RESEARCH ARTICLE

RELATIVE FREQUENCY OF HYDROCEPHALUS IN RASHT PEDIATRIC PATIENTS

Elham BIDABADI MD

Assistant Professor of Pediatric Neurology, Guilan University of Medical Sciences,Guilan,Iran

Corresponding Author:
E. Bidabadi MD
17th Shahrivar Hospital, Rasht, Iran
Email:mashoof@mail.com

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Abstract

Objective

There are few studies evaluating the relative frequency of different etiologies of hydrocephalus in pediatric population, in Iran; therefore, this topic was considered in the present study.

Material & Methods

In a prospective study, the study group consisted of all children fulfilling the imaging criteria for having hydrocephalus, aged below 12 years, admitted at Rasht 17th Shahrivar and Poursina Hospitals, between March 2006 and September 2008. Demographic data and information on the etiology and type of hydrocephalus, and surgical and pathological diagnosis were recorded for all cases. All data were analyzed with SPSS version 13 software.

Results

There were a total of 67 patients, with a mean age of 33.25 ± 43.40 months. Hydrocephalus was seen most frequently in the first 2 years of life. Thirty three patients (49.3%) were female and 34 (50.7%) were male. Mean age of the girls and boys was 30.78 ± 46.46 and 35.64 ± 40.77 months, respectively (p=0.650). Communicating and obstructive hydrocephalus was seen in 30 (44.8%) and 37 cases (55.2%), respectively. There was no statistically significant difference in the types of hydrocephalus between the two sex groups. Mean age in obstructive and communicating groups was 3.76 ± 46.17 and 35.09 ± 40.42 months, respectively (p=0.0006). The most common causes of hydrocephalus were myelomeningocele for obstructive (19.4% of total cases), and TORCH and meningitis for communicating (8.9% of total cases for each).

Conclusion

In our study, there was a mild preponderance for male sex and obstructive type. Mean age of the patients with obstructive hydrocephalus was significantly lower than those with communicating type. Overall, the most common cause of hydrocephalus was myelomeningocele.

Keywords: Child, Hydrocephalus, Etiology.

Introduction

Hydrocephalus is a disorder in which the cerebral ventricular system contains an excessive amount of cerebrospinal fluid (CSF) and is dilated because of increased pressure (1,2) . The increased pressure distinguishes hydrocephalus from atrophy, in which dilatation is due to loss of brain tissue. CSF accumulates due to an imbalance between production and absorption.

The prevalence of congenital and infantile hydrocephalus has been estimated

as 0.48 to 0.81 per 1000 live and still births (3-6). The outcome of hydrocephalus depends upon the etiology, associated abnormalities, and complications such as infection. Survival in untreated hydrocephalus is poor. Approximately 50 percent of affected patients die before three years of age, and 77 to 80 percent die before reaching adulthood (3). Treatment markedly improves the outcome for hydrocephalus not associated with tumor, with 89 and 95 percent survival in two reports (7,8).

There are few studies evaluating the relative frequency of different etiologies of hydrocephalus in pediatric population in Iran; therefore, this topic was addressed in the present study.

Material & Methods

In a prospective study, the study group consisted of all children fulfilling the imaging (brain CT scan and/or brain MRI) criteria for having hydrocephalus, aged below 12 years, admitted at Rasht 17th Shahrivar and Poursina hospitals, two referral pediatric and neurosurgical centers in Guilan province, affiliated to the department of child neurology and neurosurgery, between March 2006 and September 2008. Demographic information was collected including age, and gender. Age (in months and years) was calculated from the date of birth. Information on the etiology and type of hydrocephalus, and surgical and pathological diagnosis, if any, were recorded for all cases.

Data was analyzed with SPSS version 13 statistical software. Discrete variables were expressed as counts (%) and compared using the Chi-square tests. Continuous variables were expressed as mean ± SD and compared by means of the unpaired, two-sided t test. Adjusted odds ratios and 95% Wald confidence intervals were calculated based on these models. Statistical significance was set at $P<0.05$.

Results

There was a total of 67 patients with a mean age of 33.25 ± 43.40 months. Thirty three patients (49.3%) were female and 34 (50.7%) were male. Mean age of the girls and boys was 30.78 ± 46.46 and 35.64 ± 40.77 months, respectively. There was no statistically significant difference in mean age between genders (p=0.650).

Communicating and obstructive hydrocephalus was seen in 30 (44.8%) and 37 (55.2%) Cases, respectively. There were 15 (45.5%) communicating and 18 (54.5%) obstructive cases in the female, and 15 (44.1%) communicating and 19 (55.9%) obstructive cases in the male group (Table 1). There was no statistically significant difference in the types of hydrocephalus between sexes.

Table 1: Etiology and types of hydrocephalus

Etiology	Type	Male (% of males)	Female (% of females)	Total (% of total)
Myelomeningocele	Obstructive (55.2%)	9 (26.5%)	4 (12.1%)	13 (19.4%)
3rd ventricle tumor		2 (5.9%)	1 (3%)	3 (4.5%)
Posterior fossa tumor		3 (8.8%)	1 (3%)	4 (6%)
Other brain tumors		1 (2.9%)	2 (6.1%)	3 (4.5%)
Aqueductal stenosis		1 (2.9%)	2 (6.1%)	3 (4.5%)
Dandy Walker malformation		1 (2.9%)	0	1 (1.5%)
Brain abscess		0	1 (3%)	1 (1.5%)
Intracranial hemorrhage		0	4 (12.1%)	4 (6%)
Unknown (obstructive)		6 (17.6%)	3 (9.1%)	9 (13.4%)
TORCH	Communicating (44.8%)	5 (14.7%)	1 (3%)	6 (8.9%)
Meningitis		0	6 (18.2%)	6 (8.9%)
SAH		0	3 (9.1%)	3 (4.5%)
IVH		2 (5.9%)	1 (3%)	3 (4.5%)
History of trauma		1 (2.9%)	1 (3%)	2 (2.9%)
Neurodegenerative disease		0	1 (3%)	1 (1.5%)
Unknown (communicating)		3 (8.8%)	2 (6.1%)	5 (7.5%)
Total		34	33	67

Mean age of the participants in the obstructive and communicating groups was 3.76 ± 46.17 and 35.09 ± 40.42 months, respectively (p=0.0006). This difference was found to be significant.

As shown in the Table 1, the most common causes of hydrocephalus were myelomeningocele for obstructive (19.4% of total cases) and TORCH and meningitis for communicating (8.9% of total cases for each) hydrocephalus.

Data on the etiology of hydrocephalus based on different age groups are provided in Table 2. Hydrocephalus was seen most frequently in the first 2 years of life, as shown in the Table 2.

Table 2: Etiology of hydrocephalus in different age groups

Age group	Type		Male (% in the age group)	Female (% in the age group)	Total (% of total cases)	The most frequent cause of hydrocephalus (% in the age group)
	Obstructive (% in the age group)	Communicating (% in the age group)				
<2 years	25 (64.1%)	14 (35.9%)	18 (46.2%)	21 (53.8%)	39 (58.2%)	Myelomeningocele (33.3%)
2-4 years	1 (16.7%)	5 (83.3%)	3 (50%)	3 (50%)	6 (8.95%)	Meningitis (33.3%)
4-6 years	2 (28.6%)	5 (71.4%)	4 (57.1%)	3 (42.9%)	7 (10.45%)	Unknown (42.9%)
6-8 years	3 (60%)	2 (40%)	4 (80%)	1 (20%)	5 (7.46%)	Tumors (60%)
8-10 years	4 (57.1%)	3 (42.9%)	5 (71.4%)	2 (28.6%)	7 (10.45%)	Posterior fossa tumors (42.9%)
10-12 years	2 (66.67%)	1 (33.33%)	0	3 (100%)	3 (4.48%)	Unknown (66.67%)

Discussion

Some authors have identified no sex preponderance in hydrocephalus (9,10), but some have found a male preponderance (11); there was a mild male domination (50.7% vs. 49.3%) in our study.

Hydrocephalus can be congenital or acquired. Both categories include a diverse group of conditions.

Congenital hydrocephalus can result from CNS malformations (which include nonsyndromic and syndromic disorders), infection, trauma, and teratogens (8,12).

CNS malformations are frequently associated with hydrocephalus. In Chiari malformation, which often accompanies a neural tube defect, portions of the brain stem and cerebellum are caudally displaced into the cervical spinal canal. This obstructs the flow of CSF in the posterior fossa, leading to hydrocephalus. The majority of patients with myelomeningocele have hydrocephalus, as 19.4% of our cases. In our study, it was found to be the most common cause of obstructive hydrocephalus, especially in children under 2 years of age. The etiology is obstruction of fourth ventricular outflow or flow of CSF through the posterior fossa due to the Chiari malformation or an associated aqueductal stenosis. The Dandy-Walker malformation consists of a large posterior fossa cyst that is continuous with the fourth ventricle and defective development of the cerebellum, including partial or complete absence of the vermis. In this condition, hydrocephalus results from secondary obstruction of the foramina of Luschka and Magendie. It was seen in only 1 boy in our study.

Isolated hydrocephalus is frequently caused by aqueductal stenosis. This can be due to congenital narrowing of the aqueduct, or result from inflammation due to intrauterine infection. The most common genetic form of congenital hydrocephalus is X-linked hydrocephalus with stenosis of the aqueduct of Sylvius (12,13). Approximately 50 percent of the affected boys have adducted thumbs, which is helpful in diagnosis. Some have other CNS abnormalities such as agenesis or dysgenesis of the corpus callosum, a small brainstem, or absence of the pyramidal tract. This disorder is due to mutations in the gene encoding L1, a neuronal cell adhesion molecule that belongs to the immunoglobulin superfamily and is essential in neurodevelopment (12-15). Aqueductal stenosis was detected in 4.5% of our cases.

Intrauterine infections such as rubella, cytomegalovirus, toxoplasmosis, and syphilis can result in congenital hydrocephalus, as seen in 8.9% of our cases. The mechanism is inflammation of the ependymal lining of the ventricular system and the meninges in the subarachnoid space. This may lead to obstruction of CSF flow through the aqueduct or basal cisterns.

Common causes of acquired hydrocephalus are CNS infections such as bacterial meningitis or viral infections including mumps, and tumors, especially posterior fossa medulloblastomas, astrocytomas, and ependymomas. These conditions interfere with the flow of CSF through the ventricular system.

Obstruction caused by a CNS tumor, especially if located near the midline, is a cause of hydrocephalus; fifteen percent of our cases suffered from brain tumors, posterior fossa tumors were the most common of them.

Another important cause is hemorrhage into the subarachnoid space or, less commonly, into the ventricular system, by ruptured aneurysms, arteriovenous malformations, trauma, or systemic bleeding disorders. The hemorrhage induces an inflammatory response followed by fibrosis, obstructing the flow and/or absorption of CSF. Posthemorrhagic hydrocephalus occurs in approximately 35 percent of preterm infants with intraventricular hemorrhage (IVH). It can be obstructive, communicating, or both, and can be transient or sustained, with a slow or rapid progression.

Fifteen percent of our cases suffered from intracranial hemorrhage.

Vein of Galen malformation is a rare cause of hydrocephalus. Obstruction results from compression of the aqueduct of Sylvius by the markedly dilated and distorted vein of Galen. Presentation in the neonatal period typically includes intractable heart failure (16).

Hydrocephalus can be part of syndromes associated with dysmorphic features and other congenital abnormalities. The most frequent cytogenetic disorders associated with hydrocephalus are trisomies 13, 18, 9 and 9p, and triploidy (12-15).

In conclusion,We noted a preponderance for male sex and the obstructive type. Mean age of the patients with the obstructive type was significantly lower than the communicating type. Overall, the most common cause of hydrocephalus was myelomeningocele.

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