Surgical Treatment of Hydrocephalus: A Historical Perspective

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Hydrocephalus · Neurosurgical history · Shunts · Third ventriculostomy · Ventriculocisternostomy

Abstract
Surgical treatment for hydrocephalus has a long and eventful history. This review emphasizes the significant advances made in this century, describing the personalities involved, technical approaches attempted, and materials tested. It concludes by suggesting that better methods of treatment can and should continue to be developed.

Introduction
Surgeons have operated on human skulls since the neolithic period, and in order to fully appreciate the development of modern neurosurgery, one must look at its earlier counterpart. The ancient civilizations of the East have left writings that describe their treatment of head and spinal injuries as early as 3000–2500 BC [1]. Yet from the relatively advanced state of medicine in these ancient societies, progress in the area of neurological surgery stagnated up until the present century. Here I intend to focus on one specific pathologic state that modern neurosurgeons have to some degree successfully conquered, namely, hydrocephalus. This common condition (incidence >3/1,000 births) had uniformly tragic consequences up until the 2nd or 3rd decade of the 20th century. The purpose of this review is to give a general history of the treatment of hydrocephalus, focusing on some of the more significant recent improvements in surgical treatment, and why shunting has become the current treatment of choice in most cases of infantile hydrocephalus.

Hydrocephalus in Ancient History

Returning to the ancient pioneers in neurosurgery, we find that by 400 BC trepanning was already widely practiced [2]. Hippocrates recommended it for the treatment of epilepsy, blindness, headache, and possibly hydrocephalus. Although physicians clearly recognized hydrocephalus as a clinical entity at this time, it was not until around the 17th century that definitive evidence of surgical therapy for this condition first appeared. But one should not take this lack of treatment as an indication of lack of interest in the problem. Galen spoke of it in the 2nd century AD, and Vesalius gave the first description of the ventricles as the location of excess fluid collection in the 16th century.
During the intervening centuries between Galen and Vesalius, several cultures practiced a primitive form of neurosurgery and operated on hydrocephalics. Charaf ed Din, an Arab physician, described percutaneous ventricular drainage in 1465, following which the child rapidly succumbed to the sudden, uncontrolled reduction of pressure. The German surgeon Hildanus described the same outcome at the turn of the 17th century [3]. (Death in these cases probably followed inward collapse of the skull; early 20th century neurosurgeons describe using plaster molds to prevent this fatal occurrence.) Trepanned skulls from the Egyptians and the Aztecs [4] provide even earlier evidence of neurosurgical operations performed in widely separate locations of the ancient world. For the most part, without awareness of sterile technique or an understanding of physiology, these efforts were doomed to failure.

Given the dismal prospects of surgical therapy, many practitioners relied on conservative medical treatment. Up until the beginning of the present century, physicians used repeated percutaneous punctures, head wrapping, diuresis, ligation of the carotid arteries, and the universal blood-letting in the management of hydrocephalus, all with consistently fatal results. One idea was that external pressure might reduce the fluid accumulation, and thus various means of compression were applied to the enlarged skull [5, 6]. Rubber bandages provided stronger tension and constant pressure compared to other materials [7]. However, pressure on the skin caused decubitus ulcers, and the resulting increased intracranial pressure created decreased CSF circulation, and even seizures and skull fractures [8]. As neurosurgeon John E. Scarff determined in a review of treatment for hydrocephalus, ‘as late as the beginning of the present century, the pathology of this condition was obscure, no rational methods of therapy had been developed, and ... no successful surgical treatment had ever been achieved’ [9].

Curative surgical treatment, then, had to await the emergence of neurosurgery as a distinct medical specialty, which in turn had to await the advent of developments in other areas of medicine. The discovery of anesthesia in the 1840s, while greatly enhancing the capabilities of general surgeons, did little for the development of neurosurgery due to lack of knowledge in the area. Later, Pasteur’s and Koch’s in bacteriology, and Lister’s introduction of asepsis in surgery, brought about the technical advancements required for successful surgical invasion of the brain (table 1).

Table 1. Timeline of eras and events

<table>
<thead>
<tr>
<th>Early descriptions of hydrocephalus and its treatment</th>
<th>400 BC Hippocrates</th>
<th>200 Galen</th>
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<tr>
<td>1465 Charaf Ed Din – paracentesis of lateral ventricles</td>
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Prerequisite advances in other areas

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<th>Neuroanatomy, pathology, and physiology</th>
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<tr>
<td>1543 Vesalius – ventricles as the site of excess fluid collection</td>
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<td>1769 Morgagni – pathology associated with hydrocephalus</td>
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<td>1825 Magendie – CSF circulation and pathways</td>
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<td>1855 Lushka</td>
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<td>1854 Faivre – CSF produced by choroid plexus</td>
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<td>1876 Key and Reitzus – CSF physiology</td>
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Basis for clinical surgery

| 1840s volatile general anesthetics |
| 1869 Lister – principles of antisepsis in surgery |
| 1870s Koch – germ theory of disease |
| 1878 Pasteur – medical application of germ theory |

Neurosurgery as a discipline

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<th>Key events</th>
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<tr>
<td>1905 Kausch – first ventriculoperitoneal shunt</td>
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<td>1918 Dandy – choroid plexectomy</td>
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<td>1922 Dandy – third ventriculostomy</td>
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<td>1936 Stookey and Scarff – endoscopic ventriculostomy</td>
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<td>1939 Torkildsen – ventriculocisternostomy</td>
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<td>1942 Scarff – cauterization of choroid plexus</td>
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<td>1949 Matson – lumboperitoneal shunt</td>
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<td>1952 Nulsen and Spitz – competent ball valve, ventriculoatrial shunt</td>
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<td>1956 Holter – first Silastic slit valve</td>
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<td>1973 Portnoy – first anti-siphon device</td>
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This list is intended to give perspective to the current status of hydrocephalus treatment by placing it in the context of certain past achievements and advances. In general, the dates of events associated with names describe the year of a significant publication.

Progress in the Past Century: The Men and Their Materials

While Vesalius, Magendie, Morgagni, and others were making progress in the areas of neuroanatomy and neuropathology, in the mid to late 1800s, rapid advancements also began to appear in neurophysiology. All these discoveries led to the growth of the specialty of neurosurgery, pioneered in the early 20th century by Sir Victor Horsley in England, and Harvey Cushing, Walter Dandy, and others at Johns Hopkins in the US. The personalities involved in establishing neurosurgery had much to do with how the area developed. After graduating from Johns

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Hopkins in 1910, Dandy (fig. 1) alone published at least 17 papers dealing directly with hydrocephalus (most of them between 1914 and 1922) and a third that number on the diagnostic technique of ventriculography. Establishment of ‘the Hunterian’, a laboratory for experimental surgery at Johns Hopkins first directed by Cushing, also greatly aided progress in the etiology and treatment of hydrocephalus. The contributions of these men and their colleagues have led to the development of some successful surgical treatments for hydrocephalus.

To fully appreciate the difficulties involved in devising a successful surgical treatment for hydrocephalus, one must have a detailed understanding of ventricular neuroanatomy and the causes of hydrocephalus. In 1914, Dandy and the pediatrician Kenneth D. Blackfan published the first extensive study – a 76-page article – on the subject. In this initial study, Dandy and Blackfan divided hydrocephalus for the first time into noncommunicating (obstructive) and communicating types. Dandy emphasized that the etiology, though frequently idiopathic, was a crucial factor in selecting a patient’s treatment. They recognized that the condition could result from blockage at various sites and documented these in their paper [10].

As a result of Dandy’s work on hydrocephalic dogs, the physiology of CSF circulation and the etiology of hydrocephalus emerged in clearer light. The irony of the monumental contribution Dandy made during these years of experimenting is that his work at the Johns Hopkins Hunterian Laboratory was not a matter of choice. Rather, more correctly, not his first choice. Harvey Cushing, the first American neurosurgeon and Dandy’s mentor during his initial year at the Hunterian in 1911, had promised to take Dandy with him when he left for Boston the following year. But the time Cushing and Dandy spent together proved only that the Laboratory had more than one strong personality, and as a result of several conflicts between the two men, Cushing left for Boston without Dandy. Halstead had already filled his house staff at Johns Hopkins for the year; consequently, Dandy was left without work. Kindly, Halstead arranged for him to stay on at the Hunterian until a position became available on the house staff, during which time Dandy carried out several of his classic experiments.

Why Dandy chose to investigate hydrocephalus and not some other brain disorder is interesting to consider. In the absence of any specific evidence, it seems likely that at this time there was little else to work on; neurosurgeons simply did not have the skill, knowledge, or equipment to treat patients. Additionally, hydrocephalus was a common disease. The influence of Cushing behind Dandy’s contributions is a less obvious but equally important factor. Despite their differences, it was Cushing’s influence that ultimately persuaded Dandy to dedicate himself to neurosurgery instead of anatomy, as Dandy had once considered. And it was Cushing’s rather harsh treatment of Dandy, according to Howard Naffziger, another of Cushing’s assistant residents, that stimulated Dandy ‘to make a good showing in neurosurgery’ [11]. Dandy’s abrupt discontinuation of hydrocephalus research after 1922 is curious. It was as if he suddenly lost interest in the disease and its treatment after nearly a decade of pioneering advance [12]. Until 1945 he reported only several patients successfully treated.

During and following his years of research on hydrocephalus, Dandy devised two new diagnostic techniques. Although the clinical manifestations of advanced hydrocephalus are easily observable, these methods allowed earlier detection and determination of the site of obstruction. Ventriculography has been of general usefulness, and one historian hailed it as ‘the greatest single contribution ever made to brain surgery’ [13]. The dye or PSP test is specific for hydrocephalus and involves injecting indigo carmine dye or PSP (phenolsulfonphthalein) into a lateral ventricle and performing a delayed lumbar puncture to determine whether the dye has circulated. Both procedures were extensively used for many years prior to the widespread use of CT and MRI.
Surgical treatment for hydrocephalus began developing at a rapid rate following Dandy’s work. These procedures were based on three principles: to reduce CSF formation at its source, to relieve the obstruction in the ventricular system, and to divert the fluid to another body location where it can either be absorbed or excreted. Although present day neurosurgeons often credit Dandy and his contemporaries with introducing these techniques, a careful scrutiny of the literature points out that nearly all current surgical techniques had been attempted by around 1900 [see ref. 6, 14, 23 for detailed lists of these earlier pioneers in the surgical treatment of hydrocephalus]. The striking feature of these early publications is the high mortality rate. Over 50% immediate operative/post-operative mortality was not uncommon, and very few patients survived beyond a few months [8].

‘No form of treatment, either medical or surgical, has yet a valid claim to the cure of a single case of hydrocephalus ...’, wrote Dandy in 1918. ‘But hydrocephalus is a curable disease’ [15]. With this statement, typical of the style Cushing criticized, Dandy opened the first of his many papers on the surgical treatment of hydrocephalus. Published in the December, 1918, issue of *Annals of Surgery*, this article introduced the technique of choroid plexectomy for communicating hydrocephalus. ‘Based on simple, fundamental, physiological and surgical principles’, this procedure involved extirpation of the lateral ventricular choroid plexuses – the principal site of CSF formation – and, as in Dandy’s subsequent operations, ‘avoided the use and permanent implant of any rubber, plastic, or metallic tubes or valves’ [9]. Neurosurgeon John Scarff (fig. 2) authored this statement and was instrumental in developing endoscopic cauterization of the plexuses, the technique that soon replaced Dandy’s first procedure. Although enthusiastically received and practiced by many neurosurgeons initially, most abandoned the procedure in the 1950s after the development of ventriculoatrial shunting techniques, because of generally poor results of the former [16, 17]. Scarff, however, continued to advocate choroid plexectomy and Dandy’s later technique of third ventriculostomy as the simplest and most physiological methods of surgical treatment. This method of treatment (endoscopic coagulation) has been revived more recently for use in certain subgroups of hydrocephalic children [18–20].

Scarff, following his graduation from the Johns Hopkins Medical School in 1924, worked for 1 year as one of the last surgical research fellows in the Hunterian Laboratory. Initially intending to specialize in thoracic surgery, Scarff served as a surgical resident at the Johns Hopkins Hospital from 1925 to 1927, where he operated with Dandy and was likely exposed to Dandy’s ideas about hydrocephalus. In either case, Dandy excited Scarff’s interest in neurosurgery, and Scarff chose to pursue neurosurgical training – in Boston with Cushing. While the role that Dandy played in Scarff’s ultimate decision to pursue neurosurgery is evident, his influence in Scarff’s pursuit of hydrocephalus is less direct. It seems that Scarff turned to hydrocephalus some years later, realizing that Dandy had contributed enormously to the field but that much still remained to be accomplished. Viewing Scarff’s later contributions with Byron Stookey while at Columbia University, it is interesting to consider the influence both Dandy and Cushing have had on the development of neurosurgical treatment beyond their own contributions.

The experience gained by these and many other unsuccessful attempts to reduce CSF formation led to increased interest in means of enhancing drainage. ‘In all this there was great energy and invention ...’ points out Milhorat [17], contributor to a contemporary textbook on pediatric neurosurgery. Dandy introduced third ventriculostomy for obstructive hydrocephalus in 1922, followed shortly by several other accounts of the procedure. In 1936, Scarff and his coworker Stookey reported a new version of this operation which allowed circulation from the third ventricle to the subarachnoid cisterns. Specifically, the technique used an endoscope to drain the ventricles through openings made in the lamina terminalis and the floor of...
the third ventricle [21]. By 1951, reports indicated that, ‘on the whole, it (third ventriculostomy) has not been very successful. For the most part, the openings in the third ventricle have closed off. The indications for this procedure are rather limited’ [14]. Despite this prognosis, developments in the area continued, and recently there have been excellent results with third ventriculostomy in selected patients [22–24].

Choroid plexectomy, another technique pioneered by Dandy [15] and refined by Scarff [25], aimed at reducing the production of excess fluid at its source. While the idea was plausible, in practice it did not work as well as predicted and sometimes even resulted in fluid overproduction. The reason for this became clear only later: isotope studies of CSF flow and absorption demonstrated that the choroid plexus is not the primary source of CSF production [26]. Again, however, recent reports indicate that this technique can be useful in selected cases of communicating hydrocephalus [27].

Three years after Stookey and Scarff published their endoscopic ventriculostomy article, a Danish surgeon, Torkildsen, announced a promising new technique called triventriculoauriculostomy, consisting of the insertion of one end of a catheter into the lateral ventricle and the other into the cisterna magna. ‘In all cases the effect of the operation has been a rapid disappearance of any symptom of increased intracranial pressure’, Torkildsen [28] reported. This intracranial bypass technique was the first truly successful shunting procedure. Although several articles [16, 29–31] have indicated that it is still useful today in carefully selected cases of obstructive hydrocephalus, ventriculocisternostomy, like third ventriculostomy, cannot cure many cases. Both procedures depend on the ability of the basal and convexity CSF pathways to reabsorb fluid and thus can only correct obstructive disease.

Torkildsen’s report in 1939 sparked a resurgence of interest in hydrocephalus, and researchers devised many ingenious operations. Most of these new techniques involved diversion of the CSF to another body cavity or duct. Determined, inventive neurosurgeons attempted to shunt the excess CSF to nearly every conceivable site, including the mastoid air cells, fallopian tube, digestive tract, gall bladder, thoracic duct, urinary bladder, and Stensen’s duct [17]. As Spitz, one of the pioneers of ventriculoauriculostomy, points out, ‘the variety of surgical techniques developed ... gives silent testimony to the difficulties encountered’ [32].

One such shunt system was the lumboureteral shunt, popularized by Matson [33]. A primary advantage of this technique was that the ureter has the correct hydrodynamic resistance, which eliminated the need for a valve. It was also a simple procedure. The child lost a healthy kidney, however, and loss of electrolytes and fluid into the urine was a frequent and serious problem. Relatively few neurosurgeons describe using this operation. And yet, like the other techniques that have been mostly abandoned over the years, this one also has its advocates. The procedure no longer entails loss of a kidney, and the lower incidence of revisions makes it a suitable alternative to cases of ventriculoatrial (VA) or ventriculoperitoneal (VP) shunts that have failed multiple times [34].

The reader may be interested to know the fate of the resected kidneys in Matson’s procedure. Matson provided these to the Harvard Transplant Service, under the direction of Dr. Joseph Murray, who had already begun the work for which Murray and E. Donnall Thomas received the 1990 Nobel Prize. When the transplant service was unable to use most of them, the kidneys were passed on to Dr. John Enders, who in turn used them for his Nobel Prize winning studies on developing a non-neuronal tissue culture system for poliovirus [35].

From experience with extracranial shunts, two sites have emerged as having particular merit; these are the right atrium of the heart and the peritoneal cavity. Neurosurgeons had investigated shunting to the vascular system intermittently throughout the first half of the 20th century. In 1952, however, Nulsen and Spitz [36] published a report that marks both the modern era of hydrocephalus surgery and a temporary preoccupation with ventriculoauriculostomy. Significantly, this was the first case to employ a competent, unidirectional valve to prevent reflux. This advance allowed a more general use of shunts, which previously had no method of pressure regulation and were plagued with problems of retrograde flow and overdrainage of CSF. For the next 2 decades, VA shunts enjoyed universal popularity, and numerous reports on this procedure were published [37]. ‘In our hands ... this is by far the most effective treatment currently available for the relief of this distressing disease’ [16]. Statements such as this abound in the literature.

Subsequent improvement in shunt techniques seems to have contributed to the success of the operation, which nevertheless suffered from several serious problems. Some of these are not unique to shunts but also occur with the central venous catheters of today, such as difficulties with accurate placement (into the superior vena cava or right atrium as opposed to some other venous channel), and irritation of the nodes which can lead to arrythmias. Long-term complications include release of microemboli from the tube, leading to pulmonary hypertension;
chronic bacteremia with the threat of fatal infections; glomerulonephritis resulting in renal failure, and perforation of the right atrium by the tube, producing cardiac tamponade [38, 39].

With these developments, VP shunts began to resurface as a possible substitution for ventriculocisternostomy and have now replaced this procedure as the treatment of choice for hydrocephalus. First performed in 1905 by Kausch, a German neurosurgeon, this procedure ‘fell into disrepute’ before World War I and was virtually abandoned for the next 30 years [14]. Surgeons resisted this technique because of problems with frequent occlusion of the tube. Two factors contributed to its revival, namely the absence of the serious and often fatal cardiopulmonary complications related to VA shunts, and the introduction of silicone rubber, which enabled surgeons to develop tubes that prevented shunts from routinely occluding in the peritoneum [40]. Jackson [14], in giving his specific reasons for reversion to VP shunting, noted that the previous failure was likely mechanical and not physiological. The advantages of this type of shunt include a lower mortality rate attributable to the shunt itself, ease and lower incidence of revision, and fewer and less serious complications [38, 41]. However, conflicting evidence from several sources suggests that these claims are subject to the individual author’s experiences and/or bias.

Despite the superiority of this technique that most sources report, several unique complications occur which show that VP shunts are not the best possible treatment. These include risk of inguinal hernia, abdominal ascites with cyst formation, perforation of a viscus, and danger of spreading neoplasm or infection to the peritoneal cavity [38]. Either type of shunt may result in slit ventricles from overshunting.

The technological improvements mentioned earlier fall into two categories. The first of these is the development of a suitable shunt material. Early reports by Dandy, Scarff, and others are filled with complaints of serious reactions to foreign materials such as rubber tubes placed within the body [9, 42, 43]. Dandy tried temporary placement of a rubber tube in the brain to hold open the cerebrospinal cavities, but the severe reaction to it inevitably and ‘ultimately necessitates removal at a second operation’ [42]. Even when these tubes remained, they often occluded rapidly. Then in World War II, polyethylene and silicones developed for insulating spark plugs on bombers proved nearly unreactive in the human body [40, 44]. The first silicone rubber specifically for medical purposes emerged in 1953. The most extensive single use of silicone rubber as a subdermal prosthetic device up until 1968 had been in VA shunting; the Holter valve in Pudenz’s ventriculocirculatory shunt system was implanted into an estimated 100,000 people during the 1960s. These shunts, made entirely of Silastic brand silicone rubber, are in contact with brain tissue, bone, muscle, and blood with no adverse effects [44]. The success of this refinement may indicate one of the few positive results that war may have.

With the employment of silicone rubber came the development of effective valves. The Holter valve, mentioned earlier, has been the most widely used. John W. Holter, an engineer with a hydrocephalic child, developed this valve over a period of a few weeks in 1956 in conjunction with neurosurgeon E.B. Spitz, quoted earlier. For the first time, VA shunts became feasible, and part of the revival of interest in shunting techniques for the treatment of hydrocephalus must be credited to the valves. Until this time, some still regarded hydrocephalus surgery as a ‘futile exercise’ [17]. Neurosurgeons have since developed other valves both with silicones and other materials such as stainless steel. The merits of silicone rubber – both for valves and tubes – are extensive: they are autoclavable, they do not deteriorate, nothing sticks to them (no danger of occlusion), and they cause the least tissue reaction of any known material [44]. More recently anti-siphon devices have been added as an additional improvement to the system.

How Far Have We Come?

‘Hardly any other pathological condition has been accorded more determined attention on the part of the medical profession with the aim of finding a cure for it than has hydrocephalus. And in hardly a single other condition have cures been so illusive or so often wrecked on purely mechanical obstacles. Yet the outlook is certainly not hopeless’ [43].

Contrast this 1929 statement of a concerned neurosurgeon to that of his modern fellow physician 50 years later:

‘Without question the development of simple and effective extracranial shunts stands today as the single most important advance in the surgical treatment of hydrocephalus. So greatly have these ingenious devices improved the outlook for patients with hydrocephalus that the results of early operations ... seem as dismal in comparison as the results of 19th century surgery must have appeared to Cushing and Dandy’ [17].
Today, the mortality rate of first operations is close to zero, and the 5-year survival rate approaches 90%. Untreated congenitally hydrocephalic infants, by contrast, have a 26% chance of survival to adulthood if they live to 3 months of age [39]. In addition, relief of hydrocephalus greatly improves the potential intellectual development of the child.

The single most important contribution to this improvement in treatment is the technological advances described earlier. As Boyd points out, ‘the problem has not been lack of understanding of causes of hydrocephalus nor of measures necessary to correct the condition – rather, the problem has been one of technology’ [14]. Also important is the greater technical skill of surgeons coupled with better facilities and general surgical procedures (sterility, antibiotics). Finally – and not least of all – is the human factor; the dedication of a large number of qualified, persevering, and ingenious neurosurgeons is clearly evident in the history of surgical treatment for hydrocephalus [6, 14, 39].

The issue of why shunting is currently the most popular form of surgical treatment for hydrocephalus requires a more complex answer. First, a review of the literature does not unequivocally suggest that shunting is the most effective form of treatment. Scarff’s exhaustive 1963 review of all reported surgical procedures up until that time included over 1,700 cases treated by choroid plexectomy, third ventriculostomy, ventriculocisternostomy, and a variety of shunt techniques. This influential report, widely cited in the literature, concludes that the ‘superiority of those operations ... which do not require the use of mechanical tubes and valves ... appears to be clearly established’ [9]. Scarff’s data indicate that, while operative mortality and initial successes were similar in both types of treatment, there were 10–20 times more serious late complications after shunting than after choroid plexectomy or third ventriculostomy. In a subsequent article, Scarff noted an incidence of 2.5% serious late complications for the ‘physiological’ treatments, and a rate of 57% for shunts [45]. He concluded that the evidence indicated the results of third ventriculostomy and choroid plexectomy are ‘considerably better than the results obtained with any of the so-called CSF shunts’ [46]. Jaksche and Loew [29], in 1986, came to similar conclusions regarding the merit of ventriculocisternostomy (mortality rate less than 1%) over extracranial shunts for selected cases.

Other researchers have questioned the validity of these claims. In evaluating Scarff’s 1963 review, the British Medical Journal doubted whether the two groups of patients (shunts vs. nonshunts) were even properly comparable, noting also that ‘if the earlier operations were as satisfactory as it appears it is difficult to see why such enthusiasm has been shown for the valves’ [47]. And a later neurosurgeon concedes that ‘meaningful assessment ... (of types of treatment) is surprisingly difficult’ [39]. Variables such as interval between onset and treatment, age at treatment, the issue of spontaneous recovery (40% estimated in some series), the skill and experience of the surgeon, the selection of which patients are allowed to undergo surgery, and many others must be taken into account. In addition, different studies judge with different standards, as Scarff indicated in 1970 with regard to choroid plexectomy. The merits of this method, he notes, are in its low incidence of serious late complications and high incidence of long-term survival, rather than a low operative mortality or high rate of initial successes [45], which are the methods of determining outcomes in some reports. Scarff’s success with this method is at least partly due to the long experience he had with it. That the operations endorsed by Scarff and Dandy have still survived is certainly related to the large influence these men, particularly Dandy, have enjoyed. Both Dandy’s 1918 article on choroid plexectomy and Scarff’s 1963 and 1970 reviews appear on the first pages of the prestigious journals in which they were published.

So why has shunting become the treatment of choice for hydrocephalus? The simple answer is that nothing else has worked as well. Other factors, however, have also played a role. It is likely that the impetus the invention of silicone rubber and effective valves has given to shunting techniques is of some significance. In modern society, the general trend toward seeing all new technological developments as improvements – regardless of their intrinsic merit – no doubt has a lot to do in this case as it did in the development of surgery and modern diagnostic techniques. A similar expectation in modern medicine for doctors to do something with obvious, visible results is also involved; an extracranial shunt system which produces an apparent ridge along the side of a shunted infant’s skull is much more readily appreciated than a single hole through a thin membrane in some obscure and hidden ventricular wall. One can draw analogies to similar situations in the past. Colonial blood-letting and purging, the rapid growth of surgery in the 19th century, the medicalization of child birth and growth of obstetrics, and the anti-contagionist sanitation movement all offer evidence of society’s felt need to see dramatic effects. At the same time, the current state of medical practice and litigation may discourage the type of bold contribution made 40 years ago by John Holter in designing the first slit valve.
for his acutely ill child [48]. And the fact that manufacturers are reluctant to wait several years for the results of clinical trials may contribute to the lack of initiative on the part of industry [35]. Both of these processes would tend to inhibit the development of radical new treatments.

In summary, then, most surgeons have adopted shunting because it is the most effective treatment available at this time. However, this choice is mostly a matter of elimination, and it is clear that better techniques can and should be devised. As the title of one recent article suggests: How to keep shunts functioning, or ‘the impossible dream’ [49]. Even Milhorat, who hailed shunts as ‘the single most important advance in the surgical treatment of hydrocephalus’, states also that ‘what is clear is that the introduction of catheters and valves into the body cavities has inherent disadvantages ...’ [17]. Extracranial shunts are associated with a high incidence of complications, and in a growing child, a single shunt operation is rarely definitive; some unhappy cases have undergone seven or eight revisions in the same number of months, as any pediatric neurosurgeon can attest. Even successfully treated children are still susceptible to significant shunt-related problems as adults [50]. Also importantly, the burden of shunting procedures on the health care system should not be overlooked. A 1995 article cited the figure of 33,000 shunt placements per year, with an annual cost for such procedures of nearly $100 million [51].

Current efforts to improve the outcome of surgical treatment of hydrocephalus are directed mainly at refining the shunt systems. These advances include impedance matching, anti-siphon devices, and variable resistance valves. Such improvements should eliminate or reduce problems with over- or underdrainage and proximal occlusion, but still retain the potential for infection, distal obstruction, and the need for multiple revisions in a growing child.

Despite the continued focus on shunts, most of the other early operations have also reappeared recently. Ventriculocystoscopy, first attempted several decades before shunts became popular, has resurfaced as a method with diagnostic and therapeutic merit for hydrocephalus as well as other neurosurgical diseases [52, 53]. Studies describing coagulation of the choroid plexus [27], ventriculocysternostomy [16, 29–31, 54], third ventriculostomy [22–24], and even the use of compression bandages to promote reabsorption of CSF [55] have all recently been described. These studies provide further evidence of the continuing search for new strategies.

We can anticipate that in the future, the simpler, physiological techniques advocated by Dandy and Scarff can be developed further. There is also the prospect of treating hydrocephalus long term with drugs that would substantially reduce CSF formation without causing serious metabolic shifts. The occurrence of spontaneous arrest in some hydrocephalus patients is intriguing; the possibility of extending this process to other cases has been suggested [56]. To develop new therapies based on these principles will require a thorough understanding, even down to the molecular level, of the basic physiological and pathological processes that result in the various forms of hydrocephalus. This is especially true of the non-obstructive form of the disease, for which two mouse models have recently been described [57, 58]. These alternatives offer promise of better results, and there is need to investigate all of them if we hope to establish a more effective and useful treatment for this challenging disease.

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