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Headaches in Patients With Shunts

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Headache is one of the most common afflictions suffered by humans. Headache in patients with a shunt triggers a series of events that includes utilization of expensive technologies and often potentially dangerous surgical intervention. The purpose of this study was to determine the incidence of headaches in patients with shunts and, hopefully, the relationship of those headache disorders to the treatment of hydrocephalus. The Hydrocephalus Association maintains a self-reporting database recorded from individuals treated for hydrocephalus and their families. This database was mined to determine the incidence of severe headaches requiring treatment and interfering with normal life in patients who have been treated for hydrocephalus. There were 1,242 responders between the ages of 19 months and 45 years of age. Of these, 1,233 answered the question, "Do you or your family member suffer from (does your child complain of) frequent or chronic headaches?" This subset forms the basis of this study. Three groups were defined by age: children (19 months-12 years), adolescents (13 years-19 years), and young adults (20 years-45 years). Most respondents were initially treated during infancy (before 18 months of age); 84% of children and 69% of both adolescents and young adults were treated very early in life. Severe headaches became a more frequent problem as the age of the population treated for hydrocephalus increased. In terms of frequency and severity of headaches, direct comparisons with epidemiologic studies of normal populations are difficult because of the limitations of data available in the database. However, it is likely that this population has a higher incidence of severe headaches than normal populations. The cost of management of headaches in this population is very high, and the patients are at risk throughout life. Early treatment decisions have a significant effect on later quality of life. Strategies that lead to normalization of cerebrospinal fluid dynamics and life without shunt dependency are justified if they can be shown to improve later quality of life.
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Until well into the second half of the 20th century, the diagnosis of hydrocephalus was either a death sentence or compatible only with a life of severe disability.[1,2] The development of valve-regulated shunts led to the expectation of survival and a hope for normalcy for a large number of patients otherwise facing tragedy. However, the treatment of hydrocephalus did not lead to normal lives for these patients. Patients with a shunt live with the constant fear of shunt failure and a long litany of complications associated with dependency on an implanted foreign body such as infection, mechanical failure, and death from complications of this treatment. In most patients, effective treatment of hydrocephalus in infancy often leads to a radiographically subnormal ventricular size. A much smaller but poorly defined group of patients suffers symptoms from the overdrainage associated with radiographic slit ventricles.[3]

Severe headaches in patients who have been treated for hydrocephalus with shunts and small ventricles on imaging studies have a condition referred to as the slit ventricle syndrome (SVS).[4] We have defined 5 subsets of these patients whose headaches have different causes including intermittent proximal obstruction, low-pressure headaches, shunt failure with ventricles that do not expand (ie, normal volume hydrocephalus [NVH]),[5] cephalocranial disproportion found exclusively in patients with craniofacial syndromes, and shunt-related migraine.[6] There are numerous treatment options for patients with shunt-related headaches regardless of their cause. These treatments included a valve upgrade with devices that retard siphoning,[6,7] subtemporal decompression,[8] cranial expansion,[9] programmed shunt removal,[10] antimigraine medical management,[11,12] and shunt systems that incorporate drainage of the cortical subarachnoid space and the ventricle such as lumboperitoneal shunts[13,14] and cisterna magna-to-ventricle-to-peritoneal shunts.[15,16]

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Regardless of the cause of the SVS, these patients are always at risk for death or neurologic injury if a true shunt failure is ignored. For many patients, each headache results in a trip to the emergency room with a scan and often a surgical procedure that fails to cure their problem. Discussions among neurosurgeons vary greatly as to the incidence of this problem and whether surgical intervention is justified for headache in patients with small ventricles. Therefore, this study was performed in an attempt to define the previously unknown denominator. What is the actual likelihood of a patient undergoing treatment for hydrocephalus having severe headaches? Why is there such a disparity in the opinions of neurosurgeons about the seriousness of this problem? How can advances in understanding the problem and the development of new technologies improve the quality of life for this growing population of patients?

Methods

The Hydrocephalus Association, a support organization for patients with hydrocephalus and their families, has developed a voluntary self-reporting database. Participation in the database was solicited through the association's quarterly newsletter, biannual national conferences, and a network of neurosurgeons with a specific interest in the treatment of hydrocephalus. It is also available to anyone via the association's Web site. Patients and family members have the option to complete information online in a database compliant with the Health Insurance Portability and Accountability Act, via paper copy, or over the telephone. This database makes no claims to be representative of all patients with hydrocephalus but reaches out to all who need more information and advocacy. Despite the limitations of a voluntary database, the data presented here are the best that can be obtained at present.

Patients and their families filled out forms containing demographic information, diagnosis, complications, and questions related to their quality of life and outcomes. The present study focused on a small part of the database related to the following question: "Do you or your family member suffer from (does your child complain of) frequent or chronic headaches?" We mined the database for information related to 3 age groups of patients who have been treated for hydrocephalus: children ages 19 months to 12 years, adolescents ages 13 years-19 years, and young adults ages 20 years-45 years. We evaluated the age of patients at their initial treatment and the likelihood that they would suffer from frequent disabling headaches. We attempted to define the scope of the problem in terms of the effect of the treatment of hydrocephalus on this most chronic of problems. These data should define areas of potential study and the need for further research and technologic improvements to decrease the overall risks and costs of treatment and to improve the quality of life in these patients who will live many more decades.

Results

There were 1,242 responders in the age groups from 19 months to 45 years of age. Of these, 1,233 (99.4%) responded to the following question: "Do you or your family member suffer from (does your child complain of) frequent or chronic headaches?" The database contains information on younger infants and older adults, but it would be difficult to determine the incidence of headaches in preverbal children and the problems of older adults with hydrocephalus are thought to be distinct. Therefore, the focus was on the middle groups. Three categories were studied. Responses for AQ: 1 the childhood group aged 19 months to 12 years were primarily given by the parents. This group represented 51% (634 responses) of the total number of respondents. Children aged 13 years to 19 years accounted for 17% (213 responses) of the total, and adults aged 20 to 45 years accounted for 32% (386 responses).

Hydrocephalus needing treatment was a chronic disease that began in infancy in 90% of the children, in 73% of the adolescents, and in 74% of the adults who responded that they had undergone their first treatment for hydrocephalus in the first 18 months of their life.

Family members reported severe disabling headaches in 29% of the children. As the children aged, the problem of headaches became more serious. Severe, disabling, and frequent headaches were reported in 42% of the adolescents and 44% of the adults. These respondents reported that headaches had a significant impact on their quality of life. The incidence of headaches between children and adolescents and between children and adults was significantly different ($P < .05$). The incidence of headaches was not significantly different in adolescents and adults. Headaches that interfered with the quality of life may begin in childhood but occurred in at least 40% of adolescents and young adults.

Discussion

There is now an expectation that infants with hydrocephalus may live into middle age or later. Many shunt-dependent patients have few or no other anomalies and have the potential to live productive, independent lives. Nonetheless, patients with shunts live with a constant anxiety that their shunt may fail at any time. The failure may be an inconvenience requiring interruption of active lives, or it can lead to severe neurologic injury or even death.[17]

Many changes occur because of chronic shunting including increases in the thickness of the skull (including the skull base), asymmetry of the cerebral ventricles or radiographic slit ventricles, and the inability of the ventricles to expand at the time of shunt failure.[18,19] Which of these changes, if any, lead to SVS and to severe, disabling headaches cannot be determined for certain. The classification of slit ventricles based on monitoring of intracranial pressure (ICP) at least defines what pathophysiologies are associated with severe headaches and gives clues about why shunt-dependent patients might have severe headaches.[6] The classification does not, however, deal with the important function of the denominator. How many adolescents and young adults suffer from severe headache disorders, and how are they affected by the treatment that they receive?

The present study is only a start in dealing with this important issue. It suffers from several flaws. It is a voluntary reporting of patients who have been in contact with either the Hydrocephalus Association or with neurosurgeons who are participating in the study. This is not a population-based study even of patients who have been shunted.

Headaches are probably the most common affliction of humankind with a general worldwide incidence of chronic daily headaches (CDHs) of 4% with a 1.8:1 female-to-male ratio.[4,20] CDHs are defined as severe headaches that last at least 4 hours a day for at least 15 days per month. A CDH has many causes including migraine in both its classic and common forms, tension/muscle contraction headaches, fibromyalgia/chronic fatigue syndrome, and the well-founded medication-overuse headaches.[4,20-22] Based on the questions in the hydrocephalus questionnaire, it is not possible to equate the answers with CDH, but until further databases can be produced it will have to do as a start. It seems that severe and disabling headaches occur 10 times as frequently in the population of patients with shunts as CDH occurs in the general population.

Headaches in this population carry a great burden for both patients and the health care system. When severe headaches occur in shunted individuals, they are told that ignoring a shunt failure could lead to severe consequences and even death. Physicians who care for these patients cannot afford to ignore the symptoms and risk a shunt failure causing coma or death. Patients are sent to emergency rooms where sequential computed tomography scans are performed and often show no anatomic reason for the headache. Nonneurosurgeons who attempt to treat headaches in these patients become frustrated, and all become convinced that there is something wrong with the shunt. This frustration often leads to surgery that is found to be unnecessary and placing patients at risk for infection and other surgical complications.

How should these patients be treated if it is determined that the shunt is working properly? This problem is important and, to some extent, frustrating. First, it is essential to ascertain that patients are not suffering from severely increased ICP. To do so, it is frequently necessary to monitor ICP and to ensure that the shunt is functioning properly. In many patients, especially those whose ventricles do not expand at the time of shunt failure, ICP must be measured by a manometric shunt tap.[6] Several authors recommend the use of antimigraine management for all patients with SVS.[11,12] Some patients do indeed have migraine and a shunt. In these patients, the symptoms are usually characteristic of migraines, and there is often a strong family history of migraines. Although the overuse of antimigraine medication is associated with medication-overuse headaches, most patients with this problem have been treated with opiates. Patients with this chronic form of headache disorder must avoid the use of opiates because it leads to the potentiation of the headaches themselves and then eventually causes the headaches. Any treatment regimen for the management of this condition first requires that patients be weaned appropriately from such medication.[20]

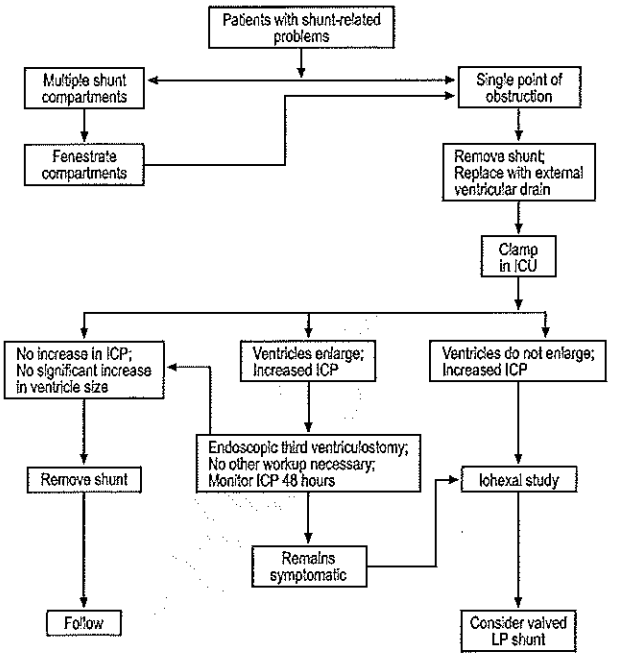

Figure 1 An algorithm for the shunt-removal protocol leading to improved treatment of patients with shunt-related headaches. (Courtesy of Barrow Neurological Institute.)

Is there a way to manage these patients rationally with fewer imaging studies, fewer trips to emergency rooms, and fewer futile surgeries? We await the development of technology that will allow shunts to contain pressure transducers that will enable clinicians to determine when a headache is just a headache and to adjust the shunt telemetrically as needed.[23] A system for closed- or open-loop control of ICP in hydrocephalus should be within reach now that there are several models of shunt systems with adjustable closing pressures or resistances.

At present, we advocate a "shunt-removal protocol" for the evaluation of these patients and to improve understanding of the condition by patients, family, and physicians.[10,24] Figure 1 is the algorithm that we use for shunted patients who have severe, incapacitating headaches. In this protocol, the shunt is removed and replaced with an external ventricular drain. The drain is closed and ICP is measured. F1

As can be seen from the algorithm, there are 3 possible outcomes. In some patients, the ventricles normalize, and the pressure is not elevated significantly. These patients can have the drains removed safely and be followed. The second possibility is that the ventricles enlarge, and the pressure becomes elevated. In these cases, we advocate performing endoscopic third ventriculostomy (ETV). Except in patients with spina bifida whose hydrocephalus is caused by the Chiari II malformation in all its complexity, patients with increases in ventricular size have had an 80% success rate with ETV. The implication is that simply seeing the ventricles expand at the time of shunt failure implies that there is no communication between the ventricular system and the cor-

tical subarachnoid space, and that is all that is needed to recommend ETV.

The third arm of the algorithm is for patients whose ICP increases substantially but whose ventricles do not expand. In these patients, the initial cause of their hydrocephalus involved the cortical subarachnoid space. These patients have all been found to have had hydrocephalus in infancy, although in some it was not treated until later in life. In some of these patients, we have performed retrograde venograms and measured venous pressure. The results indicated that the initial cause of their hydrocephalus was venous hypertension. Treatment of these patients requires shunt systems that access the cortical subarachnoid space using either lumboperitoneal shunts or, for patients who are not candidates for this procedure, a cisterna magna-to-ventricle-to-peritoneal shunt.[6,13-16,24]

Each patient who undergoes our shunt removal protocol understands the role that the shunt plays in his/her headaches. By using adjustable valves with devices that retard siphoning, it is possible to adjust the shunts to normalize intracranial dynamics. To be normal means that all cerebrospinal fluid compartments communicate with each other, recumbent ICP is between 5 mm Hg and 15 mm Hg, and erect pressure is between −5 and + 5 mm Hg. This approach has successfully relieved headaches in at least 75% of all patients treated this way. The other 25% fully understand the pathophysiology of their headaches and what steps and what medications are most useful in these difficult cases.

Finally, we now understand that hydrocephalus is a chronic condition. It may be justified to "program" the management of hydrocephalus to maximize the likelihood of life without the need for a shunt. Recent advances in the understanding of the pathophysiology of hydrocephalus and advances in technology have the potential to minimize the negative effects of this condition and its treatment on the quality of these patients' long lives.

Conclusions

Hydrocephalus is a lifelong condition, and decisions made in infancy can affect the quality of patients' lives as they age. Headaches in patients with shunts do not necessarily mean that the shunt has failed. They may mean that patients are experiencing extremely abnormal cerebrospinal fluid pressure dynamics. New technological advances may have the ability to normalize intracranial dynamics to help more of these patients to live more normal lives. Full understanding of the cerebrospinal fluid dynamics will lead to a more rational approach to the management of headaches in this challenging patient population.

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