Five-Year Comparative Study of Hydrocephalus in Children With and Without Operation (113 Cases)*

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HYDROCEPHALUS long has been recognized by neurosurgeons as a sign of serious intracranial disease involving obstructions in the pathways of the cerebrospinal fluid. Multiple etiologies of such obstructions have been identified, but some of the major pathophysiologic mechanisms of the cerebrospinal-fluid system in hydrocephalus remain in doubt and under investigation. Progressing hydrocephalus, whatever its cause, produces unequivocal loss of cerebral mass, however, and presumably results thereby in damage to the brain in addition to the damage caused by the process producing the blockage of the cerebrospinal fluid. Neurosurgical operative techniques recently have been developed to such a degree of technical excellence, however, that progressing hydrocephalus can be arrested by relatively simple surgical techniques, providing adequate postoperative follow-up of such patients is maintained. Thorough study of a significant group of such patients over a long period of time should give some information as to the value of such procedures in preventing brain damage that otherwise might produce an incompetent individual. For reliable conclusions, such a series of surgically treated patients should be compared with a parallel series of patients not operated upon, especially in regard to functional status of the patients.

With this objective in mind, a study has been carried out over a 5-year period in which two groups of patients with hydrocephalus have been followed simultaneously. In one group, operative treatment of progressing hydrocephalus was carried out as early as possible, and in the other group no operative procedure at all was carried out. This report compares the initial results in these two groups over the first 5 years of this study on 113 hydrocephalic children. The morbidity, mortality, functional status, and factors related to these are compared in the two groups.

Methods

This investigation is based on 113 patients with hydrocephalus, ranging from premature infants to children 13 years of age. All of these patients were seen on the Neurosurgery or Pediatrics Services at the University of Washington Affiliated Hospitals, including the University Hospital, the King County Hospital, and the Children's Orthopedic Hospital. In all instances the patient was worked up on the Neurosurgery Service or the Pediatrics Service initially in order to determine the etiology of the hydrocephalus, its state of progression, and its state of activity. At this point a decision concerning whether operation should be carried out or not was made by the neurosurgeon consulted or the neurosurgeon handling the patient. Certain criteria were set up initially to determine whether operation was warranted or not, but these were abandoned later. The work-up of all the children to the point of deciding about operation, however, was essentially the same in each case.

Group Without Operation. The 48 patients in this group were worked up thoroughly in the hospital because of enlarging head or enlarging head and myelomeningocele. The initial work-up consisted of complete neurological evaluation, circumference of head,
measurements of chest and body compared periodically to the standard charts, transillumination, roentgenograms of skull and spine, recordings of intraventricular pressure, followed by pneumoencephalographic or ventriculographic air studies, as indicated, to determine the etiology of the hydrocephalus and its degree of progression. In some of these instances the initial air study was all that was done in the hospital concerning the state of progression of the hydrocephalus. In other instances periodic measurements of intraventricular pressure were achieved as well as repeated air studies prior to the decision that no operation was warranted. Some of the neurosurgical staff seldom advised operation at any stage of hydrocephalus, preferring to adopt a conservative approach, and this situation automatically produced this nonoperative group.

Follow-up examinations on these children were obtained over a period of 5 years with considerable difficulty in some instances. However, every child was examined by one of the authors on periodic follow-up examinations or by local qualified professional personnel when great distances were involved. Neurological evaluations at those examinations included roentgenograms of the skull and measurements of the head. Few "progress" air studies were possible, however. The frequency of these examinations varied, but were as often as every 6 to 8 months.

Periodic psychologic testings were done by trained psychologists in both the operative and nonoperative groups in similar manner. In the very young children, developmental quotients (D.Q.) were determined using the Peabody Picture Test and Gesell Developmental Scale, depending somewhat on the particular child and its age. With appropriate age of the children, intelligence quotients (I.Q.) likewise were determined, administering the Stanford-Binet and Wechsler Intelligence Tests. The frequency of testing varied, based on three criteria: a) according to changing development—every 3 months; b) when stabilized—6 months to 1 year; c) whenever insult was suffered—during acute phase and if possible 6 weeks later.

The fate of those who succumbed during the 5-year period was determined either by observation at autopsy or by securing the death certificates or autopsy reports if the patient died outside the hospital.

Group With Operation. There are 65 patients in this group, most of them brought to the attention of the neurosurgeon very early in their hospital course. Complete neurological evaluation, critical measurements of the body, records of intraventricular pressure, roentgenograms of the skull, and specific differential air studies of the brain were carried out sequentially. A definite diagnosis of the site of block was established by air study in each instance. The progressive nature of the hydrocephalus was studied by serial ventricular pressures, air studies, and measurements of the head. In many instances a ventricular study of clearance of RISA was helpful in determining the activity of the hydrocephalus. Measurements of the width of the cerebral mantle were obtained as early as possible by the air studies. If these studies demonstrated the etiology of the hydrocephalus and indicated that a progressing, nonarrested hydrocephalus was present, operation was deemed warranted. If the children were in adequate shape for a shunting procedure, such was carried out promptly and at times very early in life, including premature babies. In those children with myelomeningocele, ventriculo-atrial shunting was established prior to any direct attack on the myelomeningocele. Direct operation on the myelomeningocele was done only when the mass of the myelomeningocele was painful or cumbersome for purposes of rehabilitation. The first operative procedure in all but a few instances was a standard ventriculo-atrial shunt using the Pudenz-Heyer valve. In the small infants, the small 1 mm. cardiac catheter-valve was used during the last 2 1/2 years, whereas the larger one (the only one available) was used earlier in the series. Flushing devices were inserted in this type of shunt only during the last 18 months.

Postoperative follow-up was prolonged and repetitive, including CO₂ "bubble"
ventriculograms4,12,17,18 to estimate width of cerebral mantle and record ventricular pressure, ventricular clearance of RISA to establish the functioning status of the shunt, and repeated neurological evaluation as well as psychological testing (Stanford-Binet and Wechsler Intelligence Tests). These evaluations occurred at 3 months, 6 months, 9 months and 12 months following operation, and then at least once or twice during each subsequent year. The follow-up examinations were done irrespective of symptoms.

Signs of inadequate functioning of the shunt were investigated promptly, and adequate studies were undertaken to determine the site of blockage in the shunt. A functioning shunt was re-established promptly. Studies to determine function of the shunt or cause of dysfunction included mediastinal venography to establish presence of thrombosis of the innominate vein as well as direct irrigation of the cardiac catheter with opaque dye to determine whether an occluding mural thrombus was present in the atrium. As the children approached $3\frac{1}{2}$ to 4 years of age, dye studies of the cardiac catheter were mandatory to demonstrate the cephalad migration of the cardiac catheter up into the occluded internal jugular vein from the superior vena cava, and when such was demonstrated, the cardiac catheter-valve was replaced promptly with a new one.

In numerous instances, occlusion of the cerebral end of the shunt was demonstrated secondary to plugging of the catheter by choroid plexus or ependymal tissue. These were replaced promptly.

Periodic intelligence and developmental tests were administered to these children during the 5-year period by trained psychologists.

It was apparent early in the study that repetitive examinations of these children were necessary in order to identify malfunctioning shunts, many of which were still working but not adequately so. Malfunctioning shunts functioned poorly for a time prior to complete obstruction, and recognition and early replacement precluded many potentially emergent situations.

Results

Group Totals. As is shown in Fig. 1, the 5-year study includes 118 hydrocephalic children. Of this group, 65 were operated on because of demonstrated progressing hydrocephalus and a ventriculo-atrial shunt was established. In this group of 65, there were 41 who had no myelomeningocele, whereas 24 did have associated myelomeningocele (41 per cent). Both of these subgroups, however, were investigated in the same manner and differential air studies were done thoroughly in order to establish the etiology of the hydrocephalus prior to operation.

There were 48 children included in the group in which no operation was performed. Of these, 28 had no myelomeningocele and 20 had myelomeningocele (37 per cent). Again, an effort was made to demonstrate the etiology of the hydrocephalus in all of these children, and the decision that no operation was to be performed was made during the first 3 to 6 months of life. In no instance was a diagnosis made of “arrested” hydrocephalus.

Initial diagnosis and inclusion in the study were at age 1 month or less for 37 infants; 2 to 3 months for 12 infants; 3 to 6 months for 9 infants; and 6 months to 6 years for 7 children in the operative group. Diagnosis was established in the nonoperative group at age 1 month or less for 26 infants; 2 to 3 months for 3 infants; 3 to 6 months for 7 infants; and 6 months to 9 years for 12 children.

Etiology of the Hydrocephalus. The anatomic cause of the progressing hydrocephalus was determined as accurately as possible in every case by means of differential air studies—i.e., pneumoencephalogram, ventriculo-
gram, or a combined pneumoencephalogram-ventriculogram. Table 1 demonstrates the similarity in etiology of the site of block of cerebrospinal fluid in the two groups. The operative and the nonoperative groups are divided again into those with myelomeningocele and those without myelomeningocele. Communicating hydrocephalus produced by a basal arachnoiditis, presumably this in turn caused earlier by either a low-grade infection or hemorrhage, predominated as the cause for hydrocephalus. Stenosis of the aqueduct was the second most common site of cerebrospinal-fluid obstruction, and the Walker-Dandy syndrome was the third most common. It was surprising that Arnold-Chiari malformation (Type II) was present in only 3 of 24 patients with myelomeningocele who were operated on and in only 5 of those with myelomeningocele in whom no operation was performed. It was postulated that possibly the early age at which the air studies were done may have resulted in classifying these as communicating hydrocephalus rather than the Arnold-Chiari disorders which had not as yet developed fully into this recognizable syndrome. Since this diagnosis was made only by air study done via lumbar puncture, possible error in this diagnosis also must be considered.

**Morbidity—Infection.** In Table 2 the types of infection in the two groups are compared.

The operative group showed 5 children who had infections of the central nervous system, 3 of whom died, whereas 4 died in the nonoperative group. Meningitis in 2 cases in the operative group was treated successfully.

Septicemia was present in 7 children who had operation. Five of these children recovered satisfactorily, a chronic septicemia developed in 1, and 1 died. No septicemias were recorded as such in the nonoperative group, though some of those who died may have had septicemia, especially those with myelomeningocele.

Infection of the wound occurred in only 2 in the operative group, both of whom re-

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**TABLE 1**

<table>
<thead>
<tr>
<th>Etiology</th>
<th>Operation</th>
<th>No Operation</th>
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<tbody>
<tr>
<td></td>
<td>Myelomeningocele</td>
<td>No Myelomeningocele</td>
</tr>
<tr>
<td>Communicating hydrocephalus (basal arachnoiditis—infeciton, blood)</td>
<td>15</td>
<td>12</td>
</tr>
<tr>
<td>Aqueduct stenosis</td>
<td>5</td>
<td>4</td>
</tr>
<tr>
<td>Mass in 3rd ventricle</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Obstruction exit 4th ventricle (Walker-Dandy syndrome)</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td>Encephalocele</td>
<td>3</td>
<td>0</td>
</tr>
<tr>
<td>Arnold-Chiari malformation</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Multiple anomalies</td>
<td>0</td>
<td>3</td>
</tr>
<tr>
<td>Cystic brain disease</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Undiagnosed</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>24</strong></td>
<td><strong>41</strong></td>
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</table>

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**TABLE 2**

<table>
<thead>
<tr>
<th></th>
<th>Operation</th>
<th>No Operation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infection of CNS</td>
<td>5</td>
<td>4</td>
</tr>
<tr>
<td></td>
<td>3 deaths</td>
<td>4 deaths</td>
</tr>
<tr>
<td></td>
<td>2 recovered</td>
<td></td>
</tr>
<tr>
<td>Septicemia</td>
<td>7</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>1 death</td>
<td></td>
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<tr>
<td></td>
<td>1 chronic</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>5 recovered</td>
<td></td>
</tr>
<tr>
<td>Pneumonia</td>
<td>0</td>
<td>3</td>
</tr>
<tr>
<td></td>
<td>3 deaths</td>
<td></td>
</tr>
<tr>
<td>Infection of wound</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>2 recovered</td>
<td></td>
</tr>
</tbody>
</table>
covered, though the shunt had to be removed. Three children had mediastinal venous obstruction; 2 recovered and 1 expired. In all these instances the ventriculo-atrial shunt was removed and a ventriculomastoid or a subarachnoidal ureteral shunt was done.

-Morbidity: Obstructions of Ventriculo-Atrial Shunt. Obstructions of the ventriculo-atrial shunt in the cardiac end occurred in the first 9 months in 22 of 52 patients. The statistics are demonstrated in Table 3 in which such early obstructions of the shunt show a relationship to the character of the cerebrospinal fluid being shunted. In 21 patients who showed absolutely normal cerebrospinal fluid, only 5 revisions were necessary. In a group of 24 patients who had either high protein (greater than 20 mg. per cent) in the ventricular fluid, or a significant number of cells when first examined before operation, 48 revisions were necessary. Six in this group died ultimately. In an additional group of 7 cases in which the spinal fluid was not reported adequately, 12 revisions were necessary though the relations to the cerebrospinal fluid are not clear. In most of these cases, the larger cardiac catheter-valve was being used at that time. Since the smaller (1 mm. in diameter) infantile type of catheter-valve has been used, the incidence of early obstruction of the cardiac end has become far less. The actual statistics in this group currently are not worked out. The results in Table 3 might indicate that patients with abnormal cerebrospinal fluid (high protein, many cells) are more likely to have obstruction of the cardiac end than those patients who have a normal spinal fluid.

A second type of obstruction of the cardiac end occurred after 9 months and was related to the factors demonstrated in Table 4. Four had broken shunts at the connector or at the outlet of the flushing device. It was not clear as to whether these were secondary to direct trauma or whether there was excessive manipulation at the time of the insertion of the flushing device.

One patient had an obstruction of the cardiac end which at the time of re-operation showed the cardiac catheter to be disconnected and lost into the jugular venous system. The catheter was nowhere to be found by open dissection of the neck, and 2 years later careful roentgenograms of the chest showed the tiny catheter coiled up in the right superior pulmonary artery, producing a mild pulmonary infarct. No symptoms had occurred secondary to this as far as we could tell.

A significant cause of late obstruction was a distracted shunt produced by growth of the body. Elongation of the structures of

<table>
<thead>
<tr>
<th>CSF</th>
<th>No. of Patients</th>
<th>No. of Revisions (Cardiac-end Obstructions)</th>
<th>Progressive</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Normal</td>
<td>21</td>
<td>18</td>
<td>1</td>
</tr>
<tr>
<td>High protein, or cells</td>
<td>24</td>
<td>5</td>
<td>1</td>
</tr>
<tr>
<td>Inadequate report</td>
<td>7</td>
<td>1</td>
<td></td>
</tr>
</tbody>
</table>


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**Table 4**

Morbidity (operative group): late (at least 9 months postoperative) obstruction of shunts (preliminary data)

<table>
<thead>
<tr>
<th>Broken shunts</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>at connector or flushing device</td>
<td>4</td>
</tr>
<tr>
<td>Disjointed shunt</td>
<td>1</td>
</tr>
<tr>
<td>cardiac catheter-valve lost into heart</td>
<td></td>
</tr>
<tr>
<td>Distracted shunt</td>
<td></td>
</tr>
<tr>
<td>1. Growth of body causing migration of cardiac end up into thrombus of internal jugular vein, 3½–4 years of age</td>
<td>14</td>
</tr>
<tr>
<td>2. Ventricular end pulled out of CSF by returning thickness of cortex or effects of growth by fixation in neck</td>
<td>6</td>
</tr>
</tbody>
</table>
head and neck served to "pull" the catheter up out of the right atrium into the thrombus formation of the internal jugular vein. If the shunt had been inserted in infancy, such effects of distraction became evident clinically at 3½ to 4 years of age. Fourteen such cases have been recognized and the patients were reoperated on successfully. Replacement of the catheter into the right atrium has been achieved by extracting the old valve and placing a new one down through that same tract after the use of appropriate dilators, or else by putting a new valve into the jugular vein and threading it down into the appropriate position.

Distraction of the ventricular end in which the ventricular catheter apparently was distracted from the ventricle by either returning thickness of the overlying mantle of brain or by elongation of the neck in growth with fixation of the shunt in the neck occurred in 6 cases. Other causes of obstruction of the ventricular catheter occurred occasionally as well (to be considered in another report).

**Morbidity—Cost.** Although cost of medical care of children with hydrocephalus often is brought up as argument for or against active treatment, actual determination of cost in any single situation is almost impossible. Nevertheless, if one should judge this by the rate of institutional care in the two groups, Table 5 shows that in the operative group only 8 of 49 patients (16 per cent) required care in a state institution, whereas 13 of 24 (54 per cent) required institutionalization in the nonoperative group. By and large, the children operated on were cared for in their own home environment. The cost per patient in a state institution runs on the average about $1700 to $1800 a year. The hospital cost for their initial treatment, however, and the follow-up examination is currently not available.

**Mortality.** The crude figures for mortality are listed in Table 6. These figures are the results in the 5-year period in an "on-going" study of patients who have varying periods of observation—i.e. patients were added to this group during the 5-year period of time, etc. In the operative group of 65 patients there were 15 deaths, or a 23 per cent mortality, whereas in the nonoperative group of 48 patients, 22 died, giving a 46 per cent mortality. It may be significant that 11 autopsies were achieved in the 15 deaths in the operative group (72 per cent) whereas only 4 autopsies out of 22 deaths were done in the nonoperative group (only 18 per cent). Of the 15 patients who died in the operative group, 6 had myelomeningocele (40 per cent of deaths) and in the nonoperative group 10 patients with myelomeningocele were included in the 22 deaths (45 per cent of deaths).

It was logical to consider that patients with myelomeningocele might have a higher risk of death, but the make-up of the group originally showed that 37 per cent of the operative group had myelomeningocele and 40 per cent of the nonoperative group had myelomeningocele, comparing very closely with the percentages shown in the mortality figures. It would appear that the presence or absence of myelomeningocele, then, had little to do in influencing survival.

**Mortality—Cause of Death.** Of the 15 children who died in the operative group, 11 were examined post mortem which established the cause of death as demonstrated in Table 7. In the nonoperative group with 22 deaths, however, only 4 had autopsy and the cause of death established adequately. Only 4 children in the operative group died without postmortem examination and the cause of death was listed only by clinical impression.

**TABLE 5**

<table>
<thead>
<tr>
<th>Group with operation</th>
<th>Group without operation</th>
</tr>
</thead>
<tbody>
<tr>
<td>8/49—16%</td>
<td>13/24—54%</td>
</tr>
</tbody>
</table>

**TABLE 6**

<table>
<thead>
<tr>
<th>Group with Operation (65 patients)</th>
<th>Group without Operation (48 patients)</th>
</tr>
</thead>
<tbody>
<tr>
<td>15 deaths—23%</td>
<td>22 deaths—46%</td>
</tr>
<tr>
<td>11 autopsies—72%</td>
<td>4 autopsies—18%</td>
</tr>
<tr>
<td>(6 myelomeningocele—40%)</td>
<td>(10 myelomeningocele—45%)</td>
</tr>
</tbody>
</table>
In the nonoperative group, on the other hand, 18 children died without autopsy and the cause of death was established only by the clinical examination, though in 6 of these children no adequate record was available. It should be pointed out that, in the nonoperative group, some of these statistics were obtained with great difficulty from the State Department of Vital Statistics, the local Coroner’s Office in the region where the children had lived, and by the attending physician’s records. In many instances these records were felt not to give a complete picture of the terminal event or cause of death.

Predicted Prognosis, Verification. In the group of children who did not have operation for their hydrocephalus, 46 had a prediction of their prognosis stated in the chart, thus allowing a correlation of this prediction with their ultimate outcome over this 5-year period. Table 8 shows the correlation for this small group. In the “hopeless” or “imminent-death” group in which 21 patients were classified, 13 of these children died, 6 were noncompetitive (I.Q. of less than 75), and 2 were competitive and alive with an intelligence quotient of 75 or higher. In a group of 12 patients who were stated to have “arrested” hydrocephalus, 2 died, 5 are still alive with an I.Q. of less than 75, and 5 are alive with an I.Q. greater than 75. In an additional group who were stated to have a “good prognosis,” it may be pertinent that none of the survivors has a competitive I.Q. (75 or higher), 9 are alive with an I.Q. of less than 75 (74 per cent) and 4 are dead (26 per cent). The predicted course for such children shows a poor correlation with the actual course that these children followed. This may indicate that such predictions are quite unreliable and inadvisable, especially if done on the basis of only a few clinical observations spaced relatively close together during infancy.

Changes in Width of Cerebral Mantle. In each instance the initial ventriculographic study, whether by pneumoencephalogram or ventriculogram with gas, was measured carefully for width of cerebral mantle in the frontal region, using as the measured area the distance between the inner table of the frontal bone and the nearest portion of the frontal horn of the ventricle anterior to the
Comparative Study of Hydrocephalus in Children

Fig. 2. Serial ventriculographic study. (A) 3 weeks of age (pre-operative pneumoencephalogram shows communicating hydrocephalus, right frontal cyst). (B) 10 weeks of age (7 weeks after ventriculo-atrial shunting, air study shows striking reduction of hydrocephalus and no cyst visible).

foramen of Monro. Intraventricular pressures or pressures at lumbar puncture were observed each time such a study was done. Repeated or follow-up ventriculographic studies were done at various periods postoperatively to show the changes in the width of the cortical mantle irrespective of the clinical status of the patient. Such studies were done to a lesser extent in the nonoperative group. In most instances of obstruction of the shunt, measurable decrease in the width of the cerebral mantle (increase in ventricular size) was present if the obstruction had been present for a significant time. At the same time, increased ventricular pressure was present (above 100 mm. cerebrospinal fluid) and ventricular clearance of RISA showed less than normal clearance. Fig. 2 shows serial ventriculographic studies, illustrating the increase in width of cerebral mantle resulting from adequate shunting of cerebrospinal fluid during a 7-week period of time in a hydrocephalic infant 3 weeks old. Such changes in thickness of cerebral mantle can occur rather dramatically in even 5 days.

Width of Cerebral Mantle and Intraventricular Pressure. Fig. 3 (operative group) shows the changes in width of cerebral mantle as related to intraventricular pressure (direct measurement). Changes in width of cerebral mantle that were recorded had occurred in the interim between serial air studies. Ven-

Fig. 3. Change in width of cerebral mantle related to ventricular pressure.
Ventricular pressures were recorded at the time of the serial air studies. Therefore, pressures at any one recording were related to the change in width of mantle which had occurred in the interval of time since the last air study. A change in cerebral mantle was recorded only if the change was greater than 0.5 cm. (5 mm.). Almost all roentgenographic examinations included two to six lateral projections in the "brow-up" position in which variations in width were noted on any one examination to be \( \pm 0.3 \) cm. The chart indicates that in all our observations in which the intraventricular pressure was 120 mm. of cerebrospinal fluid or lower, width of cerebral mantle had increased, having, of course, started as abnormally thinned. In those cases in which the intraventricular pressure was greater than 120 mm. of cerebrospinal fluid, there was loss of the width of cerebral mantle as demonstrated in the chart.

**Ventricular RISA-Clearance Studies.** Intraventricular RISA in the amount of 5 \( \mu \)c was used as a follow-up study periodically in many of the operative group of children. This was used in the manner described previously and was found reliable in differentiating functioning versus nonfunctioning or poorly functioning shunts. Children in whom the shunt was functioning adequately showed a normal type of curve of disappearance of ventricular RISA. In all instances in which the ventricular RISA showed retention, as it would in an active hydrocephalic, the shunt was found to be obstructed at operation, either partially or completely. Use of RISA in classifying these children initially has not as yet been considered reliable enough to use as the only initial diagnostic aid. However, Fig. 4 demonstrates the type of disappearance as described previously.

**Psychological Investigations.** (1) Intelligence quotient as related to width of cerebral mantle (frontal). In 63 observations, the intelligence quotient was correlated with the width of the cerebral mantle (cm.) which was present at approximately the time the intelligence quotient was determined. This is demonstrated in Fig. 5, using children from both the operative and the nonoperative groups. The chart can be interpreted as indicating that a width of cerebral mantle of less than 1.2 cm. is associated with a low-intelligence quotient. With a width of cerebral mantle greater than 1.2 cm., however, the intelligence quotient is more likely to be adequate. However, adequate mass of brain does not necessarily imply adequate cerebral function.

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**Fig. 4.** Graph of clearance of RISA (5 \( \mu \)c) from ventricles. Vertical bars indicate standard deviations. Activity of ventricular cerebrospinal fluid 1 hr. after injection used as reference and equated to unity (see Table 2).

**Fig. 5.** Intelligence quotient related to width of frontal cerebral mantle (63 observations).
(2) Intelligence quotient as related to duration of increased intracranial pressure. In 83 observations, Fig. 6 shows the relationship of the intelligence quotient to duration of increased intracranial pressure. These observations are divided into those children in the operative and nonoperative groups, as well as those who had anomalous brains and some who are now dead. Increased intracranial pressure in these groups again was considered present if the measured pressure was 100 mm. of cerebrospinal fluid or over. This chart does not have complete reliability from a statistical standpoint as demonstrated by the $X^2$ calculations for the 1- to 2-month period of time, but it does appear that prolonged increased pressure is by and large associated with an intelligence quotient of considerably less than 75 in these patients. It should be noted that 3 children who were operated on and who had increased pressure for over 2 years had intelligence quotients of around 100, but it should also be pointed out that these recordings of pressure were intermittent. It is entirely probable that prolonged pressure at this level had not been present in those 3 since the ventriculo-atrial shunts had functioned very well for at least a portion of that time, and conceivably the pressure was normal during a critical developmental phase of the brain (related to age of the child). This type of correlation however, needs considerable support and additional numbers of observation.

Survival. When the life expectancy suggested from this series of operative and nonoperative cases is computed on actuarial principles using a life-table, the life expectancy of a child in either group can be demonstrated by the life-chart in Fig. 7. The graph for the operative group and the nonoperative group in our series begins at birth, whereas the graph for the series of Laurence and Coates \(^{11}\) begins at 3 months of age for their figures. On this basis, the calculations for survival alone, irrespective of their competence, shows that in the operative group 61 per cent should survive to 10 years, whereas in the nonoperative group 32 per cent should survive. In the series reported by Laurence and Coates, of 182 hydrocephalic children not operated on, the survival expectancy is 27.5 per cent. \(^{8,10,11}\) If hydrocephalus and myelomeningocele are both present, Laurence\(^8\) stated that hydrocephalus itself is the gravest sign against survival.
**Functional Status.** A comparison of the functional status of the hydrocephalic children in this group is shown in Table 9, compared with the report of Laurence and Coates.\(^{11}\) It must be kept in mind that this chart is comparing the results currently available in a 5-year period in a group of patients who are part of an "on-going" study. This means that some patients at this time have been under observation for 5 years, some for only 3\(\frac{1}{2}\) years, some for 2\(\frac{1}{2}\) years, and some for a period of time varying between these. Such statistics do give some prediction, however, as to the ultimate outcome when the factor of time related to onset of disease and the time of study are balanced. In this chart the intelligence quotients were measured repetitively and the last measurement was taken for the achieved I.Q. in any single case. In the operative group 44 per cent have an I.Q. of 75 or above, 33 per cent are noncompetitive with an I.Q. of 75 or below, and 23 per cent have succumbed. In the nonoperative group, 11 per cent have an I.Q. of 75 or above, 41 per cent are noncompetitive with an I.Q. of 75 or below, and 48 per cent have succumbed. At present, our statistics (nonoperative group) do not parallel those of Laurence and Coates\(^{11}\) who showed a much higher incidence of competitive children in the group not operated upon. Whether this is because our group has been followed a shorter period of time or is ascribable to bias in selection of groups is difficult to say.

A chart based on the life-table is presented in Fig. 8 concerning the mortality and morbidity of the children in these two groups in our series. This life-table begins at date of birth rather than at the 3-month period of time as was done in Laurence and Coates\(^{11}\) series. The per cent of patients at risk is shown on the Y axis and the X axis shows the age. The survival rate in the operative group (calculation based on 82 observations) shows

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**TABLE 9**

*Comparison of current status of hydrocephalic children in this study (March 1963)*

<table>
<thead>
<tr>
<th></th>
<th>Operation</th>
<th>No Operation</th>
<th>Laurence and Coates(^{11})</th>
</tr>
</thead>
<tbody>
<tr>
<td>Functional</td>
<td>29 (44%)</td>
<td>5 (11%)</td>
<td>31 (23.8%)</td>
</tr>
<tr>
<td>Nonfunctional</td>
<td>9 (9%)</td>
<td>19 (41%)</td>
<td>50 (37.6%)</td>
</tr>
<tr>
<td>Dead</td>
<td>12 (8%)</td>
<td>72 (48%)</td>
<td>99 (48.8%)</td>
</tr>
<tr>
<td>Total</td>
<td>94</td>
<td>46</td>
<td>182</td>
</tr>
</tbody>
</table>

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Fig. 7. Life-chart of these hydrocephalic children compared to group of Laurence and Coates.\(^{11}\)
that 61.4 per cent of the operative group should survive to 10 years, and that survivors will be made up of 33.8 per cent competitive children and 27.6 per cent noncompetitive children. In the same group, 38.6 per cent will have died. The comparison with the nonoperative group shows that 22.2 per cent will have survived, this group being made up of only 5.5 per cent who will be competent children (I.Q. of 75 or above) and 16.7 per cent who will be incompetent (I.Q. of 75 or less). The remainder, or 77.8 per cent, will have succumbed by that time. The figure of 22.2 per cent of survivors compares surprisingly closely with that of 25 per cent given by Laurence in his group of patients not operated upon even though his life-table figures were based on a starting age of 3 months.

Discussion

Data in our study show an autopsy percentage of 18 per cent in the nonoperative group (Fig. 7). This low autopsy percentage results in many missed opportunities for study of hydrocephalic processes. The operative group, however, had an autopsy percentage of 72 per cent, indicating a considerably greater opportunity for study of the processes causing hydrocephalus. It is reasonable that with adequate and reasonably undistorted material thus available, experienced neuropathologists, neuroembryologists, and medical geneticists will maintain, thereby, interest in the problem. Study of the multiple etiologies of hydrocephalus by these disciplines in conjunction with the clinicians should solve the problem of the "how and why" of hydrocephalus and myelomeningocele. The ultimate goal of prevention could be realizable in the foreseeable future.

With this in mind, it also is our feeling that a program of the nature we are conducting should be conducted in a large enough medical center so that the disciplines mentioned can be included in the effort constantly to maximize the opportunity for basic solution of the causes of hydrocephalus. The neuropathologist, neuroembryologist, medical geneticist, obstetrician and teratologist should all be part of the team together with the essential neurosurgeon and pediatrician.

Whereas the surgical technique for control
of hydrocephalus has become relatively standardized during the past several years,\textsuperscript{2,14} its use in small hospitals where the above facilities are not available would seem contraindicated if the above attitude is accepted. Such treatment for treatment's sake only, though done with the best of intentions, would serve to delay the thorough and conclusive investigation of the hydrocephalic processes so urgently needed to achieve ultimate prevention.

This report deals with inadequate numbers of patients in any single category (Figs. 1 and 2) of hydrocephalus. Even though 113 children have been studied, significant numbers in any particular etiologic category are not available simply because of the many causes of hydrocephalus itself. Further studies in breadth and depth are needed to provide adequate numbers in single etiologic groups. Our study has numbers statistically insignificant for correlating intelligence and etiology in hydrocephalus, for instance. Since we can control hydrocephalus now, such a correlation will be revealing in regard to the capacity of the brain relative to the lesions producing hydrocephalus.

Moreover, it is evident that the long-term problem of the child with hydrocephalus and myelomeningocele is much greater than the problem of the child with hydrocephalus only.\textsuperscript{9} Laurence and Coates\textsuperscript{11} have shown already that the intelligence quotient in children who have survived 10 years is related directly to physical handicap (i.e. degree of paraplegia, etc.). It would be better, then, to study children who have only hydrocephalus separately from children with hydrocephalus and myelomeningocele.

Arrested hydrocephalus\textsuperscript{10,17} has not occurred in our operative group. To us, this term means that the process originally causing the hydrocephalus has resolved sufficiently to prevent progressive ventricular dilatation, whereas compensated hydrocephalus implies that mechanisms have been brought into action, ordinarily not present normally, which serve to stop ventricular dilatation. In the latter classification are the ventricular dilatation itself\textsuperscript{3} as well as effects of surgical operation. The almost complete lack of arrested hydrocephalus in our operative group has been unexpected and disappointing. This may imply that forces of "circulation" of cerebrospinal fluid\textsuperscript{4} are important in opening up cerebrospinal-fluid spaces closed by inflammatory processes, i.e., shunting procedures prevent this by establishing abnormal patterns of flow of cerebrospinal fluid. It appears so far that once a shunt is established in a progressing hydrocephalic, it may mean life-long need for such.

Extensive experience in the operative technique of the ventriculo-atrial shunting has been achieved, but a detailed account of this needs a separate report.

**Summary**

On the basis of a 5-year study of 113 hydrocephalic children of whom 65 were operated on early with ventriculo-atrial shunting and 48 were not operated on, all of them having been studied periodically in a similar manner to determine mortality, morbidity, intelligence, and related factors, the following conclusions are justified:

1. The program in which shunting operations were done apparently gives significant advantages to the children over the program in which no shunting operations were done (based on life-table data projected from birth to 10 years)
   a. survival: 61.8 per cent as compared to 22.2 per cent;
   b. number of competent children (I.Q. of 75 or better): 33.8 per cent vs. 5.5 per cent;
   c. number of noncompetitive children (I.Q. below 75): 27.6 per cent vs. 16.7 per cent.

2. The major cause of death and morbidity in both groups is infection.

3. In the operative group, morbidity associated with the shunting procedure is related to obstruction of the shunt and septicemia. The early occlusions show a
correlation with abnormal, inflammatory type of spinal fluid being shunted into the blood stream. The late obstructions are associated with mechanical breakage of the shunt or factors of growth causing retraction of the cardiac end from the atrium. Prompt re-establishment of a functioning shunt has been possible in all instances. Periodic follow-up examinations, irrespective of symptoms, have proved invaluable in early recognition of malfunctioning shunts. Septicemia usually required removal of the shunt.

4. From the program of periodic evaluations, including "bubble" ventriculograms, studies of clearance of RISA, and psychological testings, correlations showed:

a. intellectual ability seemed associated with width of cerebral mantle;

b. width of cerebral mantle in hydrocephalus decreases with ventricular pressures of 120 or over; it increases with pressures less than this;

c. prolonged shunting of cerebrospinal fluid can give dramatic increase in width of cerebral mantle, apparently irrespective of basic etiology of the hydrocephalus;

d. intellectual capacity could not be correlated with the basic etiology of the hydrocephalus, but did correlate inversely with duration of increased intracranial pressure.

5. The many etiologies of hydrocephalus require large numbers of patients in each group for thorough evaluation. Factors favoring such studies should be carried out in medical centers where appropriate disciplines can study in depth. Prevention should be the ultimate goal.

6. In the operative group, only 1 patient compensated his hydrocephalic process and no single patient demonstrated "arrest" of or recovery from the basic hydrocephalus-producing process in spite of numerous attempts to demonstrate this. This might imply a necessity for life-long shunting of cerebrospinal fluid in these cases.

The authors wish to thank the entire Neuro-surgical Staff of the University of Washington Affiliated Hospitals including the King County Hospital, the Children's Orthopedic Hospital and the University Hospital for their cooperation in this study. The authors also wish to thank the clinical psychologists on the staff of the University Hospital and the Children's Orthopedic Hospital for the psychological testing of these patients.

References


Discussion*

Dr. Frank E. Nulsen: Dr. Bering's studies on ventricular absorption of cerebrospinal fluid seem to point out that this absorption increases a homeostatic mechanism when the ventricular obstruction increases, pressure becoming more effective with time. This may be the basis for the contrast in behavior of the patient in suddenly versus slowly developing obstructive lesions. The gaining of their ability to tolerate obstruction may be subject to therapeutic manipulation.

His further implication that obstructed ventricles would not enlarge at all in the absence of pulsatile action of the choroid plexus is hardly in agreement with our clinical experience.

Dr. Foltz's study of hydrocephalics not operated upon is more creditable than the recent British communication. Our decisions to withhold treatment for the bad ones do not always solve the situation as he has shown. These 59 per cent survivors to date of this group not operated upon apparently are more of a burden on society than the ones operated upon, and some had neglected treatment with a potential for normal development which any simple gauge like cortical thickness does not always define.

Actually we have 4 of these children who are below Dr. Foltz's critical thickness of 12 mm., all over 5 years of age with adequate I.Q. remains. So this measurement gives one a possibility but not a certainty of retardation.

I have had time to compare these various statistics carefully: Dr. Spitz's 544 cases, Dr. Foltz's 655, and our own 93. There is really a remarkable parallel in long-term mortality, ranging between 20 and 30 per cent, and in functional status in those old enough for testing where 55 to 70 per cent of survivors are in a better than 75 to 85 per cent I.Q. group, including a good number of truly brilliant school-age children.

The good cases prove the potential of the method, while the high proportion of revisions in each series reflects the care and supervision needed to end up with this proportion of nearly normal children. The greatest limiting factor, of course, is the fact that none of the veins in the head or neck tolerate the catheters we have without thrombosis. The tip must lie in the superior cava or below but stop short of the tricuspid valve. Hence longer tubes must be inserted when growth results in pulling the tip of the catheter up to the T5 level where it will become engaged in thrombus.

With some 20,000 Holter valves already in use, to name one brand, it behooves us to educate all users of valves in the prevention of malfunction by accurate

* A third paper presented at this session was "The evolution of the ventriculo-vascular operation in the management of hydrocephalus" by Drs. Pierre L. LeRoy, Luis Schut and Eugene B. Spitz.

initial placement and planned revision for growth and to try to solve further the questions of prognosis and selection as well as the problem of occasional sepsis which I personally believe in all late cases relates to placement close to the tricuspid valve.

Dr. Foltz and his group and Dr. Spitz with his have each contributed greatly to this understanding.

Dr. Oscar Sugar: I only want to bring up Dr. Meyers' semantic difficulty. What is meant by the thickness of the cortex? Because in Chicago, the air studies on these babies show different thicknesses in different parts of the brain. So when you speak of 1 cm. or 1.2 cm., I would like to know what projection you are measuring with what.

Dr. M. N. Estridge: I would like to tell you of our experiences in 10 children who were operated upon more than 5 years ago using the Pudenz-Heyer valve. Of these, 4 are now living. The majority of deaths have been from infection. Those living are in good condition and have not required any revision of the shunt.

Recently, however, we have had two complications related to growth of the body. In one case the tubing became extruded from the vein and formed a fluctuant swelling along the course of the shunt. At operation we were unable to find the vein to perform a revision. The shunt was removed and the child has gotten along satisfactorily for 6 months without further treatment.

In the other case the shunt apparently functioned for 5 years, until the child died from an episode of acute increased intracranial pressure. At autopsy the jugular vein could not be found, nor was there any connection between the vascular system and the valve.

Because of the marked 'growth of the body in the other children we suggest that this operation may be a temporary procedure to allow spontaneous arrest.

I would like to hear further comments on the methods of testing the efficiency of the shunt, and would like to ask Dr. Foltz for suggestions of recognizing and treating children who are going to have difficulty of this sort.

Dr. Edgar A. Bering, Jr.: The measurements we have been making may be contrary to clinical intuition, but they are not, in fact, contrary to clinical experience, because measurements of formation and adsorption of cerebrospinal fluid have never been made in patients using the methods we have used.

Pulse-pressure measurements of cerebrospinal fluid have been made in infants, and the pulse-pressure is enormously high in the hydrocephalic state, but after a shunt or some procedure has been done the pulse-pressure will drop enormously.

The problem of mean pressure is a difficult one. The
increase in pulse-pressure will raise the mean pressure. The pulse-pressure does not fluctuate around some zero line with any change equal above and below, but it is a pulse on a constant base. The lowest point is probably about the same in both the normal or the hydrocephalic, but the patient with the highest pulse-pressure will have the highest mean pressure in the ventricle.

Dr. EDO L. FOLTZ: I shall answer the questions in reverse order.

[Slide] Relative to sudden obstruction of the shunt and the resulting emergent need for revision because of high intracranial pressure, I should like to emphasize that in our program we attempt to avoid such surprises by periodic, thorough evaluation of the functioning status of the shunt (irrespective of symptomatology) by: 1) measurement of ventricular pressure; 2) comparison of ventricular size ("bubble" ventriculogram) with last previous study; 3) results of psychometric tests compared with former results; 4) physical status of the flushing device (evidence of obstruction at either end). If the cerebrospinal-fluid pressure is higher than last recorded, if the ventricles are bigger than last recorded, if the I.Q. shows a "fall-off," if the flushing device is difficult to depress or fill, or if the child has passed through a rapid phase of growth during the last 18 months, direct observation of the shunt and its function probably is needed. Pressure and flow from the ventricular end of the shunt can be ascertained quickly by disconnecting the flushing device and the cardiac end of the shunt can be checked relative to pressure of transmission, as well. At times, this is inconclusive. Under these circumstances, as shown in the slide, the cardiac catheter-valve can be irrigated with 1.5 cc. of Pantopaque during which injection a roentgenogram of the chest is taken. "Hang-up" of dye at the tip of the catheter-valve indicates a mural thrombus is enveloping the tip. This is almost always associated with higher than desired transmission of pressure through the catheter-valve and possibly only intermittent function of the valve. With this evidence which may also show migration of the tip from the atrium at T6 to the superior vena cava at T4, replacement of the cardiac catheter-valve is mandatory before complete shut-off occurs. The involved catheter-valve may be removed, but if not, a second catheter-valve is placed accurately via the internal jugular or subclavian vein to the desired position in the right atrium. Although 50 per cent of such cases show thrombosis of the internal jugular vein, we have yet to fail in re-establishing the shunt on the same side as the original. Such early replacement prior to complete obstruction prevents unwarranted cerebral damage.

[Slide] In using the term "cerebral mantle," we are not referring to cerebral cortex alone. Cerebral mantle consists of the intracranial contents outside the ventricles—between the ventricles and the inner table of the skull. This, thereby, includes white matter, cortex, meninges, dura mater, blood vessels, and cerebrospinal-fluid spaces. This ventriculographic series demonstrates the dramatic change possible in the cerebral mantle. The cerebral mantle at 8 weeks of age measures about 0.8 cm. thick in the frontal region. At 10 weeks of age (7 weeks postoperatively), the mantle measures almost 3.0 cm.

[Slide] Dr. Sugar, width of cerebral mantle is measured quite arbitrarily, but in the same manner each time. The width of the frontal cerebral mantle is measured on the air study as the smallest distance from the anterior-superior ventricular wall (of the frontal horn anterior to the foramen of Monro) to the inner table of the skull in the lateral view. Roentgen-ray factors of distortion are relatively constant and can be ignored in view of the magnitude of the changes recorded.

[Slide] As to how long we follow these children, I can only say we see no end in sight. This slide demonstrates the type of correlation we can thereby achieve. The width of the cerebral mantle by ventriculogram is correlated over a 5-year period with concomitant size of head, ventricular pressure, and intelligence quotient. These studies were done at about the same time, although intermittently, in the 5-year period. Note that the size of the head is poor as an early indicator of rising intracranial pressure from a malfunctioning shunt on 2 occasions, but pressures and width of mantle did change significantly. Such follow-up studies are done irrespective of lack of symptoms.

[Slide] To answer the question as to how early one should shunt a hydrocephalic child, our opinion is that shunting is possible and reasonable as early as one can demonstrate conclusive evidence of progressing hydrocephalus and show the anatomic lesion producing it. The child demonstrated on the screen is only 8 months gestational age, and still 1 month shy of the usual birth age. He was born by cesarean section at 6½ months gestational age because intra-uterine hydrocephalus had been diagnosed by roentgen ray. Differential air study at 8 months of age demonstrated communicating hydrocephalus. It was progressive because of high pressure and enlarging head. Ventriculo-atrial shunt was accomplished, using the infantile type (1 mm. in diameter) of cardiac catheter-valve (Pudenz-Heyer). With such shunts, small-sized infants can be shunted easily and quickly. This child's cerebral mantle has increased very dramatically since operation. It is our impression that the immature hydrocephalic brain of infants only a few weeks old shows a much more rapid and greater increase in width of cerebral mantle following shunting than if shunting is delayed a few months. We suspect this may be related directly to intellectual functioning as well.