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Long-term outcomes in patients with treated childhood hydrocephalus

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Object. The goal in this study was to determine the long-term effects of childhood hydrocephalus.

Methods. A patient-reported survey completed by 1953 participants was used to collect data in a subgroup of 1459 individuals who had been treated for hydrocephalus in childhood. Data on shunt complications, including total shunt revisions and infections, were examined in those whose hydrocephalus had been diagnosed at least 10 years earlier (718 patients). Social and functional outcomes were examined in patients who were 20 years of age or older at the time of survey completion (403 individuals). Specific questions addressed the presence of depression, the patient's marital status, independent living arrangements, and the educational level attained.

Shunt complications were common; 54% of patients had four or more shunt revisions, and 9% had three or more shunt infections. Depression requiring treatment occurred in 45% of participants. Other measures of social functioning all reflected a major impact of childhood hydrocephalus. In general, a worse outcome was found in patients whose hydrocephalus was diagnosed before 18 months of age.

Conclusions. The lifelong morbidity associated with shunt placement to treat childhood hydrocephalus is substantial, and it includes shunt-related complications and comorbidities that adversely affect social functioning.

Key Words • hydrocephalus • shunt complication • Hydrocephalus Association database • pediatric neurosurgery

Hydrocephalus is the pathological condition caused by an abnormality of production or absorption of CSF in the brain. This disease is particularly common in infants and children. Hydrocephalus may be primary (idiopathic) or secondary (acquired), with the majority of congenital cases being idiopathic in origin. Common causes of secondary hydrocephalus in children include meningitis, trauma, brain tumors, intracranial hemorrhage, and developmental anomalies of the brain.[11,14] The population incidence of congenital hydrocephalus has been estimated to be between 2.5 and 8.2 per 10,000 live births.[5,13,16] The natural history of untreated congenital hydrocephalus is progressive cognitive decline and early death, usually before the third decade of life.[1,4,9]

The standard treatment for hydrocephalus is placement of a shunt for CSF diversion.[3] Advances in surgical techniques and shunt materials have reduced the risks related to the initial procedure, although long-term risks include repeated shunt malfunction, shunt infection, and death associated with shunt failure. Prospective studies have revealed that the majority of patients who receive a CSF shunt will require at least one revision within the first 2 years after the initial procedure.[2,3,8]

Assuming an incidence of 0.5 per 1000 live births, and an annual birth rate of 4.1 million in the US, we can estimate that approximately 2000 cases of congenital hydrocephalus occur each year. Based on data from a sample of pediatric patients in US hospitals, it can be estimated that at least 10,000 shunt procedures are performed annually.[15] Direct treatment-related costs for patients of all ages with hydrocephalus exceed $1 billion annually in the US.[12] However, the impact of hydrocephalus must be considered not only in terms of direct hospital-based care for treatment of shunt malfunctions but also in how frequently its related conditions require additional medical interventions. The long-term outcome for patients with childhood hydrocephalus in the modern era is poorly defined, particularly with regard to treatment complications and the impact of the disease on activities of daily living. To understand better the long-term impact of hydrocephalus, to identify treatment complications, and to define areas of future study, we examined information on diagnosis, shunt complications, and functional consequences that was collected in a large, self-

Abbreviations used in this paper: CSF = cerebrospinal fluid; VP = ventriculoperitoneal.

reported patient survey. This type of data collection has inherent limitations, such as selection bias, lack of generalizability, validation of reported end points, and inability to generate population-based data. Nevertheless, the results presented provide a compelling picture of the considerable burden experienced by individuals who have hydrocephalus.

Clinical Material and Methods

Data Collection

Starting in February 2003, the Hydrocephalus Association initiated a self-reported patient registry (The Hydrocephalus Association Database Project) that collected responses about patient demographic characteristics, familial factors, comorbidities, quality-of-life issues, and treatment complications. Surveys were distributed to providers (mainly neurosurgeons) or were available through a Web-based system. The survey, which can be viewed online at http://www.hydrocephalusdatabase.org/, was publicized through national meetings of the Hydrocephalus Association, professional organizations, and brochure mailings to physicians. All data were supplied by the patients or by their guardians who completed the surveys. The only inclusion criterion was that the patient or guardian was aware of a diagnosis of hydrocephalus, regardless of the disease's origin. No specific exclusion instructions were specified in the survey, and participants did not have to be members of the Hydrocephalus Association. The diagnosis of the condition underlying the hydrocephalus was not available to us in this data set. Although there were questions in the survey regarding the origin of the hydrocephalus, the write-in responses were too varied to be meaningful. The registry is maintained by the Hydrocephalus Association.

Data collected between February 2003 and June 2005 from surveys completed by 1953 participants were used for this study. We analyzed a subgroup of 1459 patients who had childhood hydrocephalus, defined as diagnosis made before the patient was 20 years of age. The rate of shunt complications was determined in individuals who had received a diagnosis of hydrocephalus at least 10 years before survey completion (718 patients). The time from diagnosis to entry in the registry was recorded as a series of intervals. The youngest age from a selected age range was used to calculate the length of follow up. We assumed that shunt insertion occurred at the same time as the initial diagnosis of hydrocephalus. For these reasons, the true length of the follow-up period may have been overestimated, but a more conservative measure of the shunt complication rate is obtained. Thus, the actual rate of shunt complications might be higher than what was calculated based on these data.

Statistical Analysis

We examined social and functional outcomes among patients who were adults ($\geq$ 20 years of age; 403 respondents) at the time they completed the survey, and compared these outcomes among patients who were diagnosed during infancy ($<$ 18 months old) or childhood. Comparisons were performed using chi-square tests of significance. Probability values less than 0.05 were considered significant. The database was screened for duplicate records by examining unique identifier codes for individually submitted information. All data were analyzed using commercially available software (Stata Version 8, StataCorp LP).

Results

Demographic Data

Of 1459 patients with childhood hydrocephalus, in 66% the diagnosis was made between birth and 18 months of age and in 20% it was made prenatally (Table 1). The cause of the hydrocephalus was reported as congenital in 60% of patients and acquired in 39%. A somewhat larger proportion of the patients was male (52%) than female (48%). The majority of patients (72%) were children when the survey was completed, and in most cases (67%) the survey was completed by either a parent or grandparent. Most of the patients (85%) identified their race as Caucasian. Eight percent of patients reported a family history of hydrocephalus; 3% described a first-degree relative who suffered from the disease.

TABLE 1

*Demographic characteristics of 1459 individuals with childhood hydrocephalus**

Characteristic	No. (%)
sex	
female	693 (47.5)
male	766 (52.5)
race	
Caucasian	1243 (85.2)
African–American	55 (3.8)
Hispanic	56 (3.8)
Asian	19 (1.3)
other†	54 (3.7)
missing data	32 (2.2)
country of residence at time of survey	
US	1356 (92.9)
other	103 (7.1)
age at time of survey	
birth–18 mos	227 (15.5)
19 mos–12 yrs	618 (42.4)
13 yrs–19 yrs	211 (14.5)
20 yrs–45 yrs	383 (26.2)
>46 yrs	20 (1.4)
age at diagnosis	
before birth	284 (19.5)
birth–18 mos	962 (65.9)
19 mos–12 yrs	150 (10.3)
13 yrs–19 yrs	63 (4.3)
survey completed by	
self	378 (25.9)
parent/grandparent	977 (67.0)
son/daughter	77 (5.3)
spouse	9 (0.6)
other‡	18 (1.2)
family history of hydrocephalus§	
yes	109 (7.5)
1st-degree relative	45 (3.1)
other relative	64 (4.4)
no	1350 (92.5)
total	1459 (100)

* Data obtained from the Hydrocephalus Association survey, 2003 to 2005.
† Biracial or multiracial.
‡ Aunt, friend, grandchild, or medical professional.
§ Multiple family members diagnosed in 11 patients (0.8%).

Shunt History and Complications

Among 718 individuals whose diagnosis was made at least 10 years before survey completion, 685 (95%) had received a CSF shunt (Table 2). Shunt complications occurred frequently; more than 80% of patients reported at least one revision. Among the 718 respondents, the frequency of shunt revisions was as follows: 32% required one to three revisions; 31% required four to 10 revisions; 21% required 11 or more revisions; 10% reported no revisions; and in 6% this information was not available. Although the revision rate was high in all age groups, the number of patients who needed no revisions was higher in those diagnosed when older than 18 months of age ($p = 0.002$). The frequency of shunt infections was as follows: one to two infections in 29%, three to four in 4%, and five or more in 5%. The frequency of shunt infections did not differ by age at diagnosis.

Social and Functional Outcomes

To assess long-term functional outcomes, we examined patients with childhood hydrocephalus who were 20 years of age or older at the time of survey completion (403 individuals). This group was divided into three subgroups based on age at diagnosis: Group 1, birth to 18 months; Group 2, 19 months to 12 years; and Group 3, 13 to 19 years. The mean age (31, 32, and 32 years for Groups 1, 2, and 3, respectively) at the time of survey completion did not differ appreciably between these three groups.

Several long-term comorbidities were common. The majority (71%) of patients diagnosed before 18 months of age reported a history of depression, and 45% had received treatment for this disease. In contrast, those whose hydrocephalus was diagnosed later, at 13 to 19 years of age, were less likely to report a history of depression (52%, $p = 0.01$) or to have undergone treatment for it (38%, $p = 0.43$). Similarly, social functioning was more greatly impaired in individuals who were diagnosed at an earlier age. A larger proportion of patients diagnosed in infancy were single at the time of survey completion than was reported in patients diagnosed between the ages of 13 and 19 years (68% compared with 45%, $p = 0.002$). A similar trend was seen for other measures of social functioning, such as childbearing, living independently, acquiring a driver's license, and being employed (Table 3).

TABLE 2

*Frequency of shunt placement and complications in individuals with childhood hydrocephalus diagnosed at least 10 years before survey completion**

Factor	No. of Patients (%)			
	Total	Dx ≤ 18 Mos of Age†	Dx > 18 Mos of Age†	p Value‡
no. per category	718 (100)	581 (81)	137 (19)	
ever had a VP, VA, LP, or CP shunt				0.002
yes	685 (95.4)	561 (96.6)	124 (90.5)	
no	33 (4.6)	20 (3.4)	13 (9.5)	
ever had an ETV				0.001
yes	48 (6.7)	24 (5.2)	24 (20.5)	
no	528 (73.5)	435 (94.8)	93 (79.5)	
missing or don't know	142 (19.8)	122 (—)	20 (—)	
shunt revisions				0.002
yes	605 (84.3)	504 (91.6)	101 (82.1)	
1–3 revisions	226 (31.5)	186 (33.8)	40 (32.5)	
4–10 revisions	225 (31.3)	187 (34.0)	38 (30.9)	
>11 revisions	154 (21.4)	131 (23.8)	23 (18.7)	
no	68 (9.5)	46 (8.4)	22 (17.9)	
missing or don't know	45 (6.3)	31 (—)	14 (—)	
shunt infections				0.09
yes	269 (37.5)	227 (44.8)	42 (36.2)	
1–2 infections	207 (28.8)	172 (33.9)	35 (30.2)	
3–4 infections	30 (4.2)	24 (4.7)	6 (5.2)	
>5 infections	32 (4.5)	31 (6.1)	1 (0.8)	
no	354 (49.3)	280 (55.2)	74 (63.8)	
missing or don't know	95 (13.2)	74 (—)	21 (—)	
other shunt complications§				0.20
yes	258 (35.9)	205 (36.6)	53 (42.7)	
no	426 (59.3)	355 (63.4)	71 (57.3)	
missing or don't know	34 (4.7)	21 (—)	13 (—)	

* CP = cystoperitoneal; Dx = diagnosis; ETV = endoscopic third ventriculostomy; LP = lumboperitoneal; VA = ventriculoatrial; — = percentages not calculated.

† Percentages do not represent data from missing individuals.

‡ Probability values were calculated for yes/no answers. If answers had more than one "yes" category, the categories were collapsed to "yes" or "no."

§ These shunt complications include malfunctions (such as shunt breakage, obstruction, disconnection, and leakage); overdrainage and low-pressure headaches; slit-ventricle syndrome; surgical and postoperative complications; and other complications (cysts, complications from trauma, calcification, blood clots, and psycho/motor/sensory/cognitive deficits).

Discussion

Our analysis of the Hydrocephalus Association database allowed us to examine the rate of treatment complications and functional disabilities associated with a childhood diagnosis of hydrocephalus. Although a self-reported survey has specific limitations, the significance of these data is twofold. First, it appears that the individual and social burden of this condition may be underestimated in existing studies. Second, broader measures of patient outcome and performance must be incorporated in future studies of hydrocephalus.

Disability in patients with hydrocephalus is determined in part by the severity of treatment-related complications. The complications of shunt placement include shunt infection, blockage, and mechanical failure (that is, breakage). Most of the VP shunts placed have an adequate length to allow for the growth of an infant to average adult height. Therefore, although shunt failure is anticipated, it is not an inevitable consequence of placement. Shunt failure rates are substantial in the first 2 years of life in particular, and the need for frequent revisions in the first few years of life is regarded by most practitioners as being a "complication" associated with shunt placement. The actual cause of shunt revisions reported in this study cannot be determined.

In a multicenter study, patients randomly assigned to receive different types of shunts did not demonstrate a difference in shunt failure rates: 50% of patients required a revision in the first 18 months.[2] The shunt infection rate was 8.1% during the 3-year study period. In one hospital-based study of 119 children assessed at 5 years of age, most of whom had received a shunt in infancy, 48% had undergone one to three revisions and 19% had four or more revisions.[6] These studies clearly identify early shunt complication rates, but long-term complication rates remain poorly defined. Because the majority of participants in the Hydrocephalus As-

TABLE 3

*Social and functional outcomes in 403 individuals with childhood hydrocephalus who were 20 years of age or older at the time of survey completion**

Factor	Age at Time of Dx			p Value†
	≤ 18 Mos	19 Mos–12 Yrs	13–19 Yrs	
no. of patients	305	56	42	
mean age in yrs at time of survey	31	32	32	
range	20–64	20–57	20–53	
special services in school‡				0.06
yes	137 (45.5)	24 (42.9)	11 (26.2)	
no	164 (54.5)	32 (57.1)	31 (73.8)	
missing data	4 (—)	0 (—)	0 (—)	
depression				0.04
yes	213 (71.0)	35 (63.6)	22 (52.4)	
no	87 (29.0)	20 (36.4)	20 (47.6)	
missing data	5 (—)	1 (—)	0 (—)	
treated for depression				0.43
yes	133 (45.1)	20 (37.0)	16 (38.1)	
no	162 (54.9)	34 (63.0)	26 (61.9)	
missing data	10 (—)	2 (—)	0 (—)	
headaches				0.52
yes	158 (51.8)	27 (48.2)	18 (42.9)	
no	147 (48.2)	29 (51.8)	24 (57.1)	
highest level of education				0.08§
high school	71 (24.2)	7 (12.7)	6 (14.3)	
beyond high school	222 (75.8)	48 (87.3)	36 (85.7)	
some college	94 (32.1)	13 (23.6)	15 (35.7)	
associate's degree	42 (14.3)	11 (20.0)	3 (7.1)	
bachelor's degree	57 (19.5)	16 (29.1)	12 (28.6)	
postgraduate degree	29 (9.9)	8 (14.5)	6 (14.3)	
missing data	12 (—)	1 (—)	0 (—)	
marital status				0.002
single	205 (67.7)	28 (50.0)	19 (45.2)	
ever married‖	98 (32.3)	28 (50.0)	23 (54.8)	
missing data	2 (—)	0 (—)	0 (—)	
if female, ever been pregnant				0.002
yes	39 (20.6)	12 (41.4)	15 (44.1)	
no	150 (79.4)	17 (58.6)	19 (55.9)	
are male or missing data	116 (—)	27 (—)	8 (—)	
if female, ever had offspring				0.02
yes	31 (16.7)	10 (34.5)	11 (33.3)	
no	155 (83.3)	19 (65.5)	22 (66.7)	
are male or missing data	119 (—)	27 (—)	9 (—)	
living arrangements				0.17**
independent	172 (56.8)	35 (62.5)	30 (71.4)	
alone	58 (19.1)	9 (16.1)	8 (19.0)	
roommate/friend	45 (14.8)	6 (10.7)	3 (7.1)	
spouse/children	69 (22.8)	20 (35.7)	19 (45.2)	
dependent	131 (43.2)	21 (37.5)	12 (28.6)	
parent/relative	125 (41.2)	20 (35.7)	12 (28.6)	
group home	6 (2.0)	1 (1.8)	0 (0.0)	
missing data	2 (—)	0 (—)	0 (—)	
driver's license				<0.001
yes	180 (60.0)	45 (80.4)	39 (92.9)	
no	120 (40.0)	11 (19.6)	3 (7.1)	
missing data	5 (—)	0 (—)	0 (—)	
currently employed				0.007
yes††	171 (56.8)	36 (66.7)	34 (81.0)	
no	130 (43.2)	18 (33.3)	8 (19.0)	
missing data	4 (—)	2 (—)	0 (—)	
hrs of work/wk				0.19
≤40 hrs	124 (73.4)	24 (70.6)	19 (57.6)	
>40 hrs	45 (26.6)	10 (29.4)	14 (42.4)	
missing data or not employed	136 (—)	22 (—)	9 (—)	
denied health insurance due to hydrocephalus				0.76
yes	63 (72.4)	16 (80.0)	11 (73.3)	
no	24 (27.6)	4 (20.0)	4 (26.7)	
missing data	218 (—)	36 (—)	27 (—)	
substance abuse				0.13
yes	28 (9.4)	8 (14.5)	1 (2.4)	
no	271 (90.6)	47 (85.5)	41 (97.6)	
missing data	6 (—)	1 (—)	0 (—)	

* All percentages represent these known values, with missing individuals disregarded.

† Probability values were calculated with missing data disregarded.

‡ Services such as resource specialist, special education classes, or tutoring.

§ Calculated for individuals who completed high school only compared with those who continued beyond high school.

‖ Includes individuals who are in common-law marriages, engaged, in long-term relationships, separated, or widowed.

** Calculated for individuals with independent living arrangements compared with those who had dependent living arrangements.

†† Includes retired individuals (one in the first category, two in the second, and none in third).

sociation registry were diagnosed in childhood, many years before completion of the survey, we could specifically examine shunt complication rates over a long period of time. Among our cohort, 32% of children with shunts had undergone one to three revisions, and another 52% had four or more revisions. Twenty-nine percent reported one to two shunt infections. Although these observations are consistent with other published data that highlight the poor durability of CSF shunt systems,[2,8] our data support the suggestion that the disease burden is underestimated in short-term studies. In fact, the placement of a VP shunt during infancy can be expected to require multiple surgical procedures during childhood in a majority of patients. These surgical procedures will, of course, be associated with a cumulative risk that is likely to be substantial.

There is a limited number of studies in which functional outcome has been examined beyond a few years after shunt placement. In one study, a group of 233 patients with congenital hydrocephalus who received a CSF shunt were followed for a mean of 17 years (range 5–26 years).[10] The mortality rate was 13.7% and the shunt revision rate was 71.2%. Based on a psychological examination, the details of which were not reported, intellectual ability was separated into normal (62.8%), mild retardation (29.8%), and severe retardation (7.4%). The authors of this study did not elaborate on the methods used to determine intellectual ability, and the majority of these patients (71%) had spina bifida. In another study, 129 patients who underwent shunt insertion before the age of 2 years at a single institution were followed for at least 10 years. A motor deficit was noted in 60% of the patients, visual or auditory deficit in 25%, and epilepsy in 30%.[7] Only 32% of the patients had an IQ above 90. A personality disorder was noted in 80% of pa-

tients, although the methodology used to obtain this measure was not stated.

The long-term impact of childhood hydrocephalus was considerable among patients in our cohort. Among adults (≥ 20 years of age) who had received a shunt before the age of 18 months, 45% required special services at school and 71% reported that they were or had been depressed. Furthermore, in this group 45% were treated for depression. Even in adults in whom hydrocephalus was diagnosed between 13 and 19 years of age—a group that has an overall higher level of functioning—38% reported requiring treatment for depression. From these results we infer that mental health is severely affected by childhood hydrocephalus, and that these problems should be anticipated as a part of ongoing healthcare services for these patients. Other survey items all show a general trend toward a greater impact of hydrocephalus when it is diagnosed earlier in life. In those patients in whom the disease was diagnosed before 18 months of age, two-thirds were single and 40% did not have a driver's license. From the latter observation we may infer that a large number of these individuals are dependent on family, friends, or public means for transportation.

With respect to work, although 73% of those diagnosed when they were younger than 18 months of age and who were currently employed were working 40 hours or less, 45% of the total group of 305 patients did not report data or were not employed. As expected, the actual impact on employment is likely to be major, with the anticipated effect in terms of reliance on social services. Despite being in their early 30s, most of the patients in the entire group were single and only a quarter of the women reported having had children. In those adults diagnosed before 13 years of age, approximately 40% remain dependent with respect to their living arrangements. Finally, the fact that 70 to 80% reported denial of health insurance coverage because of their diagnosis may reflect deeper problems in healthcare access for patients with chronic disorders.

Our study has a number of limitations, including selection bias. Because the primary registry is not population-based and the patients are self-selected, our findings may not be generalizable to all individuals with childhood hydrocephalus. Whether our sample represents a more or less severely affected group of individuals with childhood hydrocephalus is unknown. It is possible that patients with severe neurological impairment would be less likely to participate in the survey. On the other hand, patients with normal outcomes and no ongoing clinical problems may also be less likely to know about or participate in the registry. There are also educational and socioeconomic barriers to participating in such a study. As one example, 85% of this group of patients were Caucasian, compared with the US population, which is 74% Caucasian. Minority populations are clearly underrepresented.

The mortality rate attributed to hydrocephalus cannot be measured based on this database. Thus, we suspect that the clinical extremes of the disease may not be described in our study. The self-reported nature of the data has inherent problems with respect to validity; patient responses cannot be independently verified. Finally, the effect on patient outcome of an underlying condition (for example, neoplasm or trauma) that causes hydrocephalus is difficult to measure in this type of analysis. This should be the focus of future studies, because identification of specific diseases that are associated with worse outcomes would encourage greater attention to be paid to vulnerable populations early in childhood. Similarly, we cannot determine the relative contributions of coexisting diagnoses, such as spina bifida or prematurity, on an individual's functional ability. These other diagnoses will account for a share of the morbidity measured by a long-term functional end point.

The strengths of this study include the detailed information that is available for a large cohort of patients with childhood hydrocephalus. The patient-relevant features of the disease are likely to be included in the responses provided. The long follow-up period provides a rare opportunity to study the morbidity of treated childhood hydrocephalus, particularly in patients who are now adults.

Conclusions

The cumulative risk of shunt complications, which is probably a function of shunt revision rates, is high, and should be included in any discussion with the parent of an infant or child receiving a new CSF shunt. Our results clearly indicate that the individual and social impacts of this common condition need to be better defined using population-based longitudinal data and quantitative quality-of-life measures. This information is necessary before the long-term disease burden of childhood hydrocephalus can be accurately measured.

Disclosure

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