The prognosis of hydrocephalus overt at birth

John Mealey, Jr., M.D., Richard L. Gilmor, M.D., and Michael P. Bubb
Division of Neurological Surgery, Indiana University Medical Center, Indianapolis, Indiana

The clinical records of 79 infants with congenital hydrocephalus and excessive head size at birth were reviewed to determine the prognosis and results of surgical treatment. In 55 infants hydrocephalus was the principal malformation, and in 24 this condition occurred in association with a myelomeningocele. Follow-up was complete in 69 cases. Of 27 unoperated cases only two are living, and one of these, an achondroplastic dwarf, has normal intelligence. Only one of eight patients treated surgically before Holter valve shunts were introduced has survived, and he is moderately retarded. Of 34 infants treated by ventriculointestinal shunting, 15 have lived up to 15 years and an average of nearly 7 years; five of these survivors have normal intelligence, five are trainable, and six are grossly retarded. Ventricular shunting was accompanied by a significant persisting morbidity, and operative complications, especially sepsis, accounted for 42% of the deaths in this group. Relative head size determined at birth and the type of hydrocephalus demonstrated had no conclusive association with life or death among the uniformly megaloccephalic cases. The results showed a positive correlation between early operation for hydrocephalus and survival in the group treated by ventriculointestinal shunting, and this treatment is recommended.

Key Words - hydrocephalus at birth - myelomeningocele - ventriculointestinal shunts - intelligence - mental retardation

Infantile hydrocephalus, whether congenital in origin or acquired, does not become manifest and is not ordinarily diagnosed in the majority of affected infants until some time after birth. The natural history of this disorder and improved prognosis with current methods of ventricular drainage have been well-documented in recent years. There is, however, a paucity of information on the fate of the relatively small number of infants with congenital hydrocephalus whose heads are grossly enlarged at the time of birth. Congenital hydrocephalus is usually diagnosed before, during, or after delivery in 0.1% to 0.4% relative to live births.1,8 Obstetrical experience with hydrocephalic newborns has been notably pessimistic. In a 20-year review to 1952, Feeney and Barry5 found that only 10 of 93 live-born hydrocephalic infants survived long enough to leave the maternity hospital. In a 30-year survey to 1965 at the Mayo Clinic, Larson and Banner9 reported that only four of 35 hydrocephalic infants
born alive survived long enough for corrective surgery. Nearly half to two-thirds of the afflicted fetuses in these surveys were stillborn, and up to 40% had associated myelomeningoceles. The prognosis of congenital hydrocephalus with presentation at birth has not been defined in live-born patients of this type who are referred for possible neurosurgical treatment.

This paper analyzes the clinical findings, treatment, and outcome of a selected group of infants who were obviously hydrocephalic as newborns and were admitted to the Indiana University Medical Center from 1950 through 1969.

Clinical Material

Case Selection

Infants with congenital hydrocephalus were included in this study if the occipitofrontal circumference (OFC) was recorded at 37 cm or greater on the first day postpartum or when from the hospital records there were clear indications of abnormal head enlargement being present since birth. A head circumference of 37 cm in a newborn approximates the 98th percentile for males and females. Seventy-nine cases of this type were disclosed in a review of available medical records at this institution for the period 1950–1969. Forty-three patients were admitted during the first neonatal week and 59 before the age of 1 month; 16 other infants were under 6 months of age and four were older when first seen. Cases were excluded when there was a known history of perinatal infection or when the diagnosis of megalencephaly, toxoplasmosis, or brain tumor was established. Of the 79 cases, hydrocephalus was the primary malformation in 55, and developed in 24 others in association with a myelomeningocele. Racial and sex distributions were as follows: 75 Caucasians, 4 Negroes; 46 males, and 33 females. Follow-up was complete in 69 cases including all 24 infants with hydrocephalus associated with myelomeningocele.

Obstetrical and Family History

Gestational illnesses were recorded in only eight of the mothers bearing the hydrocephalic infants in this study, ranging from threatened abortion (5) and polyhydramnios (1) to presumed viral respiratory infections (2). Vaginal delivery was performed in 51 cases, including nine breech extractions, and 23 infants were delivered by Caesarean section. Three infants in the former group and two of those born by Caesarean section required ventricular aspiration to facilitate delivery of the enlarged head. In five cases the manner of delivery was unknown. A family history of major congenital malformation was obtained in seven cases, including congenital heart disease (1), myelomeningocele (1), and confirmed or probable hydrocephalus (5).

Head Circumference

The occipitofrontal circumference had been measured on the day of birth in 60 patients in this series and ranged from 37 to 58 cm, with a mean of 41.5 cm (Fig. 1). Average head size was slightly smaller (OFC 40.5 cm) in the 24 cases with myelomeningocele and, as expected, considerably larger (OFC 46 cm) in those infants delivered by Caesarean section. The infants whose heads were considered enlarged at birth had OFC measurements ranging from 39 to 48 cm in the first neonatal week in seven cases and from 38 to 57 cm in the second through fourth neonatal weeks in nine cases. Other patients in this category include one first admitted at 5 months of age with an OFC of 50 cm, another hospitalized at 7 weeks of age with an OFC of 51 cm, and a third admitted at 3 months of age with an OFC of 62 cm.

Type of Hydrocephalus

Pneumoencephalography and/or ventriculography confirmed the diagnosis in 51 of the 55 infants presenting with primary hydrocephalus (Table 1). Communicating hydrocephalus was demonstrated in 19 cases and in 32 cases the malformation appeared noncommunicating in nature. Aqueductal obstruction was diagnosed in 24 cases, including three who also had cystic deformities in the posterior fossa. A classic Dandy-Walker cyst was disclosed in three infants, and in five others the air studies did not clearly differentiate the exact nature of
the obstruction. The comparative thickness of the cerebral mantle was not determined in this review. Although ventriculography was not done in the 24 megaloccephalic newborns with myelomeningocele, hydrocephalus was presumed present and uniformly gross because of the excessive head circumference.\textsuperscript{12}

\begin{table}
\centering
\caption{Type of hydrocephalus in 79 cases with excessive head circumference at birth}
\begin{tabular}{|l|c|}
\hline
Type & No. of Cases \\
\hline
Primary hydrocephalus: & 55 \\
communicating & 19 \\
noncommunicating & 32 \\
aqueductal obstruction & 24* \\
Dandy-Walker cyst & 3 \\
undifferentiated & 5 \\
undetermined (no air study) & 4 \\
Associated with myelomeningocele & 24 \\
\hline
\end{tabular}
\end{table}

*Three cases also had cystic lesions in the posterior fossa.

\section*{Results: Nonsurgical Cases}

Five patients with untreated primary hydrocephalus could not be traced after being followed for periods of 1 month (3), 1 year (1), and 11 years (1) after discharge from the hospital. The 1-year-old girl had a progressive increase in the abnormal enlargement of her head and was considered retarded. The 11-year-old boy had had communicating hydrocephalus initially diagnosed by air study and survived cardiovascular surgery for correction of a tetralogy of Fallot; at 11 years, he had a greatly enlarged head (OFC 67 cm) and although retarded was educable in a special school.

Follow-up was complete in 27 patients who received supportive care after admission to the hospital and diagnosis of congenital hydrocephalus (Table 2). Only two patients are living, both with untreated communicating hydrocephalus. Only one of these is functional, with an average level of intelligence at 19 years; this girl was first seen at 4.5 months with an OFC of 46 cm, 2
Prognosis of hydrocephalus overt at birth

TABLE 2

Comparison of nonsurgical and surgical results in 69 cases of hydrocephalus manifested at birth followed over two decades, 1950-1969

<table>
<thead>
<tr>
<th>Type of Hydrocephalus</th>
<th>Alive</th>
<th>Dead</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nonsurgical cases (27):</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>communicating</td>
<td>2</td>
<td>4</td>
<td>6</td>
</tr>
<tr>
<td>noncommunicating</td>
<td>0</td>
<td>5</td>
<td>5</td>
</tr>
<tr>
<td>unknown</td>
<td>0</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>associated with myelomeningocele</td>
<td>0</td>
<td>15</td>
<td>15</td>
</tr>
<tr>
<td>total</td>
<td>2</td>
<td>25</td>
<td>27</td>
</tr>
<tr>
<td>Surgical cases (42):</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>communicating</td>
<td>5</td>
<td>3</td>
<td>8</td>
</tr>
<tr>
<td>noncommunicating</td>
<td>6</td>
<td>16</td>
<td>22</td>
</tr>
<tr>
<td>unknown</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>associated with myelomeningocele</td>
<td>4</td>
<td>5</td>
<td>9</td>
</tr>
<tr>
<td>total</td>
<td>16</td>
<td>26</td>
<td>42</td>
</tr>
<tr>
<td>Overall total</td>
<td>18</td>
<td>51</td>
<td>69</td>
</tr>
</tbody>
</table>

cm above the 98th percentile, and the diagnosis of achondroplastic dwarfism was established. Her head circumference had been recorded as 38 cm at birth. Ventriculography confirmed the symmetrical ventricular dilatation present. She made satisfactory progress throughout primary school and attended trade school. The other surviving patient remains at home in a blind, vegetative, bedfast state at 17 years of age. He is nourished by nasogastric tube feedings and is extremely megaloecephalic (OFC 83 cm at age 4 years).

Of the 25 dead patients in the follow-up category, five with primary hydrocephalus were known to have survived beyond infancy in a grossly retarded status, including one who lived 17 years before dying in an institution. No assessment of psychomotor development was made in the other 20, who died very early in life. Two infants with primary hydrocephalus did not live beyond 24 hours after birth, and three others died between 1 and 6 months of age. Of the 15 patients with excessive head circumferences and myelomeningoceles, seven died on the first day of life and five between 2 and 10 days. Three others died between 2 and 6 months of age of infections. Only four of the 15 myelomeningoceles were closed surgically, with postoperative deaths on the first day of life in two instances, within 24 hours after operation on the ninth day in a third case, and at two months in the other.

Follow-up Incomplete (5 Cases)

Five patients with surgically-treated congenital hydrocephalus manifested at birth could not be traced after being followed postoperatively for periods of 1 to 7 years. Of three children with communicating hydrocephalus treated in infancy by ventriculoatrial shunting, two were grossly retarded and epileptic at 2 and 3 years of age, and developmental progress was delayed in the third at 15 months. One patient with aqueductal obstruction treated by ventriculoatrial shunting was trainable at 7 years of age, but was handicapped by excessive head size (OFC 73 cm) and spastic paraparesis. A second case of aqueductal obstruction treated by bilateral staged choroid plexectomy, ventriculosalpingostomies (3), and a ventriculoureterostomy was known to be in a vegetative state when last observed at 2 years of age.

Follow-up Complete (42 Cases)

Operations. A total of 98 surgical procedures have been performed to date for attempted relief of the hydrocephalus initially noted at birth in these 42 cases. The primary operation for control of hydrocephalus was a ventriculoatrial shunt in 34 cases, with the Holter valve system utilized in 32 of these procedures. A ventriculocisternostomy and ventriculoparotid shunt were performed in one case each, and one other had a choroid plexectomy initially. Five infants had craniotomies (2) or suboccipital craniectomies (3) for attempted lysis of cysts obstructing the flow of ventricular fluid. The 56 subsequent operations performed in this group of patients were mostly revisions of existing shunts or different shunting procedures. Thirteen surviving patients have been subjected to 29 additional operations, including 24 revisions of the Holter shunt systems (range 1–5 per patient) and shunts to the gallbladder and pleural cavity. Two living patients with Holter valve shunts and one young adult treated in infancy by ventriculocisternostomy have had no subsequent operations for hydrocephalus. A total of 27 reoperations were performed for hydrocephalus on the 26 patients who

J. Neurosurg. / Volume 39 / September, 1973
are known dead. Of the 19 deceased patients treated initially by ventriculoatrial shunting, nine subsequently had 17 operative revisions (range, 1 to 7 per patient) with eventual diversion of cerebrospinal fluid (CSF) to the gallbladder in three of these. In 10 cases the initial ventriculoatrial shunts were never revised. Additional operations in the other seven patients who are now dead included craniotomies for lysis of cysts and shunts to the salpinx and ureter.

Mortality. Of the 42 cases followed, 16 have survived for periods up to 21 years, with an average age of over 7 years (Table 2). Of the 26 known dead, 12 died within the first 6 months of life; the other 14 died between 10 months and 5 years of age (average age, 2.4 years). All of the older infants and children except one were uniformly grossly retarded, frequently blind, and subject to recurrent seizures. The exceptional case, a patient with aqueductal obstruction and an OFC of 41 cm at birth, had normal developmental progress for 3.5 years when he died of a wound infection and ventriculitis complicating a revision of a ventriculoatrial shunt to his gallbladder. Seven of eight patients whose hydrocephalic states were treated surgically before ventriculoatrial shunts and one-way valves were introduced died in infancy (4) or early childhood (3).

Quality of Survival. Five of the 16 surgical patients who survived have normal intelligence by clinical criteria or I.Q. scores. The clinical data on this favorable subgroup are given in Table 3. All were treated by Holter ventriculoatrial shunts in infancy and, except possibly for Case 1, have remained shunt-dependent. The other 11 survivors in this group are mentally retarded. Five cases are only moderately impaired mentally, including two young children with myelomeningoceles and the oldest patient whose obstructive hydrocephalus was managed by a ventriculocisternal shunt. The less retarded patients have developed some social graces, are trainable, and several have achieved I.Q. scores of 75 to 80. Six patients are grossly retarded with essentially no demonstrable developmental potential. Three are also blind and subject to recurring seizures. Of the severely defective children, three had communicating hydrocephalus, two had obstructive hydrocephalus, and one a myelomeningocele.

Ventriculoatrial Shunts. Of the 34 infants initially treated by ventriculoatrial shunting, 15 (44%) have survived up to 15 years, for an average of nearly 7 years. One-third of the survivors are normally intelligent. Shunt complications accounted for eight of the 19 deaths in this group (42%), including six of the 12 dying in infancy and two dying later in childhood. Five of these operative deaths were the result of uncontrolled infection. Following the initial surgery, 41 revisions of 34 existing shunt systems, six shunts to the gallbladder in four cases, and three to the pleura in one (84 operative procedures), there was an operative mortality of nearly 10% with a cumulative surgical case mortality of 24%.

TABLE 3

Normally intelligent survivors treated surgically for hydrocephalus manifested at birth

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Sex, Age (yrs)</th>
<th>Type of Hydrocephalus</th>
<th>Birth OFC (cm)</th>
<th>Shunt Revisions</th>
<th>Present OFC Percentile</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M 7</td>
<td>communicating</td>
<td>“large”</td>
<td>1</td>
<td>50</td>
<td>several seizures since age 3 yrs; on anticonvulsant drugs</td>
</tr>
<tr>
<td>2</td>
<td>F 4</td>
<td>noncommunicating, aqueductal</td>
<td>46.5</td>
<td>2</td>
<td>50</td>
<td>birth OFC measured after ventricular drainage for vaginal delivery</td>
</tr>
<tr>
<td>3</td>
<td>M 5</td>
<td>noncommunicating, aqueductal</td>
<td>41</td>
<td>2</td>
<td>50</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>F 7</td>
<td>noncommunicating, Dandy-Walker associated with myelomeningocele</td>
<td>42</td>
<td>5</td>
<td>98 + 1 cm</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>F 14</td>
<td>associated with myelomeningocele</td>
<td>40</td>
<td>4</td>
<td>50</td>
<td>paraparetic but has good sphincter control</td>
</tr>
</tbody>
</table>
Prognosis of hydrocephalus overt at birth

Age at admission and the time of ventriculoatrial shunting for hydrocephalus were analyzed with reference to survival or death in 31 patients seen before 2 months of age. The 13 living were admitted at an average age of 5.5 days (S.D. ± 7.2 days) and shunted at a mean age of 23.5 days (S.D. ± 11.7 days). The corresponding mean ages in the 18 patients who died were 12.9 days (S.D. ± 16.6 days) and 31.0 days (S.D. ± 16.7 days). The average interval between hospitalization and operation for hydrocephalus was 18 days in both groups. The differences observed favoring survival of infants admitted and operated on earlier approached levels of statistical significance (student t-test, p = .06). A similar trend was noted when the normally intelligent cases were compared against the retarded ones, living and dead, but the differences observed did not prove significant. One of the normal survivors whose head measurement was unknown at birth was not shunted until nearly 6 months of age (Case 1, Table 3).

Comment

In our series the prognosis for survival and normal intelligence in infants born with hydrocephalus and excessive head size is clearly poorer than in the experience generally reported with infantile hydrocephalus. Laurence and Coates found that the hydrocephalus apparently had arrested spontaneously in 81 of 182 unoperated patients (46%) who were alive over a 20-year period. Of the survivors with arrested hydrocephalus, 73% were educable and 38% had I.Q. scores in the normal range. Yashon reported spontaneous arrest of hydrocephalus and survival without surgical treatment in 31 of 58 patients (53%), and 10 of the living patients seemed to be functioning at nearly normal levels. With current methods of surgical treatment by ventricular diversion through one-way valves, infantile hydrocephalus can be relieved with an extended survival of 60% to 80%, with 50% to 84% of the survivors having good intelligence. In contrast, among the 34 patients with overt hydrocephalus at birth treated by similar surgical procedures, in our experience, only 44%

have survived and one-third have normal intelligence. Our surgical results in extremely hydrocephalic infants with myelomeningoceles (Tables 2 and 3) closely parallel the findings of Lorber in 36 similar cases with 14 survivors, three of whom have normal intelligence. Virtually all of our cases treated nonsurgically died early or later in childhood, and only the achondroplastic dwarf reached adulthood with normal intelligence. This case was exceptional, of course, since it is known that the hydrocephalus present in infancy in some of these dwarfs can persist in a quiescent form in adulthood and intelligence can be normal. Relative head circumference did not have any decided prognostic value in predicting the eventual outcome of our cases. The mean head circumference in surviving patients was about 2 cm less than the average circumference determined in the patients who subsequently died. This finding was constant among the three groups of patients having either communicating or noncommunicating hydrocephalus or hydrocephalus and myelomeningocele. Differences of this magnitude favoring survival of the infants with smaller heads were not statistically significant, although among the infants with myelomeningoceles the data were pretty suggestive (p = .10, student t-test).

The type of hydrocephalus determined in the infants in the present series was associated with different rates of survival, being highest in patients with communicating hydrocephalus and lowest in the myelomeningocele group as a whole. Among the surgical cases followed (Table 2), however, infants with various forms of obstructive hydrocephalus were least likely to survive. The prognosis for normal mental development was similar among the main three categories of hydrocephalics and ranged from 11% to 14% of all cases treated surgically. The differences noted in the rate of survival and number of normally intelligent patients according to the type of hydrocephalus present do not reach levels of statistical significance in our total experience or among the surgical cases considered separately.

Factors responsible for the poor prognosis in infants with extreme hydrocephalus manifested around birth include the severity.
of the neural malformation itself, the potentially irreversible pressure effects of abnormal ventricular distention, the high rate of infection, and the greater chance of intracranial birth injury due to the large head. The brain may be so malformed as to preclude live birth or survival beyond a few hours or days. Fulminant meningitis and other infections are recognized as major causes of death in infants and children with hydrocephalus or myelomeningoceles. The extent of the malformation and sepsis probably accounted for most of the high early mortality noted in our cases, particularly in the myelomeningocele group where 50% died during the first neonatal week. The advanced intrauterine hydrocephalus indicated in most of our cases would also contribute to the high rate of death and mental retardation that followed. Unrelieved ventricular distention and its relatively long duration could have produced progressive destructive changes in the developing fetal and neonatal brain. Weller and Shulman have demonstrated edema, axonal degeneration and astrocytosis in white matter, ependymal disruption, and damage of cortical layers in the brains of hydrocephalic infants and puppies. They have postulated that progressive hydrocephalus with a rising CSF pressure may result in failure of brain growth and mental retardation unless ventricular drainage is instituted prior to the time neural damage is irreversible.

Ventricular shunting operations should be performed on infants with overt hydrocephalus manifested at birth to afford them a chance for survival and psychomotor development. Surgical treatment will not eliminate the high mortality in the first neonatal week and poses a continuing risk later in life. Results in patients of this type may be improved if shunting operations are performed early, around the end of the first week of life. By this time most of the inevitable natural deaths would have occurred, myelomeningoceles would be re-operated, fractional air studies carried out, and the results of CSF cultures known. Our results definitely associate a higher rate of survival with earlier ventricular shunting. Although our data are not confirmatory, there is also reason to anticipate a higher percentage of functional survivors if surgery for hydrocephalus is done early. This approach seems also quite justified since, in our experience, surgical treatment did not significantly increase the number of vegetative survivors. The average length of survival of deceased and for the most part retarded patients in the unoperated and operated groups was virtually the same, at 15 and 16 months respectively.

References

13. Lorber J, Zachary RB: Primary congenital hydrocephalus: long term results of a
Prognosis of hydrocephalus overt at birth


Address reprint requests to: John Mealey, Jr., M.D., Neurological Surgery, Indiana University School of Medicine, 1100 West Michigan Street, Indianapolis, Indiana 46202.