also required. Patients meeting these criteria then underwent a constant-rate lumbar infusion test followed by a spinal tap test in which 60 ml of CSF was removed. Shunt therapy was indicated when the patient showed a pathological resistance to outflow (R_{out}) in the infusion test and an improvement in symptoms following the spinal tap test.

The normal pressure hydrocephalus recovery rate (NPH-RR) was used to assess the postoperative development of clinical symptoms and to allow comparison between patients with varying grades of symptoms on presentation.

$$\text{NPH Recovery Rate} = \frac{\text{Kiefer Score}_{\text{preoperative}} - \text{Kiefer Score}_{\text{postoperative}}}{\text{Kiefer Score}_{\text{preoperative}}} \times 10$$

The NPH-RR scores obtained were categorised as follows; 7–10 points was classified as excellent, ≥5 as good and ≥2 as fair. NPH-RR scores of less than two points were graded as poor and were deemed to be unsatisfactory. NPH-RR scores equal to or above 2 were considered satisfactory.

Results

During the 9 year study period, 148 patients underwent shunt operations for iNPH at the Unfallkrankenhaus Berlin.

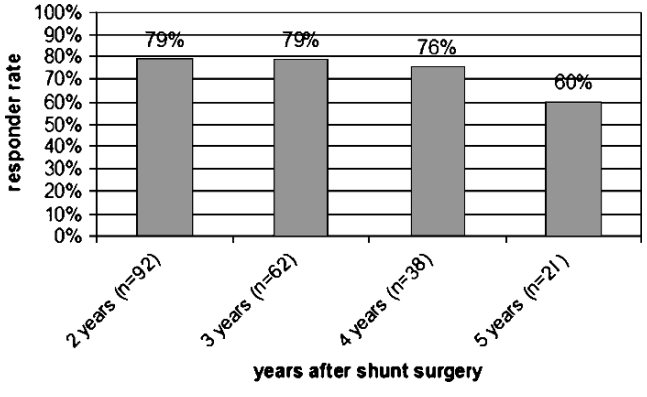

Fig. 1 Responder rate 2, 3, 4 and 5 years after shunt surgery

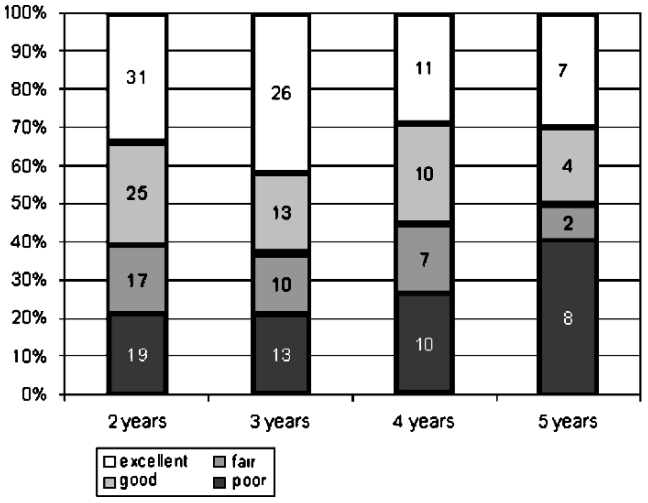

Fig. 2 Clinical outcomes measured using the NPH-RR 2, 3, 4 and 5 years after surgery. *Numbers within the boxes* represent the number of patients in that group

All patients received gravitational valves (63 dual switch valves, 42 programmable Medos® valves with a gravitational assistant valve and 43 proGAVs). Perioperative mortality was 0.7% (n=1; pulmonary embolism). A further 23 patients died during the follow-up period from causes unrelated to surgery or iNPH. The data presented here are from groups of patients followed-up at 2 years (n=92), 3 years (n=62), 4 years (n=38) and 5 years (n=21) postoperatively. The responder rate is presented in Fig. 1.

Clinical outcomes as measured using the NPH-RR (Fig. 2.) reveal that the percentage of patients experiencing a good or excellent outcome remains relatively stable after 2, 3, 4 and 5 years. At 2 years 60% of the patients had a good or excellent outcome, dropping to 50% at 5 years. By contrast, the proportion of patients suffering a poor outcome increases from 20% to 40% during this time period.

Complications 6% of patients (n=10) experienced valve independent complications postoperatively. Of these, five patients (3% of the total) had an infection and catheter displacement was recorded in a further five. Valve dependent complications occurred in 24 patients (16%), with overdrainage found in seven patients (5%) and underdrainage apparent in 17 (11%).

Discussion

The comparison of data for iNPH shunt outcome studies reported in the international literature presents several problems. The lack of distinction between patients with different pathogeneses for their iNPH often renders com-

parison of patient groups essentially meaningless [3, 10, 11] and the use of a wide range of evaluation measures and systems further complicates critical analysis of the literature [7, 9]. The frequency of follow-up periods in the literature also varies to a significant degree. It is the authors' view that 3, 6, 12 and 36 month intervals are desirable, a suggestion supported by other experts in this field [2, 4, 10, 12].

The available literature concerning iNPH outcomes following shunting reports a broad range of improvement rates. In 1984, Børgesen [2] suggested average improvement rates of 52% (range 42 – 67%) in an analysis of six studies published between 1972 and 1980. More recently, Hebb and Cusimano (2001) [5] analysed 44 publications and reported an average improvement rate of 59%, falling to 29% in the long term. Large single studies provide further figures; Romner and Zemack [12] report an improvement rate of 79% for their 146 iNPH patients at an average of 27 months (range 3 months to 9 years) postoperatively. Boon et al. [1] established average improvement rates of 53% and 47% using low pressure versus medium pressure standard valves respectively at *x* months. Mori et al. [9] determined an average improvement rate of 73% for 120 iNPH patients after 3 years while Kiefer et al. [6] observed an improvement rate of 70% in 122 iNPH patients an average of 26 months after surgery.

This study reports figures similar to those cited in the international literature. When comparing these figures with the work of others, however, it should be noted that in the classification system used in our clinical work and in this study, we consider an improvement of less than 20% (NPH-RR<2) to be a poor clinical outcome. Excellent, good and satisfactory clinical outcomes were recorded in 79% of patients 2 years postoperatively. Five years after surgery this figure had decreased to 60%. Closer examination reveals a trend towards the two extremes; the proportion of patients with good or fair outcomes decreases markedly (from 27% to 19% and from 18% to 10% respectively) whereas the poor outcome group increases considerably (20% to 40%) between year 2 and year 5, postoperatively. The percentage of patients with an excellent outcome meanwhile remains unchanged over this time period.

Conclusion

Sixty percent of patients undergoing an operation to establish a ventriculo-peritoneal shunt using a gravitational valve continue to benefit from surgery 5 years postoperatively. However, the proportion of patients with poor outcomes increases over time.

Conflict of interest statement We declare that we have no conflict of interest.

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