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Ventricular Shunt Survival in Children with Neural Tube Defects

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Summary

Ventricular shunting has dramatically improved the care of children with hydrocephalus. Yet shunt malfunctions are extremely common and cause significant morbidity. To document shunt problems in children with a neural tube defect and hydrocephalus, 67 children born since 1973 were studied via life-table analysis. 28% of the shunts failed within the first 6 months after insertion, 37% failed within the first year and 50% failed by 4½ years after insertion. Shunt survival was similar in children whether or not they had previous shunt failures. The brand of shunt system and pressure rating, the level of neurological function, the interval between closure of the neural tube lesion and shunt insertion, and head circumference percentile at the time of insertion were also not correlated with shunt failure. However, shunts inserted in the first year of life were much more likely to fail than those inserted after one year of age ($p < 0.05$). 68% of the revisions required replacement of the ventricular catheter. Shunt failure from all causes of hydrocephalus accounted for approximately 1% of paediatric admissions to Strong Memorial Hospital in 1982 with a mean cost of $ 4,543 and a mean length of stay of 9 days. Thus shunt problems remain both common and serious.

Although ventricular shunting has revolutionized the care of children with hydrocephalus, problems with shunts are extremely common. Shunt malfunctions can cause serious morbidity and mortality and are associated with loss of function and long, expensive hospital stays. Numerous studies have evaluated shunt complications[1,2,6—8,10—15,20—22]. Yet, most have grouped children with hydrocephalus together, regardless of aetiology. In addition, none has used the formal method of life-table analysis, which allows the comparison of cases followed for different lengths of time and allows statistical comparison between different subgroups. The following study was therefore undertaken to evaluate shunt dysfunction in one aetiologic group of children using the life-table method of analysis.

Methods

Children born since 1973 with neural tube defects (meningomyelocele and meningocele) and shunted hydrocephalus were identified from the records of Strong Memorial Hospital (SMH) and the Birth Defects Center (BDC) of the Department of Pediatrics at the University of Rochester School of Medicine and Dentistry (the regional referral centre for children with neural tube defects). Medical records from SMH, BDC, and private physicians were reviewed and information from them recorded. Two patients were excluded from the study because of incomplete information. Data on 67 children, who had 122 operative procedures for hydrocephalus, were analyzed using the computer facilities at the University of Rochester. SURVIVAL, the life-table analysis program of the Statistical Package for the Social Sciences[9] was used. In addition, information about hospital admissions and cost for all children with hydrocephalus were obtained from the computerized SMH clinical and financial profile statistics for 1982. Since 1978 maximum intervention has been provided to all children with neural tube defects regardless of the level of involvement. Five children with hydrocephalus were treated with acetazolamide; none were treated for longer than six weeks, and in none of these cases did the medical treatment obviate the need for surgery.

Results

62 children were diagnosed as having meningomyelocele while five had meningocele. Of the children with meningomyelocele the most caudal level of neurological function was thoracic in 17%, first to third lumbar level (L 1–L 3) in 39%, L 4–L 5 in 29%, and sacral in 15%. The median age at initial shunt insertion was 14.5 days (range 1–333). In 15% of the children the shunt was inserted prior to closure of the neural tube lesion. In 13% the shunt was inserted within a week after lesion closure, in 43% it was inserted after 7 but before 31 days had elapsed and in 29% the shunt was inserted more than 30 days after closure of the lesion.

The 67 children were followed for a mean of four years, or 268 "shunt years". During this time 55 revisions were performed. Thirty-three patients (49%) needed no revision, 20 (30%) required 1 revision, 9 (13%) had 2 revisions, 4 (6%) had 3 revisions, and 2 (3%) needed 4 revisions. Three of the 122 shunt insertions were ventriculo-atrial, the other 119 were ventriculo-peritoneal. For those patients for whom the manufacturer could be identified, 82% of the insertions were with a Pudenz valve, 7% were with a Hakim, 11% were with others (including Heyer Shulte and Holter). Fifty-four percent of the valves were medium pressure, 13% were low pressure, and for 33% the pressure was unknown. At the time of initial shunt insertion 59% of the children had a head circumference-for-weight percentile[23] of greater than 95. None of the children fell into the below 5th percentile category for head circumference. There were no deaths during the study period.

At the time of revision 15% of the children showed symptoms and signs of infection, 54% had symptoms and signs of increased intracranial pressure without infection (headache, lethargy, vomiting, weakness of lateral gaze, etc.), 16% had a rapid increase in head circumference only without symptoms, 10% had swelling around the shunt valve without symptoms, 2% had an abnormal computerized axial tomogram of the head (performed routinely once a year), 1% had elective revision, and 2% had miscellaneous reasons for revision such as the valve not pumping, and skin breakdown over the valve.

In 35% of the revisions the entire shunt system (ventricular catheter, valve and peritoneal/atrial catheter) was replaced. In 26% the abdominal catheter alone was replaced. In 6% the valve alone was replaced; in 11% the valve and ventricular catheter were replaced; and in 22% the ventricular catheter alone was replaced.

A review of the computerized hospital financial data for 1982 revealed that 0.8% (29/3539) of all paediatric admissions to SMH (excluding newborns) were for shunt revisions. The mean length of stay was nine days, with a mean hospital charge (excluding physicians' fees) of $4,453.

In order to obtain more uniformity and generalizability, the three ventriculo-atrial shunts were eliminated from the remainder of the analysis. The survival rate of the patients in whom 119 ventriculo-peritoneal shunts were inserted during this study is shown in Fig. 1. 28% of the shunts failed within the first six months after insertion, 37% failed within the first year and 50% failed by 4½ years after insertion.

To examine whether the number of prior shunt insertions affected survival of the shunt, the children were divided into five groups based on number of revisions—from 0 to 4—and compared using the life-table method of analysis[5]. Unlike an illness such as

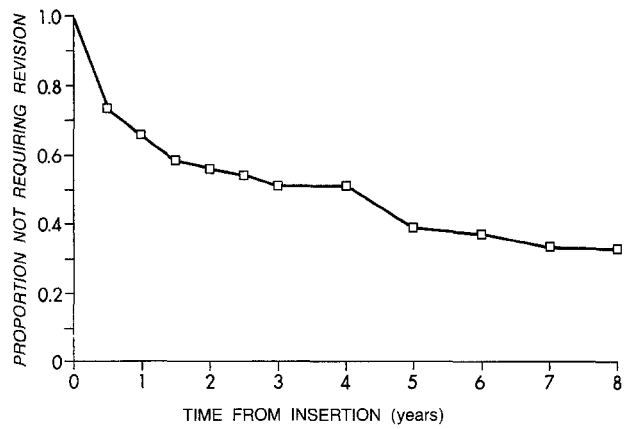

Fig. 1. Survival of 119 ventriculo-peritoneal shunts in children with neural tube defects

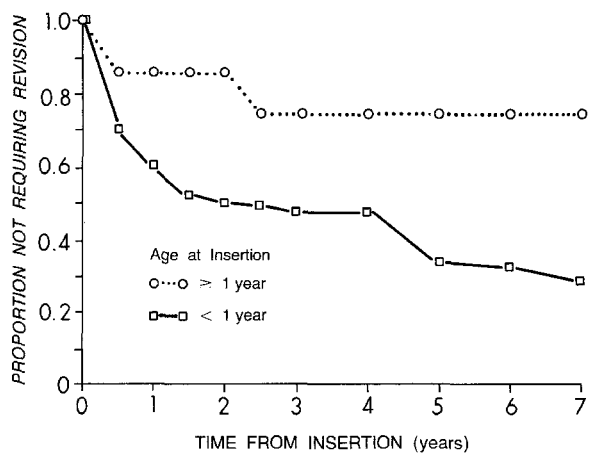

Fig. 2. Comparison of survival between ventriculo-peritoneal shunts inserted ≤ 1 year and those inserted > 1 year of age in children with neural tube defects

sickle cell anaemia, where the probability of having a sickle crisis increases significantly with the number of previous crises, the risk of shunt failure in this sample was slightly decreased in those children with previous revisions. However, these differences were not statistically significant ($p > 0.10$).

The age of the child at the time of shunt insertion was examined next. As shown in Fig. 2, shunts inserted in the first year of life were much more likely to fail than

those inserted after one year of age ($p < 0.05$). For shunts inserted in the first year of life, the failure rate was similar whether the shunt was placed in the first month, first six months or the last six months. (None of the patients in this study had their initial shunt inserted after one year of age.)

The brand of shunt (Hakim, Pudenz, other), and shunt pressure (low, medium) were not correlated with the risk of failure. The interval between closure of the lesion and shunt insertion, the level of neurological function and the head circumference percentile at the time of insertion likewise were not correlated with shunt failure.

Discussion

The findings of this study show reasonable shunt survival when contrasted with those few studies with whom they can be compared. For example, Lorber *et al.*[16] reported that his patients required 87 surgical revisions in a total of 251 shunt years, or 1 revision every 2.9 shunt years. In the current study, 55 revisions were required during 268 shunt years for an average of 1 revision per 4.8 years. Unfortunately, examination by revisions-per-shunt-year does not take into consideration the duration of follow-up or the probability of revision recurring in the same patient[5].

Several studies have published data that allow more precise comparisons. In the current study performed on children from 1973 to 1983, 72% of the shunts were functioning six months after insertion. Prior studies examining ventriculo-peritoneal shunts (with dates of study in parentheses) found six-month survival rates of 40%[22] (1970–1976) and 66%[6] (1975–1979). Six-month survival rates for ventriculo-atrial shunts have been reported to be 48% (1960–1978)[1] and 68% (1970–1976)[12]. Two year shunt survival in this study was 57%, a figure that compares favourably with the 11%[22] found for ventriculo-peritoneal shunt, and the 32%[12] and 34%[22] rates for ventriculo-atrial shunts.

Unfortunately the results of this study cannot be compared with most other evaluations of shunt function that have been published since they did not employ a life-table method of analysis. The studies that provided complication rates alone cannot be used for comparison since they do not include factors that affect risk such as the number of years of follow-up per patient or age of the patients studied. Similarly, the studies cited above that did compute their results in terms of risk by duration, included children with hydrocephalus from all aetiologies. Since children with hydrocephalus do not constitute a homogeneous group, comparing groups of children that have different distributions of types of hydrocephalus may not be valid.

The reason for the poorer survival rate of shunts inserted before 12 months of age is unclear. Children less than one year have smaller ventricles and have a much faster rate of linear growth. However, shunts inserted after one month of age or even after six months had survival rates similar to those inserted earlier. Also, head circumference percentile at the time of insertion (a reflection of ventricular size) was not correlated with survival. It is possible that the younger child's decreased activity and/or smaller proportion of time spent in an upright position affected shunt survival. As Portnoy[21] has demonstrated, the upright position alters ventricular pressure and cerebrospinal fluid flow through most valves used in ventriculo-peritoneal shunt systems. The increased activity and time in the upright position of infants over twelve months of age may prevent obstruction of the catheters and valves by means of increased flow and increased movement of the catheter. Although the production, flow and composition of cerebrospinal fluid changes from birth to 12 months[3,25], the effect of these changes on shunt function is unclear.

Surprisingly, unlike many disease processes, the occurrence of a shunt failure does not increase the risk for subsequent shunt failures. This may be related to the fact that shunt revisions are usually more limited procedures than the original insertions. It has been argued that because ventricular shunt insertion reverses cerebrospinal fluid flow and allows fluid from the lumbar region to travel to the ventricles, shunt insertion performed within a week of closure of the lesion would increase the risk or the infection. Such increased failure was not demonstrated in the current study. Although different brands of shunts may have advantages in ease of insertion of ability to detect subsequent failure, there were no differences in shunt failure rates for the various brands of valves examined. Nor were there differences in shunt survival for the different pressure ratings or for level of neurologic function. These findings must be interpreted with caution, however, since neither the timing of insertion nor the type of device used was randomized.

Although the shunt survival in this study is better than the survivals reported in earlier studies, it is still unacceptable. The external shunting of cerebrospinal fluid from the ventricles has improved markedly since Holter began producing his valves in his home

workshop[24]. Yet, in spite of this progress shunts are responsible for major morbidity and occasional mortality in children with hydrocephalus. In the current study 68% of the revisions included replacement of the ventricular catheter, a procedure involving exposure and trauma to the brain. The mean hospital cost for shunt revision (not including physician's fees) in 1982 was $ 4,543 with a mean length of stay of nine days. This does not include the discomfort and loss of function that occur before and after hospital stay, nor does it address the acute and chronic parental anxiety caused by shunts.

If one assumes that the incidence of neural tube defects (excluding anencephaly) is 6 per 10,000 live births[26], that 80% of these children will require shunts for hydrocephalus[17], that there are 3,700,000 live births in the United States annually[19], and that the survival and cost data from this study accurately reflect survival and cost for the U.S.; then $ 6,764,000 will be spent in hospital costs annually by children with spina bifida from birth to eight years of age. This does not include older children who will also require shunt revisions. If the incidence of hydrocephalus from *all* causes is 3 per 1,000 live births[18], the financial impact of this problem is even more compelling.

Because shunt failure is an important problem, efforts should be directed at improvements in the techniques used to manage hydrocephalus. In order to document improvement, however, data analysis should be performed using methods such as life-table analysis that allow comparison of different treatment regimens. Similarly comparative studies should limit their scope to children with distinct forms of hydrocephalus, even if this requires collaboration with other institutions to achieve adequate patient numbers. Finally, because studies such as this and the ones reviewed have failed to demonstrate clear differences among the various devices used, it seems prudent that future studies of new regimens should be performed using randomization and unbiased observations.

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