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Osamu Sato
Tsuyoshi Yamguchi
Mamoru Kittaka
Hiroyuki Toyama

Hydrocephalus and epilepsy

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O. Sato (✉) · T. Yamguchi · M. Kittaka
H. Toyama
Department of Neurosurgery,
Ikegami General Hospital,
1–9, 6-chome, Ikegami, Ohta-ku,
Tokyo, 146-8531, Japan
e-mail: sammy@xj8.so-net.ne.jp
Tel.: +81-42-7269384
Fax: +81-42-7269384

Abstract Since the introduction of ventriculo-atrial and/or ventriculoperitoneal shunting for hydrocephalic patients, controversies have developed regarding the likelihood of epileptic seizures developing as a result of the shunting itself and/or its complications. On the other hand, hydrocephalus is not commonly recognized as a cause of seizures in general, although epilepsy is reported to be frequently associated with shunt-treated hydrocephalus, especially in children. Several authors have reported an increased risk of epileptic seizures after shunt placement, but the underlying mechanisms are still controversial. The insult to the brain at the time of ventricular catheter insertion, the presence of the shunt tube itself as a foreign body, the burr hole location, the number of shunt revisions after malfunction, associated infection, the etiology of hydrocephalus, and associated mental retardation are thought to be related to the risk of epilepsy. Age at the time of initial shunt placement also seems to be an important factor. Early shunting is a well-known determinant of risk in shunt obstruction, and children less than 2 years old are consequently at a higher risk of developing epilepsy than older ones. It is reported that antiepileptic drug treatment is not so reliable as might be expected. Conscientious and more sophisticated EEG recording in those children may be beneficial during follow-up. The incidence of seizures in shunted children is reported to be quite high, ranging from 20% to approximately 50%, so that neurosurgeons should pay more attention to the issue of epilepsy in hydrocephalic children. Although ventriculo-extracranial shunts have been the standard treatment for hydrocephalus for decades, the long-term morbidity, including postshunt epileptic seizures, has to be taken seriously. The use of neuroendoscopic techniques when indicated may ameliorate this problem a great deal in the future.

Keywords Hydrocephalus · Epilepsy · CSF shunt · Antiepileptic drugs · Site of burr hole · Mental retardation

Introduction

Seizure disorders are relatively prevalent in children with hydrocephalus of various etiologies and often raise serious problems in their long-term outcome.

Although hydrocephalus can be caused by a variety of congenital anomalies and acquired diseases, the estimated incidence with epilepsy has been reported as much higher in children treated by shunting than in those not so treated. There are many factors that can influence the prevalence of epilepsy, and these include etiologies of hydrocephalus, age at diagnosis of hydrocephalus or shunt placement, intracranial pressure, site of burr hole, number of revisions, time-course after shunt insertion,

use of antiepileptic agents and complications such as infection and shunt tube obstruction. There are many reports in which these issues have been extensively discussed, but their results are still diverse and they remain controversial in many aspects.

This article reviews the most recent and other selected papers on epilepsy in hydrocephalic children, in the hope that it will provide some basis for guidelines on this issue.

Epilepsy in children with shunted hydrocephalus [3]

Presentation

Utilizing computed tomography (CT) and magnetic resonance (MR) imaging, 802 children were diagnosed as hydrocephalic with a need for placement of a cerebrospinal fluid (CSF) shunt. Evidence of raised intracranial pressure (ICP) accompanied by radiological evidence of enlarged ventricular system was taken to indicate necessity of shunt insertion. Children who did not survive for more than 2 years after shunt placement and those in whom the hydrocephalus was caused by tumors were excluded from the study, as were any whose data were not appropriate to the study.

Seizures were divided into the following categories: simple partial, complex partial, 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. Frequency of the seizures was also divided into four groups: daily, weekly, monthly and occasional. In analyzing interictal surface EEG, baseline activity was assessed for the presence and topography of focal spikes and the presence of generalized paroxysmal discharges, and these abnormal EEG activities were further classified in detail.

On follow-up of these children, serial neurological evaluation and developmental testing were done and the children's intelligence quotients (IQs) were categorized into normal, slightly retarded, moderately retarded and severely retarded. Then behavioral and psychosocial aspects were divided into four groups as follows: normal, psychological impairments, hyperactivity and psychosis. School performance in children was also studied. Seventy-three percent of children demonstrated markedly enlarged ventricles. The median follow-up period after shunt insertion was 7.6 years. Valves were inserted at the site of maximal ventricular dilatation, mostly in the occipital horn of the lateral ventricle (92%), preferably on the right side (85%). The first shunt placement was performed before the children reached the age of 3 months in slightly more than 50% of cases, and in only 16.7% was the procedure done when they were over 1 year of age. Within the given study period, on average two operations per patient were performed. Shunt complications requiring surgical procedure were observed in 50% of the children, and 27% of those had one complication, while 22% had two or more than two complications. Approximately one-third of the children experienced a mechanical problem with the shunt, and the infection rate for the entire series was 14% per patient and 6.8% per surgery. Postoperative CT scans indicated normal-sized ventricles in 34%, slit ventricles in 30%, and some degree of ventricular enlargement in the remainder.

During the follow-up period seizures were recorded in 32% of these 802 children, and 28.6% of them experienced their first epileptic attacks before shunting; thus, in the remaining 71.4% seizures developed after shunt insertion. The paper emphasized the fact that a peak incidence of seizure occurrence was around the time of shunt insertion and the majority developed epileptic seizures in the days preceding shunt insertion. The authors stated that the seizures were triggered by the intracranial hypertension immediately before shunt placement. Although the overall incidence of seizures in these 802 children was 32%, the majority of epileptic patients had recurrent unprovoked seizures and were poorly controlled by antiepileptic drug regimens, having several seizures per week in most cases. With regard to seizure frequency a significant difference was noted between the patients who developed seizures before shunting and those who developed them after shunt placement. In the preshunt group the frequency was much lower than in the postshunt group. In the latter, 80% of the patients had recurrent seizures. Another significant difference in seizure characteristics was observed between the pre- and postshunt groups. Generalized epilepsy was seen in 49% of patients in the preshunt group, while the incidence of partial epilepsy was 50% in the postshunt seizure group. In approximately one-third of patients in both groups, seizures were poorly controlled by antiepileptic drug regimens, and some children suffered from daily seizures (42%) and from weekly fits (18%).

Prevalence of seizures differed significantly with the etiologies of hydrocephalus. Those with a meningomyelocele experienced a significantly low incidence (7%) compared with those born prematurely or with cerebral malformations (30%). Those with prenatal hydrocephalus demonstrated a prevalence of 38%, and those with meningitis and postinfective hydrocephalus carried a high risk of epilepsy, in the order of 50%.

Central nervous system malformations were predictive variables for seizure occurrence, and these include agenesis of corpus callosum, focal migration abnormalities, encephalocele, holoprosencephaly or other posterior fossa abnormalities, such as absent or hypoplastic cerebellum and cyst. And in the presence of abnormalities on CT scans the incidence of epilepsy was 45%.

Birth injuries were another significant risk factor for the development of seizures, and the incidence of epilep-

sy was elevated among children who suffered hemorrhage (41%), infection (47%), or anoxia (58%) during the neonatal period.

A higher incidence of epilepsy was demonstrated among children who had shunt malfunction, infection, or a combination of these, and the average number of shunt revisions was significantly higher in children with seizures than in those who did not develop seizures. Without revision the incidence of epilepsy was only 20%, while it was 52% in the patients in whom more than three revisions were needed. Although the majority of children who presented with shunt malfunction had evidence of intracranial hypertension, in only 8.6% of the whole series was the experience of seizures alone the presenting symptom of shunt dysfunction.

The results of EEG were analyzed, and the role of the ventricular catheter in epilepsy and its influence on the EEG were also discussed. Preshunt EEGs disclosed a 27% incidence of a single epileptic focus, but the corresponding figure after shunts were inserted was 50%. However, there was a direct relationship between the side of the radiological abnormalities and the side of the EEG focus. This makes it difficult to know whether it was these lesions or the brain injury related to the shunt that might be responsible for the EEG abnormalities. For further analysis of the above findings, a small group of patients was selected for evaluation in detail. In this group there were no abnormal findings on CT scan or MR imaging, and the right posterior parietal ventricular catheter remained in place even after shunt revision. In this selected group of patients bilateral abnormalities were found in 76% before shunt insertion but spike-and-wave activity was recognized in only 8% before shunting, whereas an epileptic focus appeared in 30% in the right hemisphere after a shunt tube was placed. In children without cerebral parenchymal lesions on CT scan the focal side of the interictal EEG abnormalities was identical to the area of shunt insertion. No valid statistical comparison with regard to the site of shunt insertion, either frontal or posterior, has been made.

Complete withdrawal of antiepileptic medication was achieved in only 35 of 255 hydrocephalic children.

Hydrocephalic children without epilepsy showed normal intelligence in 66% of cases, while only 24% had normal intelligence when hydrocephalus was associated with epilepsy, indicating a close correlation between epilepsy and poor cognitive outcome. Normal behavior was recognized in 80% of the children without seizures, but in only 32% of hydrocephalic children with associated epilepsy.

Interpretation

Without question, this paper gives one of the most well-documented evaluations of the correlation between hydrocephalus and epilepsy, particularly with reference to shunt placement. The authors conclude that epilepsy is associated with infantile hydrocephalus in one-fourth to one-third of cases and its onset most often occurs at approximately the same time as hydrocephalus is diagnosed. The incidence of epilepsy given in this report is an average figure that has appeared in previous reports, and what is stated in the paper is in accordance with the general concept that patients with hydrocephalus are at a higher risk of having or developing a seizure disorder than the general population. It is interesting to know that seizures were recorded in 32% of the 802 children, but 28.6% of them experienced their first epileptic attacks before shunting. while in the remaining 71.4% seizures developed after insertion, suggesting a strong influence of the shunting procedure on the development of epilepsy. The frequency of seizures is lower in the preshunt than in the postshunt group. Generalized seizures were common in the preshunt group and partial epilepsy was observed in 50% of the postshunt seizure group. A peak in seizure occurrence around the time of shunt insertion, shown in the authors' Fig. 1, is described for the first time. It is also of interest that increased intracranial pressure in association with shunt malfunction may predispose to seizure occurrence in hydrocephalic children. Perinatal insult to the brain, including hemorrhage, infection and anorexia, is mentioned as one of the risk factors for the development of seizure. In the children with no radiological abnormal findings spike-and-wave activity was recognized in only 8% before shunt placement, whereas after shunt insertion an epileptic focus appeared in 30% in the right hemisphere and in 39.4% in the posterior parietal area, localized to the general region of the shunt; however, no valid statistical comparison with regard to the site of the shunt insertion, either frontal or posterior, was made.

Hydrocephalus and epileptic seizures [15]

Presentation

To answer the question of whether there is an association between shunted hydrocephalus and the development of epileptic seizures, a study on 197 shunted hydrocephalic children was made. The chart review included the age at the time of first shunting operation, total number of shunting procedures done per patient, location of shunt, etiology of hydrocephalus, types of seizures, their frequency and types of hydrocephalus according to radiological imaging. Sixty percent of 197 patients had obstructive-type hydrocephalus. The mean age of the children at the first shunt insertion was 1.5 years. Sixty-eight percent of them had their first shunt surgery before the age of 6 months, and 76% had the first shunt operation before the age of 1 year. The shunt was placed in the

right parietal region in 66% of the children and on the left side in 16%. The rest of the children had either multiple shunt insertions or lumbo-peritoneal shunts. Meningomyelocele was the most common cause of hydrocephalus(45%), but in 36% no cause was identified. The mean age of patients at the time of review was 7 years, and 52% of patients were intellectually normal, but 9% had a significant learning disorder, 14% had borderline intellectual abilities, 10% had a mild degree of mental retardation, and 15% were moderately or severely retarded. Thirty-three out of 197 patients (17%) had had at least a single episode of epileptic seizure, but no seizure occurred at the time of acute shunt obstruction or at the time of shunt malfunction. Of those who developed seizures 37% had experienced only a single seizure. Among those who developed multiple seizures, 20% had a seizure only approximately once a year, but 30% had seizures more frequently than once per month. Partial seizures were observed in 43%, while generalized seizures occurred in 40% and the rest (17%) had multiple seizure types. The mean age at first seizure was 1.35 years, and 51% of patients who developed seizures had their first seizure during the 1st year of life. Only 10% had the first seizure after the age of 3.

The clinical features of the patients who had seizures are different from the patients who did not. Intellectual status and CNS abnormalities found on examination divided the patients into two groups. The group with seizures tends to be more severely mentally retarded and to have more abnormalities seen on imaging studies than the other group.

Interpretation

In this report 17% of patients with hydrocephalus developed various types of seizures. Even though the authors point out that the patients with hydrocephalus are at a greater risk of having or developing a seizure disorder than members of the general population, the reported figure appears rather low compared with those in other reports in general. Although many reports suggest some correlation between epilepsy and shunt malfunction, the authors found no tendency for the patients to have seizures at the time of acute shunt obstruction. Also, no correlation was noted between the number of shunt revisions and seizure occurrence. Nor was any influence of age at the initial shunt procedure on epilepsy in hydrocephalic patients observed in this study. Our attention is often focused on whether the site of shunt placement correlates to the incidence of epileptic seizures or not, but this report suggests no relationship between the two. The increased risk of seizures in patients with intellectual or motor impairment indicates that underlying brain abnormalities, particularly of the cerebral cortex, are of importance in the development of seizures, and this statement is in good accordance with a number of other reports in the literature.

Hydrocephalus and epilepsy: an actuarial analysis [20]

Presentation

The prevalence of epilepsy in patients with hydrocephalus and possible risk factors was studied. The authors of this paper aimed to describe an association of epilepsy with hydrocephalus, but because of a number of intrinsic limitations it was necessary to adopt a working definition of epilepsy that would be suitable for reviewing medical records, namely an illness requiring long-term antiepileptic drug (AED) treatment. The date of onset of epilepsy was taken as the date of the first seizure that led to AED treatment, but otherwise, when the date of the first seizure was not clear, the date of initiation of AED treatment was used. Also, the date of an abnormal electroencephalogram was substituted in a few cases. No reliable information on types or frequencies of seizures is given, as the subsequent analysis of these patients was carried out at yearly intervals over a 15-year period. It was also not possible consistently to determine whether or when AED treatment was discontinued, although prophylactic AED was not prescribed for patients with shunts at the authors' institution during the study period.

The starting point of the analysis was the date of the diagnosis of hydrocephalus, which was taken to be the date on which the first shunt was placed, and the endpoint was the initiation of AED therapy. A detailed statistical analysis was done.

CSF was diverted by shunts in 464 patients as treatment for hydrocephalus in this study, and median follow-up period after the diagnosis of hydrocephalus was 66 months. The actuarial rate of initiation of AED treatment was analyzed when presented as a function of time from the diagnosis of hydrocephalus. The prevalence of AED treatment at diagnosis of hydrocephalus was 12%, and by 10 years from diagnosis 33% of patients had been treated for epilepsy. No trend toward declining annual risk of development was observed. The initial prevalence of AED treatment was very similar in all age groups.

Patients who required more than four shunting operations needed slightly less AED treatment, but this finding was not statistically significant.

There were 339 patients who had exclusively frontal burr hole sites for shunting, and 71 had exclusively parietal burr holes. A trend toward slightly less AED treatment in the parietal burr hole group was seen, and this trend was accounted for entirely by the experiences of patients first treated for hydrocephalus as neonates. No such trend was apparent in patients who were older when hydrocephalus was diagnosed. Therefore, burr hole site

had no detectable effect on the prevalence of AED treatment. However, patients with meningomyelocele who had had CSF shunts inserted at both frontal and parietal burr hole sites experienced more AED treatment than patients who had had shunts inserted at only one of these sites.

Fifty patients were treated for one or more shunt infections during the follow-up period, but no more AED treatment was required in these patients than in those who did not have infectious episodes.

Posttraumatic and postmeningitic hydrocephalus showed the highest prevalence of epilepsy, while patients with meningomyelocele experienced significantly less AED treatment. It is stated that the prevalence of AED treatment was already established at the recorded time of diagnosis of hydrocephalus.

In conclusion, this analysis extending 15 years from the diagnosis of hydrocephalus indicates that there is no point beyond which hydrocephalic patients can be considered free of the risk of epilepsy. Surgical complications were only weakly associated with the risk of epilepsy, even when such an association was sought within etiologically homogeneous subgroups of the patients studied.

Interpretation

The prevalence of epilepsy in hydrocephalic patients is one of the major concerns for pediatric neurosurgeons, and this paper has made it clear up to a point whether or not the prevalence will increase with time after shunting. In view of the time-course after shunting this paper gives us unique information. The prevalence of AED treatment at diagnosis of hydrocephalus was 12%, but by 10 years after diagnosis 33% of patients had been treated for epilepsy and no trend toward diminishing annual risk was observed. There was no age difference at the initial prevalence of AED treatment.

It is not always easy to define the criteria of epilepsy and to determine the date of the first seizure unless all the essential records are readily available throughout the follow-up period. In this report the authors applied a totally practical method to define the starting and endpoints supplemented by an extensive statistical analysis. Burr hole site is often discussed in relation to seizure prevalence in the literature, but the site demonstrated no detectable effect on the prevalence of AED treatment in this study. Shunt infection did not require more AED treatment in this study, while examination of the etiologies of hydrocephalus had decisive results, with posttraumatic and postmeningitic hydrocephalus requiring the highest incidence of AED treatment and with meningomyelocele requiring less frequent use of AED. This finding is in agreement with most reports in the literature. As the authors conclude, it is interesting to note that there is no point during the follow-up period beyond which hydrocephalic patients can be considered free of the risk of epilepsy.

Late outcome of the surgical treatment of hydrocephalus [11]

Presentation

Long-term outcomes of hydrocephalic children treated by shunt surgery are reported, with particular reference to their psycho-intellectual development. As well as the psycho-intellectual outcome, mortality and morbidity, including motor deficits, visual or auditory deficits, and also the issue of epilepsy are reported and discussed. The study was conducted in 129 children, children who harbored tumors being excluded. Then a relationship between final outcome and epilepsy was studied, as were etiology and initial ventricular size. In these children the first shunt was inserted before the age of 2 years, but in 75% of them the shunt surgery was performed before they had reached the age of 6 months. The final neurological examination revealed some motor function deficit in 60% of the patients and epilepsy in 30%. One-third of the children who developed epilepsy was classified as intractable. The presence of epilepsy is an aggravating factor on psycho-intellectual outcome, and only 13% of the children with epilepsy had an IQ above 90while the final IQ was above 90 in 32% of the children. Furthermore, IQ was below 50 in 44% of those with epilepsy.

Behavioral disorder is often a determining factor in social integration and schooling, and the children who had completely normal schooling had a mean IQ of 97, but 40% of hydrocephalic children did not fit in anywhere within the normal school system.

Interpretation

The long-term outcome in hydrocephalic children treated by shunt placement is discussed with special reference to their psycho-intellectual development. The incidence of epilepsy was 32% in the children who had their first shunt implantation before they were 2 years old. The most important message of this report is that the presence of epilepsy is definitively an aggravating factor in psycho-intellectual development. When a hydrocephalic child develops epilepsy, his or her chance of achieving an IQ above 90 is only 13%, and the possibility of having a poor IQ below 50 is 44%. Also, epilepsy was frequently associated with severe behavioral problems. The degree of ventricular dilatation or intraventricular pressure in relation to the incidence of epilepsy and to their psycho-intellectual outcome was not described, and no correlation between shunt failure and development of

epilepsy was mentioned. Neither seizure types nor frequencies were described.

Epileptic seizures as a sign of cerebrospinal fluid shunt malfunction [14]

Presentation

A cohort of 817 shunted hydrocephalic children was reviewed and analyzed in detail with reference to seizure development. The study included 544 emergency room visits or encounters for seizures and 1,831 shunt revisions. It was found that 38% of the patients with shunts had seizures and there was a significant difference in seizure incidence among the diagnostic groups: the incidence was high in the groups in which the causes were abscess (66.2%), intracranial hemorrhage (52.9%), trauma (50.5%) and prematurity (41.1%), but was relatively low in the group with spina bifida (35.4%). The postshunt seizure incidence was 59% (181 out of 307 patients), while the first seizure occurred after shunting in 181 of 817(22%). Among the patients who developed their first seizure postoperatively, the trauma patients had a significantly lower incidence (20%) than the spina bifida (77%) and malignancy groups (76%). The abscess patient group had a lower incidence (42%) than the spina bifida patients (77%) did. The first seizure occurred at a median of 1.6 years after the original shunt insertion, and only 12% had their first seizure during the 1st week after shunt surgery. More proximal revisions were done in children with seizures versus children who had no seizures. Prevalence of postoperative seizures by the sites of ventricular access was not significantly different. Among 1,831 revisions only 16 (0.9%) were preceded by seizures, and only 2.6% of the shunted children who were brought to the emergency room because of a seizure were found to have shunt malfunction requiring shunt revision. The authors conclude that shunts do cause epilepsy, but the underlying abnormalities of the brain are more important in determining whether or not the child develops epilepsy. And shunt malfunction is rarely heralded by a seizure; consequently, a seizure is seldom an indication of shunt failure.

Interpretation

This is an excellent paper, which clearly shows what the possibility of shunt malfunction is when a shunted child is brought to an emergency room with an episode of seizure. Shunts do cause epilepsy, and the reported prevalence of epilepsy in shunted hydrocephalic children, at 38%, is in the range that has appeared in other reports.

Various incidences of epilepsy of different etiologies underlying the hydrocephalic process are described. It is generally agreed that hydrocephalic children with spina bifida have a low incidence of epilepsy; however, it is rather surprising to learn that among the patients who develop their first seizure postoperatively the children with spina bifida have the highest incidence of seizure development (77%) and the head-injured children, a low incidence (20%). No explanation is given for this curious result, nor is there any speculation on the subject. It is also of interest to know that more proximal revisions were done in children with seizures than in children who had no seizures, while the prevalence of postshunt seizures by the sites of ventricular access was not significantly different. The first seizure was observed at a median of 1.6 years after the original shunt insertion. But it is noteworthy that only 12% of children who developed seizures after shunting had their first seizure during the 1st postoperative week. Seizures occurred in 16 patients who consequently required shunt revision and can be classified into several types. Postictal EEGs was recorded and analyzed in detail in 6 of these children, but their findings varied, indicating no specific pattern. The issue of revisions complicated by shunt infection in relation to the occurrence of epilepsy is actually one of our major concerns, but it is not mentioned in this paper.

Epilepsy in shunt-treated hydrocephalus [16]

Presentation

One hundred eighty-two patients who had undergone initial shunting at the authors' institution were followed up for at least 1 year (range 1–21 years). The children's average age at follow-up was 7.2 years, and they were an average of 10 years old at the time of the retrospective evaluation of the data. Data analyzed included:

1 Etiology of hydrocephalus
2 Onset and clinical presentation of epilepsy
3 Functional status such as motor deficit, mental retardation and cerebral palsy
4 EEG findings
5 Sex, shunt revisions, shunt location and burr hole sites

No patients received prophylactic anticonvulsant treatment after shunting. In 83% of the patients proximal shunt tubing was placed on the right side, and 50% of the study patients had multiple shunt revisions. Epilepsy was diagnosed in 20% (37 out of 182) of the children, and 14% (25 out of 182) had symptomatic seizures according to the authors' definition of symptomatic seizures: seizures attributable to raised intracranial pressure, postoperative edema, or shunt complications and

seizures that should not be termed epilepsy. Twenty-one patients had symptomatic seizures only, and were classified as the nonepilepsy group; consequently 4 patients were classified as the epilepsy group.

In general, patients with epilepsy had shunting surgery much earlier (mean age of 6 months) than the patients without epilepsy (mean age of 31 months), regardless of the etiology. The incidence of epilepsy was high in the patients with posthemorrhagic and postinfectious hydrocephalus, and none of the patients with tumors had epilepsy. Motor function disability, mental retardation and cerebral palsy were more common in the patients with epilepsy than in those without.

Fifteen patients out of 182 developed epilepsy before shunt insertion, and 22 developed epilepsy after shunting; the peak for the onset of seizures was within 6 months following shunt placement. The patients who had multiple shunt revisions failed to show any increase in the prevalence of epilepsy, and burr hole sites made no difference, although focal EEG abnormalities at the site of shunt insertion were recorded in some patients. Among the patients with epilepsy EEG recordings were normal in 14%.

Interpretation

The stated prevalence of epilepsy in hydrocephalic children is 20%, and this seems a little lower than those that have appeared in the literature. Their patients were followed up for from 1 year to 21 years after shunt placement, so that the follow-up period might not have been long enough in some patients. The fact is that some of the patients would develop seizures as time passed, as stated in the literature.

The authors separate symptomatic seizures, classifying patients thus affected as the nonepileptic group, from the epilepsy group. However, it is not very clear why the authors differentiate patients with symptomatic seizures from the epilepsy group, and we just wonder whether their criterion of symptomatic seizures is generally accepted. At any rate this paper should be read with these issues in mind.

It is reported that the patients with epilepsy underwent shunt surgery earlier, at a mean age of 6 months as against 31 months in the patients without epilepsy, regardless of the etiology of hydrocephalus. A mean age of 31 months in the latter group seems late to implant shunts in hydrocephalic children in general, but no specific description of these patients is given. In this study no children with hydrocephalus caused by tumors developed seizures, which is a point of difference from other reports in the literature, but otherwise the prevalence of seizures in hydrocephalus of different etiologies is in accordance with the reports of other authors.

Pattern of electroencephalograph abnormalities in children with hydrocephalus. A study of 68 children [1]

Presentation

Sixty-eight patients (age range 1 month to 17 years) with hydrocephalus had standard EEG recordings, and 48 of them had their EEG recordings exclusively after ventriculo-peritoneal shunting. Only 8.3% of traces were normal, giving an overall abnormality of 92%.

The abnormalities recorded in this study were predominantly slow-wave asynchronous generalized discharges and epileptiform generalized and focal activities. The frequency of these abnormalities is similar in the shunted (92%) and unshunted (90%) patients. In the unshunted group generalized asynchronous slow-wave activity was found in 60%, generalized epileptiform activity in 30% and focal epileptiform activity in 15%, and only 2 out of 20 patients had normal recordings. In the shunted group asynchronous slow waves were observed in 45.8%, generalized epileptiform activity in 31.3%, and focal epileptiform activity was seen in 22.9%, while 4 out of 48 demonstrated normal recordings. The frequency of epileptiform activity was higher (54%) in the shunted than in the unshunted group (46%).

Only 4 out of 68 patients had clinical seizures. The authors conclude that hydrocephalus in children may be associated with generalized or focal EEG abnormalities regardless of the cause and that this may reflect the heterogeneity of the neural generator in the underlying disease process.

Interpretation

The incidence of clinical seizure in this report is 5.9%, while no follow-up period is mentioned. If these patients were not followed up for long enough relative to the occurrence of epilepsy, this might explain this extremely low incidence of seizures. Although the present study showed the frequency of various EEG abnormalities was similar in the shunted and unshunted groups, the frequency of epileptiform activity was higher in the shunted than in the unshunted children. The number of patients studied was rather small and the significance of the results was not statistically analyzed, but it is interesting to know that epileptiform activity was slightly more common in the shunted than in the unshunted group, assuming that the epileptic activities could be related to surgical procedures in the patients with shunts.

Continuous spikes and waves during sleep in children with shunted hydrocephalus [25]

Presentation

Twenty children affected by hydrocephalus of different etiologies were studied with periodic EEG polygraph recordings while awake and during their afternoon sleep. All of them underwent ventriculo-peritoneal shunt placement during the 1st year of life, but EEG recordings were made at a mean age of 5 (range 1–16) years. The patients were divided into two subgroups: those who showed the presence of continuous spikes and waves during slow sleep (CSWS) and those who did not, where CSWS was taken to mean the presence of continuous spikes and waves during at least 85% of slow sleep. Those in whom CSWS was present during the afternoon sleep were also submitted to nocturnal or to 24-h EEG recording. Neuroradiological examination by MRI was done in all, with special attention paid to the following findings:

1 Cortical atrophy
2 Ventriculomegaly
3 Alteration of the white matter

In addition, intelligence testing (WISC-R) was performed in some patients. Partial seizures were observed in 65% of the patients, and 80% of these developed partial motor seizures. The average age at which seizures first appeared was 2 years and 9 months. EEG recordings while awake revealed focal EEG abnormalities in all the patients, and the focal abnormal activities were on the same side as the shunt in 95% of cases. These abnormalities consisted of spikes followed by slow waves, and they had amplitudes of 300 V or more in 65% of cases. In all patients in the CSWS group, for an average of 18 months there were abnormalities consisting of spikes followed by slow waves with an amplitude greater than 300 V and secondary bilateral synchrony. However, in the group without CSWS wide-amplitude abnormalities were found in only 50% of the subjects and no bilateral synchrony was found in any of them. All the patients in the CSWC group developed epileptic seizures, and most of them (85%) had partial motor seizures, while partial secondary generalized seizures were seen in only 17%. In the group without CSWS only 50% of the patients developed seizures and partial motor seizures were found in 57%, in contrast to the 85% in the patients with CSWS. Furthermore, in the patients with CSWS all three neuroradiological abnormalities mentioned previously were present in 50%, and cortical atrophy and ventriculomegaly were present in all of them. In contrast, in the group without CSWS none presented all three abnormalities all together.

The authors also state that they have demonstrated that the intellectual capability of all the subjects deteriorated in the presence of CSWS abnormalities.

Interpretation

Although the number of patients studied is limited, this paper is extremely informative on EEG findings seen in shunted hydrocephalus patients. Their method of EEG recording is truly extensive and conscientious in many ways. The patients were divided into two groups: with CSWS and without CSWS. Then a unique contrast was clearly presented between these two groups. In the group with CSWS the incidence of seizure was 100%, and 85% of the subjects in this group developed partial motor seizure, but in contrast the incidence of epilepsy was only 50% in the non-CSWS group and the incidence of partial motor seizures was significantly lower (57%) than in the CSWS group. Furthermore, EEG abnormalities observed included high amplitude of 300 V or more in the group with CSWS without exception, but such abnormal high amplitudes were seen in only in 50% of patients in the non-CSWS group. From the results described in this report it seems highly possible that a patient would definitely develop seizures after shunting when his or her EEG recording revealed CSWS and an abnormally high amplitude. As claimed by the authors, the morphological characteristics of the epileptiform abnormalities associated with wide amplitude have never been reported in the literature. Likewise the possible relationship between CSWS and hydrocephalus has also never been mentioned before. The correlation between EEG abnormalities and morphological changes seen on imaging is also of great interest. The finding that focal EEG abnormalities in awake children were on the same side as the shunt in 95% of cases is striking and highly suggestive: it seems likely that shunt implantation can be an extraordinary factor in the causation of EEG abnormalities and ultimately in the development of seizures. These results indicate that EEG recording during sleep can show that CSWS is the real cause of the deterioration in those children, and particularly in subjects who present abnormalities while awake and show a tendency to develop secondary bilateral synchrony. This paper reminds us of and impresses upon us the important role conscientious EEG recordings can have in the investigation of hydrocephalic children, though in fact EEG recordings are often disregarded or are no longer mandatory in many institutions.

Discussion

Review of these papers has made it clear that the issue of epilepsy in hydrocephalus, especially in shunted children, should have received more attention by all neurosurgeons. Many factors are implicated, which is what makes the problem so complicated. A comprehensible point is an association of CNS malformations with hydrocephalus in terms of incidence of epilepsy. It is agreed in the papers reviewed [3, 11, 15, 16, 20] and

many others [17, 24, 23] that there is an increased risk of seizures in patients with CNS malformations, while only Copeland et al. [6] were not able to convince their readers that there was a correlation between pathologic conditions causing hydrocephalus and the incidence of seizures. Mental retardation is another factor that aggravates the risk of having or developing seizures. Mental retardation was diagnosed four times as frequently in children with seizures [18]. Children with IQ scores of 70 or below were more likely to experience seizures than were children with normal intelligence [22], and furthermore in children with meningomyelocele mental retardation was statistically more common when associated with seizures, regardless of the presence or absence of hydrocephalus [17].

EEG findings and seizure types observed in hydrocephalus with shunt and without shunt were documented in the reviewed papers [1, 16], and additional findings have been well described in the literature [4, 7, 8, 13, 21]. Graebner and Celesia [9] stated that 62% of subjects in the nonshunted group and 85% in the shunted group had abnormal EEGs, and focal specific paroxysmal discharges were present in 38% in the shunted group, in contrast to only 10% in the nonshunted group. Comparing EEG recordings taken before and after shunt surgery, Varfis et al. [24] supported the view that an epileptogenic focus has developed around the place of ventricular catheter of the shunt system. Among shunted hydrocephalic patients after meningitis or intraventricular hemorrhage fits other than generalized convulsions were rare, while generalized and focal fits were equally represented in the hydrocephalic patients after Chiari malformation or aqueduct stances [22].

Although prophylactic use of AEDs is not routine on the whole, the administration of AEDs is still a controversial matter. At the time of initial CSF shunt surgery 12% of patients had already been treated with AEDs, and the hazard rate for initiation of AEDs was a constant 2% a year; in consequence the estimated prevalence of AED treatment had risen to 33% by 10 years after initial shunt insertion [20]. Unfortunately the prognosis for seizure control was not good in general [3]. No real difference was found in the severity of CNS abnormalities between those children controlled by medication and those with uncontrollable seizures [18]. EEG results at the time of the first seizure, findings on CT scan, and number of seizures before AED treatment did not correlate with seizure remission when medication was withdrawn [18]. In the same communication it was stated that 39.4% of the children with convulsions had poorly controlled seizures when the study group excluded the patients associated with meningomyelocele. However, the prognosis of the seizures is somewhat different from that reported by others in children with hydrocephalus after meningomyelocele, and in these subjects three-quarters had antiepileptic medication discontinued without recurrence of seizures and some others were well controlled with medication [17]. The debate continues on prophylactic use of AEDs, Dan and Wade [7] remaining unconvinced while Copeland et al. [6] conclude that prophylactic use of AEDs is probably indicated for a minimum of 1 year.

It is stated in many reports that the prevalence of seizures is significantly higher in the shunted than in the nonshunted group [4, 5, 13, 22, 24]. Even the presence of a shunt catheter could itself promote an epileptogenic focus [3]. Although the subjects are limited to the patients with meningomyelocele, it is striking that only 2% of nonshunted patients experienced seizures in spite of a high incidence of ventriculomegaly while the incidence was 22% in shunted patients [5]. In the same way the prevalence of seizures was significantly higher in the shunted (48%) than in the nonshunted group (20%), and the results were in good agreement with the EEG findings [9].

However, some other authors were not convinced of a correlation between shunt insertion and the risk of seizures. Postoperative onset of epilepsy, focal seizures on the contralateral side, and EEG changes ipsilateral to the localization of the shunt did not appear to be valid criteria allowing the identification of patients who will suffer from shunt-related epilepsy [16], and emphasizing the role played by underlying brain anomalies, there is support for the hypothesis that the occurrence of seizure in children with hydrocephalus is not related to the treatment of hydrocephalus [15].

Correlation between subsequent risk of seizure occurrence and location of shunt placement or burr hole site is another concern of ours. Of patients with a parietal ventricular catheterization site only 6.6% had convulsions, in contrast to 54.5% of those with a frontal site [7].

With regard to the relationship between shunt location and possible seizure initiation, Buchhalter and Dichter [4] support the view that the overwhelming majority of focal interictal EEG abnormalities in patients are lateralized to the side of shunt placement and that frontal shunt placement is associated with a significantly higher incidence of seizures than a posterior location. There was a higher incidence of abnormal EEG traces in the shunt group (95%) than in the nonshunted group (47%) and particularly slow wave foci were seen more frequently over the hemisphere which had the shunt, the foci localized to the neighborhood of the shunt itself in more than half [13]. Moreover, the epileptogenic focus always located where the catheter was first inserted, and this was not present before the diversion surgery [24]. In addition, focal specific paroxysmal discharges and slow waves were recorded most frequently over the shunted hemisphere, and the report concludes with the statement that the shunt has irritating effects on the cerebral cortex and is responsible for the findings mentioned above [9]. When studied only in the children with no evidence of cerebral parenchymal lesions on CT scanning, the focal

side of the interictal EEG abnormalities was identical to the area of shunt insertion in most cases, and the discharges of slow or spike waves started in the right parietal area where the valve was inserted and involved the anterior region [3]. In contrast, in a number of reports in the literature it is not acknowledged that there is any correlation between the site of shunt placement and seizure occurrence or EEG abnormalities. It is concluded by Piatt and Carlson [20] that burr hole site is an independent risk factor for the development of epilepsy in patients with hydrocephalus. According to the paper by Klepper et al. [16], ipsilateral changes in EEG did not result in contralateral seizure activity in most patients, although focal EEG changes correlated with the anatomical site of the shunting and this was interpreted with some reservations, as the incidence of abnormal EEG recordings in nonshunted children was already high. Burr hole location, either frontal or occipital, did not make any significant difference in seizure prevalence [16]. No statistical difference between frontal and parietal entry points was revealed with respect to the incidence of seizures by location of the shunt [5]. The number of children studied was very small, but whether or not a seizure was focal was not statistically likely to be related to the shunt site according to the report by Stellman et al. [22]. Although the subjects were limited exclusively to patients with meningomyelocele, the location of the shunt catheter was not predictive of seizures [17].

On the other hand, the implication of shunt malfunction or infection is one of the major concerns regarding the prevalence of seizures in shunted children with hydrocephalus. The average number of shunt revisions was significantly greater in children with seizures than in those who did not have seizures, and infection itself or a combination of infection and malfunction increased the risk of seizures [3]. The need for more frequent valve revisions appeared to increase that tendency [12]. And there is very persuasive evidence to suggest a significant correlation between the frequency of revisions and the occurrence of seizures, as only 5.9% of those who did not have a catheter revision developed epilepsy, compared with 24.2% of those with two or more proximal side revisions [7]. Chadduck and Adametz [5] described increased risk of seizures (22%) in the patients who had required subsequent shunt malfunction in comparison with the incidence(9%) in the patients who had not. Furthermore, a large number of seizure incidents was shown to have a specific precipitant, usually shunt infection or shunt-related problems [22]. According to Blaauw [2], infection appeared to be closely connected with late epilepsy.

Therefore, the onset of a first-time seizure, or a new seizure in a shunted patient should alert one to shunt obstruction.

However, some reports are not in agreement with the stated increased risk of seizures with associated shunt malfunction and/or infection. Although evidence of increased ICP caused by shunt obstruction has been documented in association with seizures, 9% of the patients with meningomyelocele had a seizure in the days preceding the diagnosis and treatment of shunt malfunction, but in none of these episodes was seizure the sole presenting symptom of shunt malfunction [10]. No correlation was found between the occurrence of epileptic seizures and a shunt malfunction, and none of the patients in this study had seizures attributable to acute obstruction of CSF or the number of shunt revisions [15]. When 50 patients out of 464 were treated for one or more shunt infections there was no tendency to a need for more AED treatment than in the patients who had no infectious episodes [20]. The risk of epilepsy was not influenced by the number of shunt revisions [16].

The age at initial shunt insertion seemed to influence the occurrence of the epileptogenic scar, and the younger the patient at the date of surgery, the greater the chance of epileptogenic scar's developing, usually during the 2nd year after surgery, but the delay period before the onset of clinical seizures was variable [24]. However, in children with meningomyelocele, age at the time of shunt insertion was not predictive of seizures and none of these children have had recurrent seizures in follow-up periods up ranging from 10 to 16 years [17]. Copeland et al. [6] also stated that there was no correlation between the incidence of seizures and the age of the patients. Likewise, mental retardation was the only independent predictor of seizure occurrence and the age at the time of shunt insertion had no correlation [18]. Epilepsy that occurred within 1 week after shunt insertion was called early epilepsy by Blaauw [2], and 40% of seizures fell in this category.

The mean age at the time of the first seizure was 1.35 years, and 51% of those who were going to have seizures had their first during the 1st year of life, 75% before the age of 2 years, and 90% by the age of 3 years [15]. Eight percent of the patients studied by Klepper et al. [16] developed epilepsy before shunt implantation, and 12% of them, after shunting; onset of epilepsy peaked within 6 months of the shunting procedure. On the other hand, none of the study group had experienced seizures prior to shunt placement and the risk of seizures fell from 5% in the 1st year after shunting to 1.1% after the 3rd year [7]. With respect to the time to onset of epilepsy the results varied from 24 h to 3 years, but 58% of those who developed seizures had their first seizure within the 1st month following shunt surgery [6].

Of those who developed epileptic attacks after shunt surgery, 28.6% experienced their first attack before shunt surgery and the remaining 71.4% had their first attack after shunting; there was a peak incidence of seizure occurrence around the time of shunt insertion [3].

Cerebrospinal fluid or shunt infection has a specific precipitant role in the development of seizures, and inci-

dence of fits in the postmeningitis group was 54%, a figure was significantly higher than the 27% in primary group [12]. The increased incidence of postshunt epilepsy was 46% when complicated with infection, compared with 20% in patients without infection [6]. The prevalence of seizures in the patients who required shunt implantation without infectious complications was 22%, but the likelihood of developing seizures increased to 47% with a history of shunt infection [5]. Bourgeois et al. [3] also agreed upon this tendency.

Birth injury or perinatal insult may play a significant role in development of seizures in hydrocephalic children [3], and this finding has never been reported previously.

In most of the children who suffered from hydrocephalus, the context of epilepsy and shunt surgery is complex. Although it became apparent that uncertainty persists and many issues remain controversial, seizures are more frequently reported when hydrocephalus is associated with CNS anomalies and mental retardation. In general, number of revisions, shunt malfunction and infection seem to be aggravating factors in the development of seizures. Likewise, age at the time of the initial shunt insertion appears to correlate with the risk of seizure occurrence, and the younger the child is, the greater the risk. Burr hole site seems to influence seizure development in hydrocephalic children, as seen particularly when papers on EEG recordings are reviewed.

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