Summary: To trace the history of the treatment of hydrocephalus is to document the parallel development of medicine as a whole; when one reviews the treatment of hydrocephalus, the integral relationship between basic science and therapy is reaffirmed. The treatment of hydrocephalus, over the centuries, underwent three stages of evolution. Prior to the late 19th century, treatment for “water on the brain” involved more observation than intervention. During antiquity, middle ages and renaissance, hydrocephalus was not understood. Medical treatment was useless; surgery was hopeless. The second stage extends from the 19th century to the end of the first half of the 20th century. Cerebrospinal fluid circulation was now understood; surgery however, remained inefficient, but some patients survived with arrested hydrocephalus. The third stage begins in the nineteen fifties with the development of silicone shunts with a valve. Surgery transforms the prognosis of hydrocephalus, but the number of post-operative complications creates new problems. The different attempts that have been made during these past two decades to solve these problems are reviewed. They have resulted in a reduction of the mechanical and infectious complications. CSF overdrainage has been minimized. Percutaneous ventriculo-cisternostomies have in some cases replaced shunts. In the future, to improve outcome in these hydrocephalics, surgery, when indicated, should be performed as early as possible. Knowledge and prevention of the causes of hydrocephalus should be developed.

Key words: Hydrocephalus, history, surgery, cerebrospinal fluid, shunts.

INTRODUCTION

The treatment of hydrocephalus, over the centuries, underwent three stages of evolution. During antiquity, middle ages and renaissance, hydrocephalus was not understood. Prior to the late 19th century, treatment for “water on the brain” involved more observation than intervention (1). Medical treatment was useless; surgery was hopeless. The second stage extends from the 19th century to the end of the first half of the 20th century. Cerebrospinal fluid (CFS) circulation was now understood; surgery however, remained inefficient, but some patients survived with arrested hydrocephalus. The third stage begins in the nineteen fifties with the development of silicone shunts with a valve. Surgery transforms the prognosis of hydrocephalus, but the number of post-operative complications creates new problems. The different attempts that have been made during these past two decades to solve these problems are reviewed. They have resulted in a reduction of the mechanical and infectious complications. CSF overdrainage has been minimized. Percutaneous ventriculo-cisternostomies have in some cases replaced shunts. In the future, to improve outcome in these hydrocephalics, surgery, when indicated, should be performed as early as possible. Knowledge and prevention of the causes of hydrocephalus should be developed.

ANCIENT TIME

In the ancient medical literature hydrocephalus was not often described although its existence and symptomatology were well known. Hippocrates (5th century B.C.), the father of medicine, is thought to be...
the first physician to attempt and document the treat-
ment of hydrocephalus (2, 3). He is often cited as the
first to have performed ventricular punctures as it is
possible he was merely draining the subdural or suba-
rachnoid space. Hippocrates recommended trepanning
for the treatment of epilepsy, blindness and possibly
hydrocephalus. Further description of this condition
can be found in the works of Galen (130–200 A.D.). He
believed this condition was caused by an extraxial ac-
cumulation of cerebrospinal liquid rather than enlarge-
ment of the ventricles. This belief led to many erroneo-
us diagnoses and treatments. He recounted examples
and described the thinness of the brain and skull asso-
ciated with this condition (4). He found the ventricles
to be in communication with each other and believed
that the “soul” contained within these structures under-
go a purification process with the waste being depos-
ited in the pituitary gland. The Greeks reportedly trea-
ted hydrocephalus by twisting bark around the pati-
ent’s head and inserting it into trephined openings (5).

Most detailed descriptions of hydrocephalus in-
cluding the surgical treatment are extant in the
encyclopaedic works on medicine of the physicians
Oreibasios and Aetios from Amida from the 4th
and 6th centuries A.D. Due to the lack of autopsies in ancient
times, the hydrocephalus was never linked to the path-
ology of the ventricles. All forms of hydrocephalus
were believed to be caused by improper handling of the
head by the midwife during delivery. Only the extrac-
reranal fluid collections were considered to be suitable
for surgical treatment. The surgery consisted in one or
more incisions and evacuation of the fluid. The wound
was not closed but let open for three days. The plasters
or sutures closed the incisions. The surgical technique
goes back probably to Antyllos a surgeon from the 3rd
century A.D. whose considerations were cited in the
work of Oreibasios. The early Arabic physicians took
over the surgical indications, the operative technique
and modified the Greek concept of hydrocephalus (6).

**MIDDLE AGES**

In the Middle Ages, the Arabic surgeon Abu al-Qa-
sim Khalaf ibn al-Abbas Al-Zahrawi (936–1013)
known in the medical literature as Albucasis, wrote a
the Kitab al-Tasrif (1000). In this 30-volume medical
encyclopedia which was taught at Muslim and Euro-
pean medical schools until the 17th century, he touched
on many aspects of neurosurgery, including the diag-
nosis and treatment of hydrocephalus. Evacuation of
superficial intracranial fluid in hydrocephalic children
was first described in detail by Albucasis (7). Abu Ali
al-Husain ibn Abdallah ibn Sina, is often known by his
Latin name of Avicenna, separated the traumatic haem-
atoms outside the skull from the term hydrocepha-
lus. Avicenna, had not linked hydrocephalus with the
ventricular system. Haraf 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 Ger-
man surgeon Hildananus describe the same outcome at
the turn of the 17th century (8).
Andreas Vesalius (1514–1564), a Flemish anatomist, revealed as a single pathology an extremely dilative ventricular system filled with water-like fluid which made it necessary to change completely the ancient concept of hydrocephalus (6). The autopsy of a child with an exorbitant hydrocephalus performed by the anatomist Vesalius at the University of Padua clarified many of the anatomical and pathological characteristics of hydrocephalus, noting that in one of his patients, “the water had not collected between the skull and its outer surrounding membrane, but within the ventricles of the brain” (9).

In 1664 Thomas Willis (1621–1675), neuroanatomist, considered by many to be the father of neuroscience was the first to suggest that the choroid plexuses produced CSF, contrary to the major paradigm at that time, which held that the ventricles contained a vapor during life and, after death, condensed and gravitated to the spaces in and around the brain and spinal cord (10, 11). In 1701, Antonio Pacchioni (1665–1726), an Italian scientist and anatomist, described the arachnoid granulations, which he falsely believed were the source of CSF production (12). In 1761, Giovanni Battista Morgagni (1682–1771), an Italian anatomist, wrote in Seats and Causes of Diseases that hydrocephalus could occur without accompanying head enlargement; however, he did not know the source of the excess fluid in this disease process. Monro illustrated the presence of the paired intraventricular foramen. In Observations on the Dropsy in the Brain, written in the middle 18th century, Robert Whytt (1714–1766) first described hydrocephalus as a disease, illustrating several cases of internal hydrocephalus caused by tuberculous meningitis (13).

Because of the poor understanding of the pathophysiology of hydrocephalus, initial therapeutic attempts were sporadic and generally resulted in failure. Given the dismal prospects of surgical therapy, many practitioners relied on conservative medical treatment. Attempted treatments included multiple medications and purgatives such as rhubarb, jalop, calomel and oil, as well as various diuretics, injection of intraventricular iodine, head wrapping, blood letting, and skull trephination. The use of carotid artery ligation was also reported. One can speculate that cures were rare and treatment fraught with complications (14). One idea was that external pressure may reduce fluid accumulation, and thus various means of compression were applied to the enlarged skull (15). Rubber bandages provided stronger tension and constant pressure compared to other materials. However, pressure on the skin caused decubitus ulcer and the resulting increased intracranial pressure created decreased CSF circulation and even seizures and skull fractures.

In the 19th century, the understanding of the anatomy and physiology of the ventricles and the CSF was advanced remarkably. François Magendie (1783–1855), a French physiologist, considered a pioneer of experimental physiology. In 1825 Magendie, in several important papers, illustrated the medial cerebellar foramen and eloquently described the circulation of CSF within the brain (16, 17). A German anatomist, Hubert von Luschka (1820–1875), in 1859, confirmed the presence of the foramina of Magendie and described two additional lateral foramina. A milestone in the understanding of CSF circulation was the classic anatomical atlas of Key and Retzius in which they described in detail the meninges, the subarachnoid spaces and cisterns, the ventricles and the arachnoid villi, virtually the entire circulation of the CSF from production to absorption.

In the early 20th century, Lewis Weed described the embryology of the choroid plexus and confirmed the absorptive capacity of the arachnoid villi. Weed’s research dealt largely with cerebrospinal fluid and with the development of the membranes that surround the central nervous system. He discovered the origin of the cerebrospinal fluid and mapped out its circulation, an accomplishment which led to a number of important clinical developments (18). Concurrent with the physiological advances made during this period, a new understanding of this disease process was further elucidated.
ted in the classic work *Observations on the Pathology of Hydrocephalus* Dorothy Russell provided an encyclopedic collection of hydrocephalic specimens (19). These descriptions were to have great influence on the future therapeutic modalities for this disorder.

As the 20th century progressed, more defined investigations into the physiology of CSF dynamics and hydrocephalus became possible. The introduction of radioactive tracers in the 1950s allowed for the detailed analysis of the circulatory dynamics of CSF. Papenheimer’s perfusion method helped establish the rates of CSF production and absorption, while elucidating the extrachoroidal formation of CSF. Igor Klatzo (1916–2007) demonstrated that this movement was caused by bulk flow. In 1970, Thomas Herrick Milhorat illustrated the increase in periventricular permeability and the concept of transependymal absorption inexperimental hydrocephalus (20). This was later found to correlate with periventricular low densities observed on computerized tomography scans obtained in patients with untreated hydrocephalus.

A further milestone in understanding hydrocephalus came with the discovery that acute hydrocephalus could develop within hours in contrast to weeks or months, which was the prevailing paradigm. Effective therapy requires aseptic surgery as well as pathophysiological knowledge—both unavailable before the late nineteenth century. In 1881, Carl Wernicke (1848–1905), a German physician, anatomist and neuropathologist, inaugurated sterile ventricular puncture and Cushing devised a technique in which the lumbar subarachnoid space was connected to the peritoneal cavity (23). Kausch used a rubber conduit to drain the lateral ventricle into the peritoneal cavity (23).

This concept, however, did not receive much initial enthusiasm. During this time, Heile attempted to perform spinal CSF drainage into the peritoneum by sewing the serosa of the bowel to the dura mater, connecting the subarachnoid space to the peritoneum by use of a silk suture, and by using other conduits such as veins or latex rubber tubes. He also was the first credited with CSF diversion to the urinary system (24).

Harvey Williams Cushing (1869–1939), an American neurosurgeon and a pioneer of brain surgery, paid tribute to this notable work, naming it the “third circulation”. Under the pioneering efforts of Cushing and his followers, neurosurgery emerged as a distinctive specialty. Cushing devised a technique in which the lumbar subarachnoid space was connected to the peritoneal cavity or retroperitoneum by using silver cannulas passed through apertures through the L-4 vertebral body. Cushing can also be credited with the innovative idea (for that time) that as the “third circulation”; the CSF had unique function greatly more complex than simply providing buoyancy for the brain. In 1914, Walter Edward Dandy (1886–1946) and Kenneth D. Blackfan (1883–1941) developed a technique of producing experimental obstructive hydrocephalus in dogs by placing cotton pledgets at the distal aqueduct of Sylvius, thereby causing proximal ventricular dilation (25). Dandy also reported that with unilateral choroid plexectomy and obstruction of the foramen of Monro, the plexectomized ventricle would collapse while the contralateral ventricle would dilate; he concluded that CSF was produced exclusively by the choroid plexus. This in turn led Dandy to introduce, in 1918, bilateral choroid plexectomy as a means of reducing CSF production (26).
Charles Putnam Symonds (1890–1978) and John Edwin Scarff (1898–1978) expanded on this technique by including endoscopic cauterization of the choroid plexus in the late 1930s and early 1940s (27, 28). Upon review, however, in the majority of patients the ventricles demonstrated progressive enlargement at the same or greater rate than that observed preoperatively with disappointingly poor results; thus, by the 1950s, these techniques had largely been abandoned. The placement of intracranial shunts was also investigated.

Third ventriculostomy was introduced by Dandy to bypass aqueductal stenosis, and this technique was later refined by Stookey and Scarff (29). In their procedure the lamina terminalis was approached via a subfrontal or subtemporal route through the interpeduncular cistern into the floor of the third ventricle. Although the mortality rate was somewhat high, the reported arrest of hydrocephalus in surviving patients was approximately 70%. This technique was further refined with the use of endoscopes. Arne Torkildsen devised a procedure in which a shunt was placed from the lateral ventricle to the cisterna magna (ventriculocisternostomy); initially the success rate was high but so too was surgery-related morbidity, which was subsequently reduced (30).

Efforts to divert CSF to remote body cavities were also advanced. Ureteral diversionary procedures — from the ventricles and lumbar subarachnoid spaces — were reported by Matson. Although it was associated with a very low mortality rate, this procedure did require a nephrectomy and was complicated by both infection and electrolyte abnormalities, particularly troublesome in infants. The concept of valves and flow regulation was reinforced by this procedure (although the idea had its roots in the work reported by Payr in which he used venous valves); Matson believed that the success of the technique lay in the natural valve function provided by the ureter. Other attempted spaces included the heart, jugular vein, thoracic duct, pleural space, gallbladder, fallopian tube, ileum, and salivary ducts. Over time, the right atrial and peritoneal spaces became the locations of choice for shunts. Attempts at medical cures or symptomatic arrests were made during this period. Reports of thyroid extract, vital dyes, and various diuretics found their way into the clinical practice in the early part of the 20th century but lost favor because it became more apparent that hydrocephalus was primarily a disease best treated with surgery although it was without definitive cure (9).

The development that ushered in the modern era of hydrocephalus surgery was the introduction of valve regulated shunts and biocompatible synthetic materials in 1952.

In 1952 Frank Nulsen and Eugene Spitz, residents in neurosurgery at the Hospital of the University of Pennsylvania, USA, working in conjunction with John Holter, the father of a child with hydrocephalus, reported the successful use of a ventriculo-jugular shunt regulated by a spring and ball valve. At approximately the same time Pudenz produced a one-way slit valve made of silicone (31).

The development of the valve system combined with the application of new bioavailable materials allowed for the safe and reliable diversion of CSF without many of the complications of unregulated CSF drainage. Ames (32) and Raimondi (33) resurrected the concept of ventriculoperitoneal procedures in which these new devices were used. In the 30 years since this resurgence, great advances and modifications in hardware have been realized. There are now literally hundreds of options for valves, proximal and distal catheters, antisiphon devices to prevent overdrainage, and, more recently, programmable valves for fine-tuning CSF flow rates.

NEUROENDOSCOPY

Concomitant with the advance in shunt-related materials, progress in imaging technology has further allowed clinicians to treat hydrocephalus with greater success and safety. In the 1980s and 1990s the use of an endoscope again found a role in neurosurgery, the benefits of which include more accurate placement of ventricular catheters and a resurgence of the third ventriculostomy for aqueductal stenosis (34). In the 1990, there has been a renaissance of endoscopic ventriculostomy, which is widely accepted as a method of first choice in adult patients with aquired or late-onset, occlusive hydrocephalus.

The treatment of hydrocephalus nowadays is still a challenge for neurosurgery. Neuroendoscopy is a valuable alternative of the CSF shunts in hydrocephalus management. Because of the complicated and always changeable pathophysiology of hydrocephalus, the history of the endoscopic treatment of hydrocephalus is also a history of severe frustrations, great expectations, and significant achievements. The historical milestones and state of the art of neuroendoscopic treatment of hydrocephalus are reviewed for each of its surgical techniques: choroid plexus coagulation, third ventriculostomy, aqueductoplasty, septostomy, foraminal plasty of the foramen of Monro, and foraminal plasty of the foramen of Magendie. The future trends of neuroendoscopic treatment of hydrocephalus such as robotics, image-guided neuroendoscopic surgical techniques, treatment “in utero”, application of stem cell therapy, implementation of new technological solutions, and so on are discussed in the light of the approaching end of the century of neuroendoscopy (35).
Stereotactic localization has led to more functional forms of therapy and safer approaches for the drainage of CSF. Furthermore, with the advent of prenatal ultrasonography, diagnosis of hydrocephalus in utero has led to attempts with intrauterine fetal surgery. The rationale for this intervention is that early surgery can prevent progressive injury from ongoing pathophysiology or from secondary damage in the intrauterine environment. In the realm of neurosurgery, attempts had been performed in the late 1970s and early 1980s to treat hydrocephalus diagnosed in utero. Procedures such as ventriculoamniotic shunts and serial cephalocenteses were attempted to curb the ventriculomegaly (36).

High morbidity and mortality rates, however, marked these early attempts at treatment, and outcomes were generally worse than in those in whom shunting procedures were performed in the neonatal and infant periods. Currently, there is a defacto moratorium on fetal surgery to treat hydrocephalus, as the issues of patient selection and surgical procedure remain in question. Fetal therapy for spinal dysraphism — associated hydrocephalus has seen an emergence in recent years, spurred on by advances in neuroimaging, better understanding of the pathophysiological nature of the disease, and the refinements of surgical techniques including endoscopy.

Bruner have reported the first cases of intrauterine closure of a myelomeningocele in 1997. The suspected benefits of this early intervention include decreased hind brain herniation, improvement of lower extremity function, and the decreased need for shunts (36).

CONCLUSION
Whereas great advancements and achievements have been made over the course of medical history, clinicians in the new millennium will be required to continue face the challenges of presented by hydrocephalus. It appears that treatment up to this point and time has focused on the arrest of the disease process, with further therapy focused on the complications of these treatment modalities. With current research in molecular biology, gene therapy, and neural regeneration, the concept of a functional cure may become an achievable goal. As in the past, the integration of the discoveries in basic science and clinical innovation will continue to lead the path as it has in the past.

ABBREVIATIONS
CSF — cerebrospinal fluid

Sažetak

HIDROCEFALUS — ISTORIJAT HIRURŠKOG LEĆENJA KROZ VEKOVE

Aleksandar J. Milojević, Branka S. Radojičić, Igor D. Melnjikov


Ključne reči: hidrocefalus, istorija, hirurgija, cerebrospinalna tečnost, šant.
REFERENCES


Correspondence to/Autor za korespodenciju
Branka Radojčić, MD, PhD
Clinic for Pediatric Surgery
Institute of Child and Youth Health Care of Vojvodina
Hajduk Veljkova 10
21000 Novi Sad
Serbia
Tel: +381 69 11 50 900
Fax:+381 21 6366 222
E-mail: branika.radojicic@gmail.com