Epilepsy as a Late Complication

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Contents

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18.1 Introduction

Hydrocephalus has an incidence ratio of 1–3 cases/1,000 children in the population [20, 27], while epilepsy has an incidence ratio of 4–9 cases/1,000 children in the population [27]. Nowadays, surgery is the treatment of choice for congenital or acquired hydrocephalus and

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occurrence of epilepsy after shunting procedures is a well-known problem, but our knowledge concerning its mechanism is very limited [11, 16, 20, 22, 23, 27, 34, 36, 38, 42, 43, 47].

In the present chapter, we will review the relationship between hydrocephalus and epilepsy as a complication of shunt placement in detail to provide a useful information for the families of the patients, as well as neurosurgeons and neurologists, about the outcome of postoperative hydrocephalic patients.

18.2 Incidence of Epilepsy in Patients with Hydrocephalus

It is now widely accepted that epilepsy is frequently seen in children with hydrocephalus, although its mechanism is not clear [3]. In the current literature, there are conflicting reports as to the prevalence of epilepsy in hydrocephalus, ranging from 9 to 65 % [11, 16, 20, 22, 23, 27, 34, 36, 38, 42, 43, 47].

Based on results of a retrospective analysis of a total of 200 children with hydrocephalus, Hosking [22] reported that seizures developed in 30 % of the cases during a follow-up period of 5-year. Afterwards, Blaauw [4] published a retrospective review of 323 hydrocephalic children and he found that epileptic seizures were developed in 34 % of the patients with hydrocephalus caused by various etiologies including hemorrhage and infections. Also, Leggate et al. [30] found that seizures developed in 16 % of 56 hydrocephalic patients. In similar, Saukkonen and von Wendt [42] also reported that epileptic seizures developed in 80 of 168 patients (48 %) during the follow-up period of about 9 years. In 1992, an incidence of epileptic seizures as 49 % of the patients was reported in a series of 68 patients with congenital hydrocephalus [36]. In a retrospective review of 464 patients with hydrocephalus, Piatt and Carlson [38] reported that 12 % of patients had epilepsy at the time of diagnosis of hydrocephalus (Fig. 18.1).

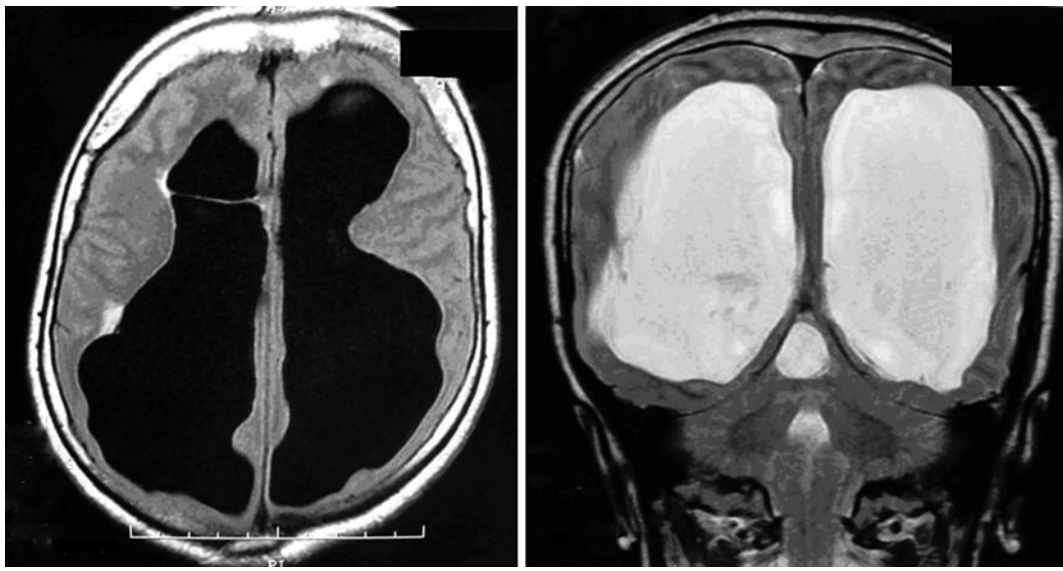

Fig. 18.1 MR scans of a 28-year-old woman with a history of learning difficulties at childhood and recent episodic diffuse headache that had lasted for almost 3 months as well as additional generalized seizure 1 month prior to admission revealed a chronic (no active) hydrocephalus. The patient was treated conservatively by antiepileptic drug (valproate) without any ventriculoperitoneal shunt insertion and had a good outcome during 15 months follow-up (Courtesy of Ali Akhaddar MD, Rabat, Morocco)

18.3 Influence of Etiology of Hydrocephalus Upon Epilepsy

In the current literature, there is general agreement that the etiology of hydrocephalus may play a critical role in the development of epilepsy, albeit the results are conflicting [4, 22, 24, 36, 38]. Piatt and Carlson [38] suggested that the cause of the hydrocephalus was correlated with the risk of development of epilepsy. Etiological categories of hydrocephalus are summarized as follows: hemorrhage, infection, intracranial tumor, myelomeningocele, other congenital malformations such as aqueduct stenosis, arachnoidal cyst, Dandy–Walker malformation, and idiopathic [20] (Fig. 18.2).

The highest incidence of epilepsy was found in cases who had posthemorrhagic and postinfectious hydrocephalus, etiologies known to be associated with complex brain pathology and low functional status. In 1974, Hosking [22] reported that hydrocephalus was developed secondary to either neonatal intracranial hemorrhage or meningitis. Afterwards, Blaauw [4] found that seizures associated with hydrocephalus were more frequent in the patients with shunt infections.

Notably, hydrocephalus associated with various congenital anomalies including myelomeningocele or arachnoidal cysts carried a far higher incidence of epilepsy [28]. In 1978, Lorber et al. [34] reported that 49 % of the hydrocephalic patients associated with a morphological lesion of the central nervous system (CNS) had epilepsy and they suggested that epileptic seizures were frequently seen in patients with physical or mental disabilities. Then, Noetzel and Blake [36] found that, in a long-term follow-up on 68 hydrocephalic patients, mental retardation and malformations of the CNS correlated with seizure occurrence. Also, Keene and Ventureyra [27] reported that the risk of seizures in hydrocephalic children associated with motor or intellectual impairment was increased due to underlying brain abnormalities.

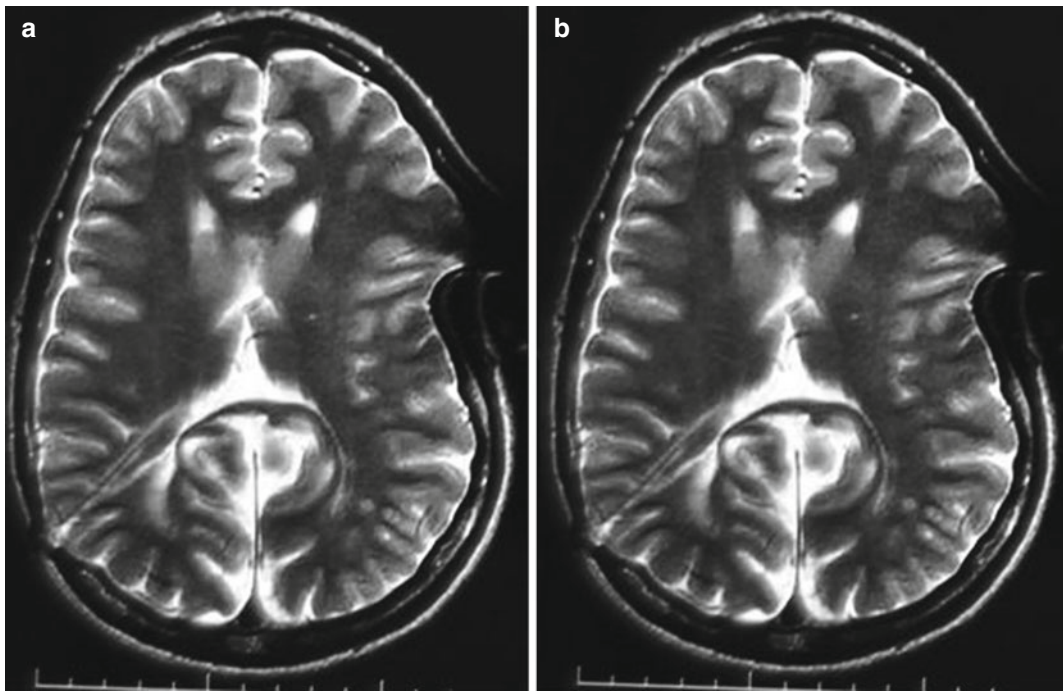

Fig. 18.2 (**a**, **b**) MR scans of a 19-year-old young lady with bilateral shunts and partial agenesis of corpus callosum with recent memory changes, decreased school performance, and automatisms. EEG identified temporal lobe epilepsy (Courtesy of Jogi V. Pattisapu, MD, Orlando, FL)

In a series of ten hydrocephalic children with tuberous sclerosis and intraventricular subependymal giant cell astrocytomas, Di Rocco et al. [14] reported that seven patients underwent direct surgical excision of the lesion, but the remaining three patients underwent a ventriculoperitoneal (VP) shunting and then removal of the intraventricular tumor. In addition, Di Rocco et al. [14] found that the surgical tumor removal was followed by a significant improvement in the epilepsy and they concluded that the surgical removal of the intraventricular tumors is the most appropriate treatment in patients with tuberous sclerosis and associated hydrocephalus.

Interestingly, as a cause of neonatal epilepsy with hydrocephalus, β-mannosidosis, which results from a deficiency of β-mannosidase, is an extremely rare disorder in humans [6]. Broomfield et al. [6] suggested that it should be considered in the differential diagnosis of neonatal seizures and subsequent hydrocephalus during follow-up, whereas others reported that there was no association between occurrence of epileptic seizures and the underlying etiology of the hydrocephalus [12, 27, 42]. In these patients, clinical findings such as altered skull morphology and intractable seizures develop in the neonatal period.

18.4 Influence of Intracranial Shunting Procedure Upon Development of Epilepsy in Hydrocephalic Patients

In neurosurgery, various shunting techniques known as ventriculo-atrial (VA) and VP are the standard treatment for hydrocephalus in both children and adults. Some authors reported that patients undergoing shunt surgery are at high risk of developing epilepsy as a surgical complication, but relation of the hydrocephalus and the shunting operation with the development of epilepsy is still controversial [28, 33, 40].

Today, it is accepted that there is an increased incidence of epilepsy risk after placement of the ventricular catheter, ranging from 5 to 58 % [5, 10, 12, 23, 24, 27, 28, 43]. To date, many authors reported large clinical series of seizure disorder following intracranial shunt insertion for hydrocephalus. In 1986, Stellman et al. [43] studied a total of 202 shunted hydrocephalic children, congenital or acquired origin, and they found an incidence of seizure disorder as 39 %. Of the 207 shunted hydrocephalic patients reported by Dan and Wade [12], 9.4 % had epilepsy. Besides, Johnson et al. [24] found that 38 % of the 817 children with shunted hydrocephalus had epilepsy. In a review of 182 patients, Klepper et al. [28] reported that shunt-related epilepsy was developed in 12 % patients. In a retrospective review of 197 patients with shunted hydrocephalus, Keene and Ventureyra [27] found that 17 % of hydrocephalic patients developed seizures.

Several authors investigated the role of shunting procedure upon the development of epilepsy in hydrocephalic patients. In a review of 92 patients with hydrocephalus, Ines and Markand [23] found that the incidence of epilepsy was high in the shunted group (65 % in the shunted group, while 18 % in the nonshunted group). Retrospectively, Venes and Deuser [47] found that 24 patients of 93 patients with hydrocephalus had epileptic seizures before the shunting procedure, but epilepsy developed following the procedure in only 5 patients. Afterwards, in a study of 168 shunt-treated hydrocephalic children, Saukkonen et al. [42] found that 48 % of the patients had epileptic seizures: 22 % of patients had epilepsy prior to the shunting procedure, and 26 % had epilepsy following the shunting procedure. Moreover, Klepper et al. [28] found that 37 (20 %) of the 182 patients developed epilepsy, 15 patients (8 %) before shunt insertion, and 22 patients (12 %) after intracranial shunting.

From an etiologic point of view, some authors investigated the effect of hydrocephalus upon the development of epilepsy in shunt-treated patients. In a retrospective study of 315 shunted hydrocephalic children, Lorber et al. [34] found that only 4 hydrocephalic patients with congenital etiology had seizures before the shunt placement, while seizures were related to the shunt device in 15 patients. Then, Klepper et al. [28] reported that epilepsy developed in 37 (20 %) of 182 patients with shunt insertion for hydrocephalus due to various etiologies including posthemorrhagic (5 %), postinfectious

(4 %), myelomeningocele (2 %), and aqueduct stenosis (0 %). In a retrospective study of 802 children with hydrocephalus, Bourgeois et al. [5] reported that 32 % of the patients had epileptic seizure, possibly owing to such episodes of raised ICP or the presence of a shunt device as an epileptogenic focus. Further, Kao et al. [26] found that postmeningitis hydrocephalic patients showed the highest incidence of epilepsy as 40 %, possibly due to its high shunt revision rate.

In clinical practice, the findings confirming the effect of shunting in development of epilepsy are: (a) the development of epilepsy following surgery; (b) focal discharges at the site of the shunt in electroencephalography (EEG); and (c) the existence of contralateral seizures [10, 23, 42, 47]. Besides, Ines and Markand [23] reported that all of the shunted patients who had epilepsy developed them after the shunting procedure and left-sided focal epilepsy was the most frequent focal motor seizures in the patients with shunt placement on the right side [23]. Based on their observation upon seizures involving the body side contralateral to the shunt placement, Copeland et al. [10] noted that development of seizures was possibly due to the surgical shunting procedure.

18.5 Causative Factors for Epilepsy in Patients Who Underwent Shunting Procedure for Hydrocephalus

Especially in shunted hydrocephalic children, it is commonly recognized that epileptic seizures occur as a result of shunting procedures, surgical complications due to these procedures, or the hydrocephalus itself. In this section, we will focus on increased risk of epilepsy following placement of shunt device, possibly related with the sex of patients and the age of patient at time of shunt placement, number of shunt revision procedures, shunt location (frontal, parietal) and shunt systems used, shunt malfunction, shunt infection, slit ventricle syndrome (SLVS), cortical malformation, intracranial hemorrhage, hyponatremia due to abdominal pseudocyst, episodes of raised intracranial pressure (ICP), and intracranial hypotension related with body posture in detail (Table 18.1).

Table 18.1 Causative factors for epilepsy in shunted hydrocephalic patients

Cause of epilepsy
Gender and age of patient
Number of shunt revision
Shunt location and shunt systems used
Shunt malfunction
Shunt infection
Slit ventricle syndrome
Cortical malformation
Intracranial malformation
Hyponatremia due to abdominal pseudocyst
Episodes of raised intracranial pressure
Intracranial hypotension related with body posture

18.5.1 Sex and Age of Patient at Time of Shunt Surgery

Studies suggested that there was no link between gender of the patients and occurrence of epilepsy in hydrocephalic patients [28]. On the other hand, there is now compelling evidence that age of the patient at the time of shunting procedure may be an important factor. It has been shown that children younger than 2 years of age have a high risk of epilepsy in contrast to older ones, possibly due to an increased risk of shunt malfunction [40]. As expected, early shunting as a well-known determinant of risk in cases with shunt obstruction was associated with a higher risk for epilepsy [10, 28, 40]. Accordingly, Dan and Wade [12] also found that postshunt seizures developed in 9 % of 207 patients with ventricular shunts, ranging from 15 % in infants to 7 % in patients over 50 years of age. Based on the results of their retrospective series, Noetzel and Blake [36] noted that risk factors for development of epileptic seizure in patients with shunted hydrocephalus included age at time of shunting. However, there was no correlation between the occurrence of epileptic seizures and the age of the patient at the time of initial shunt procedure [27, 38].

18.5.2 Number of Shunt Revisions

In a previous study, it has been reported that epilepsy developed in 24 % patients with shunt revision, in contrast to 6 % patients without shunt revision [12]. According to the results of a retrospective study, Noetzel and Blake [36] noted that risk factors for development of epilepsy in patients with shunted hydrocephalus included the total number of shunt revisions. Then, Johnson et al. [24] reported that a shunt revision was done in 3 % of admissions to the emergency unit of the hospital for epilepsy, and 1 % of shunt revisions was complicated with epilepsy.

In the existence of multiple shunt revisions, epilepsy is more common owing to traumatic injury to the brain tissue during the intracranial shunting for hydrocephalus [4, 10, 12, 22, 23, 24, 34, 36, 38, 40, 42, 43, 47]. Importantly, Heinsbergen et al. [20] found that patients with more than two shunt revisions have a high incidence of epileptic seizure. Especially in patients with postmeningitis hydrocephalus, higher shunt revision rates were reported compared with those due to other etiological types of hydrocephalus [26].

Nevertheless, others suggested that there was no correlation between risk of development of epileptic seizures and the number of shunt revisions [27, 28, 38, 42]. In a review of 168 shunt-treated children for hydrocephalus, Saukkonen et al. [42] found that there was no correlation between epileptic seizure and number of shunt revisions. They agree that multiple shunt revisions had no influence on the incidence of epilepsy and thus the total number of shunt revisions did not differ between the epileptic and nonepileptic groups [27, 28, 38, 42].

18.5.3 Shunt Location and Shunt Systems Used

Numerous studies have underlined that anatomic location of shunt insertion is important for the development of epilepsy [10, 12, 22, 23, 34, 36, 38, 40, 42, 43, 47]. In 1986, Dan and Wade [12] reported that 6 % of 168 cases who had shunt placement in the parietal region had epilepsy, in contrast to 55 % of 11 patients who had undergone shunting procedure in the frontal region. Nevertheless, others found that the location of the burr hole for the shunt insertion and shunt device, frontal and parietal areas, did not correlate with the occurrence of focal or generalized seizures [24, 27, 43, 47].

Further studies investigating the role of the shunt systems as a foreign body upon development of epilepsy revealed that there is no difference between the epileptic and nonepileptic groups [28, 40].

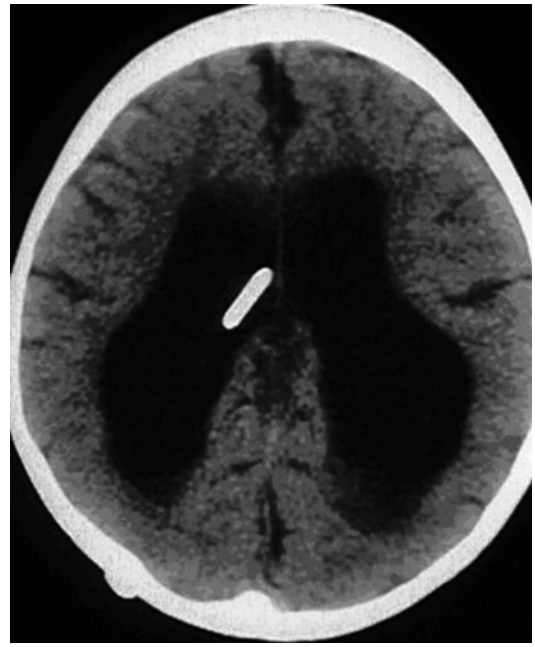

Fig. 18.3 CT scan of a 4-year-old girl with acute symptoms of headaches and generalized seizure presenting with enlarged ventricles due to shunt malfunction. The last surgical procedure was the original shunt insertion which was performed 3 years earlier (Courtesy of Jogi V. Pattisapu, MD, Orlando, FL)

18.5.4 Shunt Malfunction

It is logical to suggest that shunt malfunction may be related with epileptic seizure (Fig. 18.3). In a review of 200 hydrocephalic children, 10 patients had a seizure due to a blocked shunt device [22]. Faillace and Canady [18] retrospectively reviewed

15 patients with hydrocephalus who had an epileptic seizure at the time of shunt malfunction and they found that there had been no history of epilepsy in 8 patients. They suggest that as a rule, shunt malfunction should be considered, if a new or recurrent epileptic seizure develops after shunt insertion for hydrocephalus [18].

On the other hand, epilepsy was not generally associated with shunt malfunction in some series [4, 19, 27, 34, 36, 38, 42, 43]. Thus, they concluded that the existence of epileptic seizure alone was not a reliable indicator of a shunt malfunction [19].

18.5.5 Shunt Infection

Numerous retrospective studies reported that the risk of development of epileptic seizures was significantly increased in cases with shunt and/or cerebrospinal fluid (CSF) infection [10, 22, 24, 36, 37, 40, 43, 45]. In a review of 168 shunt-treated hydrocephalic children, however, Saukkonen et al. [42] found that there was no link between epileptic seizure and existence of shunt infection. Likewise, Piatt and Carlson [38] reported that there was no correlation between risk of development of epileptic seizures and a history of shunt infection. No matter in what way, shunt infection should be considered as a general rule, if an epileptic seizure develops after shunt insertion for hydrocephalus.

18.5.6 Slit Ventricle Syndrome

Typically, SLVS, which is characterized by very small ("slit-like") ventricles in computed tomography (CT) or magnetic response imaging (MRI), occurs as a result of collapse of the ventricles due to overdrainage of the CSF in minority of patients after shunt placement or revision (Fig. 18.4). As a cause of epilepsy after shunting, it was observed in only three of 182 patients with hydrocephalus, corresponding with the 0.9–3.3 % incidence in the current literature [41, 42]. After shunting procedure, epilepsy developed in 44 % of patients in the SLVS group, in contrast to 6 % of those in the non-SLVS group [41]. Out of 141 hydrocephalic

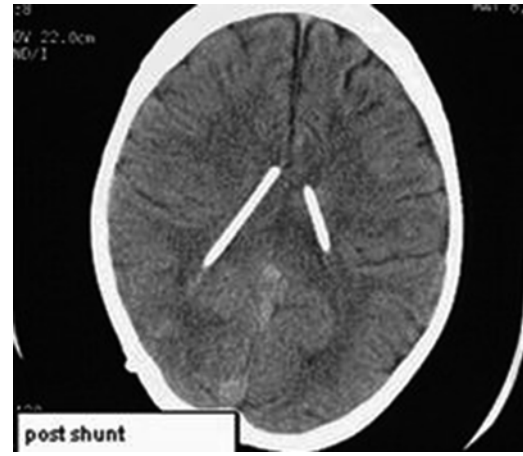

Fig. 18.4 CT scan of a 32-month-old boy with rapid ventricular decompression and absence-type seizures 5 weeks after shunt revision revealed collapsed ventricles (Courtesy of Jogi V. Pattisapu, MD, Orlando, FL)

patients treated with shunting, epilepsy developed in 31 those with SLVS, but 7 those with normal or dilated ventricles during the follow-up period [42]. More importantly, the same authors found that epilepsy decreased in patients with the SLVS after treatment [41]. Thus, one may suggest that serial EEG evaluation is useful in the follow-up of the patients after shunting.

18.5.7 Cortical Malformation

According to the results of their retrospective series, Noetzel and Blake [36] noted that risk factors for development of epileptic seizure in patients with shunted hydrocephalus included the existence of neuroradiological findings of cortical malformation. Nowadays, it is generally known that histopathological etiology of epilepsy such as cortical dysplasia, hemimegalencephaly, and Rasmussen encephalitis is the most important determinant for development of hydrocephalus [37].

18.5.8 Intracranial Hemorrhage

Epileptic seizure is believed to be a common presenting symptom in neonates, children, and adults with intracranial hemorrhage. Talwar

et al. [45] noted that 3 patients out of 81 children had epileptic seizures possibly due to intracranial hemorrhage during the VP shunt revision surgery as a surgical complication. Likewise, Johnson et al. [24] also pointed out that epilepsy was more frequent in patients with acute intracranial bleeding.

18.5.9 Hyponatremia Due to Abdominal Pseudocyst

Interestingly, Buyukyavuz et al. [7] firstly reported a case of hyponatremic seizure caused by intra-abdominal pseudocyst formation as a complication of the VP shunt. As far as we know, there is no documented case of this condition in the current literature to date.

18.5.10 Episodes of Raised Intracranial Pressure

Interestingly, in a retrospective study of 802 children with hydrocephalus, Bourgeois et al. [5] reported that 32 % of the children had epileptic seizure, possibly owing to such episodes of raised ICP or the presence of a shunt device as an epileptogenic focus.

18.5.11 Intracranial Hypotension Related with Body Posture

Interestingly, Agrawal and Durity [1] described a child with a VP shunting who presented with epileptic seizures related with posture of the child, possibly due to intracranial hypotension.

18.6 Time of Occurrence of Epileptic Seizure After Shunting Procedure

In 1981, Copeland et al. [10] reported that 58 % of the cases developed epilepsy following the shunt placement within the first month. In similar, Dan and Wade [12] found a higher incidence of epileptic seizure in patients with multiple ventricular catheter revisions with a decreasing risk of seizures from 5 % in the post-shunt 1st year to 1 % after the 3rd year of shunting. Johnson et al. [24] found that 22 % of the 817 children with shunted hydrocephalus had first epileptic attack following the initial shunt placement, whereas 38 % of all patients had at least one epileptic seizure.

Based on their long-term follow-up retrospective study, Saukkonen et al. [42] reported that 14 % of the patients had seizures following the insertion of the shunt within the first 6 months; 25 % of the patients developed epilepsy within the 1st year, 40 % of the patients had epileptic seizure within 2 years, and the remaining 61 % of the patients developed epilepsy following shunting within 2–15 years. Afterwards, in a retrospective review of 464 patients with hydrocephalus, Piatt and Carlson [38] reported that 12 % of patients developed epileptic seizures at the time of diagnosis of hydrocephalus and the risk of epilepsy was 2 % following the shunt placement for each year and 33 % of the patients by 10 years following the shunt surgery.

18.7 EEG Changes in Hydrocephalic Patients with/Without Shunting

18.7.1 EEG Findings in Hydrocephalus

To date, many authors have reported a significantly higher rate of EEG abnormalities, such as generalized slow-wave activity, unilateral or focal attenuation, and focal spike waves and/or sharp waves, in patients with hydrocephalus and seizures, in contrast to hydrocephalic patients without development of epilepsy [10, 12, 23, 36, 41, 42, 44, 45, 47] Also, Carballo et al. [8] reported a series of 9 cases with hydrocephalus and continuous spikes and waves during slow sleep (CSWS), related with epilepsy.

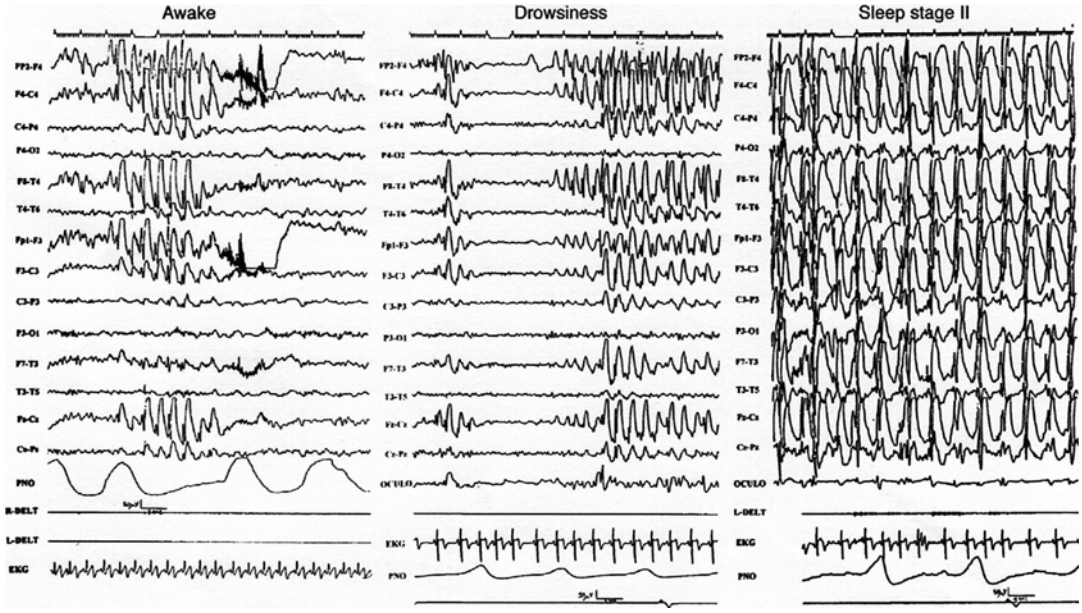

Fig. 18.5 EEG of a children with shunted hydrocephalus showing focal abnormalities (*left*), secondary bilateral synchrony (*centre*), continuous spikes and waves during sleep (*right*) (Reproduced with permission from Veggiotti et al. [46])

18.7.2 Focal EEG Changes in Hydrocephalic Patients with Shunting

So far, various focal epileptiform abnormalities have been described in children with cerebral ventricular shunting. Al-Sulaiman and Ismail [2] investigated the EEG abnormalities in 68 cases with hydrocephalus and they found focal or generalized findings in the shunted group including slow waves in 26 cases, epileptiform activity in 26, hypsarrhythmia in 4, and amplitude abnormalities in 2, giving a total ratio of abnormality above 90 %.

Besides, Veggiotti et al. [46] reported that focal EEG abnormalities were ipsilateral to the location of the shunt device in 95 % of children (Fig. 18.5). In general, it has been accepted that traumatic injury to the brain during the shunt procedure and the existence of an intracranial foreign material will result in focal epilepsy in contralateral side and EEG changes ipsilateral to the localization of the shunt [10, 12, 22, 23, 33, 34, 36, 38, 42, 43, 46, 47]. Ines and Markand [23] found that there was a high incidence of focal EEG abnormalities in the shunted hydrocephalic patients, about 50 % of the nonshunted hydrocephalic patients and almost all of the shunted hydrocephalic group, suggesting that the shunt as a kind of foreign body may be responsible for these findings. Likewise, Liguori et al. [33] evaluated the EEG findings in 40 patients with shunted hydrocephalus and epileptic seizures and they found that the frequencies of both specific and nonspecific EEG findings are higher on the shunted hemisphere (19 patients) compared to the unshunted side (8 patients), suggesting the presence of the intraventricular shunt catheter as a cause of the EEG focus.

Moreover, Saukkonen et al. [41] suggested that a kind of shunt malfunction should be suspected in patients with hydrocephalus, if any abnormal focal EEG finding develops after shunting procedure. Recently, Posar and Parmeggiani [39] described a case of an early-onset hydrocephalus causing partial epilepsy with a particular EEG finding, known as CSWS, possibly due to involvement of frontal, parietal, and occipital lobes.

In a series of 113 children with shunting, Saukkonen et al. [41] investigated the relationship between epilepsy and the EEG changes in the SLVS following shunting. The same authors found a generalized spike and sharp wave activity (SWA) in 81 % of 63 patients in the SLVS group, but in 54 % of patients of non-SLVS group following shunting procedure [41]. Importantly, the EEG findings disappeared in patients with the SLVS following treatment [41].

Nevertheless, current data is conflicting with regard to the effects of the intraventricular shunt catheter as a cause of the EEG focus. From a total of 168 shunted hydrocephalic patients, generalized SWA in EEG before shunting procedure was seen in 45 % of the patients, whereas partial epilepsy following shunting procedure was seen in 9 % of the patients [42]. Then, Veggiotti et al. [46] reported focal EEG abnormalities which were ipsilateral to the site of shunt in 95 % of children, confirming its possible role in the epilepsy. In contrast, however, Saukkonen et al. [42] reported that epileptic seizure had no correlation with the side of the shunt or with the side of the epileptic activity in the EEG.

Accordingly, Saukkonen et al. [41] reported that focal EEG changes are frequently seen within the first year of life in hydrocephalic children and a slow-wave focus may arise in an enlargement of the third ventricle or of the posterior fossa, not related with the direct effect of an intraventricular shunt catheter. Likewise, Klepper et al. [28] found that there were focal EEG abnormalities related to the anatomical location of the shunt in 14 of 16 (88 %) patients, while contralateral focal seizures and focal EEG abnormalities on the same side to the shunt device were present in only three patients (2 %). Based on their findings, Klepper et al. [28] suggested a minor effect of the surgical procedure for decision concerning epilepsy related with shunting in the presence of the following three criteria: (a) development of epilepsy in the postoperative period; (b) focal seizures contralateral to the site of shunt placement; and (c) presence of EEG changes which are ipsilateral to the site of the shunt device. In conclusion, they speculated that the epilepsy was determined by the cause of hydrocephalus rather than by the shunting procedure, an overestimated complication of intracranial shunting [28].

18.8 Relationship Between Mental or Physical Disability and Occurrence of Seizures in Shunted Hydrocephalus

Some authors pointed out various risk factors, including mental or physical disability, for development of epileptic seizure in patients with shunted hydrocephalus [29, 36, 40]. Stellman et al. [43] found that epileptic seizures frequently developed owing to various shunt-related problems in children with mental or physical disability. Later, Keene and Ventureyra [27] observed that epilepsy developed in patients with hydrocephalus associated with motor and/or cognitive disability, suggesting the importance of encephalopathy as an etiological factor instead of hydrocephalus.

18.9 Postoperative Hydrocephalus After Hemispherectomy in Patients with Epilepsy

From a surgical point of view, there are various hemispherectomy procedures – anatomical, functional, and modified – and the modified approach has some advantages in pediatric patients with hemispheric cortical dysplasia with small and/or malformed ventricles [9, 25]. In a series of 9 children with hemimegalencephaly who underwent surgical procedure for intractable epilepsy, Di Rocco et al. [15] reported a dramatic improvement in the seizures following hemispherectomy in all children. Afterwards, Di Rocco et al. [13] reported that there were a total of 5 children with a secondary hydrocephalus in a series of 15 children operated with hemimegalencephaly.

Unfortunately, a high incidence of hydrocephalus following various cerebral hemispherectomy procedures in pediatric patients with intractable seizures is a well-known entity [9, 25, 31, 32]. Phung et al. [37] suggested several mechanisms

leading to some changes in CSF bulk flow for development of hydrocephalus following various hemispherectomy procedures. In a retrospective review of their findings, Di Rocco et al. [13] found that the age factor appeared to play a critical role in the development of postoperative hydrocephalus, as all of five children with the complication were less than 9 months of age at the time of the hemispherectomy. In a recent review of the findings from a total of 736 patients who underwent hemispherectomy procedure, Lew et al. [32] reported that the hydrocephalus was seen as an early or late surgical complication in patients, ranging from the early postoperative period to 8.5 years following surgical procedure. More recently, the same authors reported that the use of Avitene caused a higher incidence of hydrocephalus following the surgery in cases with modified hemispherotomy, a safe surgical technique, in patients with epilepsy (56 % vs 18 %) [31].

18.10 Outcome of Hydrocephalic Patients with Epilepsy

As a general rule, the etiology of hydrocephalus is the decisive factor in determining the outcome in hydrocephalic patients with epilepsy. So far, numerous studies have been done upon the result of shunted hydrocephalic patients with epilepsy [20, 21, 26]. Based on the findings from their hydrocephalic patients treated with shunting, Saukkonen et al. [42] reported that epilepsy developed in all of the patients without any prophylactic antiepileptic treatment, whereas 68 % of those with prophylactic treatment remained free of seizures.

Afterwards, Heinsbergen et al. [20] found that hydrocephalic children, owing to various congenital malformation including spina bifida had better prognosis, in contrast to other ones. On the other hand, the outcome of the patients with hydrocephalus is poor in the presence of the following risk factors: (a) peri- and postnatal hemorrhage; (b) delay in drain insertion more than 1 month; (c) children less than 2 years old; and (d) concomitant pathology such as Dandy–Walker malformation, aqueduct stenosis, myelomeningocele, and arachnoid cyst ([35, 40], Heinsbergen et al. [20]).

Indeed, Bourgeois et al. [5] suggested that the presence of epilepsy itself is an important predictor of poor outcome in children who were operated using a kind of shunt device. As given above, it may be a sign of shunt malfunction in cases with hydrocephalus. Regarding the role of surgery and antiepileptic drugs, Faillace and Canady [18] observed that seizure activity stopped in patients after revision procedure for shunt malfunction and medical treatment.

18.11 Future Treatment Options

Even today, it is unfortunate to note that antiepileptic medical treatment is ineffective in most of the cases with postshunt epilepsy [40]. Furthermore, the shunting procedure to control hydrocephalus may cause a complication with high incidence [11, 17]. On the other hand, the choice of the surgical technique to prevent various shunt complications is still a subject of debate, although there is a general agreement on the effectiveness of the surgery in controlling the seizure disorder in hydrocephalic patients. As a result of understanding of the pathophysiology of hydrocephalus, it has been suggested that development of more physiological new surgical techniques such as endoscopic third ventriculostomy procedure in neurosurgical practice may be useful in the prevention of this problem in the future [17, 40]. Our management strategies based on case reports or case series are very limited to date. Therefore, it is suggested that a prospective study is needed to identify the factors predisposing to epileptic seizures in hydrocephalic patients undergoing shunt surgery and to improve the life quality of the patients [5]. As a consequence, we strongly believe that the relationship between the shunting procedure for hydrocephalus and the epilepsy may be disclosed with improved studies using animal models in future.

Conclusion

The topic of epilepsy in patients with hydrocephalus is very important because the incidence of epilepsy in children with shunting is reported to be high, up to 50 % of patients. In

addition to the etiology of the hydrocephalus, shunt dysfunction or various shunt complications may cause epileptic seizures in patients with hydrocephalus. It is our opinion that appropriate knowledge of the mechanisms responsible for the development of epilepsy in shunted hydrocephalus together with forthcoming improvements in the management of hydrocephalus will doubtless contribute to decrease these complications in future. It is now evident that every neurosurgeon should know the problem of epilepsy in hydrocephalic patients because of high incidence of seizures in shunted patients.

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