Dandy-Walker syndrome: an evaluation of surgical treatment

EDWIN G. FISCHER, M.D.
Department of Neurosurgery of the Children's Hospital Medical Center and the Massachusetts General Hospital, and the Department of Surgery of Harvard Medical School, Boston, Massachusetts

Twenty-seven patients with Dandy-Walker syndrome surgically treated between 1947 and 1972 are reviewed. The results of treatment and the ultimate functional status of the patients are evaluated. The initial treatment in 17 cases was excision of cyst membranes; 15 of these patients required further surgery to control hydrocephalus, while the other two, one of whom had normal pressure preoperatively, still have large heads. Ten patients were treated by ventricular shunting alone. Thus far 11 patients have died; the commonest causes of death have been CSF infection, uncontrolled hydrocephalus, and acute shunt obstruction. In a 1½ to 20-year follow-up, eight patients have normal or near-normal mental function; these results seem to correlate with the absence or control of increased intracranial pressure. Six patients who are alive are clinically retarded; two of these had severe, diffuse neurological deficits before treatment, two had multiple shunt problems and revisions, and two were either retarded or had severe behavior problems before treatment.

Key Words: Dandy-Walker cyst, surgical treatment, excision of cyst membrane, shunts, hydrocephalus

The "Dandy-Walker syndrome" or "atresia of the foramina of Luschka and Magendie" was one of the first causes of hydrocephalus to be treated successfully by surgery. Early reports described occasional long-term satisfactory survival of patients following extensive resection of the posterior fossa cyst membrane. Some of these patients had stormy postoperative courses with increased intracranial pressure gradually subsiding over several months, although this was not always the case. Later, Matson recommended a combination of wide membrane excision (including decompression of upwardly herniated abnormal cerebellar tissue), especially in older patients or if the ventricles were relatively small, followed by shunting, when necessary. Raimondi, et al., in a report of eight patients, felt that excision of the cyst wall was of no value and that all Dandy-Walker cysts should have simultaneous shunting of both the cyst and the lateral ventricles. This approach was evolved because two neonates in their group died immediately following ventricular shunt operations, they thought as a result of "foraminal impaction." A third patient of theirs developed symptoms of progressive medullary compression in the face of a functioning supratentorial shunt and was dramatically improved by a cyst shunt.
Background

“Dandy-Walker syndrome” was the designation applied by Benda to a group of hindbrain malformations characterized by aplasia or hypoplasia of the cerebellar vermis, often associated with anterior and/or upward dislocation of the abnormal vermis; enlargement of the fourth ventricle into a posterior fossa cyst; and, generally, hydrocephalus. This designation was in deference to the descriptions of this group of malformations as a treatable clinical entity by Dandy and Blackfan and Taggart and Walker. Initially, the primary congenital abnormality was thought to be atresia of the foramina of Luschka and Magendie, but this has been an inconstant finding. The pathogenesis and detailed pathological descriptions of these malformations have been heatedly discussed in the past, and have been the subject of extensive reviews. Associated congenital anomalies have been reported including agenesis of the corpus callosum, occipital encephalocele, congenital renal cysts, diaphragmatic hernia, and Meckel’s diverticulum.

Hydrocephalus is the usual mode of presentation in infancy, but neurological findings such as spasticity, nystagmus, ataxia, blindness and other cranial nerve palsies may also be present. Patients developing symptoms later in life may have a clinical picture indistinguishable from a posterior fossa tumor, with headache, vomiting, ataxia, and cranial nerve palsies of recent onset. Such symptoms may occur episodically or following trauma. The malformation has also been found incidentally at autopsy.

Although the skull contour in early months of life may appear normal, enlargement of the posterior fossa is generally obvious on physical examination and plain skull x-ray films. The posterior fossa cyst is associated with an apparent failure of the tentorium to descend to its normal position, and with it the transverse sinuses, an event which may also occur in posterior fossa extra-axial cysts. This elevated position of the sinuses can be detected by transillumination of the skull in the first few months of life, by the abnormally high position of the bony markings of the sinuses on the skull films after 1 year of age, by brain scanning, and by angiography. Other features to be seen on arteriography are: absent posterior inferior cerebellar arteries or an abnormal course of their vermian branches, an absent or small and anteriorly displaced cerebellar blush, and elongation of the vein of Galen.

Ventriculographic findings are symmetrical hydrocephalus with elevation of the occipital horns, a small cerebellar shadow, absence of the fourth ventricle, and an enormous posterior fossa cyst filling from the ventricular system and extending into the cervical canal as low as C-2. Occasionally air may communicate between the ventricular system and the lumbar subarachnoid space. The aqueduct may be blocked to air because of anterior compression of the tectum by the elevated and anteriorly located vermis. Subsequent upward herniation of the cyst through the tentorial notch has been the explanation in some cases for sudden death following ventriculography, and for progressive deterioration following lateral ventricular shunting.

We are reporting a review of 27 patients with the Dandy-Walker syndrome treated surgically over the past 25 years. The purpose of the study was to clarify the indications and usefulness of the three modes of therapy employed (membrane excision alone, membrane excision and lateral ventricular shunting, and lateral ventricular shunting alone) and to learn further what factors in this syndrome, or its treatment were relevant to long-term success or failure.

Clinical Material

The files of the Record Rooms and Neurosurgical Departments of the Children’s Hospital Medical Center and the Massachusetts General Hospital for the past 25 years were searched. We found 27 patients in whom the diagnosis of Dandy-Walker syndrome could be made with confidence from the available x-rays, x-ray reports or operative notes. Two patients were lost to follow-up. Twenty patients (74%) were less than 1 year old at the time of diagnosis and treatment; four were less

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**TABLE 1**
Analysis of 11 deaths (41% of series)

<table>
<thead>
<tr>
<th>Cause of Death</th>
<th>No. of Cases</th>
<th>Treatment</th>
<th>Excision Membrane (EM)</th>
<th>EM and Shunt</th>
<th>Shunt Alone</th>
</tr>
</thead>
<tbody>
<tr>
<td>meningitis or ventriculitis</td>
<td>5</td>
<td>0</td>
<td>5</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>uncontrolled hydrocephalus</td>
<td>1</td>
<td>1 (choroid plexectomy)</td>
<td>0</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>acute shunt obstruction</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>unknown</td>
<td>4</td>
<td>1 (repeat EM)</td>
<td>1</td>
<td>2</td>
<td></td>
</tr>
</tbody>
</table>

than 1 month of age. The female-to-male ratio was 1.7 to 1, a female predominance noted previously by Schreiber and Reye, and comparable to that cited for myelomeningocele and anencephaly.

**Results**

Thus far, 11 patients have died, giving an overall mortality of 41% (Table 1). Two deaths of unknown cause occurred in patients treated by shunt alone, both of whom had ultimately had ureteral shunts inserted following treatments for ventriculitis. The remaining nine deaths were in patients with symptomatic hydrocephalus following membrane excision who were treated by choroid plexectomy, multiple attempts at membrane excision, or shunting. All but one of these deaths occurred between 1947 and 1959. The commonest cause of death was cerebrospinal fluid (CSF) infection (5 patients); other causes included uncontrolled hydrocephalus (1) and acute shunt obstruction (1). The cause of death was not determined in four patients.

Excision of cyst membranes was the primary treatment in 17 patients (61% of the entire series). The outcome for this group is charted in Table 2. Air studies demonstrated failure of the operation to establish communication with the lumbar subarachnoid space in at least three patients. Two patients (11%) did not have increased intracranial pressure postoperatively and therefore received no other treatment; one of them did not have increased pressure at the time of operation, but the other had papilledema and a ventricular pressure of over 320 mm H2O; both are living (11 and 5 years respectively following operation); the first has a severe behavior disorder which existed prior to treatment, the other is doing well with an I.Q. of 95. Uncontrolled hydrocephalus occurred postoperatively in 15 patients (88%), with subsequent shunt operations being performed on 13, choroid plexectomy in one, and additional attempts at membrane excision in the last. Seven of the 13 with shunts died, as did the two who had progressive hydrocephalus treated by choroid plexectomy or repeat membrane

**TABLE 2**
Results of excision of cyst membranes as primary treatment

<table>
<thead>
<tr>
<th>Subsequent Course</th>
<th>No. of Cases</th>
<th>Mental Function</th>
<th>Lost to Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Satisfactory</td>
<td>Poor</td>
</tr>
<tr>
<td>subsequent shunt</td>
<td>13</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>choroid plexectomy</td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>repeat membrane excision</td>
<td>1</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>total</td>
<td>17</td>
<td>6</td>
<td>1</td>
</tr>
</tbody>
</table>

* Includes one patient who was severely retarded before dying of acute shunt obstruction 6 years after treatment.

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excision. Two patients with subsequent shunts were lost to follow-up. Of the four living patients who underwent excision of membranes followed by shunting, one has marked motor retardation, is blind, and his head circumference is at the 3rd percentile 18 months postoperatively. The other three are doing well; one, 4 years after treatment, is considered as bright as her siblings although her head circumference is at the 97th percentile; the second is doing “A work” in the fourth grade, 10 years following treatment, with a head circumference at the 50th percentile; and the third is of “low normal” intelligence, has graduated from high school and works as a clerk for the Internal Revenue Service.

Ten patients were initially treated by ventricular shunt alone, including one patient who had a Torkildsen shunt. The two patients in this group who died were treated in 1954 and 1959, respectively, and died of unknown cause; in each the shunt had been revised to the ureteral type following treatment for ventriculitis. One patient, who was deaf and blind and had nystagmus, strabismus, a well-functioning shunt, and a head circumference at the 3rd percentile, was lost to follow-up 3 years after treatment; he had been considered severely retarded. A second patient, whose shunt never malfunctioned, was severely retarded when treated at 3 years 4 months, and has remained so since. Two additional patients who have done poorly (one has an I.Q. in the upper 60’s, the other is grossly retarded and had an age of 3 years 10 months on the Peabody picture vocabulary test, when his chronological age was 4 years 8 months), required multiple shunt revisions because of obstruction in the early phase of their treatment. Four patients in this group (43% of survivors of the shunt group) have done well; two with normal head growth are developing completely normally 1½ and 3 years following treatment; the third patient is in her normal grade in Junior High School, 14 years following the initial treatment, although her head has always grown at about the 97th percentile, and she has required five revisions for shunt obstruction. The fourth patient had hydrocephalus, but no increased pressure at the time of treatment. She had a posterior fossa exploration and insertion of a Torkildsen shunt, without excision of membranes and ultimately finished high school and married, although her head continued to grow along the 97th percentile.

Two patients had apparent aqueductal obstruction as demonstrated by ventriculography. One was the patient mentioned above who had a Torkildsen shunt. The other patient was treated by excision of membranes followed by a ventriculoatrial (VA) shunt which has been revised several times. She also has done well, but with head growth above the 97th percentile. In neither case were symptoms of upward herniation evident.

In several patients communication between the lateral ventricles and the lumbar subarachnoid space was demonstrated by dye studies but not by air. This communication was best exemplified by one patient whose CSF production and absorption were studied by ventriculolumbar perfusion and found to be normal. At the time of this study, the ventricles were large, but the pressure was 136 mm H₂O; good communication was seen with dye study but was not visible with air. Two weeks following the study, he was readmitted to the hospital with a ventricular pressure of over 600 mm H₂O. It was thought that a small opening in the cyst membrane had been intermittently functioning as a valve.

To test the possibility of enhancement of development of the surface subarachnoid pathways by a functioning shunt as suggested by Milhorat, et al., one patient had a pneumoencephalogram (air inserted from below) at 8 months of age, 4 months following treatment with a successful VA shunt. Air did not pass beyond the basal cisterns.

Discussion

The most important single principle governing the development of adequate or good mental function in this series of patients with Dandy-Walker syndrome seemed to be control of intracranial pressure, whether by successful shunting or by natural mechanisms of compensation with or without shunting. The eight patients (30% of the total series) with the best
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**TABLE 3**

**Summary of eight patients with satisfactory result**

<table>
<thead>
<tr>
<th>Year Treated</th>
<th>Treatment</th>
<th>Age at Treatment (mos)</th>
<th>Control of Hydrocephalus</th>
<th>Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>1952</td>
<td>Torkildsen shunt</td>
<td>11</td>
<td>aqueduct block on ventriculogram, no pressure when treated, head growth parallel to but above 97th percentile</td>
<td>high school grad, married</td>
</tr>
<tr>
<td>1954</td>
<td>excised membrane and shunt</td>
<td>78</td>
<td>head growth parallel to but above 97th percentile</td>
<td>low normal intelligence, high school grad, clerk Internal Revenue Service</td>
</tr>
<tr>
<td>1959</td>
<td>shunt</td>
<td>4</td>
<td>head growth parallel to but above 97th percentile</td>
<td>passing grades in Jr. High School, aged 14 yrs</td>
</tr>
<tr>
<td>1963</td>
<td>excised membrane and shunt</td>
<td>5</td>
<td>aqueduct block on ventriculogram, head growing along 90th percentile</td>
<td>&quot;A&quot; work, 4th grade, aged 11 yrs</td>
</tr>
<tr>
<td>1967</td>
<td>excised membrane</td>
<td>24</td>
<td>no pressure when treated, head growth parallel to but above 97th percentile</td>
<td>I.Q. 95, 5 yrs postop</td>
</tr>
<tr>
<td>1967</td>
<td>excised membrane and shunt</td>
<td>6</td>
<td>head growth parallel to but above 97th percentile</td>
<td>according to mother as bright as siblings 4½ yrs postop</td>
</tr>
<tr>
<td>1969</td>
<td>shunt</td>
<td>11</td>
<td>head size normal</td>
<td>normal development at 3 yrs</td>
</tr>
<tr>
<td>1971</td>
<td>shunt</td>
<td>4</td>
<td>head growing normally, just at 97th percentile</td>
<td>normal development at 1½ yrs</td>
</tr>
</tbody>
</table>

Result had either uncomplicated, functioning shunts with normal-sized heads (3 patients), or persistently large ventricles and large heads despite excision of membranes or shunting, without signs or symptoms of increased pressure (5 patients) (Table 3).

Three of our patients whose hydrocephalus was very successfully controlled by shunting (in two cases following excision of membranes) nevertheless were severely retarded. Therefore good mental function did not always follow control of hydrocephalus. Two of these are still alive; one died of shunt obstruction 7 years following excision of membranes and shunting. All three had evidence of severe neurological abnormality at the time of treatment, with nystagmus, squint, spasticity, and, in two cases, blindness. Such findings, when marked, may therefore be a poor prognostic sign regardless of how well the hydrocephalus is treated, and may reflect a more severe degree of congenital central nervous system abnormality. Minor findings of nystagmus and ataxia are not uncommon in patients who do well and should not be considered prognostically ominous.

Excision of membranes was the most frequent primary treatment in this group. In only one of 18 patients was it thought to have contributed to the control of hydrocephalus; this patient, who was treated at 2 years of age, had papilledema and a ventricular pressure of 320 mm H2O prior to operation, and subsequently had no symptoms or signs of increased intracranial pressure, although her head growth continued along the 97th percentile. In light of the fact that hydrocephalus in this condition is variable, it must be assumed that the development of the mechanisms for circulating or absorbing cerebrospinal fluid must be equally variable. The extremes of this are exemplified by two of our patients, one of whom was a 3½-year-old who had normal CSF production and absorption as determined by ventriculolumbar perfusion. The other was an infant in whom no air passed higher than the basal cisterns on pneumoencephalography after four months of successful ventricular shunting.

In patients with normal absorptive mechanisms, patent foramina of Luschka or small openings in the cyst membrane may be
operating as a valve, which could account for the not infrequent observation of communication between the ventricles and lumbar subarachnoid space by dye, but not by air. In such a situation, membrane excision might theoretically be the appropriate treatment. Such patients would more likely be those presenting at an older age, especially if ventricular enlargement is not marked. Positive identification could be made by CSF absorption and production studies, or possibly by the isotope cisternographic or pneumoencephalographic demonstration of intact subarachnoid pathways over the hemispheres.

The problem of upward herniation of the posterior fossa cyst with brainstem compression, following air study or shunting, was conspicuously absent from this series. It can only be speculated that the high incidence of posterior fossa exploration and membrane excision might be the reason for this. In no case was shunting followed by recognized progressive posterior fossa cyst enlargement or herniation.

The high mortality in patients who had undergone membrane excision was probably due to the fact that all but one of these patients were treated before 1959, when morbidity and mortality from infection and shunting was much higher than it is now. There has been only one death in the nine patients treated since 1959; she had excision of membranes and a VA shunt and died of *Staphylococcus aureus* ventriculitis.

**Conclusions**

Successful control of increased intracranial pressure should be the therapeutic goal in the treatment of the Dandy-Walker syndrome. In our series of 28 patients, this led to adequate mental function (I.Q. of 95 or better) or normal development in 30% of all cases or in 50% of current survivors, including patients whose head continued to grow to an abnormally large size, but at a normal rate and without symptoms of increased pressure.

Ten percent of our patients suffered severe neurological deficits and retardation, despite very good control of hydrocephalus; these may represent patients with more severe malformations of the central nervous system.

The overall mortality rate was 41%; infection, uncontrolled hydrocephalus, and shunt obstruction were the three identifiable causes of death. Four patients died of unknown cause.

Membrane excision relieved symptoms of increased pressure in only one of 16 patients and even then it did not significantly retard head growth. One additional patient, who had a large head but no sign of increased pressure preoperatively, was unchanged by membrane excision. Direct ventricular shunting therefore seems to be the treatment of choice when progressive, symptomatic hydrocephalus is present. Should the posterior fossa cyst enlarge following ventricular shunting causing upward herniation, a situation not recognized in this series, then an additional direct shunt of the cyst, as recommended by Raimondi, *et al.*, might be indicated. Protection against this complication by membrane excision is a possibility that has not yet been clearly demonstrated. If good evidence exists that CSF absorptive pathways and mechanisms are intact, then wide excision of membranes to establish communication with the subarachnoid space is a theoretically rational primary method of treatment.

**References**

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Address reprint requests to: Edwin G. Fischer, M.D., Department of Neurosurgery, The Children's Hospital Medical Center, Peter Bent Brigham Hospital, 300 Longwood Avenue, Boston, Massachusetts 02115.