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Harvey Cushing's Early Management of Hydrocephalus: An historical picture of the conundrum of hydrocephalus until modern shunts after WWII

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Abstract

Purpose—Throughout his early career, Cushing proposed a variety of methods for temporary and permanent drainage and diversion of CSF in his patients, and acknowledged that certain techniques were more suited to particular subsets of hydrocephalus.

Methods—Following IRB approval, and through the courtesy of the Alan Mason Chesney Archives, the surgical records of the Johns Hopkins Hospital, from 1896 to 1912, were reviewed. Patients operated upon by Harvey Cushing were selected for further analysis. Within this cohort, we recovered all available records for a single patient with hydrocephalus and spina bifida, who was treated with a ventriculosubgaleal shunt prior to repair of the spina bifida.

Results—A 3 month-old infant presented with hydrocephalus associated with spina bifida. Cushing performed serial lumbar and ventricular punctures. Following this, Cushing took the patient to the operating room for placement of a ventriculosubgaleal shunt. The patient subsequently underwent excision of the myelomeningocele sac, with post-operative mortality due to unspecified causes.

Conclusions—Cushing's publications document a preference for translumbar-peritoneal drainage in patients with congenital hydrocephalus, particularly those with spina bifida. Although the placement of ventriculosubgaleal shunts has become an accepted practice for contemporary neurosurgeons, this case illustrates the challenges that early neurosurgeons faced in developing operative approaches for the treatment of congenital hydrocephalus.

Keywords

Cushing; Congenital Hydrocephalus; Spina Bifida; Ventriculosubgaleal Shunt

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Introduction

Congenital hydrocephalus has been a documented ailment of childhood since ancient times, with Hippocrates offering the first recorded description [1], and reported to have performed drainage of cerebrospinal fluid (CSF) through transcranial ventricular puncture [2]. Attempts at drainage or diversion of CSF for the treatment of hydrocephalus were made by physicians throughout history, culminating in a multitude of operative techniques proposed by surgeons at the turn of the twentieth century. Quincke suggested drainage through serial lumbar punctures in 1891 [3–5], and von Mikulicz and Senn documented permanent drainage into the subgaleal space in 1893 and 1908, respectively [3–5]. Although the optimal approach to draining CSF in patients with congenital hydrocephalus remained a topic for debate, the necessity of doing so was well established by the time Cushing developed his neurosurgical practice at the Johns Hopkins Hospital. Throughout his early career, Cushing proposed a variety of methods for temporary and permanent drainage and diversion of CSF in his patients, and acknowledged that certain techniques were more suited to particular subsets of hydrocephalus[5,6]. While Cushing frequently employed translumbar-peritoneal drainage in pediatric patients with hydrocephalus associated with spina bifida, he attempted a single ventriculosubgaleal shunt during his early career at the Johns Hopkins Hospital, which we report here.

Methods

Following IRB approval, and through the courtesy of the Alan Mason Chesney Archives, the surgical records of the Johns Hopkins Hospital, from 1896 to 1912, were reviewed. Patients operated upon by Harvey Cushing were selected for further analysis; within this cohort, a single patient with hydrocephalus and spina bifida, who was treated with a ventriculosubgaleal shunt prior to repair of the spina bifida, was recovered.

Case Report

On December 18th, 1908 a 3 month-old infant presented with a chief complaint of spina bifida, with concurrent hydrocephalus. A detailed birth history was taken at the time of admission:

“Full term child. Normal easy labor. No instruments at birth. The spina bifida was of course pronounced and has remained unchanged [...] Even at birth child had a large head and during the past few weeks this enlargement has considerably increased. Hydrocephalus now is well pronounced.”

On physical examination, detailed description of the child’s head were made: “Occipital-Front diam 18 cm. Circumference – Occip-Front – 43 cm. Bi-pariet (suboccip-breg) 41 cm. Fontanelles widely open – dura feels tense. Sutures of calvarium not closed.”

On December 21st, the patient’s ventricle was tapped by Cushing; no improvement was noted. On January 8th, 1909, Cushing brought the child to the operating room for “drainage left lateral ventricle c [sic] tube under scalp.” His operative note describes the procedure (Figure 1):

“A horseshoe flap of scalp measuring possibly 5 cm. in its two diameters was turned back, exposing the left parietal eminence. On retracting the base of the flap a point possibly 3 cm. from the mid line was exposed and the bone was penetrated with a perforator. Through this opening a sharp aspirating needle was introduced and then a small drainage tube measuring 2 cm. in length and possibly 2 mm. in diameter. The tube was introduced with a stilette which prevented the single terminal opening from becoming plugged. On removing the stiletto clear cerebro

spinal fluid spurted from the external opening. The flange was so placed as to lie flat upon the surface of the bone and epicranium. The scalp was then replaced and sutured without drainage.”

On the second post-operative day, the patient developed “a consid [sic] rise in temp [...] and a macular rash appeared over abdomen, neck and [illegible]. Pt was given some HgCl+. Temp went down tonight.” On the following day, January 10th, the wound was inspected and found to be clean and free of infection. In addition, the notes describe “considerable area of oedema over left half of scalp where there is definite pitting on pressure.” On January 12, the stitches were removed, and the child was described as “apparently doing very well.”

On January 19th, almost two weeks following the operation, the chart documents the state of the ventriculogaleal shunt:

“The patch of oedema in scalp has largely disappeared leaving only a local bulging (fluid) over needle and under flap. Mass behind is very full and more tense than before. Fontanelle also bulging.”

The meningocele sac was tapped at that time, and on January 20th, Cushing brought the patient to the operating room for “excision of meningo-myelocele.” Post-operatively, the child developed a temperature, which resolved a week after the operation.

The patient continued to regurgitate food, and was placed on a diet of milk and water, followed by a mixture of milk and “beef juice,” with slight improvement. On February 19th, the ventriculogaleal shunt was re-evaluated:

“The outer [illegible] of the ventricular drainage tube – edema definitely circumscribed – about 2 ½ cm in diam. No surrounding boggy of scalp – patch of fluid about end of drainage tube feels as well walled off as a cyst.”

The following day the patient died, “apparently of [illegible] [...] No convulsions. No evident increase in intra-ventricular tension.” No post-mortem examination was performed.

Discussion

Methods for draining cerebrospinal fluid in cases of congenital hydrocephalus were developed in the late nineteenth century, with Quincke advocating the serial tapping of the spinal meninges [3,4,1,5], Keen attempting external drainage[7], von Mikulicz establishing drainage into the subgaleal space by means of a gold tube [4], and various attempts made at reestablishing ventricle communication through corpus callosum puncture[1,5,6]. These methods met with limited success, and patients succumbed to complications of the operative interventions or increased intracranial pressure from incompletely treated hydrocephalus[4].

Cushing commented on these attempts in his 1908 sections for W.W. Keen’s text *Surgery: Its Principles and Practice*, noting in particular the inadvisability of ventriculogaleal shunts in patients with congenital hydrocephalus[6]. Cushing went on to describe his detailed technique for establishing retroperitoneal drainage of the lumbar meninges in patients with congenital hydrocephalus, concluding that “this combined lumbar method is especially desirable in cases of hydrocephalus complicated by spina bifida” [6].

Interestingly, less than a year after this publication, Cushing performed the operation described here: a ventriculogaleal shunt for drainage of hydrocephalus associated with spina bifida. The surgical records indicate that this was Cushing’s sole attempt at the procedure, and he was disappointed with the results, which he commented briefly upon in his 1928 address: “The efforts which were made many years ago to drain the hydrocephalic

ventricles by establishing a communication with the subaponeurotic layer of the scalp [...] led to disappointments” [5]. Cushing favored translumbar-peritoneal drainage, and had performed the procedure for seven years prior to the case reported here. However, Cushing was concerned that patients treated through that approach had experienced mortality due to intestinal intussusception, and in 1928 he stated that “the procedure came to be discarded” [5]. This unexpected complication may have led Cushing to pursue alternate methods of drainage in the patient presented here.

In the 1928 address, Cushing commented that the translumbar-peritoneal drainage was introduced because “It was thought that this loose tissue in direct proximity to the thoracic duct would be a medium more favourable for permanent drainage than the subaponeurotic layer of the scalp” [5]. The early surgical records do not indicate that Cushing attempted a ventriculosubgaleal shunt for temporary CSF diversion; however, contemporary neurosurgeons frequently employ these shunts as temporizing measures for hydrocephalus secondary to trauma [8,9], or in premature neonates for treatment of post-hemorrhagic hydrocephalus [10,11].

The development of options for the permanent drainage of CSF in cases of pediatric hydrocephalus offered patients at the turn of the twentieth century hope, but the complications from the materials available and from the operative procedure itself prevented many long-term successes. Cushing himself attempted to evade these pitfalls by employing a transplanted vein as a shunt catheter [12]; this provided a more pliable material than the traditionally used silver tubes, and offered the added benefit of having built-in valves, which prevented retrograde flow. However, this method did not become widespread, and the adequate long-term management of hydrocephalus in the pediatric population remained a challenge for neurosurgeons well into the twenty-first century. The development of plastic and silicone polymers provided improved options for shunt construction during the 1940s [13]. The development of synthetic shunts containing valves, pioneered by Holter and Spitz, marks the beginning of the contemporary management of hydrocephalus [14,13]. The continued challenges of infection, malfunction, and a myriad of other complications presented by synthetic shunt systems in pediatric hydrocephalus have led neurosurgeons to develop additional operative approaches to the disease, including the endoscopic third ventriculostomy, and to pursue the development of more sophisticated shunt catheters to reduce shunt-associated infections.

In reviewing the developments in understanding the CSF pathways, from the first lumbar puncture through the first callosal puncture, Cushing offered the insight that “contempt of these procedures has come only with familiarity, and we soon forget that they were once so novel that their performance took courage”[5]. Although the placement of ventriculosubgaleal shunts has become an accepted practice for contemporary neurosurgeons, this case illustrates the challenges that early neurosurgeons faced in developing operative approaches for the treatment of congenital hydrocephalus.

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Jan. 9, 1900, } *Drainage Left lateral*
 Operation, } *ventricle & tube under*
 Dr. Cushing, } *scalp.*
 Primary chloroform. (Davis)

A horseshoe shaped flap of scalp measuring possibly 5 cm. in its two diameters was turned back, exposing the left parietal eminence. On retracting the base of the flap a point possibly 3 cm. from the mid line was exposed and the bone was penetrated with a perforator. Through this opening a sharp aspirating needle was introduced and then a small drainage tube measuring 2 cm. in length and possibly 2 mm. in diameter. The tube was introduced with a stilette which prevented the single terminal opening from becoming plugged. On removing the stilette clear cerebro spinal fluid spurted from the external opening.

The flange was so placed as to lie flat upon the surface of the bone and epicranium.

The scalp was then replaced and sutured without drainage.

(Dr. Cushing).

Figure 1.

Cushing's operative note, documenting the approach used for placement of a ventriculosubgaleal shunt in a 3 month-old child with congenital hydrocephalus associated with spina bifida.