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Cerebrospinal Fluid Shunt Survival and Etiology of Failures: A Seven-Year Institutional Experience

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Key Words
Hydrocephalus · Ventriculoperitoneal shunt · Shunt failure

Abstract

Background: Innovations in shunt technology and neuroendoscopy have been increasingly applied to shunt management. However, the relative life span of shunts and the etiology of shunt failure have not been characterized recently. ***Methods:*** We reviewed the records of all shunting procedures at our institution between January 1992 and December 1998. Independent predictors of shunt failure were analyzed via multivariate Cox regression analysis in 836 shunting procedures. Independent predictors of the etiology of failure (infection, proximal obstruction, distal malfunction) were analyzed via multivariate logistic regression analysis in the 383 shunts which failed. ***Results:*** A total of 353 pediatric patients underwent 308 shunt placements and 528 revisions. The risk (hazard ratio; HR) of shunt failure decreased as a function of time in both primary placements and revised shunts. In failed shunts, the odds of infection decreased 4-fold per year of shunt function, while the odds of distal malfunction increased 1.45-fold per year. Increasing number of shunt revisions (HR 1.31, $p < 0.05$), decreasing patient age in years (HR 1.04, $p < 0.001$), gestational age <40 weeks (HR 2.15, $p < 0.001$) but not the etiology of hydrocephalus were associated with an increased risk of shunt failure. Revisions versus primary placements, Dandy-Walker cysts and gestational age <40 weeks were independently associated with proximal, distal and infectious causes of failure, respectively. ***Conclusions:*** The long-term shunt revision rates observed here are similar to those reported over the past 2 decades. Shunt life span remains poorer in shunt revisions and in younger patients. Patient characteristics may suggest a specific risk and mechanism of failure, aiding in the long-term management of shunted hydrocephalus.

Introduction

Hydrocephalus is a common condition of childhood that is associated with many diseases. These include a variety of congenital anomalies, head injury, brain tumors, meningitis and brain hemorrhage [1]. In the United States, hydrocephalus accounts for 70,000 hospital admissions yearly [2]. As a result, approximately 18,000 CSF

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shunts are placed per year, costing USD 100 million annually [2]. Despite the large number of affected patients, advancement in the management of hydrocephalus has remained slow.

Nulsen and Spitz [3] described the first workable CSF shunt in 1952. Ascendance of the peritoneal cavity as a receptacle for diverted CSF and the emergence of silicone as a biomaterial for shunt fabrication remained the predominant advancements in shunt technology for many decades [4–7]. Nonetheless, rates of shunt failure and underlying causes of CSF shunt failure were extensively characterized [4, 7–15]. In the past decade, ultrasound, stereotactic and neuroendoscopic techniques have been increasingly utilized in an attempt to decrease shunt failure rates [16–21]. Since these innovations, few institutional descriptions of shunt survival have subsequently been reported [1, 22].

We conducted a retrospective study of 836 shunting procedures to analyze both shunt survival and the etiology of shunt failure as a function of time, patient age, etiology of hydrocephalus and shunt revision number.

Patients and Methods

We reviewed the records of all pediatric patients who had undergone ventriculoperitoneal procedures for hydrocephalus between January 1992 and December 15, 1998. Standard follow-up consisted of clinical examination at 1 week, 1 month, 3 months and yearly thereafter. Interval head CT scans were performed on a yearly basis following shunt placement in all patients unless they were clinically symptomatic. Information on patient age and gender, etiology of hydrocephalus, date of shunt placement, new placement versus revision, use of endoscope, valve type, date of shunt failure, date of last follow-up and cause of shunt failure was collected retrospectively from patient records. All shunts were placed by one of two fellowship-trained, full-time pediatric neurosurgeons.

This study focused on the functional life of the individual shunts placed. During the observation period, all shunts were designated as meeting only one of two endpoints: (1) shunt failure requiring revision, or (2) no shunt failure at the last follow-up examination. Infection necessitating shunt removal or shunt malfunction leading to radiological or symptomatic progression of hydrocephalus constituted shunt failure. The etiology of failure was characterized as infection, valve malfunction, proximal catheter obstruction or distal catheter malfunction (obstruction or disconnection). Death of the patient was considered shunt failure only if shunt failure was the immediate cause of death. Deceased patients were otherwise included as lost to follow-up, with the last clinical examination being the last follow-up time.

Proportional hazards regression models were created to model shunt failure hazard on factors related to patient, diagnosis and surgery characteristics. Since our unit of analysis was the surgery (as opposed to the patient), it was necessary to extend the Cox model to account for the fact that individual patients may receive multiple surgeries if they experience multiple shunt failures [23]. Conditional risk set techniques described by Prentice et al. [24] and Cleves [25] and employed by Tuli et al. [26] were used for proportional hazards modeling. Stata procedures stset and stcox were used for conditional regression modeling [27]. Shunts were assigned a level according to the order of a placement versus revision surgery. The influence of the underlying diagnosis on shunt failure was assessed using indicator variables. Each resultant model was clustered on shunt order.

The independent association of patient and operative variables to the underlying etiology of shunt failure was analyzed using multivariate logistic regression models (SAS logistic procedure [28]) in the 383 failed shunts assessed during the observational period. Individual causes of failure were modeled separately with shunt order, patient age, gender, use of scope and underlying diagnosis as predictors. Odds ratios with 95% confidence intervals were obtained from these models.

Results

Patient and Shunt Data

A total of 353 patients underwent 836 shunt placements or revisions (2.37 procedures per patient) between January 1992 and December 1998 (table 1). Twelve patients died during the follow-up period (1.4%) from causes related to sequelae of shunt failure. At the end of the follow-up period, 445 shunts (53%) had failed. For the 391 remaining functional shunts, the mean follow-up was 3.2 years (range 1.0–7.5 years).

Shunt Survival and Etiology of Failure as a Function of Time

The risk (hazard) of shunt failure decreased as a function of time from placement in both newly placed and revised shunts (table 2). A 14% failure rate occurred within the first month, while a further failure rate of only 5% was observed beyond 4 years for all shunts. In failed shunts evaluated at our institution (n = 383), each increasing year of adequate shunt function independently decreased the odds that failure was attributed to infection 4-fold (odds ratio 0.26) and increased the odds that failure was attributed to distal malfunction 1.45-fold (table 3). Proximal obstruction was not associated with the duration of shunt function prior to failure. Valve malfunction contributed to 4–6% of failures of both newly placed and revised shunts and did not differ as a function of time.

Effect of Patient Age on Shunt Failure

Each increasing year of patient age was associated with a 4% [hazard ratio (HR) 0.96] decrease in the risk of shunt failure (table 4). Premature neonates demonstrated the greatest risk (HR 2.15) of shunt failure (fig. 1). Patient age in years was not an independent predictor of etiology of

Table 1. Patient, surgery and shunt characteristics of patients undergoing ventriculoperitoneal shunting

Characteristic	All shunts	Placement	Revision
Number of patients	353	193	160
Male	182 (52%)	97 (50%)	85 (53%)
Female	171 (48%)	96 (50%)	75 (47%)
Number of procedures	836	308	528
Male	434 (52%)	161 (53%)	273 (52%)
Female	402 (48%)	147 (47%)	255 (48%)
Age, years (median, IQR)	6.9 (1.7–13.1)	2.1 (0.3–7.5)	7.5 (1.9–13.7)
Follow-up, years (mean ± SD)	3.2 ± 1.5	3.2 ± 1.4	3.2 ± 1.5
Diagnosis			
Myelodysplasia	261 (31%)	93 (30%)	168 (32%)
Congenital/idiopathic	191 (23%)	68 (22%)	123 (23%)
IVH	154 (18%)	59 (19%)	95 (18%)
Tumors	84 (10%)	37 (12%)	47 (9%)
Dandy-Walker/ aqueductal stenosis	49 (6%)	17 (6%)	32 (6%)
Postmeningitic	33 (4%)	13 (4%)	20 (4%)
Other	64 (8%)	21 (7%)	43 (8%)
Valve			
MS Delta	498 (60%)	198 (64%)	300 (57%)
Hakim	125 (15%)	44 (15%)	81 (16%)
Other	48 (3%)	14 (5%)	34 (6%)
Unknown	161 (19%)	49 (16%)	112 (21%)
Placement			
Frontal	186 (23%)	65 (21%)	121 (23%)
Occipital	644 (77%)	243 (79%)	401 (76%)
Endoscopic assisted	482 (58%)	184 (60%)	298 (57%)

IQR = Interquartile range.

shunt failure; however, gestational age <40 weeks increased the odds that failure was due to infection 2.05-fold (table 3). Gender was not a predictor of shunt survival or etiology of failure (table 3).

Shunt Survival according to the Etiology of Hydrocephalus

The etiology of hydrocephalus was not an independent predictor of shunt failure (table 4). In failed shunts evaluated at our institution, Dandy-Walker cyst shunts were independently associated with a 10-fold decrease (odds ratio 0.1) in the odds of proximal obstruction being the cause of failure (table 3). Otherwise, the etiology of hydrocephalus was not associated with a particular mechanism of shunt failure.

New Shunt Placement versus Revision

The risk of shunt failure increased by 31% (HR 1.31) for each subsequent shunt inserted per patient (table 4, fig. 2). In failed shunts evaluated at our institution, revised versus primary shunts were independently associated with a 1.56-fold increase in the odds of proximal obstruction and a 1.6-fold decrease in the odds of distal malfunction being the cause of failure (table 3).

Table 2. Hazard (risk) of shunt failure as a function of time from shunt placement

Interval of shunt function, years	First placement	First revision	Second revision
0–1	11.0 ± 1	14.1 ± 2	18.2 ± 2
1–2	3.5 ± 1	2.7 ± 1	3.4 ± 1
2–3	2.1 ± 1	2.3 ± 1	3.0 ± 1
3–4	2.2 ± 1	3.0 ± 2	6.1 ± 4
4–5	0.1 ± 0.1	0.1 ± 0.01	–

Results are hazard ± SE (E^{-4}).

Table 3. HRs and confidence intervals demonstrating associations between patient characteristics and mechanism of shunt failure

Variable	Infection	Proximal obstruction	Distal malfunction
Age[1]	0.98 (0.9–1.1)	0.99 (0.9–1.1)	0.98 (0.9–1.1)
Prematurity	2.05 (1.4–2.3)	0.65 (0.2–1.7)	0.97 (0.7–1.3)
Gender	1.11 (0.8–1.2)	1.0 (0.9–1.1)	0.91 (0.8–1.2)
Placement versus revision[2]	1.09 (0.6–1.9)	1.56 (1.1–2.5)	0.64 (0.4–0.9)
Time[3]	0.26 (0.1–0.5)	1.04 (0.8–1.3)	1.45 (1.2–1.8)
Etiology of hydrocephalus			
Myelodysplasia	1.18 (0.2–3.0)	0.99 (0.4–2.6)	0.72 (0.3–1.9)
Tumor	0.96 (0.1–1.2)	1.1 (0.3–3.5)	1.75 (0.5–5.6)
IVH	2.62 (0.4–2.9)	0.61 (0.2–1.7)	0.93 (0.3–2.6)
Congenital	0.46 (0.2–1.8)	0.66 (0.2–1.8)	0.98 (0.3–3.5)
Idiopathic	1.00 (0.1–1.5)	1.55 (0.4–5.4)	0.99 (0.3–3.5)
Dandy-Walker	0.86 (0.4–5.5)	0.10 (0.1–0.8)	1.52 (0.9–2.3)
Aqueductal stenosis	0.49 (0.2–3.1)	0.45 (0.1–1.6)	0.96 (0.3–3.4)

Values are HRs with 95% confidence intervals in parentheses.
1 Increasing age in years.
2 Revision associated with increased odds of proximal obstruction.
3 Increasing years from shunt placement.

Table 4. HRs, confidence intervals and probability values of shunt failure associated with various patient characteristics

Variable	HR	95% confidence interval	p value
Patient age[1]	0.96	0.94–0.97	<0.001
Gestation age <40 weeks	2.15	1.62–3.22	<0.001
Gender	1.02	0.77–1.17	0.678
Shunt number[2]	1.31	1.05–1.63	0.018
Etiology of hydrocephalus			
Myelodysplasia	1.21	0.67–2.18	0.535
Tumor	1.25	0.76–2.02	0.375
IVH	0.80	0.41–1.57	0.516
Congenital	0.71	0.39–1.30	0.263
Idiopathic	1.29	0.80–2.09	0.289
Dandy-Walker	1.02	0.64–1.63	0.935
Aqueductal stenosis	0.73	0.42–1.27	0.265

1 Increasing age (years) associated with increased shunt survival.
2 Increasing number associated with decreased shunt survival.

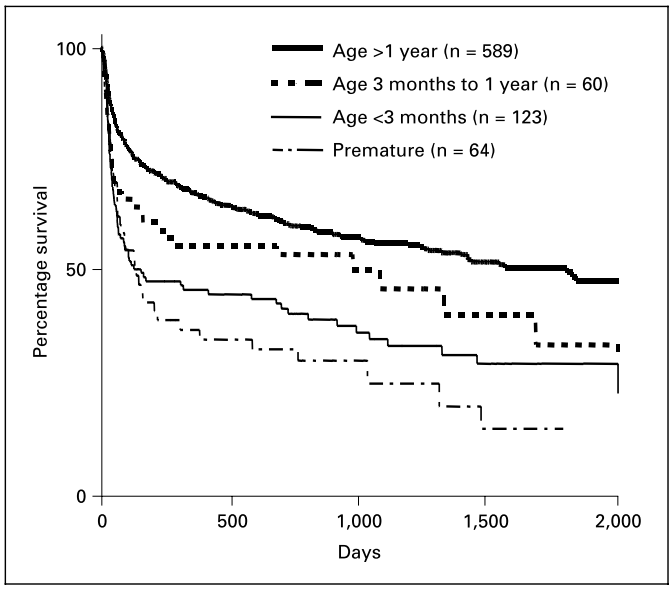

Fig. 1. Shunt survival curves categorized by patient age.

Discussion

We undertook a 7-year retrospective review of the ventriculoperitoneal shunt experience at the Duke University Medical Center. Both validating and countering published findings and 'common laws' regarding shunt survival, this study adds to the postneuroendoscopy literature on shunt management. Our aim was to attempt to identify predictors of shunt survival and to characterize how the etiology of shunt failure varied according to specific characteristics of the patient, disease and shunt procedure.

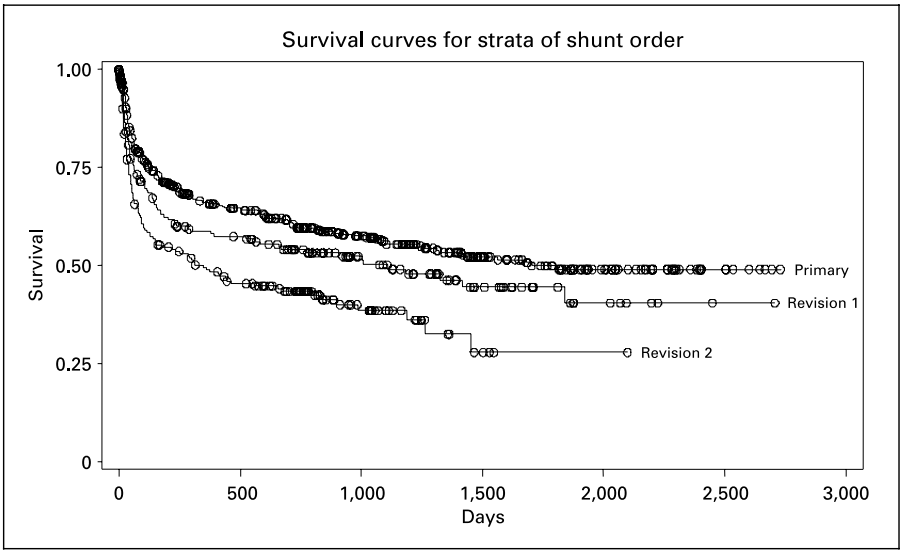

Fig. 2. Shunt survival curves for primary placement, primary revision and secondary or greater revision.

General Shunt Survival

Fifty-two percent of all newly placed or revised shunts in our review had failed by the 5-year mark. This is well within the published range in the current literature [9–12, 22, 26, 29]. We observed a 1.4% mortality rate over the study period, while recent reports suggest a mortality rate of up to 4% may be observed within 36 months in patients with shunted hydrocephalus [30]. Piatt and Carlson [11] report a median shunt survival time of 6 years, whereas 50% of shunts inserted by Liptak and McDonald [10] had failed by 23 months. Tuli et al. [26] report a 50% shunt survival at approximately 3 years in first-, second- and third-time placements, while Griebel et al. [9] state a 50% revision rate in a review spanning 7 years of experience. The probability of the occurrence of shunt malfunction in a combined review of 12 years of experience in Toronto and Paris was 81% [12]. More recently, in 1998, Lazareff et al. [22] reported that 40% of shunted patients required shunt revision, and only 35% required revision in a 5-year period in the report of Albright et al. [29] in 1999. Although revision rates appear to be lower in these more recent shunt series, possibly due to operative advancements, our long-term outcomes fail to demonstrate a decrease in shunt failure compared to earlier reports.

The notion that 'the longer a shunt functions, the less likely it is to fail' is confirmed both in our findings (table 2) and in the literature [10–12, 15, 29]. In our series, a 14% failure rate occurred in the first month, while a further failure rate of only 5% was observed beyond 2 years. It is worthwhile noting that at least two other studies have found that the interval between shunt revisions was a significant predictor of repeat failure [11, 26], and that Lazareff et al. [22] found that 'shunts that tended to fail repeatedly did so sooner than those that did not'. Major pediatric neurosurgical centers tend to have higher failure rates in the first 6 months followed by a lower rate over the next 2 years [31]. This becomes an important factor in shunt management, suggesting that shunts may need to be more closely followed within the first year after placement. Clinical follow-up may be less aggressive and progressively spaced as a shunt proves to be functional over time, reducing the cost of long-term management.

Etiology of Shunt Failure

In our series, the percentage of shunt failures attributed to distal catheter obstruction/malfunction increased as a function of time. The odds that shunt failure was attributed to distal malfunction increased 1.45-fold per year. Piatt [15] found that the incidence of distal malfunction increased proportionately among late failures. Inadequate distal catheter length resulting from child growth, a time-dependent process, may contribute, in part, to this time trend. Our findings support the literature and the common belief that distal malfunctions are less common in younger shunts. The percentage of shunt failures attributed to proximal catheter obstruction did not differ as a function of time. Proximal obstruction has been shown to contribute to a greater extent to early shunt failure than late failure [15]. It is unclear why proximal obstruction contributed equally to early and late failures in our series. Growth of the choroid plexus into the proximal catheter, which may be a time-dependent process, may explain this finding.

The percentage of shunt failures attributed to infection decreased as a function of time. Forty-five percent of shunt failures in the first month were attributed to infection, while only 6% of shunts failing after 2 years were attributed to infection. The odds that shunt failure was attributed to infection decreased 4-fold per year. Our overall infection rate approached 11%. Previously published rates ranged from 3 to 15%, and most report a steady drop-off after the first few months [1, 9, 11, 31–33]. An analysis of factors affecting infection rates is beyond the scope of this paper, which thus prevents direct comparison to other shunt series. Nevertheless, a significant decrease in the incidence of infection over time remains a consistent observation in the shunt literature. The relationship of shunt age to infection and distal catheter malfunction is valuable in the management of hydrocephalus and should influence clinical suspicion and the diagnostic protocol in the search for the etiology of shunt failure.

Patient Age

Our review revealed that patient age at the time of shunt implantation is an important predictor of shunt function and life span. In univariate analysis, premature infants demonstrated the shortest shunt survival, followed by the age groups of less than 3 months and less than 1 year at decreasing levels of statistical significance. Adjusting for the etiology of hydrocephalus, revision number and time from placement, a 4% decrease in the risk of failure was observed with each increasing year of age. Shunted premature neonates demonstrated the greatest risk of shunt failure. Piatt and Carlson [11] found an increased risk of failure in shunted patients less than 2 years of age compared with those older than 2 years. Shunt insertions in children less than 1 year old were associated with worse shunt survival in reviews by Liptak and McDonald [10] and Liptak et al. [34] regardless of the etiology of hydrocephalus. Implantation of a shunt in children aged under 40 weeks and then under 1 year was associated with worse outcome in the study of Tuli et al. [26], and nearly 50% of all failures occurred in shunted patients less than 1 month old in the report of DiRocco et al. [32].

Gestational age <40 weeks was independently associated with infection in this series, suggesting that infection may underlie the high rate of failure in these patients. Although a greater risk of shunt failure was associated with decreasing patient age in years, a particular etiology of failure was not associated with decreasing patient age. Piatt and Carlson [11] found significant differences in children under the age of 6 months, and multiple other studies have reported an association with younger age as well [1, 11, 35–38]. Premature infants have a predisposition to infections and complications in general [1]. Factors contributing to failure in premature children include a less developed immune system, an integument which is more likely to be in poorer condition and skin flora which may be denser and pathogenic in hospitalized patients [1, 35, 37].

Etiology of Hydrocephalus

The etiology of hydrocephalus was classified into seven common groups and was not associated with shunt survival in this series. This is consistent with the reports of Piatt and Carlson [11] and Griebel et al. [9], who also showed that the etiology of hydrocephalus had no significant influence on the risk of obstruction. Conversely, several reports have described increased shunt failure in the setting of intraventricular hemorrhage (IVH) [26, 32, 39–41]. Clot degeneration and subsequent proximal catheter obstruction has been described with IVH and suggested to be the reason for poor shunt survival in this setting [42]. Liptak and McDonald [10] found that patients with neural tube defects had longer shunt survival than in other etiologies, possibly due to altered CSF absorption or arrested hydrocephalus. Lazareff et al. [22] reported that tumor, IVH and meningitis were more frequent etiologies in patients who had 4 or more shunt revisions. Although Dandy-Walker cysts were not associated with altered shunt survival in this series, patients presenting with failed Dandy-Walker shunting were significantly less likely to have proximal obstruction. These findings suggest that overall follow-up may not need to be altered based on the etiology of hydrocephalus, and clinical suspicion of proximal obstruction in the setting of Dandy-Walker cysts may be relatively lowered.

New versus Revised Shunts

Failure rates increased in revised versus newly placed shunts. Tuli et al. [26] also demonstrated that shunt failure increases with an increasing number of shunt revisions. Piatt and Carlson [11] found that shunt revisions within the first 6 months have a higher chance of failure than newly placed shunts. However, Liptak et al. [34] found no difference in shunt survival between shunt revisions and new placements. Griebel et al. [9] reported no difference in the complication rate between new systems (51%) and replaced systems (62%). Our series suggests that all revised shunts may warrant more aggressive clinical follow-up given a greater risk of failure. A subgroup of patients, labeled as 'poor shunt patients', have been ob-

served to incur an extraordinary number of revisions in a lifetime, carrying an even greater risk of failure per revision number. These patients warrant more frequent clinical follow-up and may be targeted by high numbers of revisions or a short duration between shunt placement and subsequent failure [22, 26].

Failed shunt revisions versus primary placements were independently associated with significantly greater odds of proximal obstruction and decreased odds of distal malfunction. This suggests that proximal catheter pathology may underlie the high risk of failure observed in shunt revisions. Slit ventricles or CSF protein content may underlie this proximal component of revised shunt failure. Nevertheless, a controlled trial is necessary to explore the relationship between proximal obstruction and shunt revisions. Our results suggest that clinical suspicion should favor proximal obstruction in the workup of revisions of failed shunts.

Conclusions

In this retrospective review of our 7-year experience with ventriculoperitoneal CSF shunts, the high rate and etiologies of shunt failure were similar to past decades. Patient age in years, prematurity and shunt revision number were independent predictors of shunt survival, while the etiology of hydrocephalus was not associated with shunt survival. Prematurity, Dandy-Walker cysts, revision versus primary shunt placement and time from shunt placement were each independently associated with a particular etiology of shunt failure. This observational series suggests that patient characteristics may serve the neurosurgeon to predict the risk of shunt failure and the odds of specific mechanisms of failure. Management of shunted hydrocephalus patients may be improved by adjusting for a variety of patient characteristics.

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