Long-term Functional Evaluation of Congenital Hydrocephalus in Infants and Children

SHOKO SHIMOKAWA AND TAKASHI HAYASHI

Department of Neurosurgery, Institute of Neurosciences, St. Mary’s Hospital, Kurume 830-8543, Japan

Summary: In the past 20 years, 43 patients with congenital hydrocephalus underwent surgical treatment for hydrocephalus in our clinic (Myelomeningocele, Dandy-Walker syndrome, holoprosencephaly and hydranencephaly, which are frequently associated with dysgenesis of the corpus callosum, were excluded). The gestational stage at the onset of hydrocephalus was assessed on the basis of the morphology of the corpus callosum. The patients were then divided into four groups according to stage of onset. A relation between the stage of onset and the long-term functional scores was demonstrated. The earlier the gestational stage at which the hydrocephalus occurred, the lower was the long-term functional score.

Key words Congenital hydrocephalus, corpus callosum, gestational stages, long-term outcome

INTRODUCTION

The outcomes of patients with congenital hydrocephalus depend on multiple factors. It has hitherto been reported that the earlier the hydrocephalus occurs in the gestational period, the less favorable the prognosis of the patients is [3]. Although this conclusion seems plausible, no report is available in which stages of onset of congenital hydrocephalus were assessed after birth to substantiate the above conclusion. This study was designed to examine retrospectively the gestational stages of onset of hydrocephalus and to search for a possible relationship between stages of onset and results of the long-term evaluation of patients. Therefore, we analyzed images obtained by MRI in order to assess stages of onset of congenital hydrocephalus. Between 8 and 15 weeks of gestation, the precursor for ingrowth of the callosal fibers, the sulcus medianus telencephali medii and massa commissuralis, forms in the dorsal part of the lamina reuniens. Eventually, the callosal fibers themselves begin to decussate into the massa commissuralis, in the genu at approximately 11 weeks, and then in the body, splenium and rostrum at 18-20 weeks of gestation [1]. An early insult which results in incomplete formation of the callosal precursor may cause a total or partial callosal defect [5,6]. The cases in which hydrocephalus occurred after 20 weeks gestation could be further classified into two groups. Numaguchi [2] has reported that after the postnatal placement of a V-P shunt, the shape of the corpus callosum is found to be nearly normal if the ventricles have been enlarged only for a short period. If the ventricles have been kept enlarged for a long period, on the other hand, a characteristic deformation called scalloping appears on the dorsal surface of the corpus callosum. Therefore, we assessed stages of onset of congenital hydrocephalus depending on the shape of the corpus callosum, and examined the relationship between stages of onset and long-term functional prognosis.

MATERIALS AND METHODS

Materials were the patients with hydrocephalus who had been admitted to the Neonatal intensive care unit of St. Mary Hospital and treated with shunt placement from January 1980 through December 1999. (cases of meningomyelocele, Dandy-Walker syndrome, holoprosencephaly and hydranencephaly,
Fig. 1. a: Group 1 is agenesis of corpus callosum. b: Group 2 is partial callosal dysgenesis. c: Group 3 is severe deformity of corpus callosum. d: Group 4 is mild deformity of corpus callosum.
which are frequently associated with callosal dysgenesis, were excluded from this study.) During this 20-year period, the number of patients with hydrocephalus was 104. Out of these 104 cases, 43 cases underwent shunting operations. Of these 43 patients, 10 had simple hydrocephalus (a type with aqueduct stenosis), 21 had dysgenetic hydrocephalus, 10 had hydrocephalus induced by other disorders, and the remaining two had hydrocephalus of unknown origins. Of the 21 cases of dysgenetic hydrocephalus, 11 had dorsal cyst malformation, 6 had encephalocele, and 4 had arachnoid cyst. Of the 10 cases with hydrocephalus induced by other disorders, 3 had congenital posthemorrhagic hydrocephalus, 2 had neurofibromatosis type I, 3 had unilateral hydrocephalus, 1 had congenital toxoplasmosis and the remaining one had congenital malformation syndrome. We performed a clinical analysis of these 43 shunting cases in order to clearly document their long-term outcome.

Relationship between gestational stages and onset of hydrocephalus based on the morphological change of the corpus callosum

The morphological change of the corpus callosum was evaluated postoperatively with sagittal T1-weighted MR images in the 43 cases of congenital hydrocephalus. Deformations of the corpus callosum were classified into the following four types: Agenesis, partial defect, severe deformity and mild deformity. Agenesis of the corpus callosum was presumed to indicate that ventricular dilatation had interfered with the development of the corpus callosum. As a result, various abnormality on configuration according to occurrence of hydrocephalus in its grade and stage in fetus. Group 1: Agenesis of the corpus callosum was presumed to indicate that hydrocephalus had occurred before 7 weeks of gestation (Fig. 1a). Group 2: Partial callosal dysgenesis was presumed to indicate that hydrocephalus had occurred between 7 and 20 weeks gestation (Fig. 1b). Group 3 and 4: Severe (Group 3) or mild (Group 4) deformity was presumed to indicate that hydrocephalus had occurred after 20 weeks gestation (severe; Fig. 1c, mild; Fig. 1d), namely after the formation of the corpus callosum. A hydrocephalus with severe deformity suggests that the hydrocephalus had lasted for relatively long time during the development of the corpus callosum. On the other hand, when the interval between the onset of hydrocephalus and the shunting operation was short, the morphological state of the corpus callosum remained almost normal. Patients were divided into four groups according to degree of the callosum dysgenesis, and the IQ or DQ in the different groups was studied in the long-term follow-up. Long-term functional evaluation of the patients

IQ and DQ tests, performed in the outpatient clinic in this hospital, were used for the functional evaluation of the children included in this study. The IQ test to be performed in each patient was selected according to his or her chronological and mental age. When the chronological age alone was taken into consideration, the Tanaka-Binet intelligence test was used for children aged 3 years or less, the WPPSI test for those between 4 and 6 years of age, the WISCII test for those between 6 and 17 years and the WAIS-R test for those over 17. Practically, however, a test applicable to each child was selected depending on the degree of his or her disability. As for the DQ tests, the analytical developmental assessment test for infants (Tsumori and Inege, 1961) was performed. For children with pronounced disabilities, the SM social maturity scale-revised was used. When both IQ and DQ tests were applicable, the function of children was evaluated mainly on the basis of the results of an IQ test.

RESULTS

Of the 43 cases of congenital hydrocephalus, 10 had simple hydrocephalus (a type with aqueduct stenosis), 21 had dysgenetic hydrocephalus and 10 had hydrocephalus induced by other disorders. The mean long-term functional scores as evaluated by IQ and DQ tests were 44.4 in cases of congenital hydrocephalus as a whole, 63.3 in cases of simple hydrocephalus, 50.3 in cases of dysgenetic hydrocephalus. This result indicates that scores were lower in the cases of dysgenetic hydrocephalus. The number of cases having IQ scores higher than 75, namely the number of cases believed to be capable of learning is 10 (27.0%) in all of the cases of congenital hydrocephalus as a whole, 63.3 in cases of simple hydrocephalus, 50.3 in cases of dysgenetic hydrocephalus. This result indicates that scores were lower in the cases of dysgenetic hydrocephalus. The number of cases having IQ scores higher than 75, namely the number of cases believed to be capable of learning is 10 (27.0%) in all of the cases of congenital hydrocephalus, 63.3 in cases of simple hydrocephalus, 50.3 in cases of dysgenetic hydrocephalus and 6 (31.6%) in cases of dysgenetic hydrocephalus. All of the 43 cases of congenital hydrocephalus were examined with MRI. They were divided into four groups according to the shape of the corpus callosum (Table 1). Agenesis of the corpus callosum was found in 12 cases, partial defect in 6 cases, severe deformity in 8 cases, and mild deformity in 17 cases. In the 10 cases of simple hydrocephalus, agenesis of the corpus callosum was never found,
TABLE 1.
The relationship between the Group and the long-term functional score

<table>
<thead>
<tr>
<th>Group</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>total cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Simple hydrocephalus</td>
<td>0</td>
<td>1</td>
<td>3</td>
<td>6</td>
<td>10</td>
</tr>
<tr>
<td>Dysgenetic hydrocephalus</td>
<td>7</td>
<td>5</td>
<td>3</td>
<td>6</td>
<td>21</td>
</tr>
<tr>
<td>Other hydrocephalus</td>
<td>5</td>
<td>0</td>
<td>2</td>
<td>5</td>
<td>12</td>
</tr>
<tr>
<td>Total cases</td>
<td>12</td>
<td>6</td>
<td>8</td>
<td>17</td>
<td>43</td>
</tr>
</tbody>
</table>

Cases of congenital hydrocephalus were divided into Group 1 to 4 according to the shape of the corpus callosum (Group 1: agenesis of corpus callosum. Group 2: partial callosal dysgenesis. Group 3: severe deformity of corpus callosum. Group 4: mild deformity of corpus callosum.).

Whereas partial defect, severe and mild deformities were found in 1, 3 and 6 cases, respectively. This result suggests that simple hydrocephalus occurred in relatively later stages of gestation. In the 21 cases of dysgenetic hydrocephalus, agenesis of the corpus callosum was found in 7 cases, partial defect in 5 cases, severe deformity in 3 cases and mild deformity in 6 cases. This result suggests that dysgenetic hydrocephalus occurred in early stages of gestation.

The relationship between the shape of the corpus callosum and the functional evaluation was examined in the 43 subjects. The mean functional scores were 26.7 in cases with agenesis, 23.5 in cases with partial defect, 32.0 in cases with severe deformity, and 65.3 in cases with mild deformity. This suggests that the earlier the hydrocephalus occurred, the more pronounced were the disabilities (Fig. 2).

In addition to the relation between stage of onset of hydrocephalus and gestational period, other factors related to long-term functional prognosis were examined. As hitherto reported, a past history of fetal distress and respiratory distress syndrome was found to be an unfavorable factor. Other unfavorable factors were low birth-weight, abnormal head circumference, symptomatic epilepsy (especially uncontrollable type) and number of operations for hydrocephalus.

**DISCUSSION**

In the past 20 years, 207 patients with hydrocephalus were admitted to this hospital. Of these, patients with myelomeningocele, Dandy-Walker syndrome, holoprosencephaly and hydranencephaly were excluded from this study. There are relatively numerous patients with myelomeningocele. In our previous study, we have found that since myelomeningocele is often accompanied by dysfunction of the lower limbs, their functional scores may be evaluated unduly low by assessment tests which include measurement of their movement ability [4]. Patients with myelomeningocele were excluded, therefore, because their IQ and DQ scores could not be simply compared with those of patients having other subtypes of hydrocephalus. Since patients with Dandy-Walker syndrome, holoprosencephaly and hydranencephaly often show hypogenesis of the corpus callosum, they were also excluded. Besides, patients with mild hydrocephalus who were diagnosed as having the disease but were kept under observation without surgery were excluded. Therefore, 43 cases of hydrocephalus which underwent a surgical treatment were subjects of this study.

The shape of the corpus callosum remained almost unaffected in many patients with simple hydrocephalus. This suggests that hydrocephalus occurred in later stages of gestation. In the 21 cases of dysgenetic hydrocephalus, agenesis of the corpus callosum was found in 7 cases, partial defect in 5 cases, severe deformity in 3 cases and mild deformity in 6 cases. This result suggests that dysgenetic hydrocephalus occurred in early stages of gestation.

When the relationship between the shape of the corpus callosum and the long-term functional score
was studied, it was found that the more marked was the callosal deformity, the poorer was the score. If hydrocephalus may be assumed to have occurred in earlier stages in patients with more marked deformity, the above result indicates that the score of patients in whom hydrocephalus had occurred earlier tended to be worse. The results of this study suggest, therefore, that long-term prognosis of patients can be predicted by scrutiny of the shape of the corpus callosum by MRI postnatally. Some patients with total defect of the corpus callosum, however, had IQ scores higher than 75. Therefore, the hope in future should not be abandoned even if the corpus callosum is markedly deformed.

CONCLUSIONS

The earlier the hydrocephalus occurred in gestation, the poorer were results of the long-term functional evaluation. A propensity was noted for the functional score to be lower in cases of dysgenetic hydrocephalus than in those of simple hydrocephalus (hydrocephalus occurred earlier in the former than in the latter). To determine which is a more prominent risk factor, a complication with malformations of the brain or an earlier occurrence of hydrocephalus, seems to contribute to decide the appropriate postnatal time for a surgical treatment of patients with congenital hydrocephalus.

REFERENCES