30th YEAR ANNIVERSARY SUPPLEMENT

THE SURGICAL MANAGEMENT OF PEDIATRIC HYDROCEPHALUS

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ONLINE DIGITAL VIDEO

THE SURGICAL MANAGEMENT of hydrocephalus has undergone incredible changes over the past generation of neurosurgeons, including dramatic improvements in imaging, especially computed tomographic scanning and magnetic resonance imaging, and remarkably innovative advances in cerebrospinal fluid valve technology, complex computer models, and endoscopic equipment and techniques. In terms of overall patient outcomes, however, one could conclude that things are a little better, but "not much." This frustrating yet fascinating dichotomy between technological advancements and clinical outcomes makes hydrocephalus, first described by the ancients, as one of the most understated and complex disorders that neurosurgeons treat. The challenge to the next generation of neurosurgeons is to solve this vexing problem through better understanding of the basic science, improved computer models, additional technological advances, and, most importantly, a broad-based, concerted multidisciplinary attack on this disorder. This review focuses on the evolution of surgery for hydrocephalus over the last 30 years, the current state of the art of hydrocephalus treatment, and what appear to be the most promising future directions.

KEY WORDS: Cerebrospinal fluid shunt, Endoscopic third ventriculostomy, Hydrocephalus, Outcomes, Surgery

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Incidence

The incidence of pediatric hydrocephalus has declined in many developed countries (13, 14). The incidence of open neural tube defects has dropped precipitously as a result of maternal folate supplementation (61), antenatal screening, and termination of pregnancy based on superior antenatal imaging with ultrasound and magnetic resonance imaging. The incidence of cerebrospinal fluid (CSF) shunting in open neural tube defects, formerly reported to be as high as 90%, has also declined, possibly as a result of a general, more conservative approach, and also the selection of lower-grade lesions for delivery with a lower requirement for shunting (87). A prospective, randomized trial of in utero repair is also under way (10, 23), in which the primary outcome is the incidence of shunt-dependent hydrocephalus, thought to be possibly reduced by in utero repair (87). Similarly, better perinatal management of prematurity has reduced the incidence of intraventricular hemorrhage (25). Antenatal screening, genetic testing, and pregnancy termination have also reduced the incidence of other congenital malformations of the brain that cause hydrocephalus.

Diagnosis

Perhaps the greatest impact in hydrocephalus management has been that of imaging. Antenatal ultrasound and magnetic resonance imaging provide reasonably detailed fetal brain anatomy, detect malformations, have normative data for ventricular size, and allow serial investigation during gestation (*Fig. 1*) (65). Although attempts at in utero shunting have largely been abandoned because of what appeared to be a lack of efficacy or failure to appreciate the most appropriate cases, the quest for this type of surgery lingers (6, 8). In utero genetic testing by amniocentesis has largely been directed toward detecting chromosomal abnormalities in patients who are identified as having a brain abnormality such as hydrocephalus; however, this is clearly an area of future growth. X-linked hydrocephalus is now known to be part of the Crash spectrum with a mutation on chromosome Xq28 (92).

High-resolution magnetic resonance imaging of postnatal life has provided clues to the etiology of hydrocephalus, which in the past would have been labeled as idiopathic; these include intraventricular hemorrhage, aqueduct stenosis, migrational abnormalities, and so on. Serial imaging has allowed the evolution of hydrocephalus to be followed carefully, with the recognition that not only is ventriculomegaly often stable, but, particularly in intraventricular hemorrhage of prematurity, often resolves (32). Extraventricular obstructive hydrocephalus is now recognized to represent, almost universally, benign pericerebral collections of infancy that are usually familial, resolve with time, and almost never require treatment (56).

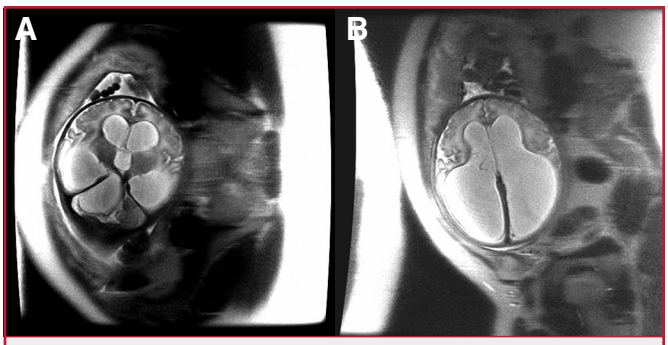

FIGURE 1. **A**, *in utero magnetic resonance imaging scan showing a fetus at 20 weeks gestation with hydrocephalus secondary to a posterior fossa arachnoid cyst. Axial images reveal a posterior fossa arachnoid cyst displacing structures to the left, occluding the aqueduct.* **B**, *more superiorly, there is significant ventricular dilation.*

The complex relationships among cysts, tumors, abscesses, and the ventricular system are now easily appreciated, as are the relationships between the ventricular walls and vascular structures in the cranial base, as must be considered when contemplating an endoscopic third ventriculostomy. Changes in ventricular size using standardized measurements or computer-generated, three-dimensional volumetric volume calculations can be used to detect small differences in size (46, 59). Similarly, the resolution of periventricular edema, persistence of flow voids through the floor of the third ventricle after endoscopic third ventriculostomy (ETV), is also accomplished easily.

Treatment

The major changes in treatment have been the use of advanced CSF valve design, broad application of endoscopic techniques, including ETV, and a general, more conservative approach given the recognition of the scale of CSF shunt complications. The important recognition that CSF shunt failure is a time-related event (68) and, if accounted for by actuarial techniques, was depressingly poor, allowed for a much easier comparison of shunt failure between centers.

CSF Shunts

The first spring, ball, and diaphragm valves were superseded by remarkably clever and innovative valve designs including anti-siphon devices (63), horizontal–vertical valves (21), flow-controlled valves (67), adjustable valves (5), and many more. Although each new valve was heralded as a significant advance that was more physiological in function with improved results in uncontrolled studies, the prospective and randomized trials told a very different story; they were all negative. When the standard valves, a novel flow-controlled valve (Orbis Sigma; Cordis, Miami, FL), and an updated anti-siphon valve (Delta Valve; Medtronic PS Medical, Goleta, CA) were compared in a prospective, randomized trial, there was no difference (*Fig. 2*) (20, 40). Similarly, when the adjustable (programmable) Medos valve (Codman/Johnson & Johnson, Raynham, MA) was compared with all other designs, and

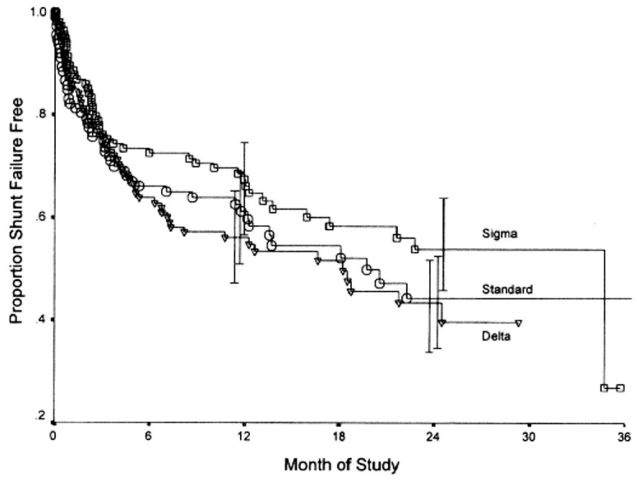

FIGURE 2. *Shunt complication-free survival curves showing a prospective, randomized shunt design trial, which compared standard valves with the Orbis Sigma Valve (Cordis, Miami, FL) and the Delta valve (Medtronic, Goleta, CA). There were no differences between the three valve designs (*from, *Drake JM, Kestle J, Milner R, Cinalli G, Boop F, Piatt J, Haines S, Schiff SJ, Cochrane DD, Steinbok P, MacNeil N: Randomized trial of cerebrospinal fluid shunt valve design in pediatric hydrocephalus.* ***Neurosurgery*** *43:294–305, 1998 [20]).*

despite the ability to adjust the valve for over- or underdrainage, avoiding a shunt revision for that reason, the failure rates were no different (*Fig. 3*) (62). A single-arm prospective study of an adjustable Delta valve, the Strata valve, suggested its failure rate was no different than any of the other previously studied valves in clinical trials (*Fig. 4*) (43). Perhaps most astounding, when ventricular size was measured pre- and postoperatively, comparing three very different valves, there was no difference over time, which indicates that the valves were not performing as the engineers had predicted (85). The prospective studies have allowed accurate measurement of anticipated shunt failure—40% at 1 year, including an approximate 10% shunt infection rate (20, 41, 43, 62).

New valve designs continue to appear with similar claims unsubstantiated by controlled data (60, 80). That neurosurgeons would embrace these reports, or use these valves, reflects a probable mixture of a "hope springs eternal" mentality, gullibility, and a failure of the United States Food and Drug Administration to regulate devices with the same scrutiny as novel pharmaceuticals.

CSF Shunt Infection

Shunt infection remains an intractable and serious complication of CSF shunting. Although the initial reports in prospective studies of shunt infection rates of 8% were greeted with disbelief, or comments about participating surgeons "not washing their hands," innumerable studies since that time have reported rates as high or higher (20, 41, 43, 62). Meta-analyses indicate that prophylactic antibiotics reduce the rate of shunt infection (31, 51). Identification of risk factors has suggested

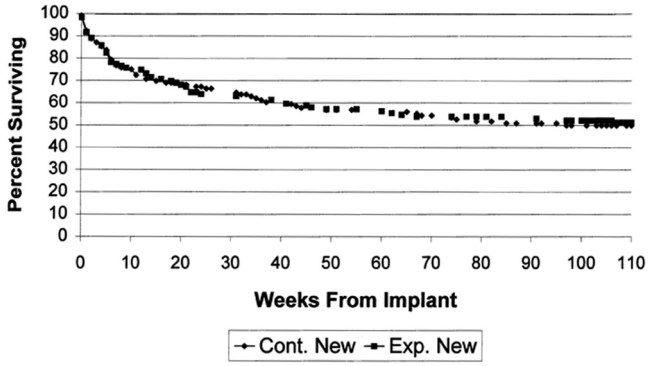

FIGURE 3. *Shunt complication-free survival curves showing the prospective, randomized Medos trial comparing the Medos valve (Codman, Raynam, MA) with all other valve designs. There was no difference in outcome, and the curves overlap (*from, *Pollack IF, Albright AL, Adelson PD: A randomized, controlled study of a programmable shunt valve versus a conventional valve for patients with hydrocephalus.* ***Neurosurgery*** *45:1399–1411, 1999 [62]).*

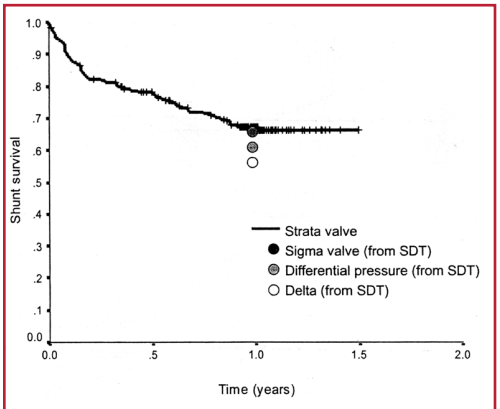

FIGURE 4. *Prospective cohort study showing the Strata valve (Medtronic, Goleta, CA), a programmable version of the Delta valve. Complication-free survival rates appeared very similar to other prospective, randomized studies, suggesting that the valve offered little if anything in terms of improved outcome (*from, *Kestle JR, Drake JM, Cochrane DD, Milner R, Walker ML, Abbott R 3rd, Boop FA: Endoscopic Shunt Insertion Trial participants: Lack of benefit of endoscopic ventriculoperitoneal shunt insertion: A multicenter randomized trial.* ***J Neurosurg*** *98:284–290, 2003 [41]).*

that age, holes in the surgical gloves, and postoperative CSF leak may be important (47). A follow-up study has shown some evidence that double gloving may help reduce shunt infection (18, 86). Shunts impregnated with antibiotics, typically rifampin and clindamycin, have been reported to reduce infection in one prospective, randomized trial, although the trial had a remarkably small number of patients and a very high infection rate in the control group (~16%) (28). Uncontrolled studies have had conflicting results (39, 73). An ongoing, but as yet unsubstantiated, concern has been the selection of gram-negative organisms or antibiotic-resistant organisms.

The treatment of shunt infection has generally been standardized to removal of shunt hardware, insertion of a temporary external ventricular drain allowed by reinternalization of the shunt system. There is a great deal of variability in terms of duration of antibiotic therapy, use of intrathecal antibiotics, and CSF parameters consistent with sterilization. A recent report reinvigorated the debate by reporting a high success rate when leaving the shunt hardware in place and using intrathecal plus systemic antibiotics, albeit through a separate CSF reservoir, which in some cases needed to be implanted before treatment, thereby negating the potential reduced surgical impact of this technique (7). Alarmingly, the reinfection rate using the standard protocols is approximately 25% when measured by prospective observation (42, 50). CSF shunt infection remains perhaps the most vexing of complications. Antibiotic impregnation seems to be a promising technique, but the long-term results are pending.

Shunt Insertion Technique

Several techniques for inserting shunts more accurately, including the use of stereotaxis (53, 93), endoscopy, and ultrasound (75), have been developed, largely to prevent errant catheters (which can end up in astonishing locations). The only technique subjected to a prospective, randomized trial was endoscopic insertion using miniature scopes, which were passed down the lumen of the ventricular catheter (41). The technique was of no benefit, and if anything, the failure rates were higher (*Fig. 5*). Interestingly, the final location of the shunt catheter was no better when using the endoscope than when using traditional landmarks, suggesting that either the catheter moved after removal of the shunt scope, or surgeons were unable to take advantage of the technology. In a *post hoc* analysis, a catheter placed away from the choroid plexus had a lower failure rate, suggesting that final catheter position within the ventricular system remains an important concept. Stereotaxis has been used primarily for patients with small ventricles. Frame-based and frameless systems and systems using magnetic field technology (in which the tip of the ventricular catheter can be tracked) have all been described. An advantage of ultrasound is that the trajectory of the catheter is observed in real time, and changes in ventricular size, indentation of the ependymal wall, and even position of the ventricular catheter holes can be visualized.

The widespread use of laparoscopy in general surgery has gradually migrated to shunt surgery, in which distal shunt catheters can be implanted under laparoscopic vision and fractured catheters can be removed (35, 44, 72, 94). This technique is perhaps best used in patients with dense peritoneal adhesions or obese patients, and routine use may allow neurosurgeons to develop the expertise to use these techniques on their own. Similarly, for the rarely used cardiac catheters, percutaneous fluoroscopic controlled access is relatively straightforward, and it reduces the extent and difficulty of cutting down and securing a tributary of the internal jugular vein (9, 15).

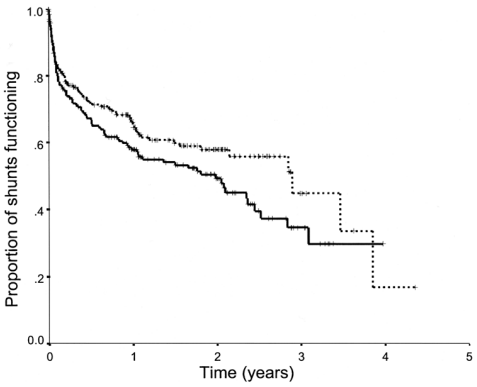

FIGURE 5. *Prospective, randomized trial showing the use of an endoscope to place the intraventricular catheter during shunt insertion. Complication-free survival rate was not different with the use of the endoscope (*from, *Kestle JR, Drake JM, Cochrane DD, Milner R, Walker ML, Abbott R 3rd, Boop FA: Lack of benefit of endoscopic ventriculoperitoneal shunt insertion: A multicenter randomized trial.* ***J Neurosurg*** *98:284–290, 2003 [41]).*

Outcomes

Shunt failure has typically been defined as a return to the operating room to treat shunt obstruction or infection or another complication, i.e., subdural hematoma (20). Repeated shunt failure has been less studied but appears to be at least a frequent event in children, and the fourth shunt failure may happen at an even shorter time interval (84). The risk factors for repeated shunt failure are not clearly defined but may be age, etiology, and, interestingly, the time interval from the previous shunt failure. This raises the specter of some intrinsic reaction to the shunt operation or hardware in some patients.

There is a poorly defined group of patients familiar to all neurosurgeons who are sometimes called "difficult shunt patients." These are patients who, for no particular reason, reappear at the emergency department and return to the operating room for what seem like interminable shunt problems. Many of them have slit-like ventricles and chronic headache and are categorized within the equally poorly defined slit ventricle syndrome (64). These patients are challenging, and no universal treatment seems helpful. Intensive investigation to rule out an unrecognized but lingering infection, and monitoring to characterize the intracranial pressure and its relationship to symptoms are very important. Surgical restraint, when the solution is not obvious, also seems wise because changing the valve to determine whether that helps seems rarely to work. These patients may be candidates for shunt removal and placement of an ETV, as discussed subsequently, although the long-term efficacy of this therapy is not established (3).

The obvious devastation in the quality of life for chronically ill shunt patients and their families has been a clarion call for an objective measure of this domain. Recently, such a measure has been developed for children older than 5 years, validated, and applied to a reasonably large cohort of shunt-dependent patients (48, 49). Shunt infection and epilepsy appear to have the greatest negative impact on quality of life, but there are also factors that might not have been anticipated, such as distance from the treatment center. Additional work in this area, for patients with ETV as well, is clearly important.

Endoscopic Techniques

(see video at web site)

The advent of high-resolution, charge-coupled device cameras combined with improved light sources and the proliferation of endoscopic techniques in other areas of surgery has led to widespread application of these techniques in neurosurgery. Germane to pediatric hydrocephalus are the ETV and cyst marsupialization. Although ETV was first attempted in the 1920s, it fell out of favor with the advent of CSF shunts in the 1950s (1). A combination of the realization of the extent of shunt complications, improved imaging to identify patients with obstructive hydrocephalus, and improved endoscopic technology has led to its reintroduction.

The operation is typically performed using a rigid endoscope, entering through a precoronal burr hole, and opening the floor of the third ventricle anterior to the mamillary bodies and the basilar bifurcation posterior to the dorsum sellae. Typically, the floor is perforated with a blunt instrument, and the opening is expanded with either a balloon or forceps. On occasion, in patients with a transparent floor of the third ventricle, an opening is made directly onto the dorsum sellae using electrocautery. This technique is used particularly when there is little space between the basilar artery and the clivus. Patients with large ventricles, an occluded aqueduct, a brainstem, and a basilar artery displaced posteriorly from the clivus seem to be the best candidates. All of these tenets have been challenged, however, as surgeons have pushed the limits in identifying surgical candidates. For example, patients with posterior fossa tumors in which the brainstem is applied to the clivus, patients with slit ventricles and shunts, and even patients with communicating hydrocephalus have all been reported to have successful outcomes after this procedure.

This wide range of potential candidates has made patient selection difficult. In children, the effects of age and etiology on outcome have been particularly controversial, with literature to support age as the most important (38, 45, 88), etiology as most important (4, 22, 24), and neither or both being important in terms of successful outcome (11, 26, 76). In a large multicenter study from Canada, when multivariate analysis was used, age was the predominant determinant of outcome for young patients, particularly neonates, having very high failure rates (*Figs. 6* and *7*) (19). This finding was supported by a large series from a single center in Australia (38). In the Canadian ETV series, 22% of the patients had preexisting shunts. The presence of a shunt did not affect the failure rate, indicating that shunt independence is a reasonable outcome in selected patients. Some have argued that even with a high failure rate, an ETV is

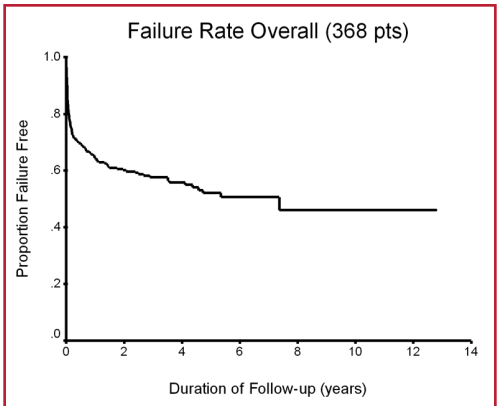

FIGURE 6. *Overall outcome from the Canadian collaborative study of endoscopic third ventriculostomy defined failure as any subsequent operation for cerebrospinal fluid diversion (*from*, Drake JM; the Canadian Pediatric Neurosurgery Study Group: Endoscopic third ventriculostomy in pediatric patients: The Canadian experience.* ***Neurosurgery*** *60:881–886, 2007 [19]).*

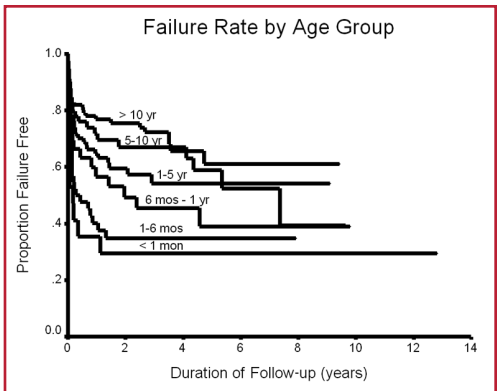

FIGURE 7. *Outcome from the Canadian collaborative study was broken down by age. There is a progressive decline in outcome with decreasing age, being lowest in children younger than 1 month of age (*from*, Drake JM; the Canadian Pediatric Neurosurgery Study Group: Endoscopic third ventriculostomy in pediatric patients: The Canadian experience.* ***Neurosurgery*** *60:881–886, 2007 [19]).*

always better than a shunt and is, therefore, a reasonable approach. Given that the failure rates in younger patients are high, the risks of the surgery itself are higher than with a ventriculoperitoneal shunt, and failure requires insertion of a ventriculoperitoneal shunt or repeat ETV, this recommendation requires reflection. As an indication of just how wide open the indications for ETV are, a recent report from a single surgeon and center from Africa reported good success with ETV in children when they used a flexible endoscope and coagulated the choroid plexus at the same time on both sides of the ventricular system, even when this technique was applied to children with such disorders as myelomeningocele (90, 91).

The complications of ETV are not infrequent and may be serious, with perforation of the basilar artery being the most dreaded (55). CSF leak, meningitis, hypothalamic injury, and cranial nerve injury have all been reported (83). The overall surgical complication rate appears to be approximately 10 to 15%. A rare and initially unrecognized complication is late rapid deterioration. In this event, patients appear to be doing well after the procedure (30, 34). They begin to complain of headache and then rapidly deteriorate; without immediate access to neurosurgery, they lapse into unconsciousness, and most die (*Table 1*). A recent report compiled 15 cases from the literature and around the world (17). The patients were operated on at an average of 8.4 years and had been followed for an average of 2.8 years. Thirteen patients died and, in all who had an autopsy or repeat ETV performed, the opening in the floor of the third ventricle was closed. The assumption that this closure was a recent event was disputed by several patients, one of whom had evidence of ETV obstruction for more than 4 years before late rapid deterioration. These findings have prompted some to suggest that an access reservoir should be implanted in all patients having this procedure, and that patients with imaging evidence of obstruction should be reoperated even if they are asymptomatic (2, 57). Whether these suggestions will reduce the incidence of this unfortunate event is unknown.

Endoscopic cyst fenestration for loculated ventricles, intraventricular cysts, extracerebral arachnoid cysts, and tumor excision has been reported to be an effective way of treating the accompanying hydrocephalus (29). Perhaps the most clearly documented cyst fenestration has been for suprasellar arachnoid cysts (58, 77, 89). High-resolution endoscopy has often revealed a "valve" opening in the base of the cyst adjacent to the basilar artery (69). Perforation of the roof of these cysts, with or without an accompanying opening in the floor, has been reported to be successful. For patients with loculated hydrocephalus and trapped ventricles, endoscopic fenestration has either obviated the need for a shunt or reduced the need for multiple shunts (27). Endoscopic cannulation of the aqueduct for a trapped fourth ventricle, usually with a shunt stent, is an example (12, 66, 70, 74). Although colloid cysts are the most common tumor to be excised endoscopically (33, 52, 71, 79), other small tumors have been removed endoscopically in the presence of hydrocephalus and without the need for subsequent shunting (78). Endoscopic biopsy, performed at the same time as a CSF diversion procedure, i.e., an ETV, has also obviated the need for an open approach to the tumors, depending on the tumor type (16).

SUMMARY AND FUTURE DIRECTIONS

It is unlikely that dramatic improvement in the treatment of pediatric hydrocephalus will occur without a better under-

TABLE 1. Summary of late rapid deterioration endoscopic third ventriculostomy cases[a]

Patient no.[b]	Country	Age (yr)/sex	Etiology	Previous shunt	Follow-up (mo)	ETV occluded	Outcome
1	UK	3/M	IVH	No	4.0	Unknown	Death
2	UK	11/M	AS	No	4.0	Unknown	Death
3	Australia	4/M	CH	No	84.0	Yes	Death
4	Canada	13/F	TG	No	42.0	Yes	Death
5	UK	0.83/M	AS	Yes	7.0	Unknown	Death
6	Canada	0.33/M	AS	No	24.0	Yes	Death
7	France	13/F	TG	Yes	24.0	Yes	Normal
8	USA	10.83/F	TG	No	30.0	Yes	Death
9	UK	13.6/F	TG	No	60.0	Yes	Death
10	UK	12.92/M	CH	Yes	57.0	Yes	Death
11	France	3.25/M	AS	Yes	9.0	Yes	Death
12	France	7.8/F	AS	No	94.0	Yes	Disability
13	UK	4.83/M	CH	No	1.4	Yes	Death
14	USA	9.92/M	AS	No	5.0	Yes	Mild disability
15	USA	5.83/F	AS	Yes	1.2	Yes	Death
16	Japan	7.92/F	AS	No	22.0	Yes	Death
	Average	7.6		Average	29.8		

[a] ETV, endoscopic third ventriculostomy; UK, United Kingdom; USA, United States; IVH, intraventricular hemorrhage; AS, aqueductal stenosis; CH, congenital hydrocephalus; TG, tectal glioma.
[b] Patients 1 through 7 were previously reported. Means for patient age and follow-up period were 7.6 years and 29.8 months, respectively.

standing of the fundamental pathogenesis of the disorder and the surgical devices that are used to treat it. This will require the challenging of conventional thinking. Perhaps the best and most recent example is the emerging evidence that arachnoid granulations have only a secondary role in CSF absorption, the primary site being the nasal lymphatics (36, 37). Behind every CSF shunt design is a mathematical model that specifies and predicts the device performance (54). These models have failed to produce devices that have had any significant effect on clinical outcome when subjected to prospective, randomized studies (20, 62) and have been unable to accurately predict such fundamentals as the change in the size of the ventricles (85). This is either because they have been overly simplistic or unrealistic (81). Developing better mathematical models is of paramount importance if better shunt designs are to emerge, but this will take a concerted and multidisciplinary effort (82). Perhaps the fundamental lesson from the last 30 years is that hydrocephalus, which appears to be an alluringly simple problem of CSF accumulation, is anything but.

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COMMENTS

This article provides a concise yet comprehensive overview of hydrocephalus as it relates to neurosurgical practice today. In reading this thoughtfully composed article, I am reminded of how far we have come in the investigation and management of this condition over the past 35 years of my own practice. This is, of course, not to discount the continuing frustrations that plague our efforts to manage patients with hydrocephalus, particularly children. To my mind, shunt infection, especially its prevention, represents the single most important problem with which we have made excruciatingly little progress. Reported infection rates are virtually unchanged over the past three decades. This continues to have enormous implications for patients, physicians, and society at large. The other continuing nemesis is, of course, shunt obstruction. Similarly, we have made only modest progress with this problem over the same three decades, despite notable improvements in shunt design and surgical technique.

I fear that as long as we require hardware of any sort to effectively treat so many cases of hydrocephalus, these two problems will continue to plague our best efforts. Over the years, attempts have been made to find a treatment that does not require implanting hardware. Choroid plexectomy and pharmacological reduction in cerebrospinal fluid (CSF) production came to naught, for sound physiological reasons. The refinement of effective endoscopic techniques along with advances in imaging have led to a revival of ventriculocisternostomy. Endoscopic third ventriculostomy (ETV) is now widely practiced and has been a godsend for many patients and their neurosurgeons. Although ETV has its limitations, in my opinion, this procedure has had a more profound effect on hydrocephalus management than any other single advance over the past 35 years.

Paul H. Chapman
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This article represents an unbiased assessment of the state of the art and science of hydrocephalus and its treatment. James Drake, arguably one of the most knowledgeable authorities on this subject, has again delineated what is known, what is myth, and what remains to be done in this field. Unfortunately, we do not understand the precise pathophysiology of each etiology of hydrocephalus; therefore, uniformly successful treatments have eluded us. Thus, we are reminded that our current treatment paradigms are mired in uncertainty and habit. Although mathematical modeling may yield better mechanical devices, thus far, randomized studies have demonstrated no frontrunner in the hardware competition for "best shunt design." Furthermore, a rather stark and honest delivery of the facts on shunt infection, shunt failure, and ETV failure are delivered. This article essentially sums up approximately 50 years of scientific knowledge about hydrocephalus and its treatments. It does not tell you what shunt to use or when to perform an ETV, but it will provide you with a scientific basis with which to make those decisions. For those who treat hydrocephalus

(i.e., all neurosurgeons), this will become a must-read reference article. And for some, it will provide an impetus to push the field forward.

Richard G. Ellenbogen
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James Drake ends his overview by stating that "hydrocephalus, which appears to be an alluringly simple problem of CSF accumulation, is anything but." It is and it isn't. CSF drains in response to a pressure differential. If the resistance to drainage increases beyond a point, which probably varies with time in any given individual and by individual, CSF accumulates. That point is simple. Why and how much the resistance changes, the physiological response to pressure change, and the treatment of CSF accumulation is not so simple.

Although there have been many new technological innovations in shunt hardware, the effectiveness of the shunting systems has not dramatically improved over the past 30 years. There have been stepwise gains, such as inserting enough peritoneal tubing at the time of the initial insertion, even in a neonate, to avoid a lengthening procedure with growth. Another is the encasement of a barium stripe within the wall of the tubing, making the tube radio-opaque, but preventing the barium in the silicone from coming in contact with the subcutaneous tissues and thereby preventing the reaction that accelerates biodegradation and subsequent tubing fracture. The major problems of obstruction and infection still remain, the reduction of which would greatly reduce shunt failure rates. Approximately four-fifths of our revisions are to replace an obstructed ventricular catheter. A considerable focus of shunt hardware has and continues to be the valve, in an attempt to regulate CSF drainage to as close to normal physiological parameters as possible. In our practice, over- or under-drainage is rarely a problem, with shunt revisions to change the pressure of the valve accounting for, at most, 1% of our revision patients. As it is the ventricular catheter that becomes partially obstructed before becoming completely blocked and, therefore, becoming the site of increased resistance to CSF flow, what happens downstream at the valve is far less relevant. An advance would be a valve that could decrease the incidence of ventricular catheter obstruction. The addition of an antisiphon device should, theoretically, help in this regard, but there is no definitive evidence to support such devices that decrease proximal obstruction.

The major advances in the past 30 years have been in imaging. To be able to noninvasively make the diagnosis, determine the anatomic abnormality (if any), surgically intervene, and then monitor the effect of treatment, be it with CSF shunt diversion, opening of an obstructed pathway such as fenestration of a loculated ventricle, or bypassing an aqueductal stenosis with an ETV, all have been made possible by the advances in imaging. The best way to avoid shunt problems is not to insert one. Being able to noninvasively monitor ventricular size has aided immeasurably in the continuing care for those with hydrocephalus.

The introduction of fetal ultrasound and magnetic resonance imaging have added a new dimension, and now one can be aware of the presence of hydrocephalus fairly early in gestation. In our practice, we routinely see families who want to know the prognosis of their fetus, which has been diagnosed as having hydrocephalus. This is a little like asking about the prognosis of having a fever. Attention is often focused on the size of the ventricles, which can easily be measured, and any change in size is noted. Overwhelmingly, the prognosis is determined by what caused the hydrocephalus rather than the size of the ventricles. As an example, it is possible to control progressive hydrocephalus that is subsequent to intraventricular hemorrhage in preterm neonates. The neurological outcome for this patient population is, by far, determined by the extent of the hemorrhage and the amount of parenchyma destruction rather than ventricular size and the control of the hydrocephalus (6). In utero treatment of hydrocephalus, as noted by Dr. Drake, has been abandoned, because the risks of the procedure outweigh the benefits. Directing therapy at ventricular size does not address the underlying problem that caused the hydrocephalus and appears to have limited benefit.

Regarding the future, advances in imaging are likely to lead the way. We should soon be able to noninvasively observe and monitor CSF movement, determine intracranial pressure, and automatically obtain the volume of CSF in the ventricles. There are, no doubt, techniques awaiting discovery that we have yet to imagine, such as MRI was 30 years ago.

Hydrocephalus is a plumbing problem: this is the simple part. Understanding the pathophysiology of CSF drainage is not so simple. For some time, the only CSF drainage pathway was thought to be via one-way valves directly into the major venous sinuses. As cited by Dr. Drake, Johnston (3) and Johnston et al. (4) have done excellent work in addressing the role of the lymphatics in the CSF drainage pathways. We have also examined those pathways and their responses to pressure (1, 2, 5, 7). Being able to manipulate CSF drainage pharmacologically might make it possible to treat hydrocephalus without hardware.

Although we still have a long way to go in the practice of pediatric neurosurgery, we do more good for more patients by treating their hydrocephalus optimally than anything else.

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Given James Drake's expertise in the arena of intracranial CSF dynamics and valve technology, he is perfectly suited to offer an authoritative overview on the treatment of hydrocephalus. His comments are concise and cover a spectrum of theoretical and practical topics. Troubling to the neurosurgeon, however, are his conclusions that underscore the disassociation between shunt technology and improvements in therapy. He justifiably points out that valve mechanics, catheter design, and programmable devices have all fallen short of predictive modeling or theoretical claims when studied in well-designed clinical trials. Although that news is sobering, it does not distract from what I believe to be the greatest advancement in the treatment of hydrocephalus over the past three decades, namely, the use of ETV for

the treatment for noncommunicating hydrocephalus, a procedure I learned from him. I am appreciative not only of Dr. Drake's overview on a vexing problem in neurosurgery, but also his continual objective assessment of purported advancements in CSF diversion.

Mark M. Souweidane
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James Drake has provided us with an evidence-based review of the current treatment of hydrocephalus. For some, his conclusions may engender a feeling of hopelessness. Lacking is the usual unfettered enthusiasm for unproven but popular technical innovations: programmable valves, endoscopic shunt placement, antibiotic-impregnated catheters, telesensors, gravity accumulators, and even ETV and choroid plexectomy. None of these devices and procedures has been proven to be a real advance by a rigorous scientific trial. Their use is often driven by poor studies, patient wishes, competition among neurosurgeons, and industry-driven marketing. Dr. Drake has done a real service in putting all of this into perspective. He did not indulge in "how I do it" cookbook recipes, which are abundant elsewhere.

The reality is that the biggest advance in the treatment of hydrocephalus since the original Spitz Holter valve was the computed tomographic scanner. The management of hydrocephalus remains extremely complex, requiring judgment and experience rather than a new gizmo. The so-called slit ventricle syndrome remains a serious diagnostic and therapeutic challenge, despite programmable valves, flow-regulated valves, and on-off valves. For diagnosis, one must resort to shunt exploration, Diamox (Lederle Pharmaceuticals, Pearl River, NY) trial, intracranial pressure monitoring, migraine medication trials, and even subtemporal craniectomy. For treatment, one may have to consider lumboperitoneal shunt, fronto-orbital advancement, or psychotherapy, depending on the presumed cause.

Another challenge is the long-term patient who has run out of distal shunt sites, or the complex patient with congenital heart disease and an abdominal event that precludes the usual ventriculoperitoneal or ventriculoatrial shunt. One must resort to unusual procedures, such as ventriculofemoralatrial shunts, gallbladder shunts, or sagittal sinus shunts.

The biggest problem remains recurring proximal obstruction that develops as a result of multicavitary hydrocephalus or choroid plexus. This may require cyst fenestration, which is most effective if performed via craniotomy with microscopic techniques rather than endoscopy.

Despite the initial promise that ETV would eliminate the need for shunts, this is clearly not the case. As we enter a new period in medicine in which we will be accountable for cost, and only scientifically proven technology will be reimbursed, all of us will be required to know what works, and what does not. Dr Drake deserves our thanks.

Leslie N. Sutton
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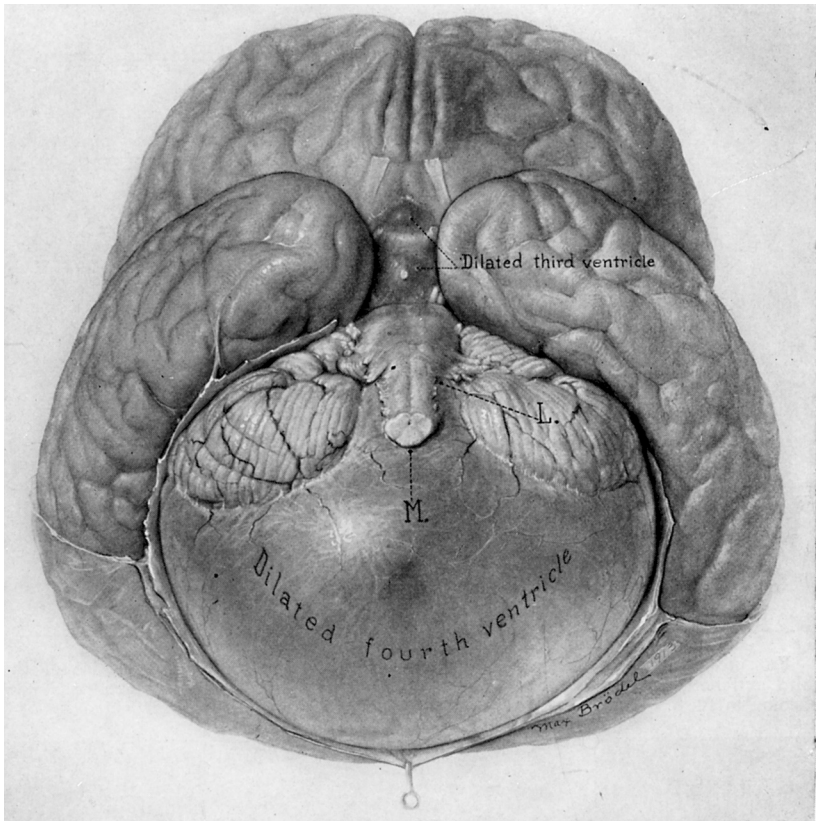

Illustration showing congenital hydrocephalus due to failure of the foramina of Luschka and Magendie to open. (From: Dandy WE: *The Brain*. New York, Harper & Row, Publishers, 1969).