ADJUSTABLE VALVES IN NORMAL-PRESSURE HYDROCEPHALUS: A RETROSPECTIVE STUDY OF 218 PATIENTS

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OBJECTIVE: We sought to assess the value of adjusting shunt valve opening pressure, complications, and outcomes with the use of an adjustable shunt valve in the treatment of patients with normal-pressure hydrocephalus (NPH).

METHODS: In a single-center retrospective study, 231 adjustable valves (range, 30–200 mm H_2O) were the first shunt implantations in 147 patients with idiopathic NPH (INPH) and 71 patients with secondary NPH (SNPH). The effect of adjustment on gait disturbance, cognitive impairment, urinary incontinence and other symptoms were evaluated, and an improvement index was created.

RESULTS: In the INPH group, 138 adjustments were performed in 49.0% of the patients (average, 0.94 adjustments/patient). For the SNPH group, 49 adjustments were performed in 32.4% of the patients (average, 0.69 adjustments/patient). The reasons for adjustment were overdrainage in 48 patients (25.7%), underdrainage in 98 patients (52.4%), subdural hematoma in 37 patients (19.8%), and other reasons in 2 patients (2.1%). Clinical status improved after 56 (49.1%) of all 114 adjustments, whereas 23 (42.6%) of 54 minor (≤20 mm H_2O) and 33 (66.0%) of 50 larger adjustments improved the patient's clinical status. The correlation of the improvement index with the size of the individual adjustments was not significant. Complications occurred in 43 (19.7%) of 218 patients, valve malfunction occurred in 3 patients (1.3%), infection occurred in 14 patients (6.4%), and nontraumatic subdural effusion occurred in 15 patients (6.9%; 8 were treated by adjustment alone). The 5-year shunt survival rate was 80.2%. Outcomes were excellent or good in 71 (78.9%) of 90 patients with INPH and in 30 (69.8%) of 43 patients with SNPH.

CONCLUSION: Noninvasive, particularly consecutive, minor or single larger adjustments to the valve opening pressure can further improve outcome in patients with NPH who undergo shunting.

KEY WORDS: Adjustable shunt valve, Cerebrospinal fluid shunt, Hydrocephalus, Normal-pressure hydrocephalus, Programmable valve

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Rendering an accurate diagnosis, predicting shunt responsiveness, and evaluating outcomes in patients with normal-pressure hydrocephalus (NPH) are a challenge. Characteristic findings of gait disturbance, cognitive impairment, urinary incontinence, and evidence of disturbed cerebrospinal fluid (CSF) reabsorption are not invariably present. The substantial presence of co-morbidities (e.g., cerebrovascular disease, Parkinsonism, cognitive impairment) and the process of normal aging must be taken into account.

Valve-regulated CSF shunt systems are used in the treatment of patients with NPH. Although catheter-related complications and shunt-related infections are the major causes of shunt failure, the type of valve implanted is also of importance. A differential pressure valve opens and begins to drain CSF at a certain pressure. Because there are no means by which to predict the most appropriate pressure setting at the time of implantation in an individual patient, adjustment without surgical shunt revision is desirable because it enables opening pressure to be adjusted in accordance with clinical or radiological findings in the postoperative period (6, 11, 13, 15, 24, 25, 32, 34–36). The adjustable valve used in this study enables adjustments to be made at 18 levels between 30 and 200 mm H_2O and has been described elsewhere (32, 41).

Since the publication of early reports of an NPH syndrome (1, 12, 14), the incidence of this entity has grown in the increasing population of older adult patients. The evaluation of variations in treatment is important in enabling optimal treatment. At the Department of Neurosurgery at Lund University Hospital, the Codman Hakim Programmable Valve (Codman & Shurtleff, Inc./Johnson & Johnson Co., Raynham, MA) has been used since 1992. Although some data on some of the patients described in this article were reported previously (41), this article focuses on NPH and adjustment strategies in the postoperative period, and the data presented are unique.

PATIENTS AND METHODS

Patient Population and Diagnosis

This single-center retrospective study identified 248 Codman Hakim Programmable Valves implanted between January 1992 and December 2000 in 230 patients diagnosed with NPH. Twelve patients (17 valves) were excluded because they either had undergone a previous shunting procedure before receiving their first shunt incorporating a Codman Hakim Programmable Valve or had received a lumboperitoneal shunt. Age, severe co-morbidity, and restricted life expectancy were not criteria for exclusion. The remaining 218 patients (94.8%) received 231 valves and comprised the sample population in this study. Ventriculoperitoneal shunts were implanted in 195 cases (89.5%) and ventriculoatrial shunts were placed in 23 cases (10.5%).

Patients were divided into idiopathic NPH (INPH) and secondary NPH (SNPH) groups on the basis of their NPH etiology (*Table 1*). NPH secondary to other causes included previous intracranial operation, radiotherapy, cerebellar hemorrhage, and Paget's disease. The patients' age at the time of the first valve implantation procedure ranged from 15 to 89 years.

The diagnosis of NPH was rendered on the basis of clinical symptoms and signs of gait disturbance, cognitive impairment, urinary incontinence, a computed tomographic (CT) scan demonstrating ventriculomegaly out of proportion to apparent atrophy, and a constant manometric lumbar infusion test (21). Ventricular dilation visualized on a CT scan was not quantified. Two or more symptoms of the NPH triad were present in 201 patients (92%). A lumbar infusion test was performed in all 147 INPH patients and in 59 of the 71 SNPH patients. If the infusion test was inconclusive, a supplementary tap test (i.e., removal of 50 ml CSF) was performed. No continuous tap tests (i.e., 1–3 d drainage) were performed. Symptoms of SNPH that developed within 1 month of the identification of the causative factor were excluded. All 19 neurosurgeons were involved in the implantation procedures. The technique for shunt insertion was a standardized retroauricular placement of the valve. Each adjustment was followed by cranial x-ray to verify the selected setting.

Valve Opening Pressure

Opening Pressure at Implantation

The opening pressure settings at implantation were decided on the basis of the patient's age, the duration of the underlying disease, the size of the ventricles, and the curve profile, amplitude, and opening pressure findings derived from a constant manometric lumbar infusion test. All infusion tests that led to shunt insertion were considered pathological. The initial baseline pressure before the infusion test was started was used in deciding on the opening pressure at the time of implantation. A low (90–130 mm H_2O) opening pressure setting was chosen when the initial pressure was high, while a high (140–180 mm H_2O) opening pressure setting was chosen when the initial pressure was low and/or was combined with low amplitude and a less pathological curve profile. In older adult patients (older than age 75 yr), the high (140–180 mm H_2O) opening pressure level was selected to avoid the occurrence of subdural hematomas.

Outcome Evaluation after Adjustment

Adjustments were evaluated as to their effect on patients' gait disturbance, cognitive impairment, and urinary incontinence with the use of a rating scale of worse, unchanged, good, or excellent based on objective and subjective observations. Other relevant symptoms or signs were included when

TABLE 1. Summary of diagnosis, sex, and age statistics in 218 patients requiring 231 Codman Hakim Programmable Valve implantations

Type of normal-pressure hydrocephalus	No. of patients (no. of valves)	Male-female ratio (M/F)	Mean age (yr)	Age range (yr)
Idiopathic	147 (152)	90/57	74.3	40.7–89.4
Secondary presentation	71 (79)	32/39	61.8	15.5–82.8
Subarachnoid hemorrhage	37 (41)	11/26	62.2	32.4–82.8
Meningitis	2 (2)	0/2	72.5	69.9–75.0
Trauma	21 (25)	17/4	57.2	15.5–80.6
Tumor	4 (4)	2/2	74.6	68.7–80.6
Other	7 (7)	2/5	68.0	57.0–77.2
All patients	218 (231)	122/96	70.4	15.5–89.4

they were the reason or one of the reasons to adjust the opening pressure. Evaluations were not taken into account (i.e., excluded from evaluation calculations) if 1) data were insufficient; 2) the gait disorder, cognitive impairment, or incontinence did not exist preoperatively; 3) the adjustment or evaluation took place less than 10 days before surgical revision because of a shunt complication; or 4) the evaluation took place less than 1 week after adjustment. Adjustments performed after subdural hematomas or hygromas were evaluated separately, and adjustments made to reset the opening pressure after magnetic resonance imaging was performed were excluded. The adjustment evaluations took place at the first visit after the adjustment.

Improvement Index

To quantify improvement, an adjustment evaluation improvement index was created (22). Four symptoms were evaluated: gait disturbance, cognitive impairment, urinary incontinence, and other symptoms such as headache, nausea, or dizziness. Each symptom was evaluated separately and was assigned a grade of 0 if there was no improvement, 1 if improvement was good or fair, or 2 if improvement was excellent. A fraction was then derived from these data, with the numerator corresponding to the sum of all improvement grades of symptoms (when there were sufficient data for grading) and the denominator corresponding to the maximum possible sum of all improvement grades of symptoms (when there were sufficient data for grading). For example, we derived a fraction between 0/6 and 6/6 in patients for whom there were data for three symptoms before adjustment, and we derived a fraction between 0/8 and 8/8 in patients for whom there were data for the whole classical triad plus one other symptom. The fraction was then expressed as an improvement index between 0 (no meaningful improvement) and 1 (excellent improvement of all symptoms).

Complications and Shunt Survival

Once a shunt was revised, it was defined to have reached its survival end point. Patients who died were censored. Because our department is the only neurosurgical center in our region, we received all referrals for shunt complications. The survival time in patients who did not reach an end point stretched from the date of implantation until May 2001.

Clinical Outcome at Follow-up

Outcome assessment was based on an evaluation performed at the patient's last contact with our department (last contact May 2001). Outcome was determined by the effect of treatment on symptoms, signs, and radiological findings. It was graded as excellent if there was a clear improvement with no or minor residual symptoms or signs and return to independent living, good if there was improvement but moderate residual symptoms or signs, unchanged if there was slight but not good improvement, and "worse." The minimum time until follow-up revision was 3 months. The average time until follow-up revision for patients who reached this minimum time point was 26.7 months (maximum, 8.8 yr) among the patients with INPH and 30.8 months (maximum, 9.1 yr) among the patients with SNPH.

Statistics

The statistical analysis tests used were the χ^2 test for binary data, the t test for sample means, and the log-rank test to compare survival times calculated by Kaplan-Meier analysis. Correlations were linear. $P < 0.05$ was considered significant. Only significant results are indicated in the Results.

RESULTS

Valve Opening Pressure

Opening Pressure at Implantation

The average opening pressure selected at implantation for both INPH and SNPH patients was 132 mm H_2O (both groups: range, 90–180; median, 130 mm H_2O).

Opening Pressure Adjustment

In the INPH group, 138 adjustments were performed (average, 0.94; maximum, 8 adjustments/patient). For the SNPH group, 49 adjustments were performed (average, 0.69; maximum, 6 adjustments/patient). Adjustments were performed at least once in 72 (49.0%) of 147 patients with INPH and in 23 (32.4%) of 71 patients with SNPH. The reasons for performing the adjustments are presented in *Table 2*. The average time after shunt insertion until the first adjustment was 231 days (median, 66 d) in the INPH group and 154 days (median, 87 d) in the SNPH group, excluding the adjustments performed for subdural effusion. A majority of adjustments were performed within 5 months of implantation. Most of the adjustments made later than 6 months after implantation were performed in the INPH patients (*Fig. 1*). On average, the individual adjustments were 28.8 mm H_2O in the INPH group as compared with 30.5 mm H_2O in the SNPH group, excluding the adjustments performed for subdural effusion.

Outcome Evaluation after Adjustment

In cases in which adjustment was evaluated, the patient's clinical status improved after 56 (49.1%) of 114 adjustments, not excluding the negative impact of co-morbidity. The calculated improvement index for these adjustments is presented in *Table 3*. The average time until adjustment evaluation was 73 days (median, 60 d) in the INPH group and 84 days (median, 68 d) in the SNPH group. Adjustments that were performed to treat overdrainage improved the clinical status in a higher percentage of patients than those that were performed to treat underdrainage (*Table 4*), and adjustments improved gait disturbance and other symptoms more than they improved cognitive impairment and urinary incontinence (*Table 5*).

Late adjustments improved patients' clinical status in 14 (33.3%) of 42 cases, and early adjustments improved patients'

TABLE 2. Reasons for adjusting the opening pressure[a]

Reason for adjustment	Idiopathic NPH (n = 138)	Secondary NPH (n = 49)	All patients (n = 187)
Overdrainage	23.9%	30.6%	25.7%
Underdrainage	53.6%	49.0%	52.4%
Underdrainage after SDH or SDHy treatment[b]	9.4%	12.2%	10.2%
Instead of shunt ligation	8.0%	2.0%	6.4%
To avoid overdrainage at removal of shunt ligature	2.9%	4.1%	3.2%
Altered by MRI	0.7%	2.0%	1.1%
Miscellaneous	1.4%	0.0%	1.1%
Total	100%	100%	100%

[a] NPH, normal-pressure hydrocephalus; SDH, subdural hematoma; SDHy, subdural hygroma; MRI, magnetic resonance imaging.
[b] Adjustments made to decrease the opening pressure back to its original value after it had been increased because of the treatment of a subdural hematoma or hygroma.

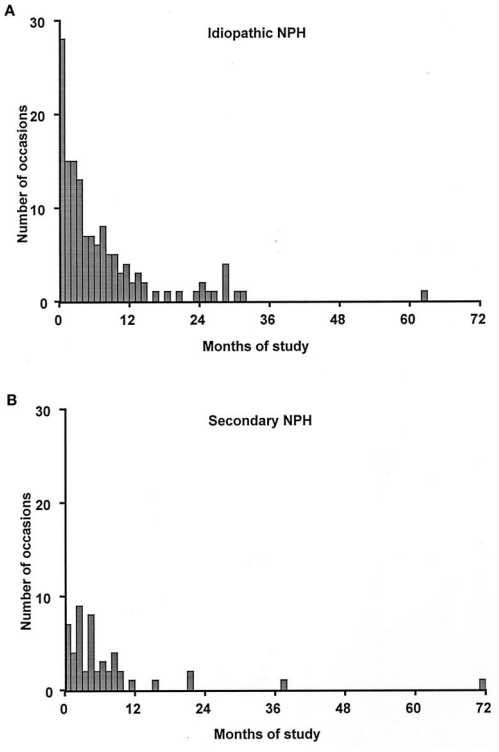

FIGURE 1. *Bar graphs depicting the time after shunt implantation until adjustment was performed in patients with idiopathic normal-pressure hydrocephalus* (A) *and patients with secondary normal-pressure hydrocephalus* (B). NPH, *normal-pressure hydrocephalus.*

clinical status in 42 (58.3%) of 72 cases ($P = 0.010$). A minority (8 of 34) of adjustments made to treat overdrainage were early, but the distribution of early (45) and late (34) adjustments for underdrainage was more equal. Minor adjustments of as much as ±20 mm H_2O improved the patient's clinical status in 23 (42.6%) of 54 cases, whereas adjustments of at least ±30 mm H_2O improved the patient's status in 33 (66.0%) of 50 cases ($P = 0.016$) (*Table 6*). However, the correlation of the improvement index with the size of the individual adjustments was not significant. The improvement index also showed no significant correlation with age or with time after implantation until adjustment. The average opening pressure levels selected at the time of the follow-up examination were 130 mm H_2O for patients with INPH (range, 40–200 mm H_2O; median, 130 mm H_2O) and 129 for patients with SNPH (range, 50–200 mm H_2O; median, 120 mm H_2O).

Complications

A total of 69 complications occurred in 43 (19.7%) of 218 patients. Of these patients, 58 required surgical revision (*Table 7*). The overall infection rate was 14 (6.4%) of 218 patients for first shunt implantations. In three of these cases, laboratory investigation did not confirm the clinical diagnosis. Six patients did not accept or did not require a new shunt after removal of the first shunt, and four of these patients proved to be clinically shunt independent. Valve-related complications were the cause of shunt malfunction in three implanted valves (1.3%). Two of these valves were obstructed, and one could not be adjusted in two attempts.

Subdural Hematomas and Hygromas

Nontraumatic subdural hematomas or hygromas were found in 14 (9.5%) of 147 patients with INPH and in 1 (1.4%) of 71 patients with SNPH (*Table 8*). Six additional traumatic subdural hematomas or hygromas occurred. Eight nontraumatic subdural hematomas or hygromas and one traumatic subdural hematoma or hygroma were treated only by increasing the valve's opening pressure; no surgery was necessary. Seven of the nontraumatic cases showed complete resorption of the hematoma or hygroma on follow-up CT scans. Of the remaining nontraumatic cases, 2 required no treatment and 10 required surgical drainage and/or ligation of the shunt system. The adjustment replaced shunt ligation in three cases, and adjustments were performed in six cases to avoid overdrainage at ligature removal. The average time after shunt

insertion until the development of a nontraumatic subdural hematoma or hygroma was 4.5 months (range, 3 d–37.5 mo).

Shunt Survival

At 3 and 5 years after implantation, the shunt survival rates were 83.7 and 83.7% in the INPH group and 76.5 and 73.7% in the SNPH group, respectively (*Fig. 2*).

Clinical Outcome at Follow-up

At a minimum of 3 months after the shunting procedures were performed, follow-up data were obtained in 191 patients. Outcomes were excellent or good in 71 (78.9%) of 90 patients with INPH and in 30 (69.8%) of 43 patients with SNPH (*Table 9*). Improvement was seen in 12 (66.7%) of 18 of patients older than 80 years of age. Forty-three patients with INPH and 14 patients with SNPH, including 1 patient who died 12 days after shunt insertion, died as a result of either progression of the primary disease or causes not related to shunt complication or infection.

DISCUSSION

A CSF shunt valve with an adjustable opening pressure level was originally proposed in 1973 (13). It not only reduces the uncertainty of having to assume preoperatively the optimal opening pressure for NPH patients (4, 7, 23, 25) but also allows for noninvasive matching of the opening pressure with the patient's intracranial hydrodynamics as the clinical course changes after implantation. The existing publications on the use of an adjustable valve in the treatment of patients with NPH are in favor of using such a valve, and the possibility of adjusting the opening pressure after implantation seems beneficial to patients' clinical outcomes (3, 6, 8, 9, 20, 24, 28, 29, 32, 34, 35, 40). We previously reported our overall experience with the use of adjustable valves. This article focuses on adjustment strategies and the evaluation of adjustments in patients with NPH (41).

Patient Population and Diagnosis

Accurate diagnosis of NPH is a challenge (38), and separating patients with NPH with co-morbidity who are shunt-responsive from those with only co-morbidity (i.e. not NPH) is not easy. No single diagnostic test has yet proved to be highly accurate (17).

Valve Opening Pressure

Opening Pressure at Implantation

To avoid causing subdural hematomas, especially in older patients, we selected a reasonably high opening pressure setting (140–180 mm H_2O) at shunt insertion and decreased it step-by-step in the postoperative period when necessary. A high initial pressure reading obtained during the preoperative infusion test indicates that a low (90–130 mm H_2O) opening pressure should be chosen at the time of implantation, and low initial pressure indicates a high (140–180 mm H_2O) should be chosen. In patients with more apparent preoperative symptoms, a low opening pressure should be chosen. The distribution of the opening pressure settings at follow-up indicates that an adjustable valve is useful, because even in a homogenous group of patients, no recommendation can be made as to which setting is optimal.

TABLE 3. Distribution of outcomes for the different ranges of the improvement index

Improvement index	Evaluated adjustments (%)(n = 114)
0.0	50.9%
>0.0–0.20	8.8%
>0.20–0.40	21.9%
>0.40–0.60	14.9%
>0.6–0.80	2.6%
>0.80–1.00	0.9%
Total	100%

Opening Pressure Adjustment and Outcome Evaluation

The question arises whether adjustments in general can improve patient outcome and whether minor adjustments specifically influence symptoms and outcome. Small, consecutive adjustments and fine-tuning or titrating the opening pressure more precisely than the traditional low-, medium-, or high-pressure settings offered by nonadjustable valves optimizes treatment by allowing for adaptation and evaluation (19, 36).

Determining the best presentation of the adjustment evaluation data is difficult. For the adjustment evaluation, a method similar to that used by Krauss et al. (22) was used. In this method, a fractional score is derived, with 0 representing no meaningful improvement and 1 representing excellent improvement of all symptoms. Because the maximum possible sum of all improvement grades of symptoms represents an excellent improvement in every symptom, which is an unattainable goal in most cases, the fractions tend to be small even though the patient outcome was very good. Radiological findings were not evaluated after each adjustment, because the change in ventricular size, in our experience, does not always correlate with the clinical findings.

Our results also demonstrate that although alterations to the opening pressure may not be effective in a defined subgroup of patients, they may be very effective in individuals within that subgroup. The treatment of gait disturbance was slightly more successful than that of urinary incontinence, which in turn was more successful than the treatment of cognitive impairment. The treatment of other symptoms such as headache, nausea, and dizziness was relatively successful.

Clinical outcome was improved after 49.1% of adjustments. Interestingly, 18.3% of patients whose pressure was not adjusted did not improve. One would have expected that at-

TABLE 4. Improvement according to reason for adjustment[a]

Reason for adjustment	% improved (no. improved/total no. of adjustments)					
	Idiopathic NPH		Secondary NPH		All patients	
Overdrainage	58.3%	(14/24)	70.0%	(7/10)	61.8%	(21/34)
Underdrainage	46.0%	(29/63)	37.5%	(6/16)	44.3%	(35/79)
Underdrainage after SDH or SDHy treatment[b]	44.4%	(4/9)	50.0%	(2/4)	46.2%	(6/13)
Treating a SDH or SDHy	36.4%	(4/11)	—	(0/0)	36.4%	(4/11)

[a] NPH, normal-pressure hydrocephalus; SDH, subdural hematoma; SDHy, subdural hygroma.
[b] Adjustments performed to decrease the opening pressure to its original value after it was increased because of the treatment of a subdural hematoma or hygroma.

TABLE 5. Outcomes with regard to the three symptoms of the normal-pressure hydrocephalus triad and for other symptoms of over- or underdrainage

Symptom	No. of evaluated adjustments	Worse (%)	Unchanged (%)	Good (%)	Excellent (%)
Gait disturbance	107	4.7%	64.5%	25.2%	5.6%
Cognitive impairment	90	0%	77.8%	21.1%	1.1%
Urinary incontinence	52	7.7%	69.2%	11.5%	11.5%
Other symptoms	50	6.0%	48.0%	36.0%	10.0%

TABLE 6. Improvement at each adjustment size

Size of adjustment (in mm H_2O)	Patients demonstrating improvement (%) (n = 114)
10	36.4%
20	41.2%
30	56.7%
40	50.0%
≥50	60.0%

tempts would have been made to adjust the opening pressure. The possible reasons for the lack of an adjustment attempt in these cases could be 1) insufficient follow-up, 2) the surgeon opted not to perform an adjustment at follow-up because of the risks of overdrainage and subsequent complications, or 3) the presence of co-morbidity.

Overdrainage

Adjusting the opening pressure can compensate for symptoms and signs of overdrainage caused by mismatching the valve's opening pressure with the patient's needs. The occurrence of symptoms and signs of overdrainage in this study and experiments suggest that siphoning is of less importance than expected with the use of a differential pressure valve (18).

TABLE 7. Summary of 58 complications that required shunt revision[a]

Type of complication	No. of occurrences (n = 43)		Incidence (%)
	INPH	SNPH	
Proximal catheter			13.8%
Obstruction	2	2	
Disconnection	0	0	
Suboptimal position	2	1	
Additional catheter insertion		1	
Valve-related	1	2	5.2%
Atrial catheter			1.7%
Suboptimal position	1	0	
Peritoneal catheter			19.0%
Obstruction	1	4	
Disconnection	0	0	
Suboptimal position	5	1	
Subdural fluid collection	7	3	17.2%
Infection	6	5	20.7%
Suspected infection	1	2	3.4%
Miscellaneous	9	2	19.0%
Total	35	23	100%

[a] INPH, idiopathic normal-pressure hydrocephalus; SNPH, secondary normal-pressure hydrocephalus.

TABLE 8. Summary of subdural hematomas and hygromas[a]

Diagnostic group	Type of hematoma	Age (yr)	Initial OP (mm H_2O)	OP at time of SDH or hygroma (mm H_2O)	Treatment			OP at time of follow-up (mm H_2O)
					Adjusted OP (mm H_2O)	Shunt ligature	Surgical evacuation	
INPH	Nontraumatic SDH	73	120	120	170	No	No	130
	Nontraumatic SDH	76	130	130	160	No	No	160
	Nontraumatic SDH	75	140	140	200	No	No	180
	Nontraumatic SDH	74	120	120	200	No	No	200
	Nontraumatic SDH	76	140	140	160	No	No	180
	Nontraumatic SDH	72	100	80	200	No	No	80
	Nontraumatic SDH	74	130	130	180	Ligature	Evacuation	180
	Nontraumatic SDH	74	140	140	200[b]	Ligature	Evacuation	180
	Nontraumatic SDH	73	90	90	No	No	Evacuation	90
	Nontraumatic SDH	78	120	160	No	No	No	160
	Nontraumatic SDH	56	120	40	No	No	No	40
	Nontraumatic hygroma	73	130	130	180	No	No	180
	Nontraumatic hygroma	81	110	110	180	No	No	180
	Nontraumatic hygroma	62	140	100	190	No	Evacuation	100
	Traumatic SDH	70	120	120	190	No	Evacuation	190
	Traumatic SDH	74	120	120	200	Ligature	Evacuation	200
	Traumatic SDH	68	130	130	190[b]	Ligature	Evacuation	190
	Traumatic hygroma	80	130	130	170	No	No	170
SNPH	Nontraumatic SDH	56	120	120	200	No	Evacuation	200
	Traumatic SDH	57	130	130	200[b]	Ligature	No	120
	Traumatic SDH	39	120	120	170[b]	Ligature	Evacuation	120

[a] Nontraumatic, chronic or subacute, not due to trauma; SDH, subdural hematoma; initial OP, valve opening pressure.
[b] Adjustment to the opening pressure prior to ligature removal.

A siphon-reducing device is an option that might improve outcomes in selected patients.

Minor Adjustments

The possibility of adjusting the opening pressure in small steps is beneficial (9, 29). It allows additional improvement of the patient's clinical status after shunt insertion. Minor adjustments led to improvement of the patient's clinical status in less often than did larger adjustments. Certain patients do respond to adjustments of as little as 10 mm H_2O, however, which is why the possibility of performing minor adjustments must exist. Every improvement in a patient's quality of life, albeit a short time or a transient improvement, must be seen as a benefit.

Adjustment Strategies

Various strategies for determining the opening pressure setting and adjustment have been presented. Some have used a high initial opening pressure that can be decreased until there is improvement of symptoms (32), whereas others recommend a low opening pressure that is increased in the postoperative period (37). A third option would be to start with a reasonably high setting at the time of shunt insertion, reduce the opening pressure to a level at which the brain parenchyma gives way to ventricular shrinkage, and then increase the pressure slightly once the clinical symptoms begin to improve to avoid complications of overdrainage (16, 37). This approach was used only in selected cases in this study because of the theoretical risk of the patient's developing subdural effusion. Whether the very low pressure needed to induce ventricular shrinkage in some patients in other

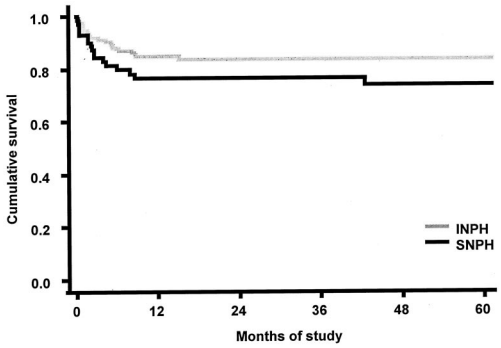

FIGURE 2. *Graph depicting the results of Kaplan-Meier statistical analysis of shunt survival in 218 patients with idiopathic and secondary normal-pressure hydrocephalus.* INPH, *idiopathic normal-pressure hydrocephalus;* SNPH, *secondary normal-pressure hydrocephalus.*

TABLE 9. Patient outcome at follow-up revision for patients who had sufficient outcome data and were followed for a minimum of 3 months, excluding deceased patients, grouped by diagnosis[a]

NPH Diagnosis	No. of patients	Outcome (%)		
		Unchanged	Good	Excellent
Idiopathic	90	21.1%	50.0%	28.9%
Primary diagnosis	43	30.2%	39.5%	30.2%
Subarachnoid hemorrhage	24	29.2%	33.3%	37.5%
Meningitis	0	—	—	—
Trauma	11	18.2%	54.5%	27.3%
Tumor	2	50.0%	0.0%	50.0%
Other	6	50.0%	50.0%	0.0%

[a] NPH, normal-pressure hydrocephalus.

studies (5, 10, 30) can be accomplished with the use of an adjustable valve alone is not known.

Complications

The complication rates in larger series of patients with NPH who undergo shunting are substantial (17, 39), a finding that is supported by the present study. Furthermore, a randomized study did not reveal a significant difference in complication and survival rates between adjustable and other types of valves (31). The overall infection rate of 6.4% and other complication rates did not differ significantly from those reported in other studies, considering the long follow-up time in this study (17). A majority of revisions were necessitated by catheter-related complications and shunt-related infections. The valve itself was the site or the cause of shunt malfunction in only 3 (1.3%) of all 231 implanted valves.

A nontraumatic subdural hematoma or hygroma was seen in 14 (9.5%) of 147 patients with INPH and in 1 (1.4%) of 71 patients with SNPH. Adjustment of the valve to a higher opening pressure was performed in the treatment of eight nontraumatic subdural hematomas or hygromas. Other larger series showed an incidence of subdural hematomas or hygromas of between 4 and 16% after shunting was performed in patients with NPH (4, 9, 22, 25, 39). In the Dutch Normal-Pressure Hydrocephalus Study (7), routine CT screening indicated that subdural effusion occurred in 71% of patients with low-pressure valves and in only 34% of patients with medium-pressure shunt systems. The ability to treat subdural fluid collection in patients with NPH by adjusting the valve to a higher setting is a clear advantage of adjustable valves (2, 3, 6, 9, 19, 24, 33–36). Adjustment replaced shunt ligation in some cases, and in other cases, it was performed to avoid overdrainage when the shunt ligature was removed. Once fluid collection has been resorbed, the opening pressure can slowly be lowered in response to symptoms and signs of underdrainage and to minimize the risk of the redevelopment of a hematoma or a hygroma.

Clinical Outcome at Follow-up

Defining improvement after shunting is difficult. We think that a small improvement must be considered when judging outcome; however, this improvement must not last too short a time. Transient improvement that does not last 1 month after the operation was not recorded, because it was considered too early to judge the patient's outcome.

CONCLUSIONS

No single diagnostic test for NPH has yet proved to be highly accurate (17). Shunting is the only widely accepted treatment for patients with NPH, even though third ventriculostomy has been discussed in some recent publications (26, 27). The use of an adjustable valve as part of the implanted shunt allows for the early treatment of overdrainage, underdrainage, and complications such as subdural hematomas and hygromas. Widely expandable opening pressure settings at follow-up indicate that an adjustable valve is useful because, even in a homogenous group of patients, no recommendation can be made as to which setting is optimal. The uncertainty in defining the optimal opening pressure at the time of implantation for patients with NPH is overcome by the use of an adjustable valve. Noninvasive, particularly consecutive, minor adjustments to the valve opening pressure can further improve outcome in patients with NPH who undergo shunting. Neuropsychological and functionality tests also would add to the accuracy and efficacy of the adjustment evaluation.

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COMMENTS

For this study, the Lund group has extracted a subgroup of 218 patients with normal-pressure hydrocephalus (NPH) from the much larger study already published in the *Journal of Neurosurgery* in 2000. They previously published a cost-effectiveness report of adjustable valves in the *British Journal of Neurosurgery* (2). In 2000, the United Kingdom and Ireland Medos Shunt Audit Group published their findings in the *British Journal of Neurosurgery* (1). It is interesting to compare some of the differences in the conclusions drawn in these reports. I am concerned that Zemack and Romner do not distinguish more accurately between solid subdural hematomas and subdural hygromas. What do they mean by *subdural hematoma*? Do they mean pure blood, and, if so,

with what degree of midline shift? Do they include simply some blood within a predominantly subdural hygroma? A number of their patients with subdural hematomas did require evacuation. How many of their patients deteriorated, in terms of Glasgow Coma Score or focal deficit before evacuation, once the pressure setting was increased and the shunt was tied off? The United Kingdom and Ireland Medos Shunt Audit Group (1) firmly concluded that it is very dangerous in the context of a solid hematoma to increase the pressure setting, because one of their patients in whom that technique was used died. My current practice is to scan patients routinely after the insertion of a subdural valve to check for asymptomatic subdural fluid collection and to reduce the pressure setting only if there is no such fluid collection.

With regard to the definition of NPH, no threshold for the outflow resistance used is provided: "All infusion tests that led to shunt insertion were considered pathological." The results of the tap tests are not provided. The definition of the choice of initial setting requires further clarification. It would have been useful for the authors to comment about the value of resetting the valve for headache. It would have been helpful if *Table 9* had included deaths in reporting patient outcomes. I was unsure whether the cumulative shunt survival curves in *Figure 2* included or excluded deaths and whether the causes of death were known to exclude shunt-related complications. This important article contributes to the worldwide debate regarding the value of programmable valves, but I am alarmed by the suggestion that the pressure setting should be increased in attempting to treat a solid subdural hematoma!

John D. Pickard
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1. Kaye AD, Fisher AJ, O'Kane C, Richards HK, Pickard JD: A clinical audit of the Hakim programmable valve in patients with complex hydrocephalus: United Kingdom and Ireland Medos Shunt Audit Group. **Br J Neurosurg** 14:535–542, 2000.
2. Zemack G, Romner B: Do adjustable shunt valves pressure our budget? A retrospective analysis of 541 implanted Codman Hakim programmable valves. **Br J Neurosurg** 15:221–227, 2001.

The authors have conducted a serious and rigorous retrospective study of the use of an adjustable valve (Codman Hakim Programmable Valve; Codman & Shurtleff, Inc./Johnson & Johnson Co., Raynham, MA) in the treatment of patients with NPH syndromes. Beyond the well-documented aspect of this very detailed article, it is interesting to note in the Results that fewer complications were observed in the secondary NPH (SNPH) group than in the idiopathic NPH (INPH) group. As a matter of fact, patient outcomes were excellent or good in 78.9 and 69.8% of the patients in the SNPH and INPH groups, respectively. Subdural hematomas and hydromas complicated evaluation in 1.4 and 9.5% of the patients in the SNPH and INPH groups, respectively. In addition, adjustments of the valve were necessary less frequently in the SNPH group (32.4%; average, 0.69; shorter period of time) than in the INPH group (49.0%; average, 0.94; longer period of time). The better results achieved in patients with SNPH are classic and in accordance with the data in the literature.

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This article describes the outcomes of 218 patients with NPH who were treated with the Codman Hakim Programmable Valve. Five-year shunt survival was 80.2%. Nontraumatic subdural effusion occurred in 6.8% of the patients. The infection rate was 6.4%. Outcomes were excellent or good in 78.9% of patients with SNPH and in 69.8% of patients with INPH. Death as a result of causes unrelated to shunt surgery or failure occurred in 26% of the patients during the study. The authors recommend the use of this valve in patients with NPH because it affords the clinician the ability to adjust the opening pressure according to the patient's clinical response and to treat subdural effusion.

The study design is retrospective, and no controls were included. Although the study's objective is described as an evaluation of first-shunt implantation, 231 valves were implanted in 218 patients, so that some patients received an additional valve and were reentered into the same study. The authors created an improvement index to assess the effect of small and large valve adjustments. This index was not validated for inter- and intraobserver reliability, and, according to the authors, it was not very sensitive. The authors also were aware of the timing, magnitude, and direction of the valve adjustment. The clinical outcome assessment was performed with the use of a scale that has not otherwise been validated. The sole prospective, randomized trial to compare this valve with all others in a predominantly pediatric population demonstrated shunt revision rates that were equivalent, despite the ability to reprogram the valve (1). The results of the use of this valve in patients with NPH seem to be promising, but whether this valve is a better choice than other alternatives remains an unanswered question.

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1. Pollack IF, Albright AL, Adelson PD: A randomized, controlled study of a programmable shunt valve versus a conventional valve for patients with hydrocephalus: Hakim-Medos Investigator Group. **Neurosurgery** 45:1399–1411, 1999.

Among the potential advantages of programmable valve-regulated shunts is the ability to adjust cerebrospinal fluid (CSF) pressure (and presumably CSF flow rate) in a noninvasive and repetitive manner. This flexibility seems to be useful in the management of postoperative problems such as CSF underdrainage, CSF overdrainage, and subdural CSF collection. Despite this apparent advantage, however, it remains to be determined whether programmable valves perform as advertised and whether they are any better than more traditional differential-pressure valves in the long run.

In the current study, the authors provide a retrospective analysis of 218 patients with NPH who underwent ventricular

shunting procedures in which a Codman-Hakim Programmable Valve was used. The strengths of the report include its large database, comparatively long follow-up, and reasonable complication rates. Its weaknesses include the combination of data from 19 neurosurgeons, varying operative techniques (e.g., ventriculoperitoneal versus ventriculoatrial shunts), and the less-than-precise criteria for the opening pressure setting and reprogramming.

As the authors point out, the diagnosis of NPH remains tenuous, and no single diagnostic test reliably predicts the outcome of surgical treatment. The results of CSF infusion tests that the authors provide are of interest. It is the shared opinion of many neurosurgeons that a favorable response to lumbar CSF drainage in patients with NPH is the most reliable predictor of successful shunt outcome.

Overall, the results of the current study add to a growing body of literature suggesting that adjustable valves provide flexibility in the treatment of patients with NPH. Subdural CSF collection remains a major complication, and it is advisable in most cases to select a programmable valve that has an incorporated antisiphon device. More experience during a longer period is required before the absolute advantages of adjustable valves over differential-pressure valves can be established.

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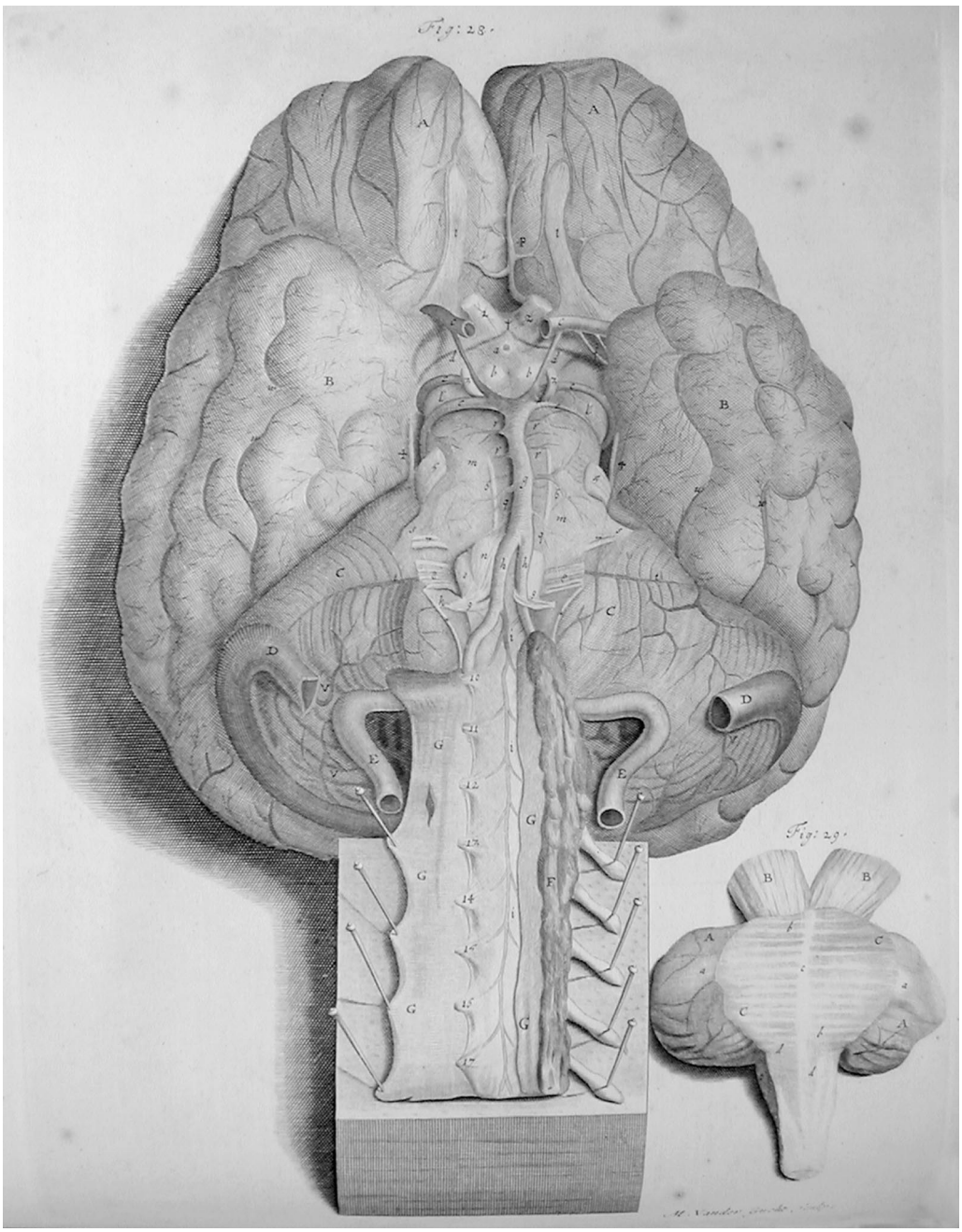

Plate from Cowper's The Anatomy of Humane Bodies. *(Also see pages 1364, 1372, 1476.) (Courtesy, Rare Book Room, Norris Medical Library, Keck School of Medicine, University of Southern California, Los Angeles, California.)*