Analysis of a Nationwide Survey on Treatment and Outcomes of Congenital Hydrocephalus in Japan

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Abstract

A nationwide questionnaire survey of congenital hydrocephalus in 2000 investigated the treatment and clinical outcomes for congenital hydrocephalus in Japan to evaluate the factors influencing clinical outcome. Surgical treatment was performed in 341 of 380 patients who survived the early neonatal period. Of 321 patients who had shunt operations, 295 (91.9%) underwent ventriculoperitoneal shunting and nine (2.8%) ventriculoatrial shunting. Programmable valves were used in 83 (33.6%) of the 247 patients at the first shunting and in 97 (39.3%) at the last shunting. The incidence of complications after the first shunting was 55.4% (46 of 83 patients) in the programmable and 61.6% (101 of 164) in the non-programmable valve groups. The types of shunt complication differed significantly between these groups (p < 0.001), as the incidence of shunt infection and malfunction was lower in the programmable valve group. Clinical outcome was generally better with later delivery stage during gestation (p < 0.02). The clinical outcome was statistically significantly better in term patients who underwent early shunt placement than in those who underwent late shunt placement (p < 0.05).

Key words: congenital hydrocephalus, nationwide survey, Japan, shunt, clinical outcome

Introduction

Congenital hydrocephalus is an etiologically heterogeneous central nervous system (CNS) abnormality characterized by an imbalance in the production and absorption of cerebrospinal fluid (CSF) leading primarily to enlargement of the ventricles, and may be associated with cerebral atrophy and mental retardation.10 The incidence of hydrocephalus in newborns is about 1:1000, and congenital hydrocephalus remains a major social and medical problem in infants.1,25 Surgical intervention is the treatment of choice for hydrocephalus, and CSF shunting is the most common and reliable method. Various treatment protocols for congenital hydrocephalus have been proposed, but many difficult problems remain unsolved, especially regarding the management of prenatally diagnosed hydrocephalus.27,28

The present analysis of the nationwide survey on congenital hydrocephalus in Japan in 2000 investigated the postnatal clinical features of children with congenital hydrocephalus, especially the relationship between treatment and clinical outcome, and the clinical features related to outcome.

Materials and Methods

A nationwide cooperative study on congenital hydrocephalus in Japan was performed in 2000 under the sponsorship of the Ministry of Health, Labor and Welfare of Japan. Congenital hydrocephalus was defined as increased retention of CSF in the ventricles associated with ventricular dilation diagnosed up to 12 months after birth.21 Congenital hydrocephalus was classified into three types: antenatally diagnosed (fetal hydrocephalus), postnatally diagnosed (infantile hydrocephalus), and diagno-
sis at unknown time (unspecified hydrocephalus). An increase in biparietal diameter was not essential for the diagnosis of fetal hydrocephalus at any stage of gestation.20)

This study consisted of a cross-sectional survey in two stages. The first survey was undertaken using a postcard questionnaire asking about the number of patients diagnosed as having congenital hydrocephalus who had visited the respective outpatient clinics or had been hospitalized during the period from January to December 1999. In January 2000, the questionnaire was sent to 2440 departments (pediatrics, neurosurgery, and obstetrics and gynecology) in Japan. An individual questionnaire, asking for epidemiological and clinical details, was sent to the 1861 departments (76.3%) that had reported cases of congenital hydrocephalus. Three hundred ninety-three patients with congenital hydrocephalus, 193 with fetal hydrocephalus, 181 with infantile hydrocephalus, and 19 with unspecified hydrocephalus, were identified by individual questionnaire. Thirteen patients were stillborn or died in the 1st week of life, and none of them received surgical treatment. The remaining 380 patients who survived 1 week or more were included in this study.

Clinical outcome was analyzed in relation to the timing of delivery and shunt placement. If a patient had undergone multiple shunt procedures, the first was taken as the datum point. Factors influencing clinical outcome were also investigated. All personal data items were subject to privacy protection.

Statistical analysis used the chi-squared ($\chi^2$) test, Mann-Whitney test, Kruskal-Wallis H test, and Spearman’s test, and were conducted with Excel 2004 (version 11.2.3; Microsoft, Redmond, Wash., U.S.A.) and SPSS (version 8.0 for Windows; SPSS, Chicago, Ill., U.S.A.) with significance accepted at the 5% level.

Results

I. Surgical treatment for hydrocephalus

Cephalocentesis, i.e. fetal head puncture, was performed in two cases at 30 and 35 weeks of gestation, respectively. Both patients received ventriculoperitoneal (VP) shunting after birth and were alive at 14 and 131 months, respectively. In this study, cephalocentesis was not included among the surgical treatments for hydrocephalus. None of the patients with hydrocephalus underwent fetal surgery such as ventriculooamniotic shunting or repair of meningocele.

Of the 380 patients who survived the early neonatal period, 341 (89.7%) had surgical treatment for hydrocephalus, 35 (9.2%) received no surgical treatment (non-surgical group), and four (1.1%) had unreported treatment. Of the 341 surgical cases, CSF shunt placement was done in 321 (94.1%) (shunt group) and only surgical treatments other than shunt procedures in 20 (5.9%) (non-shunt surgical group).

(i) CSF shunts

Timing of shunt placement: CSF shunting was classified into three types: early shunting (less than 1 week after birth), midterm shunting (from 1 week to less than 1 month), and late shunting (1 month or more). The time of delivery of the affected infants was classified into three gestational stages: before 32 weeks of gestation when the respiratory function of the fetus has not yet fully developed (pulmonary premature stage), from 32 to 36 weeks (fetal premature stage), and from 37 weeks onward (fetal mature stage).

Of the 10 patients delivered in the pulmonary premature stage, two (20%) underwent midterm shunting and eight (80%) late shunting. Of the 90 patients delivered in the fetal premature stage, 12 (13.3%) underwent early shunting, 31 (34.4%) midterm shunting, and 43 (47.8%) late shunting, and the remaining four (4.4%) were unreported. Of the 192 patients delivered in the fetal mature stage, 25 (13.0%) underwent early shunting, 62 (32.3%) midterm shunting, and 95 (49.5%) late shunting, and the remaining 10 (5.2%) were unreported.

Types of shunting and pressure valve system: Of the 321 patients who underwent shunt surgery, a VP shunt was inserted in 295 (91.9%), a ventriculooatrial (VA) shunt in nine (2.8%), a cyst-peritoneal shunt in eight (2.5%), and other types of shunt in nine (2.8%). Six of the nine patients with VA shunt received the procedure as revision surgery.

CSF shunting was classified into four types according to the pressure valve system used: programmable pressure (non-invasively pressure-adjustable), low-pressure, medium-pressure, and high-pressure valves. Table 1 shows the proportions of cases categorized by the type of pressure valve used in the first and the last CSF shunting. More cases used the programmable valve in the last shunting (39.3%) than in the first shunting (33.6%), though not statistically different ($p = 0.386, \chi^2$ test). Table 2 shows the transition of shunt types according to the pressure valve system in 20 patients who underwent multiple shunting. A programmable valve was adopted at first shunt implantation in one of 20 and at final shunt placement in 15 of 20.

Shunt complications: Of the 321 patients who underwent CSF shunt placement, 174 (54.2%) experienced operative complications and shunt revision was performed in 182 (56.7%). Among the 247
Table 1  Pressure valve system utilized for the first and the last cerebrospinal fluid shunting for congenital hydrocephalus

<table>
<thead>
<tr>
<th></th>
<th>First shunting</th>
<th>Last shunting</th>
</tr>
</thead>
<tbody>
<tr>
<td>Programmable pressure valve</td>
<td>83 (33.6)</td>
<td>97 (39.3)</td>
</tr>
<tr>
<td>Low-pressure valve</td>
<td>114 (46.2)</td>
<td>113 (45.7)</td>
</tr>
<tr>
<td>Medium-pressure valve</td>
<td>46 (18.6)</td>
<td>34 (13.8)</td>
</tr>
<tr>
<td>High-pressure valve</td>
<td>4 (1.6)</td>
<td>3 (1.2)</td>
</tr>
<tr>
<td>Total</td>
<td>247 (100)</td>
<td>247 (100)</td>
</tr>
</tbody>
</table>

Numerals show numbers of cases (%).

Table 2  Transition of the pressure valve system in 20 patients who underwent multiple shunting

<table>
<thead>
<tr>
<th>Transition of pressure valve system</th>
<th>Number of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>program -→ low</td>
<td>1</td>
</tr>
<tr>
<td>medium -→ low</td>
<td>3</td>
</tr>
<tr>
<td>low -→ medium</td>
<td>1</td>
</tr>
<tr>
<td>low -→ program</td>
<td>4</td>
</tr>
<tr>
<td>medium -→ program</td>
<td>10</td>
</tr>
<tr>
<td>high -→ program</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>20</td>
</tr>
</tbody>
</table>

high: high-pressure valve system, low: low-pressure valve system, medium: medium-pressure valve system, program: programmable pressure valve system.

patients with known valve system in the first CSF shunting, the incidence and each type of shunt complication were compared between patients who were treated with programmable pressure valves (programmable valve group) and those treated with non-programmable valves (non-programmable valve group). Shunt complications occurring within 3 months after first shunt placement were analyzed because of the cumulative nature of the rate of occurrence. Shunt complications occurred in 46 (55.4%) of the 83 patients in the programmable valve group, and in 101 (61.6%) of 164 patients in the non-programmable valve group. The incidence of shunt complications was slightly lower in the programmable valve group than in the non-programmable valve group (p = 0.35).

Table 3  Types of complications occurring within 3 months after first shunt placement for congenital hydrocephalus using a programmable valve and a non-programmable valve

<table>
<thead>
<tr>
<th></th>
<th>Programmable valve</th>
<th>Non-programmable valve</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infection</td>
<td>11 (23.9)</td>
<td>38 (37.6)</td>
</tr>
<tr>
<td>Shunt malfunction</td>
<td>13 (28.3)</td>
<td>44 (43.6)</td>
</tr>
<tr>
<td>Subdural hematoma/fluid collection</td>
<td>10 (21.7)</td>
<td>4 (4.0)</td>
</tr>
<tr>
<td>Slit ventricle</td>
<td>2 (4.3)</td>
<td>9 (8.9)</td>
</tr>
<tr>
<td>Others</td>
<td>10 (21.7)</td>
<td>6 (5.9)</td>
</tr>
<tr>
<td>Total</td>
<td>46 (100)</td>
<td>101 (100)</td>
</tr>
</tbody>
</table>

Numerals show numbers of cases (%).

Table 3 shows the rate of types of complication in CSF shunting for congenital hydrocephalus occurring within 3 months after shunt placement using a programmable valve and a non-programmable valve. The types of shunt complication differed significantly between the groups, that is, the incidences of shunt infection and shunt malfunction were significantly lower in the programmable than in the non-programmable valve group (p < 0.001, χ² test).

(ii) Operations other than shunting

Of the 341 surgically treated infants who survived the early neonatal period, 176 (51.6%) underwent operations other than shunt insertion as the initial surgical treatment for hydrocephalus. Forty-six (26.1%) of these 176 patients underwent insertion of external ventricular drainage (EVD), 40 (22.7%) placement of an Ommaya reservoir, 19 (10.8%) endoscopic third ventriculostomy (ETV), 40 (22.7%) other operations, and in the remaining 31 (17.6%) unknown procedure.

A shunt was later inserted in 45 (97.8%) of the 46 patients who underwent EVD, 38 (95%) of 40 patients who underwent placement of an Ommaya reservoir, and nine (47.4%) of 19 patients who underwent ETV.

II. Clinical outcomes

The clinical outcomes in terms of intelligence quotient or developmental quotient and the extent of physical disability in daily life were classified into four categories using modified official guidelines utilized for the comprehensive assessment of mental and physical handicaps in Japan (Table 4): normal (work or study possible), slightly disturbed (independent daily life possible), moderately disturbed (partial care required for activities of daily life), and severely disturbed (care required for all activities of daily life).

(i) Lower age limit for clinical outcome evaluation

In the present study which involved a retrospec-
Table 4 Criteria for comprehensive assessment of the degree of mental (developmental) retardation and physical disability in daily life used officially in Japan*

<table>
<thead>
<tr>
<th>Physical disability</th>
<th>Mental retardation (IQ/DQ)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0 Normal</td>
<td>≥85 84–75 74–50 49–25 &lt;24</td>
</tr>
<tr>
<td>I Slight disability</td>
<td></td>
</tr>
<tr>
<td>II Mild disability</td>
<td></td>
</tr>
<tr>
<td>III Moderate disability</td>
<td></td>
</tr>
<tr>
<td>IV Severe disability</td>
<td></td>
</tr>
</tbody>
</table>

*Modified from Watanabe and Negoro.34) [Normal, | slightly disturbed, | moderately disturbed, | severely disturbed. IQ/DQ: intelligence quotient or developmental quotient.

Table 5 Clinical outcome in three patient groups categorized by treatment for congenital hydrocephalus

<table>
<thead>
<tr>
<th></th>
<th>Shunted group</th>
<th>Non-shunted surgical group</th>
<th>Non-surgical group</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal range/ slightly disturbed</td>
<td>99 (52.7)</td>
<td>7 (58.3)</td>
<td>5 (38.5)</td>
</tr>
<tr>
<td>Moderately disturbed</td>
<td>43 (22.9)</td>
<td>2 (16.7)</td>
<td>2 (15.4)</td>
</tr>
<tr>
<td>Severely disturbed/ Dead</td>
<td>46 (24.5)</td>
<td>3 (25.0)</td>
<td>6 (46.2)</td>
</tr>
</tbody>
</table>

Total 188 (100) 12 (100) 13 (100)

Numerals show numbers of cases (%).

**Table 5 Clinical outcome in three patient groups categorized by treatment for congenital hydrocephalus**

Table 5 shows clinical outcome in three patient groups categorized by treatment for congenital hydrocephalus (shunted, non-shunted surgical, and non-surgical groups). Although the clinical outcome was better in the surgical group than in the non-surgical group, there was no significant difference in clinical outcome between the shunted, non-shunted surgical, and non-surgical groups (p = 0.235, Kruskal-Wallis H test), or between the surgical (shunted and non-shunted surgical) and non-surgical groups (p = 0.919, Mann-Whitney test). Two patients who underwent Ommaya reservoir implantation and 10 who were treated by ETV had good clinical outcomes without requiring shunts.

**Table 5 Clinical outcome in three patient groups categorized by treatment for congenital hydrocephalus**

(i) Clinical outcome of cohort

The follow-up time for evaluation of outcome varied greatly. Therefore, as data analysis requires a lower age limit for patients with congenital hydrocephalus to allow reasonable evaluation of clinical outcomes, semi-quantitative analysis was performed. A score for the grade of clinical outcome was assigned to each patient as follows: normal range 5 points, slightly disturbed 4 points, moderately disturbed 3 points, severely disturbed 1 point, and died 0 point. Then, both mean outcome score and rate of indeterminate reply regarding clinical outcome were examined as the patient aged. As shown in Fig. 1, the mean outcome score increased rapidly from around 1 to 3 until about 12 months of age, and thereafter stabilized at around 3. The rate of indeterminate reply was about 70% or more in patients aged less than 12 months, became less than 50% in patients aged 12 months or more, and was almost 0% thereafter. These features seem to reflect the low patient age, or difficulty in assessing outcomes in patients aged less than 12 months. Taking these facts into consideration, final clinical outcomes of congenital hydrocephalus were adopted in the present study only in patients with last assessment at the age of 12 months or more.

(ii) Comparison of clinical outcomes between three groups divided by the way of treatment

Of the 214 patients with a valid reply about clinical outcome at the age of 12 months or more, 188 (87.9%) belonged to the shunted group, 12 (5.6%) to the non-shunted surgical group, 13 (6.1%) the non-surgical group, and one (0.5%) to the unknown group. Table 5 shows clinical outcome in three patient groups categorized by treatment for congenital hydrocephalus (shunted, non-shunted surgical, and non-surgical groups). Although the clinical outcome was better in the surgical group than in the non-surgical group, there was no significant difference in clinical outcome between the shunted, non-shunted surgical, and non-surgical groups (p = 0.235, Kruskal-Wallis H test), or between the surgical (shunted and non-shunted surgical) and non-surgical groups (p = 0.919, Mann-Whitney test). Two patients who underwent Ommaya reservoir implantation and 10 who were treated by ETV had good clinical outcomes without requiring shunts.

(iii) Clinical outcomes in relation to timing of delivery and shunt placement

The following analysis of clinical outcome was performed only on shunted patients because the number of patients without shunting was low and
such patients were considered to have various associated factors that might have affected clinical outcome, either deteriorating factors such as intracranial hemorrhage or infection and improving factors such as only minor hydrocephalus, either of which might have contraindicated shunt placement.

Figure 2 shows the clinical outcomes of 159 patients with valid replies for time of delivery, time of shunt operation, and clinical outcome. The patients comprised seven (4.4%) who were born before 32 weeks of gestation, 38 (23.9%) from 32 to 36 weeks of gestation, and 114 (71.7%) at the fetal mature stage. Later delivery during gestation is associated with better clinical outcome. The clinical outcome was significantly correlated with gestational age at birth ($p < 0.02$, Spearman’s test). Although none of the seven patients born before 32 weeks of gestation died, good outcome was obtained in only one patient who underwent late shunt placement (Fig. 2 upper). Of 38 patients born from 32 to 36 weeks of gestation, only five (13.2%) underwent shunt placement in the 1st week after birth, and there was no evident trend in the relationship between timing of shunt placement and clinical outcome ($p = 0.370$, Spearman’s test) (Fig. 2 middle). On the other hand, for the 114 term patients, the clinical outcome was statistically significantly better in those who underwent early shunt placement than in those who underwent late shunt placement ($p < 0.05$, Spearman’s test) (Fig. 2 lower).

Discussion

I. CSF shunting

Although neuroendoscopy has been increasingly applied to the treatment of hydrocephalus, CSF shunting is still the most effective and reliable method from the practical viewpoint.32) Despite progress in early detection of congenital hydrocephalus, shunt technology and therapeutic strategies, the overall outcome of patients remains unsatisfactory. A nationwide survey of current treatment for hydrocephalus in Japan in 1996 observed a trend for simplification of the CSF shunting system by concentrating on programmable pressure valves and open end-type peritoneal catheters, and for increasing use of programmable pressure valves.19) The present survey revealed that VP shunting was used in almost all patients undergoing shunt placement, whereas VA shunting was employed in very few and as revision surgery in two thirds of cases. In more than 80% of the patients surveyed, either a programmable or low-pressure valve was used, the latter being the first-choice device. However, a programmable valve was finally adopted in 15 of the 20 patients who underwent multiple shunting as shown in Table 2.

The rate of shunt complication is reported to be highest in post-hemorrhagic preterm children.5,6,10,26] A retrospective review of pediatric patients undergoing shunt placements and revisions with VP shunts demonstrated that the rate and etiologies of shunt failure were similar to those in previous decades, and that shunt life span remained shorter in patients who undergo shunt revisions and in younger patients, but the etiology of hydrocephalus was not associated with an increased risk of shunt failure.18] Although the present survey revealed that the rate of shunt complications in patients who were treated with a programmable valve (55.4%) was
almost the same as that in those treated with a non-programmable valve (61.6%), the types of complication significantly differed between the groups. Infection and shunt malfunction, the most serious among shunt complications, were less frequent in the programmable valve group than in the non-programmable valve group. This result suggests use of a programmable valve may reduce the incidence of the most difficult shunt complications.\textsuperscript{13,14,19,35} However, the intrinsic programmable valve malfunction rate was 11.1%, whereas no intrinsic valve malfunction occurred in non-programmable valve systems used during the same period for similar causes of hydrocephalus.\textsuperscript{10} Further prospective, randomized evaluation will elucidate specific indications for programmable valve systems and better determine the reliability of these valves in the pediatric population.

II. Treatments other than shunting

EVD or implantation of an Ommaya reservoir\textsuperscript{26} were the treatment of choice for babies with insufficient body weight, or with posthemorrhagic or postinfectious hydrocephalus. In almost all our patients treated with EVD or an implanted Ommaya reservoir, shunt surgery was subsequently performed. These results suggest that these treatments are a temporary measure to gain time before CSF shunt insertion. Although primary shunting has been recommended in very low birth weight infants with high protein concentrations resulting from intraventricular hemorrhage, temporary measures are generally most desirable until the child weighs between 1200 and 2000 g and the protein level falls below 500 mg/dL.\textsuperscript{27} A small number of patients underwent ETV because at the time of this nationwide survey ETV had not become commonly used. In about a half of patients undergoing ETV, shunt surgery was not subsequently performed. About a half of the patients who were treated by ETV had good clinical outcomes without shunts. These observations suggest that ETV is excellent for treatment of hydrocephalus providing the surgical indications are correctly determined. Although the failure rate and complication rate following ETV in infants younger than 1 year of age, in whom the CSF absorption system is not yet fully developed, is higher than that in older children,\textsuperscript{22,30} the success or failure of ETV is related to etiology rather than age.\textsuperscript{5} As a result of technical and medical developments, ETV will continue to be the best choice because the procedure is safe with few complications and has a high success rate at least for non-communicating hydrocephalus in pediatric patients aged 1 or 2 years or more.\textsuperscript{23} Recently, ETV has become the treatment of choice, even in secondary groups where hydrocephalus arises because of intraventricular hemorrhage and meningitis,\textsuperscript{4,24,33} or for children aged less than 1 year.\textsuperscript{6,27}

III. Outcome

Although many studies have investigated the clinical outcomes of children with hydrocephalus after shunting, there is a surprising paucity of long-term follow-up studies.\textsuperscript{1,2,9,10,12,15,17} The present study excluded patients whose age at the last assessment of clinical outcome was less than 1 year. The 10-year survival of surgically treated patients is more than 60%, half of whom live independently and have normal intelligence.\textsuperscript{15,17} An analysis of long-term outcome in hydrocephalic children found that mortality was far from satisfactory, although postoperative mortality was virtually nil and the overall mortality rate after 10 years of follow up was as little as 5% in those who underwent shunt placement.\textsuperscript{10}

The criteria shown in Table 4, modified from the classification used officially in Japan,\textsuperscript{34} were used to comprehensively score both physical and developmental (mental) aspects because of the complex nature of impairment of the musculoskeletal system in spina bifida patients. Various methods for scoring disability associated with hydrocephalus have been proposed, such as the international classification of impairments, disabilities, and handicaps.\textsuperscript{31} The ultimate goal of the treatment of infants with hydrocephalus is to allow the patient to eventually be able to assume full responsibility for life and live independently.\textsuperscript{21} The significant highest shunt revision rate and highest incidence of epilepsy occur in post-meningitis hydrocephalus patients.\textsuperscript{12} Mental retardation, cerebral palsy, and epilepsy are significantly more frequent in children with infantile hydrocephalus in comparison with those with meningomyelocele.\textsuperscript{25} Comparable outcome studies have used intelligence quotient as a measure of mental development and the proportion of hydrocephalus patients with normal intelligence was reported to vary from 50% to 63%. However, not all studies have used the same definition of normal intelligence and the composition of the study populations has varied considerably.\textsuperscript{1,17,19} Another good measure might be whether the child can attend a normal school or not, as integration into the normal school system was possible for 60% of the children but half of them were 1–2 years behind their age group or having difficulties.\textsuperscript{10}

The present study could not show better clinical outcome in the shunt group than in the non-shunt group, but suggested that early surgery is optimum for term infants and delay in surgical treatment.
increase the risk of poor outcome. However, any
decision about surgery is made by considering the
prognosis on the basis of related factors in each in-
dividual case. Serious cases such as those with in-
tracranial hemorrhage or infection might not be
eligible for surgery, especially early shunting, be-
cause of the poor prognosis related to the serious-
ness and/or complexity of the disease, associated
diseases both in and outside the CNS, and high risk
of surgical complications including shunt malfunc-
tion or infection.9,10,21) On the other hand, surgery
might be contraindicated because of the mildness of
the condition or likelihood of a good outcome. Most
reports on the effects of CSF shunting on
hydrocephalus, including the present one, have in-
cluded few data on decision making about the timing
of shunt implantation, except in children with
spina bifida and with hemorrhage.

Age at initial shunt placement is known to affect the
outcome.1,7,8,11,15,27) Delay in surgical treatment
might also be a risk factor for poor outcome in
hydrocephalic children.3) Delay in drain implan-
tation until more than 1 month after diagnosis appar-
ently increased the probability of impairment of
mental development, such as speech and language,
and possibly learning.21) Mental and linguistic de-
velopment was poorer in children who underwent
surgery more than 1 month after diagnosis, suggest-
ing that delay in surgical treatment might be a risk
factor for poor outcome in hydrocephalic chil-
dren.9,15,27) The highest incidence of shunt inde-
dependence occurred in patients treated in early in-
fancy, possibly due to delayed maturation of the
CSF absorption mechanism, followed by late nor-
malization in these patients.11)

The pathology, in addition to the hydrocephalus,
has a negative influence on the prognosis.7,9,12,29) The
etiology may be a major determinant of outcome in
children with early postnatal surgical treatment of
fetal hydrocephalus.7) A practical clinical classifica-
tion was proposed based on the onset of
hydrocephalus and its etiology, and demonstrated
that children with congenital causes of hydroceph-
alus such as spina bifida appeared to have a better
outcome in several developmental aspects compared
with children with acquired causes.21) This feature is
compatible with other studies.1,2,11) The relationship
between the etiological facts of hydrocephalus and
clinical outcomes of infants in our survey will be
analyzed in following studies.

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Commentary

The authors have done excellent work collecting cases of congenital hydrocephalus in a Japanese nationwide investigation relating to the newborns of year 2000. The problem they address is a serious one, and they try to give answers to a series of difficult questions. The data collection is very accurate and the grouping of patients seems to me correct and sharable. The methods are careful and thorough. The results are very interesting. On one side they confirm the general feeling that, for these difficult patients, the “cure” for congenital hydrocephalus is far from being achieved. The authors report no global difference in outcome between operated and nonoperated patients (but see the limitation to this result posed by the etiology of...
hydrocephalus). On the other hand, they prove that, among the surgical group, the best results have been reached by patients operated upon earlier than the others, so the shunt, when indicated, should be carried out as soon as possible. In the Discussion, the authors correctly point out that the neuropsychological outcome of patients depend primarily on the etiology of hydrocephalus, more than on the ventricular dilation. Other data are relevant, for instance, the better performance of adjustable valves in terms of infection and mechanical obstruction is at the moment compensated by the intrinsic damageability of this type of shunt. Altogether, this is really a work worth reading.

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Moritake et al. present their Analysis of a Nationwide Survey on Treatment and Outcomes of Congenital Hydrocephalus in Japan. They analyzed the data obtained from a nationwide cooperative study performed in Japan in 2000. The paper shows a very detailed data collection of the most relevant aspects of this subject with an excellent and comprehensive discussion. The lower frequency of serious complications, infection and shunt malfunction, in the programmable valve group is very interesting. Endoscopic third ventriculostomy (ETV) was indicated only in 19 cases (10.8%), but the authors explain this fact because at the time of this nationwide survey ETV had not become commonly used. The authors pointed out as well, that ETV has become the treatment of choice, even in secondary groups where hydrocephalus arises because of intraventricular hemorrhage and meningitis or for children aged less than 1 year. The authors must be congratulated for their effort. We’ll look forward to the future publication about the relationship between the etiological facts of hydrocephalus and clinical outcomes of infants.

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