In developed countries, the incidence of congenital hydrocephalus is estimated at three to five cases per 1000 live births.9,31,34,50 The causes of congenital hydrocephalus can be divided into primary (idiopathic) or secondary (acquired) causes, with the majority being idiopathic in origin. Excluding secondary hydrocephalus and that associated with spina bifida, the incidence of primary congenital hydrocephalus has been estimated at 0.2 to 0.8 per 1000 births in several population-based studies.2,6,30,36,43 A 2:1 male predominance has been reported.14,50 The natural history of untreated, congenital hydrocephalus is progressive cognitive decline and early death, usually before the third decade of life.22–24,32 Improvements in surgical and diagnostic techniques have led to a perception that the long-term outcome for patients with congenital hydrocephalus has also improved. Few demographic or epidemiological data exist to support this perception.21,45

A clearly defined origin is identified in some cases of congenital hydrocephalus. Primary aqueductal stenosis accounts for approximately 5% of congenital hydrocephalus, whereas aqueductal stenosis secondary to neoplasm, infection, or hemorrhage accounts for another 5%.9 The coincident anatomical malformations frequently observed with idiopathic congenital hydrocephalus include Chiari malformation, Adams–Bicker syndrome, Dandy–Walker malformation, and others.6,36,41,44 In utero infection (for example, toxoplasmosis, cytomegalovirus, rubella, and bacterial infections), intraventricular hemorrhage, and congenital brain tumors account for other causes.9,32,35 Hydrocephalus occurs in approximately 80 to 90% of patients with myelomeningocele.34,38,45 Of these cases, 50% are obvious at birth, and although congenital by definition, they are usually considered as a separate entity.

The standard treatment for congenital hydrocephalus is
the placement of a diversionary CSF shunt. Advances in surgical techniques and shunt materials have reduced the risk of the initial surgical procedure, although long-term risks include repeated shunt malfunction, shunt infection, and death associated with shunt failure. Several studies have demonstrated good outcomes in children undergoing CSF shunt placement procedures for congenital hydrocephalus at single institutions. To understand better the demographics and time trends of death from congenital hydrocephalus, we used death certificate data from the NCHS to perform a population-based evaluation of deaths attributed to hydrocephalus in US children over a 20-year period.

Clinical Material and Methods

Mortality databases of the NCHS were searched to determine the death rate from childhood hydrocephalus from 1979 to 1998 (http://wonder.cdc.gov/mortICD9J.html). These databases contain information on the primary cause of death as listed on death certificates in the US. Deaths from hydrocephalus were identified using the following ICD-9 codes: for congenital hydrocephalus, 742.3 (including aqueductal stenosis and idiopathic hydrocephalus); for spina bifida with hydrocephalus, 741.0; and for unspecified communicating and obstructive hydrocephalus, 331.3 and 331.4, respectively. In our analysis, communicating and obstructive hydrocephalus coded in this manner are defined as “acquired” because coding guides specifically exclude congenital hydrocephalus from the definition of these two codes. For comparison, spina bifida without hydrocephalus (coded 741.9) was also queried. Deaths from hydrocephalus related to congenital toxoplasmosis (771.2), syphilis (090.49), and tuberculosis (013.8) have separate ICD-9 codes and were excluded from the analysis.

Our definition of “childhood” included the neonatal period (the 1st month of life) to an age of 20 years. Individuals were classified into one of four ethnic groups according to the 1977 ethnicity classification system of the NCHS: white, black, American Indian or Alaska Native, and Asian or Pacific Islander. The latter two categories were grouped together as “other” by the NCHS. Persons of Hispanic origin were considered to be “of any race” by this classification system; therefore, white Hispanics were classified as white and black Hispanics classified as black.

Death rates for infants (defined as < 12 months of age) were calculated as the number of deaths per 100,000 live births. Death rates for all other ages were calculated as the number of deaths per 100,000 person years in that age group. Age-, race-, and sex-specific population data were obtained from the US Census Bureau. Person-years were defined as the average number of persons in a category multiplied by the number of years of study.

Death rates were age-adjusted using the direct method, whereby a standard age distribution was chosen and the age-specific death rates were weighted according to the standard. The NCHS adopted the Year 2000 projected population of the US as the standard population. This new standard replaces the 1940 standard used previously by the NCHS.

To test the significance of the comparisons of two mortality rates, exact binomial confidence intervals were calculated using a described method of interval estimation for incidence rate data. Rates of decline were defined as the percentage change between observations in 1979 and observations in 1998. STATA (version 7.0; Stata Corp., College Station, TX) was used for statistical calculations.

Results

A total of 1,457,848,237 person-years were included in the study; the sex and racial distributions of the population are shown in Table 1. We identified a total of 10,406 deaths attributed to childhood hydrocephalus in the US from 1979 to 1998, yielding an overall mortality of 0.71 per 100,000 person-years. Congenital hydrocephalus accounted for 57.7% of these deaths, whereas acquired hydrocephalus and spina bifida with hydrocephalus accounted for 23.2 and 19.1%, respectively.

Death from childhood hydrocephalus is highest for infants (1–12 months of age; Fig. 1). Although infants represent only 5% of the total population of children recorded, they accounted for 57.2% of all deaths. Congenital hydrocephalus accounted for the majority of the deaths (Table 2). Age-adjusted mortality rates for black infants were
Deaths caused by congenital hydrocephalus in children

higher for congenital and acquired hydrocephalus than for white infants (Fig. 2). Black infants have a statistically significant higher relative risk of death than do white infants for congenital and acquired hydrocephalus (Table 3). Deaths due to spina bifida with hydrocephalus did not differ according to race. No gender differences in mortality rates were observed.

Between 1979 and 1998, mortality rates from hydrocephalus in all children declined 60%, from 1 to 0.4 per 100,000 person-years. For infants during the same time period, mortality rates declined 74%, from 16.1 to 4.2 per 100,000 live births (Fig. 3). A reduction in deaths from each subtype of infant hydrocephalus was observed. For congenital hydrocephalus, mortality rates declined from 8.9 to 3.1 per 100,000 live births, representing a 66% reduction. For spina bifida with hydrocephalus, mortality rates declined from 4.9 to 0.6 per 100,000 live births, representing an 88% reduction. For acquired hydrocephalus, mortality rates declined 2.3 to 0.5 per 100,000 live births, representing a 78% reduction.

Mortality rates in adults for communicating and obstructive hydrocephalus were also compared with those for childhood hydrocephalus. There were 8121 deaths from communicating and obstructive hydrocephalus in adults from 1979 to 1998. No age-adjusted reduction in mortality rates occurred for all age groups during this 20-year period; in fact, mortality rates from hydrocephalus in adults older than 65 years of age actually have increased 30% from 1 to 1.3 deaths per 100,000 person-years (data not shown).

Discussion

An estimated 15,000 cases of childhood hydrocephalus occur annually in developed countries.34 The treatment of choice for virtually all of these cases is CSF shunt treatment and revision surgery is commonplace. The management of hydrocephalus and hydrocephalus-related complications in children is perhaps the largest segment of a pediatric neurosurgical practice. Since the earliest CSF shunts placed in the 1950s, improvements in surgical technique and shunt materials have led to more durable success and have reduced rates of infection and malfunction.32,39,51 Abundant literature exists on the surgical morbidity rates and infectious complications of childhood hydrocephalus,3,7,16,17,20,21,25,32,37,45–47 but information regarding changes in mortality rates, demographics, and patterns are sparse.

The natural history of untreated hydrocephalus involves either early death or poor intellectual development in survivors. In 1962, Laurence and Coates22,24 reviewed 182 patients with infantile hydrocephalus and found that nearly 66% of untreated patients died by 18 months, and 80% died by 20 to 25 years of age. Only 46% of untreated patients survived infancy, and only 38% had normal intelligence in childhood. Mortality rates in untreated patients with hydrocephalus and myelomeningocele were even higher, reaching 85 to 96%, in contrast to 35% in treated groups.13 Even in light of conditions such as pulmonary dysplasia and infection, these high death rates prompted aggressive efforts toward early treatment of childhood hydrocephalus. Early reports clearly demonstrated improved mortality and morbidity rates in the pediatric population after CSF shunt treatment.7,38 In a review of 454 patients, only 9% of children untreated for hydrocephalus survived to age 10 years, whereas 60% survived in the group that received shunts.38 In this same group, all patients surviving with hydrocephalus and myelodysplasia were mentally disabled, whereas 40 to 60% of children with shunts had normal intellects.38

Shunt procedures are not curative, nor are they free of significant risks. Death rates after shunt placement ranged from 7.5 to 35% in several early reports from the 1970s.18,28,38 More recently, a study of 357 patients showed that the likelihood of death from hydrocephalus-related causes 15 months after the first shunt placement was ap-

![FIG. 2. Bar graph showing age-adjusted mortality from hydrocephalus subtypes for children in the US younger than 1 year of age by race from 1979 to 1998. Black bars indicate black children; white bars, white children.](image)
proximately 3% for all patients. Patients with myelomeningocele had a slightly higher risk, at 7%, whereas premature infants with posthemorrhagic hydrocephalus had a slightly lower risk, at 2%. A retrospective cohort study of 155 patients with shunts demonstrated an overall 10-year mortality rate of 10% and a 10-year shunt-infection risk of 12%. Age is a risk factor for development of a shunt infection, with almost twice the shunt infection rate for infants younger than 6 months of age. Although fundamental pathophysiological differences exist between different types of congenital hydrocephalus, its origin has only loosely been associated with differences in outcome.

On the basis of data from the NCHS, we found that mortality related to childhood hydrocephalus declined between 1979 and 1998. The change is most striking for congenital hydrocephalus compared with hydrocephalus related to spina bifida and with acquired hydrocephalus. Improvements in pediatric emergency care, widespread availability of computerized tomography scanners, rapid transport, access to pediatric critical care units, and improved shunt technology all may play a part in the observed change. It is important to note that randomized studies have not shown an improvement in shunt failure rates in relation to the type of shunt used. Newer surgical techniques, such as endoscopic-guided catheter placement, do not affect long-term shunt survival. Improved prenatal screening may lead to earlier detection of fetal hydrocephalus and to therapeutic abortions, but these data are generally not recorded.

Congenital hydrocephalus does not demonstrate any racial predilection, but spina bifida occurs less frequently in blacks compared with other ethnicities. We found that in congenital and acquired hydrocephalus, mortality rates for black infants were higher compared with those for white infants and those of other ethnicities. The relative risk of death for black infants compared with that for white infants with congenital and acquired hydrocephalus was statistically higher for each year between 1979 and 1998. Factors leading to this racial discrepancy in mortality are difficult to determine, but they could arise from differences in access to both pre- and postnatal health care and from higher rates of preterm births as well as the lower birth rate among black infants. On the other hand, deaths from spina bifida with and without hydrocephalus did not differ with race and in fact may have been slightly lower among black infants. A racial difference in population-based mortality rates could either derive from a parallel difference in case fatality (black children with spina bifida and hydrocephalus are less likely to die than are white children with these conditions) or from incidence (black children are less likely to have spina bifida than are white children; therefore, they are less likely to die of it). Although one population-based study in 1990 showed that the incidence of spina bifida is lower in blacks than in whites, how black infants with spina bifida might have lower mortality rates in comparison with white infants is difficult to explain.

The strength of our study lies in its large, population-based numbers and in the statistical power it confers. The database includes all children and adults in the US and derives from death certificate data collected by the NCHS. A population-based study of hydrocephalus in children has not been previously reported, and few databases exist to attempt such an investigation. The clinical and radiological features distinguishing congenital, obstructive, and spina bifida–related hydrocephalus are easily identifiable and were well established by 1979. In addition, hydrocephalus would have been a diagnosis easily recognized in 1979 and onward, having been made usually after an imaging study (an ultrasonography procedure or a computerized tomography scan) or when the clinical diagnosis was clear (as in a massive macroencephaly procedure). If reasonable reliability in an accurate diagnosis of the cause of death can be attributed, then the true mortality rate attributed to hydrocephalus is potentially underestimated in the NCHS database.

Our study has several limitations. Any such study using administrative databases is inherently limited because of the imperfect sensitivity and specificity of diagnostic coding. We recognize that death certificate ICD-9 coding is often fraught with inaccuracies and false diagnoses. Although misclassification bias is a possibility, we expect such a bias to be nondifferential: errors in ICD coding likely occur systematically throughout the entire database as well as throughout the time period of the study. Although absolute mortality rates may not be accurate, therefore, comparisons and time trends within the study are likely to be meaningful; if anything, a nondifferential classification bias would be expected to bias the results toward the null. On the other hand, evidence exists demonstrating the accuracy of death certificate data for subsets of diagnoses. Death certificate diagnosis of stroke was recently shown to have high specificity, with high positive predictive values. Authors of another study reported that the sensitivity of death certificate diagnosis accuracy was highest for neurological diseases, at 0.9, compared with 0.64 for cardiovascular causes of death.

Another limitation of the NCHS database is the absence of information regarding age at diagnosis, age at death, treatment undertaken, and age at treatment. These considerations have obvious impacts on incidence, prevalence, case fatality, and survival that can cause disparities in mortality rates and make interpretation of our data imperfect and difficult. Nevertheless, these limitations underscore the need for better population-based data collection. Compiling
a national database of statistics for both childhood and adult hydrocephalus would be an extremely powerful tool in studying this common neurosurgical problem.

Conclusions

Although our study suggests that modern treatment of congenital hydrocephalus has resulted in declining mortality rates for children in the US, no definitive statement is possible in the absence of prospective mortality data controlled for rates and reasons of abortion and for accurate diagnosis of cause of death. Nevertheless, our results are compelling enough to recognize that congenital hydrocephalus is a treatable and survivable disease, more so than two decades ago. Moreover, the difference in mortality rates between the types of hydrocephalus analyzed suggests that fundamental differences may exist in the pathophysiology and biology of hydrocephalus and that these differences may actually affect clinical outcomes.

References


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