Epilepsy in children with shunted hydrocephalus


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The incidence of epilepsy among children with hydrocephalus and its relation to shunts and their complications, raised intracranial pressure (ICP), and developmental outcome are explored in a retrospective study.

The authors studied a series of 802 children with hydrocephalus due to varying causes, who were treated by ventriculoperitoneal shunt placement between 1980 and 1990, with a mean follow-up period of 8 years. Patients who had tumoral hydrocephalus and those whose files lacked significant data were excluded. Data extracted from medical records, including history of the hydrocephalus and history of seizures, if any, were analyzed.

Thirty-two percent of the children had epilepsy, the onset of which frequently occurred at approximately the same time that the diagnosis of hydrocephalus was made. The majority of the affected children had severe uncontrolled epilepsy. The incidence of epilepsy was significantly affected by the original cause of the hydrocephalus. The presence of radiological abnormalities was also found to be a significant predictor of epilepsy. Similarly, shunt complications predisposed to epilepsy. Episodes of raised ICP related to hydrocephalus or in association with shunt malfunction may also predispose to epileptic seizures. Furthermore, the presence of a shunt by itself seems able to promote an epileptogenic focus. Finally, epilepsy appears to be an important predictor of poor intellectual outcome in hydrocephalic children with shunts.

A prospective study is needed to identify clearly and confirm avoidable factors predisposing to seizures in these children so that we can strive to reduce the incidence of these seizures and, subsequently, improve quality of life.

Key Words * epilepsy * hydrocephalus * ventriculoperitoneal shunt

Children requiring ventricular shunts have a higher incidence of seizures than the general population, varying from 6 to 59% in the reported literature.[2,4,5,9,11,13,15,17,22-25,29,32-35] There are many factors that may account for this, including the cause of the hydrocephalus, its subsequent treatment, and
any resulting complications. A few studies have indicated that presence of a shunt,[1,10,13,21] number of shunt revisions,[3-5,10,13,23,33] history of shunt infection,[1-4,13,23,32] and, perhaps, shunt location[5] increase the risk of developing seizures. However, long-term follow-up review has rarely been provided and a number of important questions remain unanswered. For example, the relationship between raised intracranial pressure (ICP) and seizures and the way in which seizures in hydrocephalic children may affect developmental outcome have been poorly documented. In addition, there is little information available concerning the type and frequency of seizures or electroencephalography (EEG) findings. To address all of these questions, we have analyzed data obtained in a series of children with hydrocephalus to gain a more complete picture of epilepsy in this population.

CLINICAL MATERIAL AND METHODS

Patient Population

In the period between 1980 and 1990, the cases of 802 children with hydrocephalus who had been treated with ventricular shunts at the Hospital Necker-Enfants Malades were studied and followed. Hydrocephalus requiring placement of a cerebrospinal fluid (CSF) shunt was always diagnosed using computerized tomography (CT) and/or magnetic resonance (MR) imaging. The criteria used to determine the need for shunt placement included evidence of raised ICP in combination with radiological studies revealing evidence of either hydrocephalus at the first examination or progressive ventricular enlargement.

For the purpose of this study, 45 children who did not survive for longer than 2 years after shunt implantation were excluded because it was believed that the information available in those cases would be inadequate. Two hundred fifty-seven patients who exhibited hydrocephalus in association with a tumor were excluded to avoid potential effects that the tumor may have had on any epileptic activity. Also excluded were 216 children in whom data were incomplete, resulting in 802 children entering into the study.

Compilation and Analysis of Data

Data extracted from the patients' medical records included information on gender, cause of the hydrocephalus, prenatal and perinatal history, developmental history, school performance, and family history of nonfebrile seizures. In addition, we also recorded the presence or absence of seizures, patient age at onset of seizures, EEG findings, age at first shunt placement, type of shunt procedure, number of shunt revisions, and complications.

Seizures were divided into different categories: simple partial seizures, complex partial seizures, simple partial seizures accompanied by secondary generalized tonic-clonic seizures, complex partial seizures accompanied by generalized tonic-clonic seizures, and mixed simple partial-complex seizures (focal clonic or tonic seizures with complex symptomatology), that is, unilateral motor seizures in addition to complex partial seizures. Seizure frequency was also divided into four groups: daily (at least one seizure per day [and sometimes many more]), weekly, monthly, and occasional (febrile convulsions without consequent epilepsy).

Interictal surface EEG data were assessed for baseline activity, the presence and topography of focal spikes, and the presence of generalized paroxysmal discharges in each child. Baseline activity was classified as normal, asymmetrical, or slow. Focal spikes were localized, regional, or multifocal, with or without focal, regional, or diffuse slow waves. Generalized paroxysmal discharges included diffuse
irregular spike-and-wave activity or the presence of multiple spikes and waves but excluded diffuse high-voltage slow bursts.

During the follow-up review, serial neurological evaluations and developmental testing were performed. The type of evaluation depended on the age of the child. Generally evaluations were performed every 6 months for the first 2 years of life and yearly thereafter. The children's intelligence quotients (IQs), which were determined according to the Brunet Lezine or the Wechsler test, were categorized into: "normal," either verbal or performance IQ greater than 90; "slightly retarded," either verbal or performance IQ between 70 and 90; "moderately retarded," IQ between 50 and 70; and "severely retarded," IQ less than 50. The last group was characterized by learning disabilities that included deficits in memory, attention, and speed, and problem-solving ability. Behavioral and psychosocial aspects were divided into four groups: normal behavior, psychological impairment (such as inhibition or obsession), hyperactivity, and psychosis. School performance was also studied and the children were grouped as follows: normal schooling; moderate difficulties with a school delay lasting less than 2 years; significant difficulties with a school delay lasting 2 years or more; children with no possibility of schooling; and children who were too young (< 6 years) for school. In addition, some children who did not receive formal testing were judged to be of average intelligence on the basis of their academic performance and progress in regular school, even though some patients in this group may have had unrecognized learning disabilities.

Standard statistical methods were used to analyze the different variables.

**RESULTS**

During the specified period, all 802 children included in the study were treated for hydrocephalus with CSF shunt placement. The median follow-up period after shunt insertion was 7.6 years (range 1-26 years). The distribution of the different causes of the hydrocephalus is summarized in Table 1 and is characteristic of the usual causes of hydrocephalus in children living in Western countries. The majority of children (73%) were revealed to have severely enlarged ventricles on preoperative CT scans.

<table>
<thead>
<tr>
<th>TABLE 1</th>
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<tbody>
<tr>
<td>CAUSES OF HYDROCEPHALUS IN 802 CHILDREN</td>
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<tr>
<td>Cause</td>
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</tr>
<tr>
<td>myelomeningocele</td>
</tr>
<tr>
<td>hemorrhage</td>
</tr>
<tr>
<td>infection</td>
</tr>
<tr>
<td>malformation</td>
</tr>
<tr>
<td>prenatal</td>
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<tr>
<td>other</td>
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<tr>
<td>unknown</td>
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</table>

**Shunt History**

All children were treated with conventional differential, medium-pressure valves. The location chosen for ventricular catheter placement was based on the site of maximum ventricular dilation. The location was usually in the occipital horn of the lateral ventricle (92%). The right side was the preferred approach (85%). In 51.3% of the children, the age at first shunt insertion was younger than 3 months, in 32% the age ranged from 3 months to 1 year, and in 16.7% of the children the age was older than 1 year. A total of 1637 operations were performed with an average of two operations (range 1-18 operations) per patient within the given study period.
Thus far, one-half of the children have exhibited no complications. Shunt complications requiring surgical intervention occurred in 401 (50%) of the 802 children, of whom 220 (27%) had one complication, 90 (11%) two complications, and 91 (11%) more than two complications. Approximately one-third (36%) of the children experienced a mechanical shunt malfunction, 4.4% suffered infective complications, and another 9.6% a combination of both mechanical and infective complications. Thus the infection rate for the entire series was 14% per patient and 6.8% per procedure. Shunt revisions were equally distributed among proximal, distal, and complete replacement. On postoperative CT scans the majority of the patients had either normal-sized ventricles (34%) or slit ventricles (30%), with the remainder showing some degree of enlargement.

**General Observations on Seizures**

Seizures occurred in 255 (32%) of the 802 children at some time during the follow-up period. Of these 255 patients, 73 patients (28.6%) experienced their first epileptic seizures before initial shunt placement, whereas seizures developed in the remaining 180 patients (71.4%) after shunt placement. Analyzing the time interval between the onset of epilepsy and the time of shunt insertion, there was a peak incidence of seizure occurrence around the time of shunt insertion, with the majority being observed in the days preceding shunt insertion (Fig. 1).
Fig. 1. Upper: Graph demonstrating a peak in seizure occurrence around the time of shunt insertion. Lower: Graph showing a more detailed analysis of this peak, in which an increase in seizure activity precedes shunt insertion by a few days. These graphs suggest two different patterns of seizure provocation in hydrocephalic patients. In the immediate preshunt implantation period, an increase in seizure activity is possibly triggered by the intracranial hypertension of the decompensated hydrocephalus (as suggested by the reduction in seizure activity in the weeks following shunt insertion [lower]), whereas after shunt insertion numerous previously nonepileptic children developed partial recurrent seizures (as demonstrated in the upper graph in which 71.4% of patients are shown to have developed their seizures after the shunt had been placed). N = number; Std. Dev = standard deviation.

The overall incidence of seizures in children with shunts was 32%, with two types of patients emerging. The minority (35 children [4.4% of the 802 patients]) were those who had only occasional seizures and infrequent epileptic episodes, often with recognized associated causes such as a high fever or meningitis. Most epileptic patients (220 children [27.4% of the 802 patients]), however, had recurrent unprovoked seizures for at least 2 years and their conditions were poorly controlled with medication. Recurrences
were observed even after institution of antiepileptic medication and most of these patients had seizures that did not respond well to antiepileptic drug regimens consisting, for the most part, of two or three anticonvulsants. The seizure frequency in these children was at least one prolonged convulsion per month; unfortunately, most experienced several seizures per week.

There was a significant difference (p < 0.001, chi-square test) in the severity of epilepsy between patients in whom seizures developed before shunt implantation (preshunt group) and those in whom seizures developed afterwards (postshunt group). In the preshunt group, a high incidence (86%) of occasional seizures was observed, whereas in the postshunt group most (80%) of the children had recurrent epilepsy. Of the 72 patients in the preshunt group, 33% had a seizure during the 1st month of life and 80% before 1 year.

Of the 255 children with shunts who experienced seizures, 82 (32.2%) had secondary generalized tonic-clonic seizures and 111 (43.5%) had simple partial seizures, of whom 95 (37.3%) had focal clonic seizures and 16 (6.3%) had focal seizures with complex symptomatology. An additional 39 patients (15.3%) have manifested a combination of several types of epilepsy and the remaining 23 children (9%) had catastrophic epilepsy with infantile spasms.

Significant differences in seizure characteristics were observed between the pre- and postshunt groups. In the preshunt group a high incidence of generalized epilepsy was observed, whereas in the postshunt group a high incidence of partial epilepsy was noted (Fig. 2).

![Pie charts depicting seizure characteristics in the pre- (left) and postshunt (right) groups.](image)

In both pre- and postshunt groups, in approximately one-third of the patients the epilepsy was not controlled by an adequate trial of a "first-line" conventional antiepileptic drug, used either as the sole therapy or in combination with other treatment; this proved to be severe epilepsy. Seizure frequency varied among patients but in the most cases was high: 108 (42%) suffered from daily seizures, 47 (18%) from weekly seizures, 53 (21%) from monthly seizures, and 47 (18%) had rare attacks of seizures. Common to the children with daily seizures was the occurrence of attacks of clusters of seizures lasting a few days at a time with an average seizure frequency of 10 to 20 attacks per day. Episodes of partial status epilepticus were documented in 126 patients (49.4%); these episodes required admittance to the
hospital.

**Original Cause of the Hydrocephalus**

In comparing the proportion of children with seizures, a significant difference emerged between groups with different etiologies (p < 0.001; chi-square test). Children with a myelomeningocele experienced a significantly lower overall prevalence of epilepsy (7%), compared with those born prematurely or with cerebral malformations (30%) and those with prenatal hydrocephalus (38%). Moreover, meningitis and postinfective hydrocephalus carried a high risk for epilepsy, on the order of 50%.

**Radiological Abnormalities and Central Nervous System Malformations**

The presence of radiological abnormalities on CT scanning (33% of the patients) was associated with a high incidence of epilepsy (45%) (Fig. 3).

![Fig. 3. Pie chart depicting the relative frequency of epilepsy in relation to abnormalities seen on neuroradiological examination. CC = corpus callosum.](image)

A central nervous system (CNS) malformation observed on a CT scan, such as agenesis of the corpus callosum, focal migration abnormalities, encephalocele, holoprosencephaly, absent or hypoplastic cerebellum, and posterior fossa cyst, was a statistically significant variable in predicting seizure occurrence with a higher incidence of epilepsy. Malformations not known to involve the CNS, such as polymalformative syndrome, were also found to be predisposing factors for the occurrence of seizures in hydrocephalic children.

**Birth Insults**

One-third of the children had suffered one or more injuries at birth (anoxia 8%, hemorrhage 9%, infection 10%, or multiple injuries 8%). The existence of birth injuries was another significant risk factor for the development of seizures (p = 0.001; chi-square test). There was a higher incidence of epilepsy among children who suffered hemorrhage (41%), infection (47%), or anoxia (68%) during the neonatal period than among children who did not have a positive history of birth injury (27%).

**Influence of Shunt Complications and Raised ICP on Epilepsy**
Table 2 shows the relative frequency of shunt complications in three groups, divided into those patients who had no fits, those who had occasional seizures, and those who had recurrent epilepsy. The average number of shunt revisions in children with seizures was significantly greater than in those who did not have seizures. There was a higher incidence of epilepsy among children who had shunt malfunction, infection, or a combination of both. Moreover, there was a direct correlation between the incidence of epilepsy and the number of shunt revisions, ranging from only 20% (no revisions) to 52% (three or more revisions).

<table>
<thead>
<tr>
<th>Shunt Problem</th>
<th>No Seizures</th>
<th>Occasional Seizures</th>
<th>Epilepsy</th>
</tr>
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<tbody>
<tr>
<td>Shunt malfunction</td>
<td>76</td>
<td>4</td>
<td>20</td>
</tr>
<tr>
<td>Infection</td>
<td>63</td>
<td>4</td>
<td>33</td>
</tr>
<tr>
<td>Shunt malfunction &amp; Infection</td>
<td>51</td>
<td>12</td>
<td>37</td>
</tr>
<tr>
<td>Shunt malfunction &amp; Infection &amp; Infection</td>
<td>53</td>
<td>4</td>
<td>43</td>
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In this study we also attempted to determine whether the onset of seizures or the increased frequency of seizures is a reliable indicator of increased ICP secondary to shunt dysfunction. Common presenting symptoms or signs of shunt dysfunction that were recognized as indicators of increased ICP included: headache, vomiting, increased head circumference, lethargy, coma, irritability, neck pain, opisthotonos, and impairment of visual acuity (papilledema, amblyopia, and hemianopia). The temporal relationship between seizures and shunt dysfunction was defined as seizure activity that occurred within a few days preceding the diagnosis of shunt malfunction and was not accounted for by an obvious precipitating event such as a febrile illness, an intercurrent infection, or a seizure disorder with deranged therapeutic blood anticonvulsant levels. As shown in Table 3, the majority of the 407 children admitted for shunt dysfunction presented with clinical evidence of intracranial hypertension; however, seizures alone were the presenting symptom of shunt dysfunction in 8.6% of the whole series. Furthermore, seizures were the sole presenting symptom of shunt dysfunction in 28.3% of the children with a previous history of epilepsy.

<table>
<thead>
<tr>
<th>Presenting Symptom</th>
<th>Patients W/ History of Seizures (%)</th>
<th>Patients W/O History of Seizures (%)</th>
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<tr>
<td>Intracranial hypertension</td>
<td>56.6</td>
<td>94.8</td>
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<tr>
<td>Seizures</td>
<td>26.3</td>
<td>3.5</td>
</tr>
<tr>
<td>Seizures &amp; intracranial hypertension</td>
<td>9.4</td>
<td>1.0</td>
</tr>
<tr>
<td>Other</td>
<td>5.7</td>
<td>0.7</td>
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Electroencephalography was performed in all of these patients; the results showed abnormal electrical activity, either diffuse or focal. In some of the children infantile spasms were diagnosed, whereas in the remaining children the diagnosis was either generalized seizures, partial motor seizures (sometimes in clusters or of the type "hemiconvulsion-hemiplegia"), or complex partial seizures.
Role of the Ventricular Catheter in Epilepsy and EEG Findings

The trauma of catheter placement and the subsequent presence of a foreign body within the substance of the brain are potential causes of focal cerebral dysfunction. For this reason we elected to examine children before and after shunt placement procedures by using EEG. Normal and abnormal EEG findings for the preshunt and postshunt groups are summarized in Fig. 4.

![Fig. 4. Pie charts demonstrating EEG findings in the pre- (left) and postshunt (right) groups.](image)

Paroxysmal discharges were tabulated as focal if only one focus (spikes isolated, continuous or in bursts) was demonstrated on all records for a given child. If two or more foci of spikes were demonstrated, the tracing was classified as a multifocal paroxysmal discharge record. It is interesting that most children, including some with CNS malformations, were reported as having generalized seizures, although the common EEG finding was of focal paroxysmal features, suggesting that a focal onset of the seizure may not have been recognized.

In EEG examinations performed before shunt insertion we found a 27% incidence of a single epileptic focus. This increased to a 50% incidence on postoperative EEG. However, a significant number of patients (45%) in this series had radiological abnormalities, which in their own right could be an epileptic focus, because we found a direct relationship between the side of the abnormalities and the side of the EEG focus (Table 4). This made it difficult to know whether it was these lesions or the brain injury related to the shunt that was responsible for the focal EEG paroxysmal discharges or slow waves.

<table>
<thead>
<tr>
<th>Location of Lesion</th>
<th>EEG Abnormalities (percentage of patients)</th>
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<tr>
<td></td>
<td>Bilat</td>
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<tr>
<td>bilat</td>
<td>67</td>
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<tr>
<td>rt</td>
<td>17</td>
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<td>lt</td>
<td>3</td>
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To explore this further, we selected from our patients a small subgroup of children with epilepsy, in whom no radiological abnormalities were found on CT or MR imaging and in whom right posterior parietal ventricular catheters remained on site even after shunt revision. In these 33 patients, a more
detailed analysis of the nature of the EEG abnormalities showed that bilateral abnormalities occurred in 76% of the patients before shunt implantation. In only 8% of the 33 children, spike-and-wave activity was found on EEG prior to the initial shunt placement, whereas in the postshunt group an epileptic focus appeared in 30% in the right hemisphere (Table 5) and in 39.4% on the posterior parietal area, localized to the general region of the shunt (Table 6). Thus, in most of the children with no evidence of a cerebral parenchymal lesion on CT scanning, the focal side of the interictal EEG anomalies was identical to the area of shunt insertion.

In some children who displayed hemiconvulsions followed by a transient hemiplegia, the discharges of slow or spike waves started in the right parietal area where the valve was inserted and involved the anterior regions. Therefore, in most of the cases, the spikes were present at the onset of the first seizure, but were preceded by a focus of slow waves.

With regard to shunt site and its influence on epilepsy, because nearly all our patients have been treated via a posterior approach, a valid statistical comparison with patients in whom a frontal entry point was used could not be drawn.

Seizures and Prognosis

Overall, the prognosis for seizure control was not good. Only 70 of the 255 hydrocephalic children had their antiepileptic medication discontinued for any period of time. Withdrawal of antiepileptic drugs was successful in 35 children who had been free of seizure for at least 3 years and in whom the seizures were attributed to transient conditions. None of these 35 children has had recurrent seizures at follow-up review. Thirty-five other children whose seizures were considered to represent a more chronic seizure disorder were treated for longer seizure-free periods before medication was discontinued. Fourteen of them have remained free of seizure, but 21 suffered a relapse within 6 months of cessation of antiepileptic drug therapy. Thus in these children it was concluded that antiepileptic drug monotherapy was necessary to provide an acceptable degree of seizure control.

To summarize seizure outcome, two different patterns could be observed between those patients who developed seizures before shunt implantation and those who developed them afterward. The nature of the

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<th>TABLE 5</th>
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<td><strong>SIDE OF EEG ABNORMALITIES IN THE PREOPERATIVE AND THE POSTOPERATIVE PERIODS</strong></td>
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<tr>
<td><strong>EEG Abnormalities (percentage of patients)</strong></td>
</tr>
<tr>
<td><strong>Timing</strong></td>
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<tr>
<td>before shunt placement</td>
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<td>after shunt placement</td>
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<table>
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<th>TABLE 6</th>
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<tr>
<td><strong>LOCATION OF POSTOPERATIVE EEG ABNORMALITIES</strong></td>
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<tr>
<td><strong>Location</strong></td>
</tr>
<tr>
<td>anterior</td>
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<tr>
<td>posterior</td>
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<td>multilobar</td>
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<td>diffuse</td>
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seizures in the preshunt group was often generalized and outcome was clearly related to shunt insertion, with 63% of patients improving after implantation and the remaining patients continuing to suffer from severe epilepsy. On the other hand, the nature of the seizures in the postshunt group was mainly of a focal type and was refractory to medical treatment in most cases, with only 35% of patients improving with antiepileptic medication.

**Developmental Evaluations**

Of the 547 hydrocephalic children without epilepsy, 409 underwent a developmental IQ assessment. Twelve percent of these children were mildly retarded, 13% were moderately retarded, and 9% were severely retarded. The majority (66%) of the patients had normal intelligence. In contrast, of the 255 hydrocephalic children with epilepsy, only 24% had normal intelligence with 23% being mildly, 18% moderately, and 35% severely retarded. Mental retardation was statistically significantly higher in children continuing to require medicine for their seizures compared with those whose seizures were in remission and for whom treatment was discontinued. This indicates that epilepsy correlated strongly with poor cognitive outcome. With regard to psychological outcome, there was a striking difference between hydrocephalic patients with seizures and those without seizures. In those patients with seizures 32% displayed normal behavior and 20% were psychotic; in those without seizures 80% demonstrated normal behavior. Schooling outcomes also demonstrated the dramatic influence that seizures have on academic achievement in these children. The majority of the hydrocephalic children without seizures were enrolled in a normal school curriculum as opposed to those with seizures, of whom one-half were institutionalized (Fig. 5).

![Fig. 5. Bar graphs showing schooling outcomes in hydrocephalic patients with and without epilepsy.](image)

**DISCUSSION**

There have been many reports and publications, in particular with regard to epilepsy, concerning the postoperative course and complications associated with the treatment of hydrocephalus by insertion of ventricular distal shunts. However, the relationship between shunts and their complications and patients' seizures, the interaction between intracranial hypertension and seizures, and the influence that these may
have on developmental outcome have been poorly documented.

There is general agreement in most of the literature that the incidence of epilepsy in children with hydrocephalus stemming from all causes is approximately 30%, which agrees with the incidence of 32% observed in our series. However, in contrast with most publications in which only the overall frequency of seizures is reported, in this study we differentiate between transient seizures and epilepsy, finding an incidence of occasional or transient seizures in 4.5% of the children and recurrent seizures in 27.5%.

With regards to the influence that the original cause of the hydrocephalus may have on the incidence of epilepsy, there remains a diversity of opinion in the literature. Contrary to the findings of the majority of authors[1,2,13,15,23,24,26] who have noted a correlation, others[4,29,32] have found no correlation between the cause of hydrocephalus and the incidence of epilepsy. Lorber and colleagues[20] and Hosking[13] attributed the high prevalence of epilepsy to underlying brain anomalies or disease. In most reports, children with a myelomeningocele experienced a significantly lower overall prevalence of epilepsy,[1-3,15,24] whereas meningitis[2,13] and cerebral malformations[1,23,24,32,33] are strongly correlated with epilepsy. Analyzing our results has shown quite clearly a correlation, not only for those patients with postinfective hydrocephalus but also for those whose hydrocephalus had other causes. Overall we found that myelomeningocele carries a low risk at 7%, cerebral malformations and intraventricular hemorrhage a moderate risk of approximately 30%, and infection a high risk in the order of 50%.

We have also found that other factors, such as a history of birth injury or insult, may play a significant role in the development of seizures in hydrocephalic children. Such a history has not been previously found to correlate with development of epilepsy in such children.[23,24,32] However, in contrast, we found a high correlation with children sustaining a birth injury and subsequent epilepsy, with anoxia showing the highest incidence (68%), followed by infection (47%) and hemorrhage (41%).

The patient's age at shunt insertion is reported to influence occurrence of seizures, the risk being highest in the youngest patients.[4,34] As reported by other authors, we were not able to find such a correlation in our series.[5,20,23,24]

With respect to how shunt complications may affect the subsequent development of seizures, one must consider the roles of shunt infection and shunt dysfunction separately. With a few exceptions,[20,26,29] most reports agree that shunt infection predisposes to epilepsy.[1-4,13,23,32,33] Disagreement remains on the influence that shunt malfunction may have on the risk of epilepsy. In some series[2,20,24,26,29,32] epilepsy was not commonly associated with shunt malfunction, thus not confirming the results of other investigators.[1,3-5,13,15,33] In our series, however, there was a higher incidence of epilepsy among children who had shunt malfunctions, infection, or a combination of both. Moreover, and perhaps surprisingly, there was a direct correlation between the incidence of epilepsy and the number of shunt dysfunctions requiring revisions. These results contradict those previously reported in the literature.[2,32]

Another important issue that has attracted debate in the past is whether seizure activity could be the presenting symptom of shunt dysfunction. In some reports an increased number of seizures or appearance of seizures is mentioned when a shunt is not working well.[8,15,21,31] Faillace and Canady[8] emphasized the importance of the onset of a first time seizure or new seizure activity in a child in whom a shunt had previously been implanted, despite a relatively low incidence of 2%. A relationship between seizure and shunt dysfunction or increased ICP was proposed but not clearly identified. Others do not
agree with these findings,[2,23] whereas some take an intermediate view and conclude that although the presence of seizures alone is not a good predictor of shunt malfunction, it may be seen as one of the presenting symptoms.[7,12,13,32] We found that, in patients with a previous history of epilepsy, seizures (either new onset after a long time or increased activity) were the presenting symptom of malfunction in 28%, whereas seizure as a presenting symptom only occurred in 3% of children without such a history.

Considerable controversies surround the influence of a ventricular catheter on the total risk of epilepsy. In contrast to hydrocephalic children in whom the overall reported rate is approximately 30%, the incidence of epilepsy following placement of subdural shunts and ventriculostomy is very low.[19,30] Moreover, the development of epilepsy occurring specifically after shunt placement for hydrocephalus could be directly related to shunt implantation, as evidenced by the widely varying values of reported newly occurring postshunt epilepsy, ranging from 7.2% reported by Venes and Dauser[35] to 18% and 24% quoted by Marossero and colleagues[22] and Copeland and associates.[4] respectively. In another study, Johnson, et al.,[15] suggested that the incidence of epilepsy could increase proportionally with the duration that the brain is exposed to the catheter because the majority of seizures occur months after the shunt is inserted. Collectively these reports suggest that the occurrence of cortical injury at the time of shunt placement could be a factor in the development of seizures.

The influence of the ventricular catheter has been further explored by studying EEG abnormalities, which have been frequently found in patients in whom shunts have been placed.[9,11,14,18,33,34] Most interestingly, focal paroxysmal discharges have been reported to be significantly more frequent in patients with shunts than in patients without shunts.[16,25] This has been further discussed by Graebner and Celesia,[11] who found focal specific paroxysmal discharges and slow waves in the area of the ventricular catheter in 85% of hydrocephalic children with shunts as compared with 62% of hydrocephalic children without shunts. These authors concluded that shunts could be responsible for a cortical injury.

It is difficult to relate the onset of epilepsy to the insertion of a ventricular catheter through the cerebral mantle and not to the cause of the hydrocephalus or to a focal cerebral lesion as identified on neuroradiological examination.[3,17,23,24,26-29,32,35] In clarification of this point, we selected a small group of 33 patients with postshunt epilepsy who demonstrated no radiological abnormalities and had right posterior parietal ventricular catheters that remained on site even after revisions. In this subgroup there was a right posterior parietal focus in as many as 39.4%. Moreover, the seizures in this subgroup of children without radiological abnormalities were partial and originated at the irritative "shunt-linked" focus. Of particular interest, this irritative focus was always located where the catheter was first inserted, was not present before the operation, and developed approximately 1 or 2 years after insertion of the shunt, often on a focus of slow waves.

Having dealt with all the factors that may cause the seizures, it is important to identify any factors that could predict seizure remission and subsequent cessation of medical therapy. Noetzel and Blake[23,24] have attempted to determine this. Their study indicated that mental retardation or CNS malformations are poor predictors of poor seizure control, whereas children older than 3 years of age at seizure onset, who have no EEG abnormalities and have been seizure free for 3 years while receiving anticonvulsant medications, have a good chance of remaining free of seizure after withdrawal of medical treatment. Our results have led us to similar conclusions: cessation of the antiepileptic drug regimen, albeit in a small proportion of our patients, was successful in 35 children who had been seizure free for at least 3 years and in whom seizures were attributed to transient conditions. Perhaps one of the most important points is
the universally accepted fact that the presence of seizures in children predisposes to poor intellectual outcome.[1,6,23,24,32] In our series, we not only found that epilepsy correlated strongly with poor cognitive outcome but that, in addition, there were similar strong negative correlations between epilepsy and psychological and educational outcomes as well.

Because we are now aware of some predisposing factors that are associated with seizure development in these children, we must focus on ways to reduce their occurrence. One way is to avoid shunts altogether by using alternative procedures, such as neuroendoscopic third ventriculostomy to treat hydrocephalus when anatomically possible, both as a primary treatment and in cases of shunt malfunction.[30] Another is to avoid shunt complications by taking meticulous steps to reduce infection and also subsequent shunt malfunction by striving to improve surgical technique and to develop improved materials and valves. In this way, some impact may be made on improving intellectual and psychological outcomes in these patients by reducing their overall incidence of epilepsy.

CONCLUSIONS

We conclude from our data that epilepsy is associated with infantile hydrocephalus in one-fourth to one-third of hydrocephalic children and its onset occurs at approximately the same time that diagnosis of hydrocephalus is made.

The cause of the hydrocephalus and the presence of radiological abnormalities were found to be strong predictors of epilepsy. In addition, episodes of intracranial hypertension related to hydrocephalus or shunt dysfunction may predispose a child further to have epileptic seizures. We believe it is essential to emphasize the importance of the onset of a first-time seizure or new seizure activity in a child who has a shunt.

Shunt complications, both infective and mechanical, predispose to epilepsy, but even the presence of a shunt catheter on its own could promote an epileptogenic focus.

Finally, epilepsy appears to be an important predictor of poor intellectual outcome in hydrocephalic children in whom shunts have been placed.

A prospective study is needed to identify clearly and confirm avoidable factors predisposing to seizures in these children so that we can strive to reduce the incidence of these seizures and, subsequently, improve these children's quality of life.

References


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